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Possible Cases of Leprosy and Tuberculosis in Medieval Sigtuna, Sweden

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ABSTRACT In Sigtuna, Sweden, a medieval cemetery, including 227 skeletons, was analysed in 2006. On the outskirts of the churchyard, six skeletons with bone changes indicating systemic inflammatory disease were observed. Two out of three individuals with well-preserved facial bone regions displayed signs of rhinomaxillary remodelling. Four of the afflicted exhibit severe bilateral alterations of the lower legs and phalanges of the feet and concentric atrophy of the metatarsals. In addition, one of the individuals exhibited a kyphosis in the lumbar vertebrae. In a discussion about alternative diagnoses, lepromatous leprosy and tuberculosis were identified as the causes of the destructive lesions in two individuals. Though the skeletal changes of the lower legs and feet in four cases demonstrate a close resemblance to secondary lesions of leprosy, the disease could not be confirmed. The skeletal changes of the last individual were unspecific and the possible causes several, rendering diagnosis difficult.

The burial locations imply that the afflicted persons belonged to a lower social stratum. Due to the significantly higher frequency of pathological changes in the cemetery compared to other cemeteries in the town, the individuals could be regarded as fellow sufferers among others with various medical conditions. The bioarchaeological identification of systemic infectious diseases of a group of individuals of this size is unique to north^{Q1} of Scania in Sweden, where only a few cases of leprosy and tuberculosis have previously been diagnosed. The significance of the present study is emphasised by the interconnection between the afflicted, the archaeological context and the knowledge of the medieval society in Sigtuna. Copyright © 2010 John Wiley & Sons, Ltd.

Key words: leprosy; social status; systemic disease; tuberculosis

Introduction

Leprosy or Hansen's disease is a chronic infectious disease caused by the bacteria *Mycobacterium leprae*, which create granulation affecting foremost the skin and nerves but also lymph nodes, eyes, mucous membranes and internal organs. The infection is believed to spread mainly through respiratory droplets or possibly via prolonged direct contact with ulcers or open wounds in a person with leprosy (Resnick & Niwayama, 1988: 2688). While leprosy has been diagnosed among all age groups, it is often manifested in individuals less than 20 years of age and there seems to exist a sex-specific prevalence where men are afflicted more often than women (Resnick & Niwayama, 1988: 2688). The long incubation time, often 3–6 years, and the low virulence and pathogen-

icity of the bacteria may result in its taking two decades before there are visible signs and symptoms of the disease.

The term 'leprosy' has been used widely and imprecisely throughout history and came to represent a broad spectrum of aspects in the history of medicine and cultural history, thereby complicating the discussion about the clinical disease (e.g. Heller *et al.*, 2003; Millner & Smith-Savage, 2006; Demaitre, 2007). Comparative genomics indicate that the disease [al-ready^{Q2}](#) spread from the African continent during the Pleistocene era (Monot *et al.*, 2005), although this hypothesis has been criticised (Pinhasi *et al.*, 2006). A recent comparative genomic and phylogenetic analysis of *Mycobacterium leprae* shows a strong correlation between the spread of the pathogen and early human migration patterns (Monot *et al.*, 2009). Written records arguably dealing with the disease go back to 2000–1500 BC while more convincing descriptions are dated to 600 BC (for a review, see Zias, 2002). Possibly brought from eastern Asia to the Mediterranean by the

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troops of Alexander the Great, leprosy spread on a larger scale to Europe during the fourth century BC (Roberts & Manchester, 2007). The disease must have been considered a serious health problem since *leprosaria* were founded in Rome already in the 4th century (Browne, 1975) and in France around AD 460 (Møller-Christensen, 1961). During the Middle Ages, as the disease became endemic in Europe, the awareness of the disorder increased. In the 13th century, the English physician Gilbertus Anglicus (1180–1250 AD) describes leprosy in his work *Compendium Medicinæ* as a contagious disease (Rawcliffe, 2006). The identification and description of two types of leprosy (corresponding to the tuberculoid and lepromatous form) by bishop and surgeon Theodoric of Cervia (1205–1298) show that interest in the disease was extensive (Steger & Barrett, 1994: 324). During the same century the numbers of *leprosaria* in Scandinavia increased dramatically. In Denmark alone, about 31 institutions dedicated to Saint George were founded (Bennike, 2008). With time, the frequency of institutions exclusively focused on treatment of people with leprosy dropped in Europe; whether from a decline in leprosy or a more accepting society, business closed down or merged with houses of the Holy Ghost (Ljung, 1961). A combination of several demographic, environmental, social and genetic factors is believed to have caused the decline (Rawcliffe, 2006; Magilton^{Q3}, 2008; Rubini & Zaio, 2009). The Black Death (c. 1347–1350) killed millions of Europeans: those disabled or ill were most likely the first victims (Steger & Barrett, 1994). There seems to have been a negative correlation between leprosy and tuberculosis after the Middle Ages where the number of leprosy sufferers declined and the number of those with tuberculosis rose. Bio-molecular studies have demonstrated a close relationship between *Mycobacterium tuberculosis* and *M. lepræ* (Donoghues *et al.*, 2005). It has been suggested that exposure at an early age to the more easily transmitted pathogen *M. tuberculosis* could, to a certain degree, prevent the establishment of clinical leprosy (the cross-immunity hypothesis) unless the immune system of the host was compromised by undernourishment or other infections (Manchester, 1991; Lechat, 2002). Though this hypothesis has not been completely verified (Wilbur *et al.*, 2002), several studies from the early and mid-20th century have demonstrated a protective effect for leprosy when using the BCG (*Bacille Calmette-Guérin*) vaccination to prevent progression to active tuberculosis (for a review, see Donoghues *et al.*, 2005; Roberts & Manchester, 2007). Furthermore, social factors in the form of increased population density and urbanisation may have generated a greater incidence of a virulence

disease such as tuberculosis than a disease of low pathogenicity such as leprosy, as pointed out by for example Rubini & Zaio (2009). With time, the numbers of leprosy infected decreased immensely and the disease almost disappeared in Europe, although in some parts of Scandinavia it remained a health issue even in the 19th century.

In medical history when dealing with leprosy, the bioarchaeological data seems to precede the historical sources. Previously, the earliest example of skeletal remains with signs of leprosy was dated to 2nd century BC Egypt (Dzierzykray Rogalski, 1980). However, during recent investigations in Balathal, India, a skeleton with convincing lepromatous changes dated to c. 2000 BC have been reported (Robbins *et al.*, 2009). In Europe, a skeleton from Scotland dated to 2300–2000 BC is believed to show signs of leprosy, though the diagnosis is uncertain due to the early date in a European context (Roberts, 2007). Apart from this uncertain case, the earliest European examples are from the Roman period (Roberts & Cox, 2003; Belcastro *et al.*, 2005). During the last decade, several new European cases have been published. In Ireland, the medieval historical records and place names suggest a vast distribution of leprosy; however, only a few definite cases have been diagnosed (Murphy & Manchester, 2002; Buckley, 2008). Due to a long tradition in the field of paleopathology, the number of cases with leprosy in the archaeological record in Britain is extensive (e.g. Roberts, 2002; Roberts & Manchester, 2007; Magilton *et al.*, 2008). The youngest bioarchaeological sample is a skeleton from St. Marylebone, London, which is from the early 19th century (Walker, 2008). In central and southern Europe, early medieval evidence of leprosy has been reported from Germany (Boldsen, 2008), Hungary (Pálfi, 1991; Pálfi *et al.*, 2002), the Czech Republic (Strouhal *et al.*, 2002; Likovsky *et al.*, 2006) and central Italy (Belcastro *et al.*, 2005; Rubini & Zaio, 2009). In Scandinavia, the largest skeletal assemblages with examples of leprosy are Danish, which is significant since one of the pioneers in the study of leprosy bone changes (in dry skeletons), the Danish professor Vilhem Møller-Christensen, was the first to draw attention to the potential of these skeletal assemblages (Møller-Christensen, 1953, 1958, 1961). In recent years, new discoveries of leprosy at Danish sites have been made (Brander & Lynneryp, 2002; Boldsen, 2005, 2006; Rasmussen *et al.*, 2008).

Most of the diagnosed cases with leprosy in Sweden are Medieval but the oldest is dated to the Roman Iron Age (0–400 AD) (Arcini & Artelius, 1993). The majority of those afflicted are from sites located in

Scania (during the Middle Ages, a part of Denmark). One interesting churchyard is a common cemetery (i.e. not a leprosy hospital) in Lund with 43 individuals of both sexes and all ages (all but one dated to 990–1100 AD). It has been suggested that the location of the burials in the outer zone of the churchyard reflects the low social status of the afflicted (Arcini, 1999: 130). According to medieval Norwegian law, *Eidsivatingslagen*, among other sources, the outer burial area was assigned to the poor or socially undesirable in society (Gejvall, 1960; Nilsson, 1994). A comparable 'spiritual geography' existed in other European countries (e.g. Daniell, 1997). This study also demonstrated, at least in the south of Sweden, that before the foundation of hospitals, people suffering from leprosy were buried in groups in common cemeteries. The results of an aDNA analysis in Björned in north-eastern Sweden implied that one individual suffered from leprosy but no skeletal changes associated with the disease were observed (Nuorala *et al.*, 2004). Apart from Björned, leprosy has been observed at only five or six sites north of Scania (Kopparsvik, Tierp, Öberga, Västerhus, Skänninge and Klåstad), whereof a publication is in full progress (Caroline Arcini, pers. comm.). The low number of sites could be due to the fact that few hospitals or similar institutions have been excavated. The only larger cemetery belonging to a house of the Holy Ghost (1300–1531 AD) that has been excavated

in central Sweden included 1359 skeletons *in situ* (Dahlbäck, 1982). A few cases of tuberculosis and syphilis were identified but none of leprosy.

The present study considers the skeletal changes indicative of a systemic disease on six individuals from a medieval churchyard in east-central Sweden. The bilateral skeletal changes observed foremost in the lower legs and feet and, in two individuals, also in the nasal aperture region made leprosy a possible cause. If the diagnosis could be confirmed this would be unique since no similar paleopathologic discovery have been made in this part of Sweden. Furthermore, the pathological changes combined with the burial location of the individuals are suggestive of hierarchical arrangements of social classes in the early medieval society.

Historical background

Sigtuna is located on the Baltic Sea. It was founded in the 10th century A.D. and remains the oldest surviving town in Sweden (Figure 1). From its beginning, the town was well planned and highly organised (Tesch, 1990, 2001; Petterson, 1995). Sigtuna's dwellings were constructed of wood and the artefacts found within the city limits are non-agrarian in nature (Högrell, 1990; Petterson, 1990). The churches were built of stone

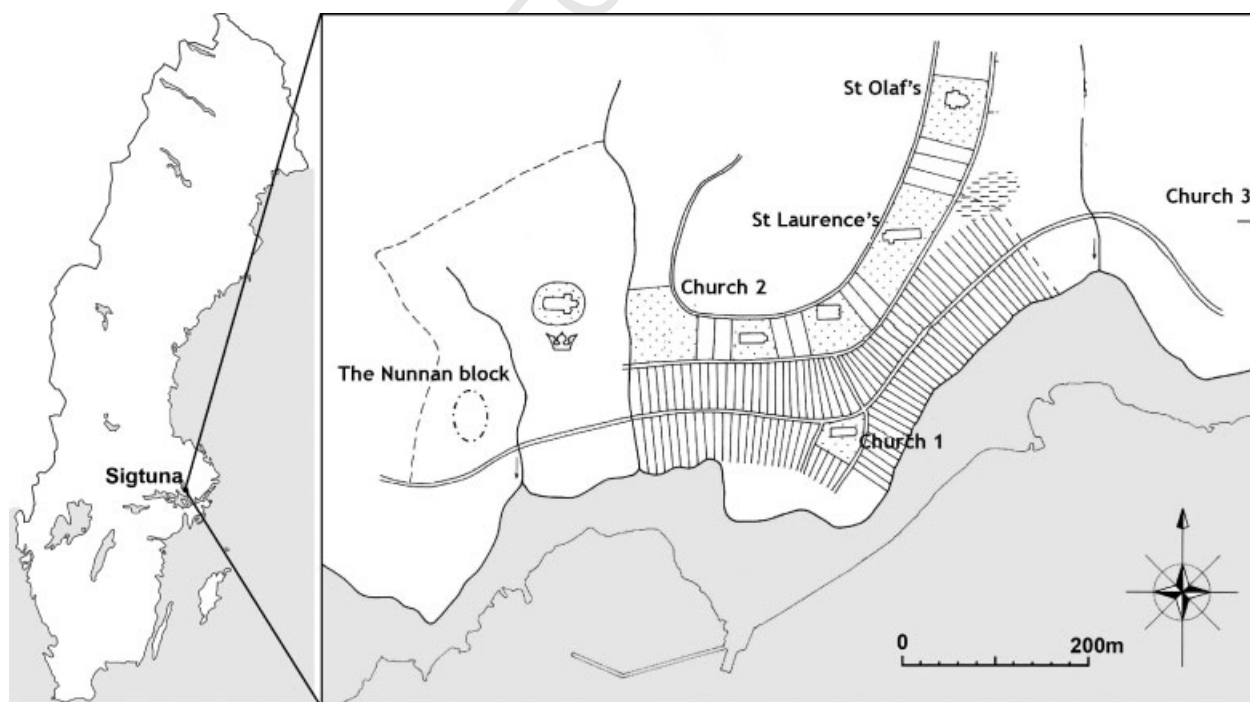


Figure 1. Map of Sweden and Sigtuna with the location of some of the excavated churchyards (the Humlegården block = Church 2).

from the initial phase. During the 13th century AD, a brick building was constructed to house the Dominican Friary and in AD 1287, a hospital was founded. Throughout its history, a sharp delineation was maintained in Sigtuna between the common and the ruling classes, which included some of the first Christian kings in Sweden (Tesch, 1990, 2000, 2001; Zachrisson, 1998; Kjellström *et al.*, 2005, 2009).

Objects and craft styles demonstrate an extensive trade with other areas in Europe such as northern Germany, Denmark, England, Holland, France, Belgium, Germany, Byzantium and Kiev (Karlsson, 1989; Larsson, 1990; Roslund, 1990, 2001). Sigtuna's households were largely self-supporting, with associated farms near by providing supplies. Archaeofloral remains indicate a diverse diet of cultivars (Hansson, 1997) and zooarchaeological analysis indicates the diet included meat from domesticated cattle, sheep, goats and pigs (Hårding, 1990), wild game, fowl and fish (Jonsson, 1989; Hårding, 1990; Vretemark, 1997). Stable isotope analyses have confirmed the diet included protein sources of terrestrial and marine origin (Kjellström *et al.*, 2009).

Approximately 800 skeletons from different cemeteries dated to the Middle Ages (970–1527 AD) have been analysed. Based on a combination of carbon dating and stratigraphy, the graves have been subdivided into burial phases corresponding to the establishment of the town (Phase 1 c. 900–1100), its peak of prosperity (Phase 2 c. 1100–1300) and its decline (Phase 3 c. 1300–1530) (Kjellström *et al.*, 2005; Wikström, 2008). The skeletons have been excavated both from burial grounds without connection to a church and from six ordinary churchyards located in

the town. Generally the graves are homogeneous: single, east–west oriented and with no or few grave goods. All age groups are represented, though the assemblage reflects a young population where about 37% are below the age of 20. The sex distribution is biased, with more men than women (Kjellström, 2005; Kjellström & Wikström, 2008).

Material and methods

In 2006, an excavation was carried out in the southern parts of the churchyard of the Humlegården block by Sigtuna Museum (Wikström, 2008). The name of the church, dated to phase 2 (c. 1100–1300), is not known. In^{Q4} a previous study of 99 skeletons from the same cemetery, the church was called 'Church 2' (Kjellström, 2005). The 220 graves included 227 skeletons. The skeletal preservation *in situ* varied from excellent (complete skeletons) to poor (a few elements with a porous character). In the skeletal assemblage higher frequencies of a variety of pathological skeletal alterations were observed than in previously analysed samples from Sigtuna (Kjellström & Wikström, 2008). For example, in the complete sample, 31 skeletons showed bilateral subperiosteal changes in the tibiae. Six skeletons excavated in the outskirts of the cemetery could be distinguished due to the fact that they either combined bilateral subperiosteal changes of the tibiae with systemic inflammatory alterations in the fibulae and feet or exhibited changes in the rhinomaxillary region. These individuals will be discussed from a detailed paleopathological point of view (Table 1) (Figure 2).

Table 1. Individuals within the Humlegården block with destructive changes indicative of leprosy. (Ph 1 = Proximal phalanges, Ph 2 = intermediate phalanges, Ph 3 = distal phalanges)

	Case 1 Id. no. 3159		Case 2 Id. no. 3077		Case 3 Id. no. 3093		Case 4 Id. no. 3320		Case 5 Id. no. 3092		Case 6 Id. no. 3401	
Sex	♂		♀		♂		—		—		♂	
Age	20–23		20–30		20–25		Adult		11–12		35–50	
Maxillary region	—		—		No changes		—		Erosive changes		Erosive changes	
Destructive inflammatory changes:	Right	Left	Right	Left	Right	Left	Right	Left	Right	Left	Right	Left
Tibia	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	No	No	No	No
Fibula	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	No	No
Metacarpals	No	No	Yes	Yes	Yes	Yes	—	—	No	No	No	No
Ph 1	No	No	Yes	Yes	Yes	Yes	—	—	No	No	No	No
Ph 2	No	No	Yes	Yes	Yes	Yes	—	—	No	No	No	No
Ph 3	No	No	Yes	Yes	No	No	—	—	No	No	No	No
Metatarsals	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	No	No	Yes	Yes
Ph 1	Yes	Yes	Yes	Yes	—	—	Yes	Yes	No	No	Yes	Yes
Ph 2	Yes	Yes	Yes	Yes	—	—	Yes	Yes	No	No	No	No
Ph 3	Yes	Yes	No	No	—	—	—	—	No	No	No	No

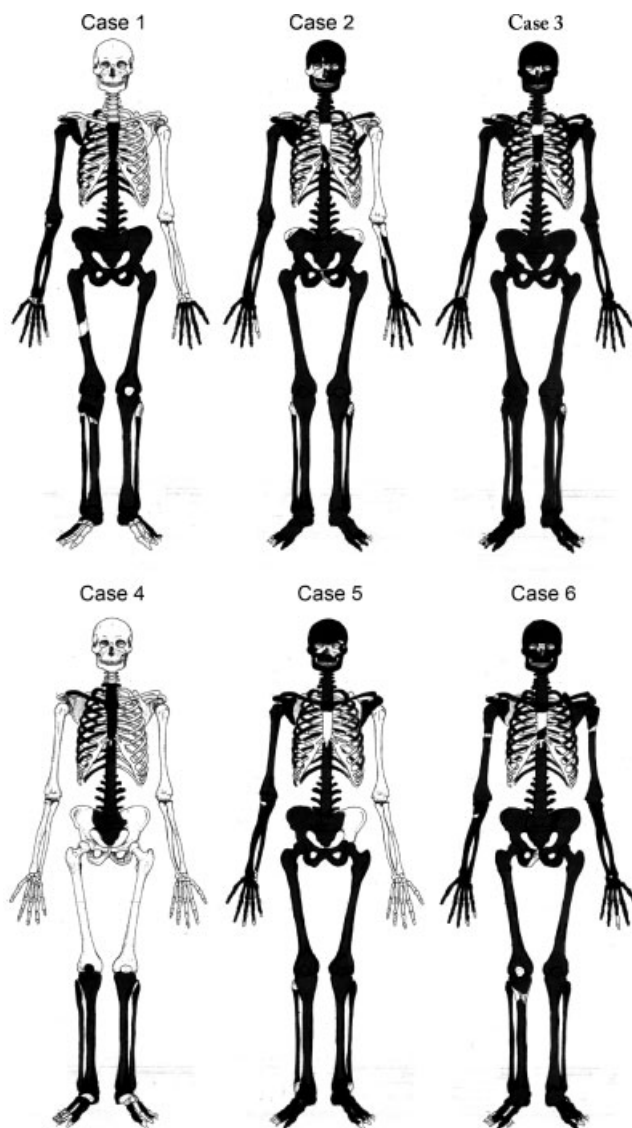


Figure 2. The preservation (in black) of the six skeletons from the Humlegården block, Sigtuna.

Age estimations in younger individuals were made using dental development and epiphyseal fusion following the work of Moorrees *et al.* (1963a and 1963b), Stloukal & Hanáková (1978), Ubelaker (1989) and Scheuer & Black (2000). For adults, age was assessed using morphologic characteristics of the skull and pelvis following Brothwell (1981), Lovejoy *et al.* (1985), Meindl & Lovejoy (1985) and Brooks & Suchey (1990). Using morphological features of the pelvis and skull in addition to measurements of the femora and humeri, sex was estimated according to Phenice (1969), Stewart (1979), Novotný (1982), Pearson and Bell, 1917–1918 in Bass (1987: 219), Buikstra & Ubelaker (1994) and Bruzek (2002).

Description of the skeletons

Case 1 (Id. no. 3159)

The skeleton of a 20–23-year-old individual was analysed. Ankylosis between an intermediate and a distal phalanx was recorded among the finger bones. No other alterations attributable to pathologies were noted in the hands.

The most severe changes were observed in the lower extremities where signs of subperiosteal bone reaction in the lower legs and pathological destruction of several bones in the feet were documented. Bilaterally both the diaphyses of the tibiae and fibulae showed longitudinally irregular, striated bone deposits foremost on the medial and lateral lower third. The deposits had a pitted surface, which ranged from smooth to rugged. In the feet, the tuber calcanei of the right calcaneus was enlarged posteriorly with a roughened exostosis. Minor exostoses were also observed on the dorsal side of the left talus and lateral cuneiform. The distal part of the left third, fourth and fifth metatarsal (the only remaining in the left foot), were atrophied and pointed (Figure 3). Only a third of the diaphyses remained and the concentric bone resorption was most probably progressing proximally. On the right side, the same type of destruction could be seen in the fifth metatarsal (the only metatarsal preserved). The proximal joint of the remaining four proximal phalanges were totally resorbed and the diaphyses were pointed.

Case 2 (Id. no. 3077)

The complete skeleton of a 20–30-year-old female individual exhibited multiple skeletal changes in the hands, legs and feet. No destructive changes were discovered in the facial skeleton due to postmortem destruction.

The changes in the hands were mild. Two of the proximal and two of the intermediate phalanges were flat in the dorso–volar direction. Discrete signs of resorption were found on two (of eight preserved) distal phalanges where the ungual tuberosity had a slightly pointed appearance.

Both tibiae displayed slight subperiosteal bone reaction on the distal lateral diaphyseal surface while more prominent new bone formation was seen bilaterally on the distal medial diaphyseal side of the fibulae. The surface of the cortex was pitted and longitudinally striated. In the feet both navicular bones

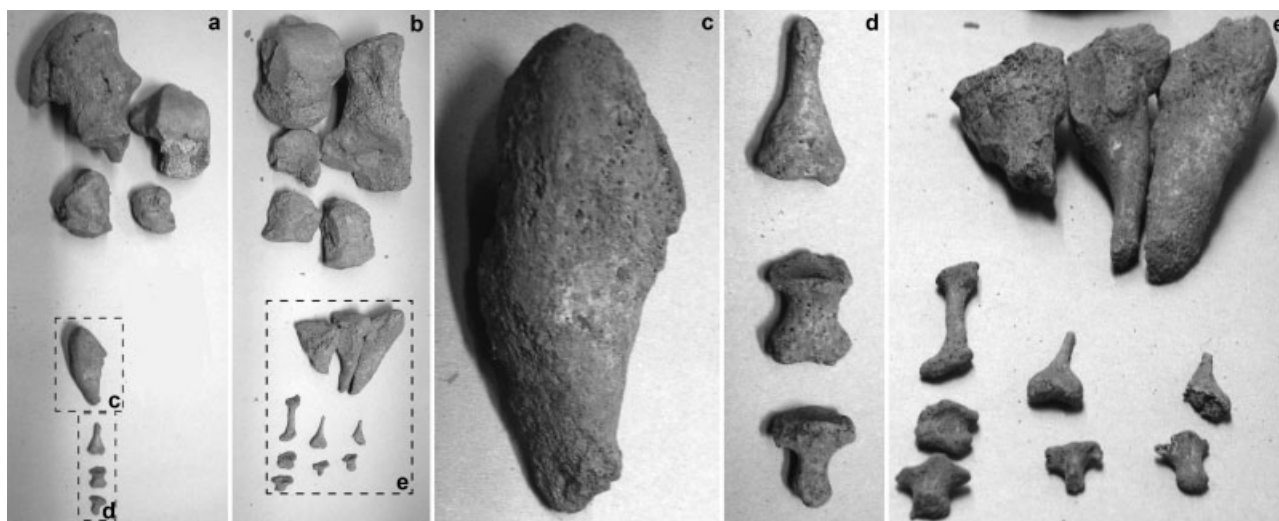


Figure 3. (a-b) The feet of an adult male (Case 1); (c) Acroosteolysis of the right fifth metatarsal; (d) Mid-diaphyseal remodelling of a proximal phalanx; (e) The remaining left third, fourth and fifth metatarsals and phalanges with 'pencil' deformities.

exhibited dorsal exostosis. The first metatarsal on both sides had unaffected metatarso-cuneiform joints but exhibited substantial bone resorption in the distal parts, where the heads were completely destroyed (Figure 4). The right first metatarsal had a cloaca on the lateral side of the diaphysis and the distal part ended in a blunt process. The diaphyses of the second, third and fourth metatarsals exhibited slight subperiosteal bone reaction. The fifth metatarsals had unaffected bases proximally, but the heads were completely resorbed and the bones end in a point. The diaphysis of the bones displayed new bone formation, especially on

their lateral sides. The proximal phalanx of the first digit on both sides showed a destroyed proximal joint surface, most notably on the right side. The joint surface articulating with the first metatarsal was placed dorsally from its original position, so that the bone must have had a vertical position when articulated (please note the similarities with the same joint in Case 4). The distal joint seemed unaffected. Among the rest of the proximal phalanges in the feet, two out of eight showed signs of bone resorption in the proximal end, though this was limited to porosity. A small, unidentified piece of bone was found among the

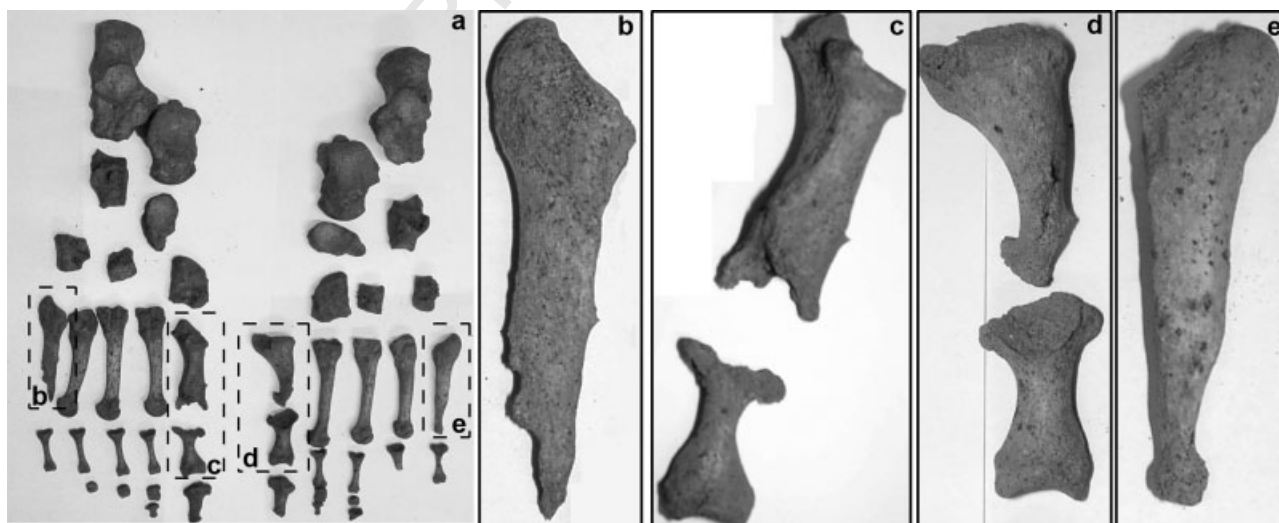


Figure 4. (a) The feet of an adult woman (Case 2); (b) Acroosteolysis of the right fifth metatarsal; (c) A 'pencil and cup' deformity of the first metatarsal and proximal phalanx on the right side; (d) A 'pencil and cup' deformity of the right first metatarsal and proximal phalanx on the left side; (e) Acroosteolysis of the left fifth metatarsal.

phalanges, most likely a severely remodelled second or third phalanx.

Case 3 (*Id. no. 3093*)

The complete skeleton of a 20–25-year-old male showed destructive remodelling on the long bones of the lower extremities and in the bones of the feet. An examination of the facial skeleton revealed no signs of rhinomaxillary remodelling in the maxilla, hard palate or piriform aperture.

In the bones of the hands only minor changes were observed. One out of five proximal phalanges from the right side had a swollen corpus and a cortically atrophied distal joint (Figure 5). The articulating intermediate phalanx had a deformed proximal articular surface with new bone deposits and porosity.

Both femora showed longitudinally striated periosteal bone formation along the dorso-medial diaphyses. The diaphyses of both tibiae and especially the fibulae were thickened, with vascular grooves and new bone deposits (Figure 6). In the right foot, the corpus of the calcaneus was deformed, caused by a combination of bone resorption and new bone formation. Within the posterior articular surface of the talus, the calcaneus exhibited new bone deposits and macroporosity. Similar to the calcaneus the articular surface of the talus was altered by osteoclastic and osteoblastic processes. The cuboid, the lateral and intermediate cuneiform on the right side and the talus, cuboid, the navicular, and lateral and intermediate cuneiform on the left side show dorsal exostoses. The first metatarsal on both sides had unaffected metatarsocuneiform joints but the diaphyses showed subperiosteal bone reaction and their heads were severely remodelled distally (Figure 7). The second, third, fourth and fifth metatarsals on both sides had a similar appearance, with seemingly unaffected metatarso-cuneiform joints, medio-lateral cortical bone loss and partially destroyed



Figure 5. The resorbed and atrophied distal joint of a proximal phalanx of an adult male (Case 3).

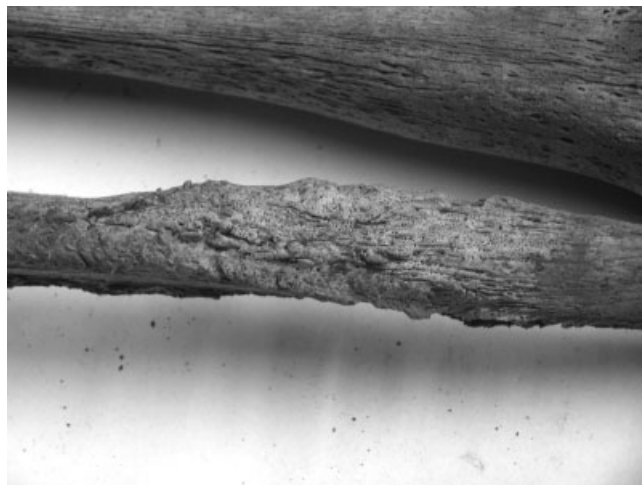


Figure 6. Proliferative bone changes on the lateral shafts of the left tibia and fibula of an adult man (Case 3).

pointed distal ends. Two separate, unidentified bone pieces from the left foot were interpreted as remains from the destroyed metatarsals. The proximal phalanges preserved for analysis had a swollen corpus with signs of resorption proximally in the margins of the metatarsophalangeal joints. Two (out of three) intermediate and two distal phalanges had fused.

Case 4 (*Id. no. 3320*)

A fragmented skeleton of an adult individual of unknown sex displayed severe pathological changes both in the spine and the feet.

In the spine, which was almost complete (C7–L5), the first three thoracic vertebrae did not reveal any pathological changes at gross examination. The corpus of the remaining thoracic vertebrae (T4–T12) showed signs of circumferential pitting: i.e. smooth-walled resorption on their ventral surfaces. The lumbosacral region exhibited collapse of vertebral bodies, kyphosis and ankylosis of the first three lumbar vertebrae (L1–3) (Figure 8). The wedge-shaped vertebral block resulted in an approximately 50° angling of the spinal column. The corpus of the first lumbar (L1) and third to fifth lumbar vertebrae (L3–5) had large, smooth walled, remodelled lytic lesions ventrally and only the vertebral arch remains of the second lumbar vertebra (L2), which, due to the complete destruction of the body, was most likely the initial focus of the destructive process. Only minor new bone formation could be observed on the surface of the remaining fragments, which formed an amorphous mass. The margins of the lesions had an almost sclerotic appearance.



Figure 7. (a) The feet of an adult man (Case 3). (b) Close-up view of the right metatarsals displaying resorption and atrophy. Note the loose osseous fragment; (c) Close-up view of the left metatarsals displaying resorption and atrophy.

Approximately 25% remained of the seemingly melted body of the third lumbar vertebra (L3). Except for arthritis between the first and second, and an ankylosis between the second and third, no change in shape of the first three lumbar neural arches (L1–3) could be observed. The spinal canal did not seem to have been affected. Severe lytic lesions were also observed on the upper ventral surface of the sacrum. The circular erosion showed signs of both resorption and peripheral remodelling, however, with an almost absence of new bone formation. No inflammatory changes were seen on the visceral surface of the costae.

In the lower extremities, both tibiae showed minor longitudinally striated, subperiosteal, new bone formation along the distal lateral surface of the diaphysis. The diaphyses of the fibulae were also thickened and also exhibited deposits of striated, subperiosteal bone reactions. In the feet, the metatarsophalangeal joint of the first metatarsal on both sides had fused with the proximal phalanx, which pointed in an axial direction (similar to Case 2) (Figure 9). The atrophied and

deformed joints suggested that the fusion was secondary to an infection. The right, second metatarsal had fused with the intermediate cuneiform, and the dorsal, porotic, cortical thickening of its diaphysis, in addition to a cloaca, indicated evidence of osteomyelitis. The remaining preserved metatarsals on the right side, the fourth and fifth, did not display any changes. Except for in the hallux, ankylosis was additionally noted in the only remaining proximal phalanx on the right side. The distal end is completely resorbed and fused with an intermediate phalanx. On the left side, irregular subperiosteal bone reaction was present on the diaphyses on the third and fourth (the only remaining) metatarsals. No phalanges were preserved from the left side.

Case 5 (*Id. no.* 3092)

The complete skeleton of an 11–12-year-old child with rhinomaxillary and postcranial inflammatory skeletal manifestations.

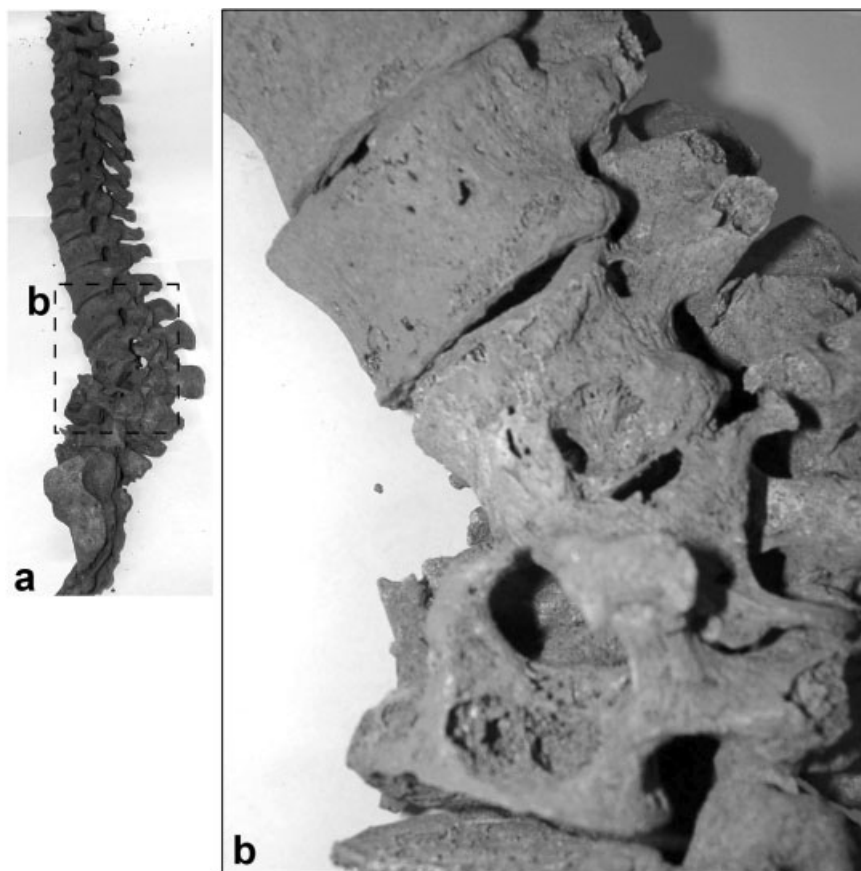


Figure 8. (a) The spine of an adult individual (Case 4). (b) Close-up view of the first to third lumbar vertebrae with lytic lesions.

In the facial region, atrophy of the anterior nasal spine and of the central part of the alveolar bone of the maxilla was observed (Figure 10). The piriform aperture exhibited a rounded margin. In the nasal passage, the palatine process was pitted and porous and the opening to the incisive canal was enlarged. The incisive alveolar sockets were shallow but the anterior surface was damaged due to poor *post mortem* preservation. Nevertheless, it is likely that some degree of alveolar resorption did occur and affected the dental development, since the roots of the central incisors are very short and slightly deformed (Figure 11).

Postcranially, of the hands only the bones of the right side were preserved and no absorptive changes were noted. The tibiae were without macroscopic changes. The left fibula showed a low degree of fine, striated, subperiosteal bone deposits on the distal diaphysis. The right fibula had a 15-mm-long and 4.5-mm-deep, delimited lytic lesion close to the metaphyseal area on the lateral surface (Figure 12). No changes were observed in the metatarsals and proximal phalanges and the intermediate phalanges were missing. The first distal phalanx (the only distal

phalanx preserved) showed concentric atrophy of the distal end, resulting in a tapered point.

Case 6 (*Id. no. 3401*)

The complete skeleton of a middle-aged male (35–50 years) with erosive changes in his facial bones and feet. The hands were almost complete, and no signs of bone remodelling could be noted.

The anterior nasal spine and the central alveolar bone of the maxilla had been lost *post-mortem*. However, pitting and remodelling was observed in the bone surrounding the piriform aperture (Figure 13). Bilateral erosive and proliferative bone reactions were present. Furthermore, the anterior palatine foramen was enlarged and inflammatory changes with new bone deposits were shown in the maxillary sinuses and on both surfaces of the palatine process (maxillary sinusitis).

The long bones of the lower extremities displayed only minor alterations. Slight longitudinally striated, subperiosteal bone formation on the distal end of the

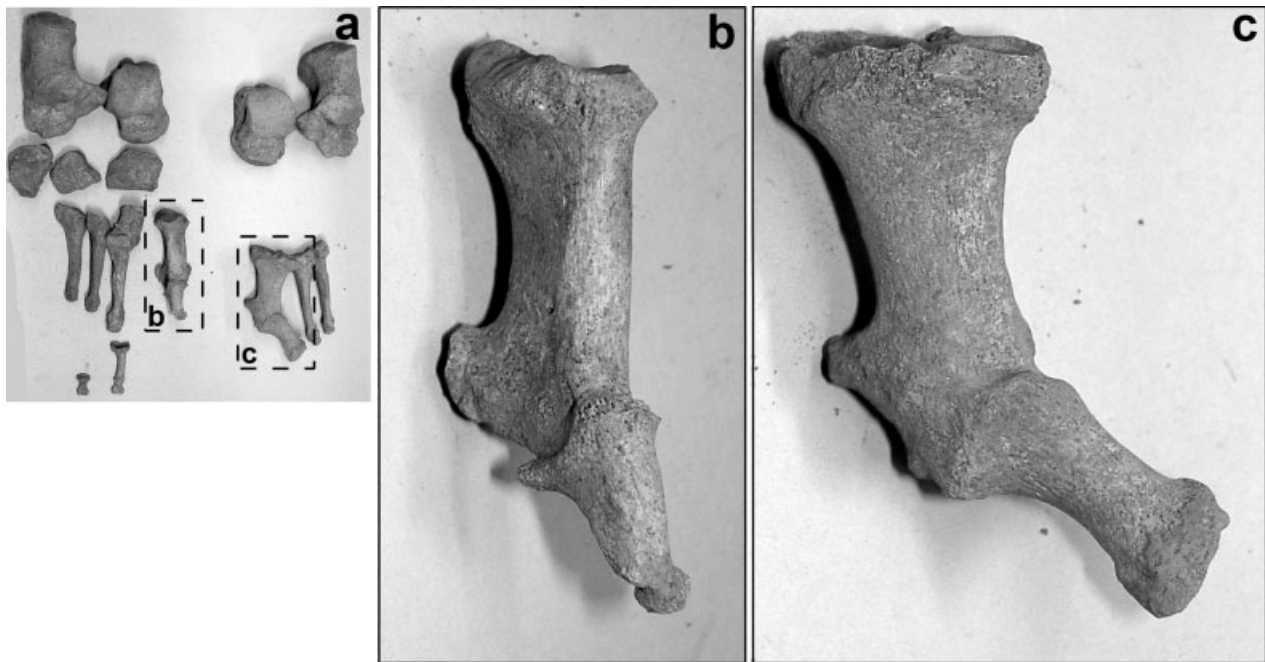


Figure 9. (a) The feet of an adult individual (Case 4). (b) Close-up view of the ankylosis between the first metatarsal and the proximal phalanx on the right side. (c) Close-up view of the ankylosis between the first metatarsal and the proximal phalanx on the left side.

diaphysis was seen on the left fibula. In the feet, destructive changes were observed on the metatarsophalangeal joint of the fifth metatarsal on the right side (Figure 14). The metatarsal capitulum showed signs of osteolysis and the adjoining proximal phalanx had a 'cupped' appearance. Inflammatory skeletal changes were also seen on the articular surface of the distal joint

surface of the proximal phalanx of the left great toe. Bone destruction and formation had severely destroyed the joint (Figure 15). The adjoining phalanx was missing.

Osseous changes in leprosy

The symmetrical, bilateral distribution of lesions primarily found on the lower legs and feet or the rhinomaxillary region made leprosy a possibility when trying to identify the cause.

In leprosy, the severity of signs and symptoms depends on the strength of the immune system of the host (Roche *et al.*, 1992; Andersen *et al.*, 1994; Ooi & Srinivasan, 2004). The bacteria themselves make little damage since they do not secrete toxins; however, a non-resistant immune system leads to tissue changes in the body. The bacteria thrive in cool areas of the body and the commonly pathologic changes are seen in the skin, peripheral nerves and in oral and nasal mucous membranes where the bacteria live and grow. Infective bone lesions in leprosy are of two types: those due to the direct presence of *Mycobacterium leprae* and those occurring secondarily as a result of nerve damage and ulceration (which are difficult to differentiate from lesions with a non-leprosy origin) (Andersen *et al.*, 1994). Depending on the immunological response and

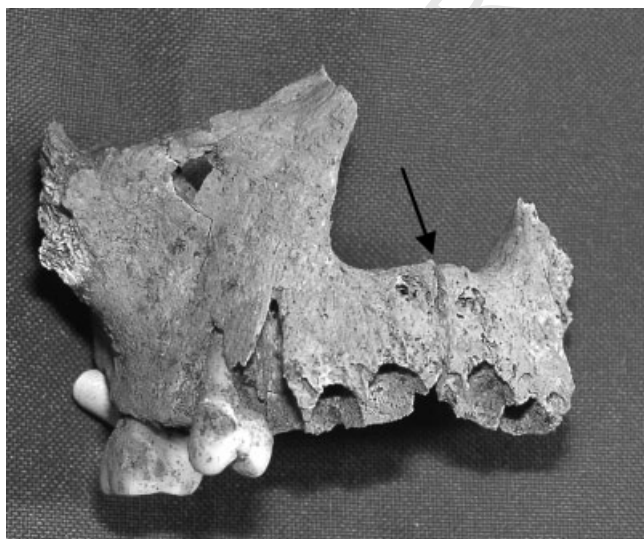


Figure 10. Atrophy of the anterior nasal spine and of the central alveolar bone in the maxilla (Case 5).

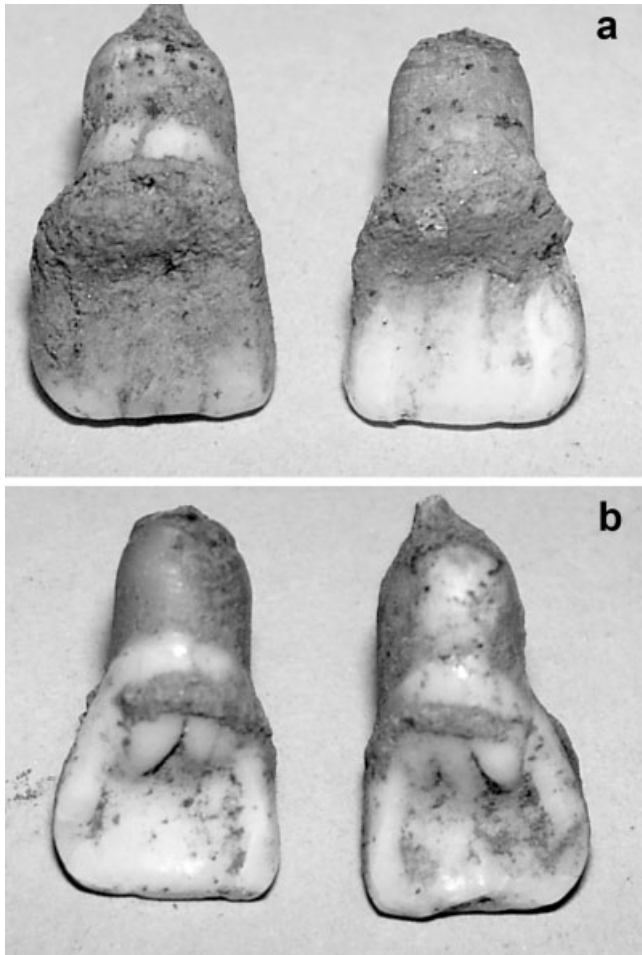


Figure 11. (a-b) Shortening of the roots of the central incisors of an 11–12-year-old child (Case 5).

clinical changes, and following the Ridley–Jopling classification, infected persons are subdivided into groups, where tuberculoid leprosy and lepromatous leprosy are the two extremes (Evans & Brachman, 1998: 386; Moschella, 2004). An infected individual with generally good health may develop the tuberculoid variety whereas an impaired immune system can lead to lepromatous leprosy, which is considered to be a more severe form (Manchester, 1991; Evans & Brachman, 1998: 387). Both types can, however, result in permanent nerve damage.

In a person with the tuberculoid type, the disease may be limited to one or a couple of discoloured, well-defined skin patches (Resnick & Niwayama, 1988: 2687). The skin is thickened and sensory, motor and autonomic nerves are damaged by the rather few encapsulated bacteria. Damage to the autonomic nerves may lead to an impaired ability to perspire, resulting in a cracked and dry skin, infections and, in



Figure 12. Close-up view of the right fibula showing an erosive lesion close to the metaphyseal area on the lateral surface (Case 5).

the worst cases, sepsis, together with necrosis and destruction of soft tissue (Vivier, 2002). Motor nerve involvement leads to contraction of fingers ('claw hand') and toes (Aufderheide & Rodríguez-Martín, 1998). As a result of this progressive paralysis and flexion contracture of muscles, grooves may be formed on the volar side of the proximal phalanges of the hands (Andersen & Manchester, 1987). The sensory nerve involvement in the hands and feet predispose the individual to trauma (Judd & Roberts, 1998; Manchester, 2002). Secondary ulceration and non-specific infections such as osteomyelitis and septic arthritis may lead to bone destruction. The low degree of *Mycobacterium leprae* present in the tissue of an individual with tuberculoid leprosy makes symptoms of the disease less aggressive, with milder pathological reactions, and most often only a single or two limbs are involved.

The lepromatous type is characterised by a slow course of events (Aufderheide & Rodríguez-Martín, 1998: 144). The process causes a bacillary multiplication through several tissues due to a defective, cell-mediated immune response (Vivier, 2002). Hence, several organs are exposed to the bacilli and may be affected, but due to the 'bacillus' preference for colder body areas, most changes are observed in the superficial subcutaneous structures. In the skin and

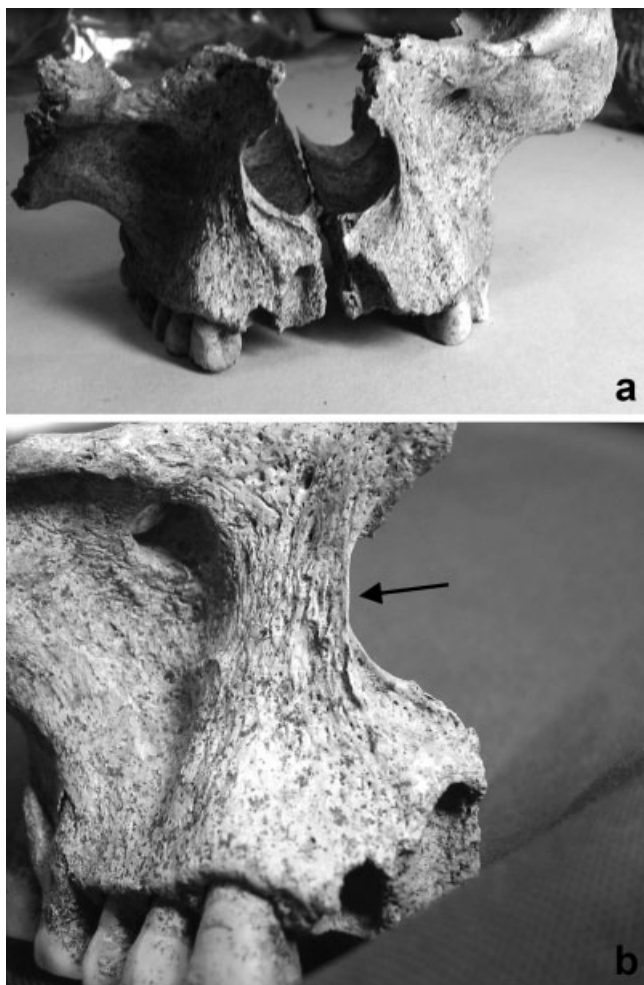


Figure 13. (a-b) Remodelling in the region of the piriform aperture with signs of erosive and proliferative bone reaction (Case 6).

mucous membranes of the face, nodules and papules are formed. The facial bones show changes of *facies leprosa* (Møller-Christensen, 1961) or *rhinomaxillary syndrome* (Andersen & Manchester, 1992), which are



Figure 14. Lytic changes in the right metatarsophalangeal joint of the fifth metatarsal (Case 6).



Figure 15. Inflammatory changes in the distal joint of the proximal phalanx of the left hallux (Case 6).

considered pathognomonic of lepromatous leprosy. [However^{Q5}](#), it has been put forward recently that other diseases may leave similar changes (Collins Cook, 2002; Ortner, 2008a, b). In the initial stage, before the infection has affected any facial bones, the disease may be seen as a chronic rhinitis (Aufderheide & Rodríguez-Martín, 1998: 144). The subsequent destructive bone alterations include atrophy of the anterior nasal spine of the maxilla and remodelling of the margins of the nasal aperture, sometimes together with resorption of the central part of the maxillary alveolar process, causing loosening of the incisors and perforation of the palate. The latter occurs at an advanced stage of the pathological process (Andersen & Manchester, 1992). Andersen & Manchester (1992) point out that the pathological changes may occur singularly or in combination. Even though *rhinomaxillary syndrome* is highly diagnostic for leprosy, milder manifestations of the syndrome must be considered (Andersen *et al.*, 1994). Postcranially, symmetrical periostitis of the tibia and fibula may be observed, most commonly on the lower two-thirds of the diaphysis of the tibia and fibula (Resnick & Niwayama, 1988; Aufderheide & Rodríguez-Martín, 1998). The majority of skeletons with other signs of leprosy examined by Møller-Christensen showed signs of periostitis on these bones and he

implied that the term 'leprosy periostitis' should be used (Møller-Christensen, 1953, 1961). The, commonly bilateral, subperiosteal changes on the diaphyses are shown as deposits of new bone with vascular grooves and a pitted surface. The most characteristic post-cranial changes are the resorption of the distal aspects of the distal phalanges of the hands and in the metatarsophalangeal joints of the feet (e.g. Møller-Christensen, 1961; Andersen *et al.*, 1994; Aufderheide & Rodríguez-Martín, 1998; Ortner, 2003). Due to deprived blood supply and nerve inflammation, cortical bone is resorbed coincidentally with some endosteal new bone formation (which diminishes the medullar cavity), resulting in a concentric narrowing of the diaphyses proximal ends (Andersen *et al.*, 1992). In less severely affected areas, subluxation and osteophytes may cause alteration in the proximal joints of phalanges resulting in a cup-shaped appearance (Ortner, 2003). As in tuberculoid leprosy, volar grooves may be seen in the phalanges of the hands. In addition to the bone resorption seen in the metatarsals and phalanges tarsal, bars may develop in the feet (Andersen & Manchester, 1988). These are hyperthrophic bone bridges, enthesopathies, on the dorsal surface of the tarsals that emerge as a result of arch collapse and subsequent mechanical stress on the foot. Most of the time the disease affects the skeleton bilaterally but the bone changes are seldom completely symmetrical. The inactivity caused by the destruction of the nerves may lead to osteoporosis, which is an important factor in the bone remodelling of the hands and feet. Clinical studies have shown that even 7-year-old children with leprosy may demonstrate generalised osteoporosis (Newman *et al.*, 1972). The most pronounced osteoporotic changes seen in skeletons from Næstved were observed in the metatarsal heads (Møller-Christensen, 1961).

Differential diagnosis

Since the pathological bone alterations in leprosy may mimic other conditions, alternative disorders must be considered for the six individuals from Sigtuna. As an alternative diagnosis to leprosy, Steinbock (1976) mentions pyogenic osteomyelitis, treponemal disease, tuberculosis, fungal infections, frostbite and diabetes mellitus. In addition, Belcastro *et al.* (2005) considers sarcoidosis, psoriatic arthritis and peripheral neuropathy. Furthermore, leishmaniasis, rheumatoid arthritis, rhinoscleroma, actinomycosis, trauma and non-specific infections are discussed. Due to the extensive skeletal changes of the spine in one of the individuals,

tuberculosis and alternative disorders of this disease are also considered.

Pyogenic osteomyelitis, a non-specific, direct or haematogenous-spread infection, can affect for example bones in the hands and feet secondarily to trauma. In contrast to leprosy, sequestrum and reactive new bone formation is seen. The destruction seldom involves several bones in a symmetrical pattern (Aufderheide & Rodríguez-Martín, 1998: 164). Pyogenic osteomyelitis may be subsequent to leprosy due to ulceration and secondary infection but does not cause rhinomaxillary syndrome.

Bone resorption due to *treponemal disease* may result in the destruction of bones in the face, lower legs, hands and feet. The bacteria of the genus *Treponema* may lead to the diseases pinta, yaws, endemic syphilis and venereal syphilis of which the former three are primarily distributed south of Europe (Roberts & Manchester, 2007). In venereal syphilis, the most characteristic changes are gummatous lesions (*caries sicca*) most often distributed on the ectocranial surface of the frontal and parietal bones (Ortner, 2003: 280). These depressed, circular clusters of 'worm-eaten' bone have clear sclerotic borders and are considered pathognomonic for treponemal disease (Aufderheide & Rodríguez-Martín, 1998). In the facial region the palate, the maxilla (but not necessarily the anterior nasal spine) and the nasal bones may succumb to necrosis. When the fingers are affected, sufferers of treponemal diseases display phalangeal expansion due to subperiosteal bone formation instead of concentric atrophy most often seen in leprosy (Ortner, 2003: 275, 296). The most affected long bone is the tibia with the typical saber shin deformity where the anterior surface of the bone is especially thickened by new periosteal bone. *Syphilis* rarely affects the spine and when it does, it is almost exclusively concentrated on the cervical region (Ortner, 2003: 313).

Bone lesions can develop due to different *fungal infections*, e.g. cryptococcosis, sporotrichosis and mucormycosis. Cryptococcosis and sporotrichosis may result in similar lesions only distinguishable from each other by anatomical location: however, only cryptococcosis is (although rarely) found in Sweden (Ortner & Putschar, 1981: 224; Edebo *et al.*, 2009). Any bone can be affected but most often they are found in bone eminences, cranial bones and vertebrae (Ortner, 2003: 326). Sporotrichosis may cause primarily bone lesions in the form of localised bone abscesses and periostitis on the tibia and the skull. Mucormycosis is a rare fungal disease, which in severe cases of facial sinus infections may lead to unilateral perforation of the hard palate (Ortner & Putschar, 1981: 227). In general, a

1 fungal infection may involve the spine and cause
2 vertebral collapse (Ortner, 2003: 325ff). If so, both the
3 body and the posterior processes of the vertebrae may
4 be involved.

5 *Actinomycosis* is an uncommon bacterial infection
6 causing multiple abscesses which mainly strike cattle
7 but which also may be found in humans (Resnick &
8 Niwayama, 1988: 2705; Rothschild *et al.*, 2006). The
9 abscesses may lead to destruction of both soft tissue
10 and bone. The most characteristic feature is distinct
11 resorptive foci located in the cerviofacial area where
12 changes of the mandible are the most affected bone.
13 Postcranially rounded lesions with sclerotic margins in
14 varying sizes (c. 0.2–0.7 cm) may be exhibited in
15 trabecular, cortical or subchondral bone (Rothschild
16 *et al.*, 2006). In the spine several vertebrae may be
17 involved exhibiting lytic and sclerotic damage. The
18 vertebral body as well as the transverse and spinous
19 processes are affected but a subsequent collapse of the
20 spine is uncommon (Resnick & Niwayama, 1988:
21 2705). The intervertebral discs are not involved.

22 Severe *frostbite* may cause bilateral absorption in the
23 hands or feet when sensory nerves are damaged due to
24 depressed vital processes in low temperatures (Bor-
25 ovikov, 1993). The toes seem to be affected more often
26 than the fingers (Lehmuskallio, 2001). The resorptive
27 changes start in the tip of the distal phalanges (Cauchy
28 *et al.*, 2001).

29 Damage of *peripheral nerves* due to a variety of causes
30 could also lead to atrophy of the bones in the hands or
31 feet. Belcastro *et al.* (2005) mention *diabetes mellitus* but
32 conclude that it affects only the feet, not the hands or
33 face. Furthermore, Steinbock (1976: 209) find it
34 unlikely that individuals with diabetes, living in the
35 pre-modern-medicine era, would survive long enough
36 for bone changes to appear. Another peripheral
37 neuropathy is Thevenard's disease, which could be
38 mistaken for leprosy (Thami *et al.*, 1993). In this very
39 uncommon, hereditary disease acroosteolysis may
40 occur when subcutaneous ulcers at the pressure points
41 of the feet affect bones and exhibit a strong
42 resemblance to leprosy bone changes. It is foremost
43 the metatarsophalangeal joints of the feet that are
44 affected bilaterally, but the hands may also be involved
45 (Banna & Foster, 1972). In late stages, the ankle and
46 knee may be affected but never the face.

47 *Psoriatic arthritis* affects multiple joints and the axial
48 skeleton, for example the sacroiliac joint, is commonly
49 affected (Ortner, 2003: 580). The disease may also
50 cause bone resorption in addition to proliferative new
51 bone reaction in the fingers and toes, foremost in
52 the distal interphalangeal joints (Ortner, 2003: 580).
53 The terminal phalanges may be totally resorbed. The

54 resulting 'cup-and-pencil' appearance may resemble
55 leprosy although it is asymmetric in distribution.

56 *Rheumatoid arthritis* is a chronic inflammatory poly-
57 articular autoimmune disease exhibited as erosive
58 arthritis. The small joints in the hands are most
59 commonly affected but the feet and larger joints may
60 also be involved. The disease causes symmetrical
61 erosive damage in primarily the margins of peripheral
62 synovial joints. The lesions exhibit round, smooth
63 edges with no new bone formation (Rothschild &
64 Martin, 1993).

65 *Sarcoidosis* is an uncommon, granulomatous, inflam-
66 matory disease which results in small (1–10 mm)
67 circular lytic lesions with little or no periosteal bone
68 formation (Ortner & Putschar, 1981: 233). The uni- or
69 bilateral bone lesions most often affect the phalanges
70 in the hands or feet (Ortner, 2003: 341). The lesion
71 pattern tends not to be symmetrical. The lesions are
72 commonly concentrated to the intermediate and distal
73 phalanges but do not include the interphalangeal
74 joints. The nasal bones may also show destruction, but
75 the maxilla and nasal spine do not need to be affected.
76 In sarcoidosis, the destruction of the spine is most often
77 located in the vertebral body, sparing the pedicles. The
78 lesions, which can result in vertebral collapse, may be
79 multifocal and discontinuous (Ortner & Putschar, 1981:
80 233).

81 *Rhinoscleroma* is a granulomatous disease caused by
82 the bacteria *Klebsiella rhinoscleromatis*. The infection,
83 which is rarely seen in temperate parts of the world,
84 may in a final stage lead to destruction of the nasal
85 cartilage and cavity (nasopharynx, larynx, trachea, and
86 bronchi) (Talwar *et al.*, 2008). Sometimes the upper
87 respiratory tract is affected. Documented bone changes
88 foremost include a dilatation of nasal cavity and a
89 thinning of nasal bones and atrophy of the pyriform
90 crest of the maxilla, but absorptive changes have also
91 been documented on the inferior turbinate bone, the
92 ethmoid air cells, the hard palate and the region behind
93 the frontal (Badrawy, 1966).

94 *Leishmaniasis* is a vector-born infection caused by the
95 protozoan parasites of the genus *Leishmania* (sandfly)
96 and is most often found in subtropical and tropical areas
97 in the world (Sharma & Singh, 2008). The infection has
98 several clinical manifestations, most often including
99 nodular lesions and chronic ulcers in the skin and
100 around the mouth and nose (mucocutaneous leishma-
101 niasis). The infection may lead to severe destruction of
102 bone and cartilage. The development and outcome of
103 the infection is regulated by the immunoinflammatory
104 response of the host (Murray *et al.*, 2005).

105 Due to a variety of causes not connected to sensory
106 loss, deep cuts may be left untreated which in turn leads

to infections such as *periostitis* and *osteomyelitis*, where the pathogenic factor cannot be identified. However, other non-infectious and non-inflammatory factors such as haemorrhage, chronic irritation or trauma may also cause a subperiosteal reaction (Schultz, 2001; Ortner, 2008a).

Tuberculosis, when affecting the skeleton, is characterised by kyphosis, an often monofocal osteomyelitis involving the spine, a low degree of hypertrophic new bone formation and a nearly non-involvement of the vertebral arches (Ortner, 1999, 2008a). The focus of the destruction is located in the first to the third lumbar vertebra, which is the most exposed site in vertebral tuberculosis (Ortner, 2003: 230). Within the vertebral body, abscesses cause it to collapse. The ventral surface of the sacrum may also be affected. Even though most of the characteristic alterations are observed in the spine, the bone destruction may be multifocal. On the visceral surface of the ribs, new bone formation may be formed as a response to a pulmonary infection. Septic arthritis may develop in a joint, most commonly in a hip or a knee. The lytic bone destruction is infrequently bilateral and, when the phalanges in the hand or metatarsals are involved, *periostitis* results in expansion rather than bone resorption—*dactylitis* (Ortner, 2003: 242). Similar to leprosy, tuberculosis may lead to bone changes in the facial region (*lupus vulgaris*) (Ortner, 2003: 253, 2008a). This relatively uncommon form of extrapulmonary tuberculosis can be observed as cutaneous lesions, sometimes located near the nose, which, if left untreated, may lead to secondary inflammatory changes of the skeletal elements (e.g. Roberts & Cox, 2003: 230f). In the lower extremities, the tibiotalar joint and the tarsal bones may be affected. The destruction may extend to the metatarsals. Abscesses and fistulae may develop in the body of the talus and calcaneus. Ortner (1999) presented a list of pathological changes that in addition to tuberculosis may lead to destructive lesions in the spine: *actinomycosis*, *brucellosis*, *echinococcosis*, *mycosis*, *sarcoidosis*, *staphylococcosis*, *syphilis*, *aneurysm*, *trauma* and *tumors*.

In *brucellosis* bone involvement is rare but when the skeleton is affected, it most often occurs in the spine (Aufderheide & Rodríguez-Martín, 1998: 192). The vertebral injuries are often multiple. *Brucellar spondylitis* most usually affects the lumbar region but multiple sites may be involved. The infection triggers the formation of abscesses causing lytic lesions followed by sclerotic reaction and new bone formation, i.e. both reparative and destructive processes (Aufderheide & Rodríguez-Martín, 1998: 193; Resnick & Niwayama, 1988: 2659).

Echinococcus is a parasite that causes the development of cysts in the abdominal region of the host. When the skeleton is affected, which is uncommon, the lower part of the spine is the most exposed area, where both the body and the posterior elements may be involved (Aufderheide & Rodríguez-Martín, 1998: 242). The bone changes consist of osteolytic lesions that may, in a very late stage, cause vertebral fractures and secondary infections (Aufderheide & Rodríguez-Martín, 1998: 242).

Aneurysms may cause pressure and erosive lesion on several vertebrae anteriorly. The resulting scalloped defects are concentrated in the centre of the bodies, sparing the endplates of the bodies (Ortner & Putschar, 1981: 247).

Vertebral *fractures* on rare occasions involve several bodies (e.g. Steinbock, 1976: 183; Bennike, 1999). Furthermore, compression fractures often result in a wedge-shaped appearance and may show signs of a callus formation (Aufderheide & Rodríguez-Martín, 1998: 140f, 315).

In addition, malignant bone tumours as well as a non-specific inflammation such as *osteomyelitis* will result in destructive bone lesions on both the vertebral bodies and the pedicles (Mays & Taylor, 2003).

Discussion

None of the six skeletons from the Humlegården block demonstrates a complete pathognomonic set of lepromatous processes (i.e. rhinomaxillary syndrome, 'leprous periostitis', erosive changes in the distal phalanges of the hands and metatarsophalangeal joints of the feet and phalanges).

Case 1 demonstrated ankylosis of the finger bones, symmetrical and bilateral subperiosteal bone reaction in the lower legs, severe changes in the feet, including exostoses in the calcanei, talus and cuneiform, and acroosteolysis in the metatarsals and phalanges. The skull was missing and could not be examined. Though the postcranial skeletal changes display a close resemblance to leprosy, possible changes in the rhinomaxillary region (pathognomonic of lepromatous leprosy) could not be investigated and differential diagnoses must be contemplated. *Periostitis* or *pyogenic osteomyelitis* may explain some bone resorption but the bilateral symmetrical subperiosteal distribution suggests a systemic disease. Also, no sequestrum has been noted. Skeletal changes due to treponemal disease include, except for *caries sicca*, a hypertrophic expansion of the tubular bones and a vast new bone formation, neither of which were observed in

Case 1. The anatomical distributions of lesions in Case 1 are not typical for frostbites or rare fungal infections such as cryptococcosis and mucormycosis. Also, the usual geographic location for rhinoscleroma and leishmaniasis in tropic or subtropic areas makes the probability of their occurring in central Sweden minimal. Peripheral nerve damage due to Thevenard's disease may result in similar bone destruction of the feet as observed in Case 1. The process with subsequent bone destruction of infected ulcers in pressure points of the feet is the same as in secondary leprosy. Psoriatic arthritis foremost affect the axial skeleton and when the hands are involved, the destructive changes are asymmetrical. The lesion pattern in sarcoidosis tends not to be symmetrical and metatarsals are not typically involved. The rounded lesions with sclerotic borders associated with actinomycosis do not resemble the acroosteolysis of Case 1. Since the individual did not exhibit any changes in the spine, diseases that characteristically affect vertebrae are not considered.

Case 2 was a complete skeleton but the region around the nose was destroyed post-mortem. Mild resorptive changes were seen in the hands. As in Case 1, symmetrical and bilateral subperiosteal bone reaction in the lower legs together with acroosteolysis was observed. The feet exhibited cloacae of the first metatarsal and pencilling of the metatarsals in addition to some resorption of phalanges. Due to the similarities in skeletal destruction to Case 1, the same discussion of alternative diagnoses is valid. The cloacae imply osteomyelitis, i.e. a severe inflammatory condition but from an unknown cause.

Case 3 exhibited atrophied phalanges in a hand and subperiosteal changes of the long bones in the legs where the tibiae and fibulae were thickened by new bone deposits. In the feet several tarsal bones, metatarsals and phalanges exhibited dorsal exostoses or were deformed by inflammatory processes. No skeletal changes were observed in the maxillary region. Again, the same differential diagnosis as with Case 1 could be used. However, in Case 3 it was possible to examine the cranium and no rhinomaxillary destruction was observed.

Cases 1, 2 and 3 demonstrate evidence of systemic infection in the lower extremities. The pencilling of mainly the first and fifth metatarsal and changes of the calcanei suggest impaired arterial circulatory supply at pressure points of the feet. Some kind of peripheral neuropathy such as leprosy or Thevenard's disease, where anaesthesia leads secondarily to ulceration, infection and inflammation of bones and joints, could be a possible cause. However, the latter is a rare

disorder: the general incidence of all motor and sensory neuropathies is 30:100 000 (Nicholson, 2007). In lepromatous leprosy, it is expected that rhinomaxillary changes will be found as well. In Cases 1 and 2, it is impossible to confirm or eliminate this alteration and Case 3 did not display any changes. This makes it hazardous to diagnose the skeletal changes as being caused by leprosy. There is no clear line between tuberculoid leprosy and lepromatous leprosy. A gradual progression of one form to the other depending on the immunology of the individual seems to exist (Manchester, 2002). Even though tuberculoid leprosy is believed to cause injuries due to loss of sensation in the hands and feet, the lesions are usually single, making this form of the disease difficult to diagnose (Aufderheide & Rodríguez-Martín, 1998: 145; Manchester, 2002). Aufderheide & Rodríguez-Martín (1998: 146) further state that bone destruction in the form of neglected trauma may 'occur earlier and more intensely' in tuberculoid leprosy. The same authors affirm that anaesthesia is more prominent in tuberculoid and borderline leprosy than in lepromatous leprosy. Some scholars suggest that a systemic, concentric bone loss of phalanges and metaphorical together with signs of neuropathic change in tarsals could be used as an operational definition for leprosy as well as rhinomaxillary syndrome (Andersen *et al.*, 1994; Ortner 2008a, b; Waldron 2009). In the present study, except for lack of information about rhinomaxillary changes (Cases 1 and 2) or lack of rhinomaxillary destruction (Case 3), the skeletal lesions and locations fulfill the criteria for leprosy. However, since the observed skeletal changes are believed to be secondary to an infection and similar to other infectious diseases, it is not possible to confirm that the individuals suffered from the disease.

Case 4 demonstrates severe pathological bone destruction of the body of three lumbar vertebrae in the spine, thickening of the tibiae and fibulae and changes in the feet. The skull was missing. The facts that the destruction in the spine was unifocal, located in the lumbar region where the surface of the remaining fragments in the bodies of L1–L3 show little reactive bone or bone repair, and the neural arches were spared suggest that the individual suffered from tuberculosis spondylitis. Alternative diagnoses as suggested by Ortner (1999) are not consistent with the osseous changes observed in Case 4. When spinal changes occur in actinomycosis, the destruction involves the neural arch but spares the intervertebral discs. In brucellosis reactive new bone formation and sclerotic bone may be observed and the lesions are often multiple. Both echinococcosis and mycosis involve the

posterior elements of the vertebrae and the former also includes fractures of the body with secondary infections. Sarcoidosis may result in multifocal destruction of the spine. Staphylococcosis and other bacteria leading to pyogenic osteomyelitis result in vast new bone formation and sequestra. In syphilis, the cervical region is involved. Non-infectious conditions such as trauma, aortic aneurysm and tumours result in other types of changes: i.e. wedge-shaped bodies or destruction that includes the pedicles and neural arch, which is not the case in this individual. Additionally, the erosion on the ventral surface of the sacrum shows a close resemblance both in anatomical position and surface appearance to previously examined skeletons diagnosed with tuberculosis (Blondiaux *et al.*, 1999; Ortner, 2003: 236).

Tuberculosis could also explain the inflammatory changes including osteomyelitis in the lower legs and feet. However, knee involvement is rather common in skeletal tuberculosis (16% cases) (Aufderheide & Rodríguez-Martín, 1998: 139). The surfaces of the proximal joints of the tibiae of Case 4 are unaffected. Furthermore, the focus of the inflammation in the feet caused by tuberculosis is not the joints of the tarsal bones. In addition, the symmetric, concentric atrophy of the proximal phalanges is not characteristic of tuberculosis, frostbite, rheumatoid arthritis or psoriasis arthritis. The osteomyelitis and ankylosis could therefore be the result of some other systemic infection, including leprosy. An individual with both leprosy and tuberculosis is neither extraordinary nor unique. The two diseases are closely related since they are caused by the same genus *Mycobacterium* and they have coexisted for centuries in many societies. The occurrence of both diseases in the same individual has been demonstrated both in archaeological samples and modern clinical examples (e.g. Vachharajani *et al.*, 1977; Agarwal *et al.*, 2000; Molto, 2002; Ortner, 2002; Brutzer 1898 in Ortner, 2003: 265; Donoghue *et al.*, 2005). In a modern study, Vachharajani *et al.* (1977) observed patients with pulmonary tuberculosis in combination with both types of leprosy, i.e. the tuberculoid and the lepromatous form, although the tuberculoid type was more common. According to Aufderheide and Rodríguez-Martín leprosaria patients often died as a result of complications of tuberculosis (1998: 147). The severe destructive changes in the spine of Case 4 indicate that the individual, if infected by both diseases, either had contracted the infection *M. tuberculosis* a long time in advance or that the more virulent character of this bacteria resulted in more severe skeletal changes than the contemporaneous infection of *M. leprae*. The skeletal changes in the spine

of this individual are characteristic for tuberculosis but the indications of a second leprosy infection are weak and are considered unconfirmed.

Case 5 was the skeleton from an 11–12-year-old child who exhibited changes to the face in the rhinomaxillary region resulting in a shortening of the roots of the central incisors, a lytic lesion of a fibula and concentric atrophy of the distal phalange.

The facial changes with a resorbed nasal spine in Case 5 meet the criteria for lepromatous and near-lepromatous leprosy (Andersen & Manchester, 1992). Due to the large numbers of bacilli in the nose of sufferers of lepromatous leprosy, the nasal spine, septum and aperture are often resorbed (Ortner 2003: 264). Possible differential diagnoses include rhinoscleroma, leishmaniasis, treponemal disease, tuberculosis and non-specific infection. Rhinoscleroma and leishmaniasis are for geographical reasons not likely to have caused the lesions. *Caries sicca* and *saber tibia*, the characteristic lesions in syphilis, are not observed in the individual. In tuberculosis, *lupus vulgaris* may lead to secondary inflammations of the facial bones with both bone resorption and new bone formation. The same reactive processes are observed in a non-specific infection. The lytic lesion of the fibula show some resemblance to actinomycosis but in this disease the lesions are multiple and foremost located to the cerviofacial region or the spine. In Case 5, there is little reactive bone in the nasal aperture implying that none of the alternative syndromes could explain the bone alterations. Furthermore, the shortening of the roots of the central incisors observed in the same individual are comparable to changes in the teeth that have been documented in other children diagnosed with the disease (*leprogenic odontodysplasia*) (Danielson, 1968; Møller-Christensen 1978). This odontological defect is rare and in bioarchaeology it has, at date, been seen only in Danish juvenile skeletons (C. Roberts 2009, pers. comm.). It has been suggested that *leprogenic odontodysplasia* may develop in children younger than 9 years, if the roots of the teeth are deformed due to the close contact between incisor germs and a pathological process such as an early development of rhinomaxillary syndrome (Danielson, 1968).

As with many other infections, young individuals are more likely to contract leprosy. Today, in areas where leprosy is endemic, the disease is typically manifested in children and juveniles between the ages of 10 and 20 (Newman *et al.*, 1972; Resnick & Niwayama, 1988: 2688; Mitchell, 2000: 249). In afflicted families, up to 50% of the children under 5 years of age develop clinical signs of the disease and by the age of 13, changes in the nasal region may be observed (for a

review of childhood leprosy, see Lewis, 2002). Using X-rays, absorption of the distal ends of the distal phalanx is seen in 9-year-old children (Newman *et al.*, 1972). In the Swedish records from a 19th-century leprosy sanatorium, 11% of the patients were below 20. In a photograph from 1906 a 13-year-old boy shows a severely disfigured face where the nose is completely destroyed and the nasal cavity is an open hole (Sundelin & Sörman, 2004). The same records describe the sufferings of a 13–14-year-old girl, exhibiting lesions in the face, legs and feet in addition to a left claw-hand deformity. Hence, even though children have less time to develop clear signs of leprosy, clinical and historical sources show that both primary and secondary changes to the skeleton at a young age occur and may be observed. In bioarchaeological settings, several examples of affected children have been documented (e.g. Møller-Christensen, 1961, 1978; Arcini, 1999; Roberts, 2002). For instance, in the leprosaria at Næstved, 10 out of 12 children in the age category *Infantile II* (with a mean of 10 years) exhibited nasal spine atrophy (Møller-Christensen, 1961: tab. 5). Postcranial bone lesions associated with neuropathy were also observed, though in fewer numbers.

Case 6 showed remodelling in the area surrounding the piriform aperture, although the anterior nasal spine and the central alveolar bone of the maxilla were lost post-mortem. The lower extremities displayed minor subperiosteal bone formation and in the feet, changes were observed in the metatarsals and phalanges. Fragmentation and bone changes of an unspecific character made it difficult to diagnose the cause of the alterations. As with Cases 1–5, leprosy, rhinoscleroma, leishmaniasis, fungal infections, treponemal disease, frostbite, peripheral neuropathy, sarcoidosis, actinomycosis, psoriatic arthritis, rheumatoid arthritis can all serve as possible diagnoses. Clinical studies have indicated that people with leprosy also suffered from maxillaris sinusitis to a higher degree than others although this has not been verified in skeletal assemblages (Boocock *et al.*, 1995). The remodelling of the piriform aperture in Case 6 with signs of both erosive and proliferation bone reaction resembles skeletal manifestations of leprosy but also can be caused by many diseases. The porotic remodelling is of a general character and could be the result of a common chronic inflammation in the maxillary sinus (sinusitis). Likewise, the localised lytic necrosis in one metatarsal and two phalanges could be caused by various non-specific or specific causes in addition to leprosy. Due to a different geographical and anatomical distribution of lesions, rhinoscleroma, leishmaniasis, cryptococcosis,

mucormycosis, treponemal disease and frostbite may be considered less likely. In contrast to Cases 1–4, the bone destruction is not consistent with secondary destruction due to peripheral nerve damage. The changes do not meet the criteria of small cyst-like structures described for sarcoidosis or the multiple spheroid lesions typical for actinomycosis. Similar to skeletal changes caused by psoriatic arthritis, the lesion pattern is asymmetrical but the sacroiliac joint and the distal phalanges are unaffected. The round-edged, lytic changes in the margin of the metatarsophalangeal joint of the fifth metatarsal in Case 6 resemble rheumatoid arthritis: however, since this a systemic polyarthritis one would expect several metatarsals to be involved in a symmetrical pattern. Since localised trauma with a subsequent infection and necrotising processes could explain the bone changes in the feet, the alterations in the face and the lower extremities need not to be related. Hence, the skeletal changes in Case 6 are diffused and could not be confidently associated with a specific disease.

The fact that six individuals with a more or less comparable pathologic pattern (not observed in any other individual in the same sample of 227 skeletons) were buried close to each other in the peripheral southern part of the churchyard is not in itself evidence of an identical disease. However, it is compelling that four skeletons (Cases 1–4) share an anatomical, symmetrical, bilateral distribution of subperiosteal bone reaction in the lower legs and characteristic, concentric atrophy in the metatarsals and phalanges of the feet, which has been described, albeit together with rhinomaxillary syndrome, as signs of leprosy. The skeletal changes of Case 5 imply that the individual suffered from lepromatous leprosy while Case 6 is difficult to diagnose. The group demonstrates at least that they suffered from a severe systemic disease and the burial location in what is believed to be a parish cemetery may suggest that they shared a comparable social status. The identification of infectious diseases such as leprosy and tuberculosis in the present study is extraordinary but not unique in Sigtuna. In another cemetery excavated in 1995, a skeleton suggestive of leprosy was exhumed (Arcini, unpublished report in Kjellström, 2005). Interestingly, this skeleton was buried in a location similar to the present individuals, in the periphery of the churchyard. Together these discoveries suggest that the six individuals, being severely disabled, belonged to the lowest group in the social hierarchy, illustrating the social stratification most often presented in traditional historical textbooks (for a review, see Rawcliffe, 2006). Hospitals or organised institutions segregating the disabled from

the rest of the community are not believed to have existed in Sigtuna before the end of the 13th century. Hence, persons affected by leprosy or other severe diseases had to be buried in the same churchyard as healthy citizens. According to written records a hospital was founded in AD 1287, close in time to the abandonment of the churchyard, though the location of this institution is not known.

During the analysis of the complete skeletal assemblage a variety of different pathological conditions was documented (Kjellström & Wikström, 2008). In addition to the most commonly observed (e.g. trauma and osteoarthritis), cases suggestive of ankylosing spondylitis, DISH and gout were documented. Furthermore, in comparison with 239 skeletons from contemporary Sigtuna, significantly higher frequencies of fractures, degenerative joint disease, unspecific subperiosteal bone infections and enamel hypoplasia were observed (Kjellström & Wikström, 2008). This implies that the quality of health of the individuals buried at the Humlegården block was rather poor. The high frequency of degenerative joint disease, unspecific subperiosteal bone infections and enamel hypoplasia indicates a more physically stressful living already from an early age, which corresponds to the idea that the group buried in this location of the churchyard was of low social status. On the other hand, one of the several causes for the development of gout could be a high intake of alcohol together with rich food, and obesity has been connected to DISH (Julkunen *et al.*, 1971), suggesting that people suffering from the effects of overindulgence were also buried in the same area. This diverse collection of afflictions indicates that at this early stage in Sigtuna, until the founding of a hospital, people affected with serious diseases as leprosy and tuberculosis were still considered to be a part of society.

Conclusions

In short, one of the examined individuals (Case 5) displayed atrophy of the nasal spine, which is interpreted as rhinomaxillary syndrome pathognomonic for lepromatous leprosy. Additionally one individual (Case 4) demonstrated characteristic changes associated with tuberculosis. Four skeletons (Cases 1 to 4) had severe bone atrophy in their feet (and three of them showed mild bone alterations in their hands). The bone alterations imply a systemic disease such as leprosy but other diseases could not be ruled out. A sixth individual (Case 6) exhibited inflammatory changes in the facial area as well as atrophy in the hands and lytic lesions in

the feet. The pathological pattern is of a general character and it is difficult to provide a clear-cut diagnosis, but in this case leprosy could also be involved.

The grouping of people suffering from severe disorders, together with data from the rest of the churchyard, presents some clues about the treatment and attitude towards the sick and disabled in Sigtuna in the period before the foundation of a hospital.

The present results will be followed up. To confirm whether leprosy or tuberculosis indeed was the cause of the bone lesions observed in the individuals without pathognomonic alterations, an aDNA analysis is planned.

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