Introduction

Leprogenic odontodysplasia (LO) or *dens leprosus* is an anomalous root development of the upper permanent incisors (Danielsen 1970, Roberts 1986). This malformation was first observed in 1966 by V. Møller-Christensen and K. Danielsen in an 8-9 years old individual exhumed from the St. Jørgen’s medieval leprosarium cemetery in Naestved, Denmark (Danielsen 1970). Later, Danielsen (1970) reported a total of 4 juveniles (8 to 11 years old) amongst the nearly 1000 skeletons unearthed from Danish medieval leprosaria cemeteries. The co-occurrence of LO with *facies leprous* led Danielsen to conclude that it was “caused by and [...] specific to low-resistance [lepromatous] leprosy in childhood” (p. 19).

This study aims to present new evidence of LO from the medieval Scandinavian archeological record and to discuss the paleopathological challenges underlying this rare condition.

Findings

Rhinomaxillary changes

Enlargement and destructive remodeling of the pyriform aperture (Fig. 2A), including the anterior nasal spine (Fig. 2B, C).

Pitting and new bone formation in the nasal surface of the palatine process of the maxilla.

“Empty nose” due to destruction of the intranasal structures.

LO was found in a loose upper right central incisor presenting a shortening of the root length that was concentrically constricted from 1.5 mm above the neck until the apex (Fig. 2D, E). The left central upper incisor is not present.

The alveolar sockets of the upper incisors were almost completely reabsorbed (Fig. 2C) and difficult to be noticed due to the accentuated destructive remodeling of the anterior maxillary alveolar process (Fig. 2A, C).

Postcranial changes

Hand bones: No lesions were noticed on the 19 bones preserved.

Lower legs: Bilateral new bone formation on the tibial and fibular diaphyses.

Foot bones: 22 bones were preserved (11 tarsal, 10 metatarsals and 1 proximal phalanx).

Woven bone and enlarged foramina at the medial surface of calcanei.

Woven bone on metatarsals diaphyses: 1st, 2nd, 3rd left and 2nd, 3rd, 4th right.

Achroosteolysis with nicking of the distal end on a foot proximal phalanx.

Discussion and Conclusions

This individual represents an important paleopathological case because:

As Danielsen’s (1970) report, this skeleton presents LO in association with rhinomaxillary changes ascribed to *rhinomaxillary syndrome* (Andersen 1992) or *facies leprous* (Møller-Christensen 1961) and occurs in a non-adult individual.

It exhibits a combination of lesions, rarely observed in children (Lewis 2008), namely rhinomaxillary changes and achroosteolysis in a foot phalanx, which are compatible with a diagnosis of lepromatous leprosy (Ortner 2008, Waldron 2009).

Based on the current state of knowledge, LO occurrence is always concomitant with:

1) rhinomaxillary changes;
2) non-adults (<20 year old) individuals;
3) Scandinavian leprosaria cemeteries.

LO and atrophy of the anterior maxillary alveolar process are thought to be caused by bacilli invasion into the pulp cavity (Sakai and Matsumoto 1968, Danielsen 1970) and surrounding premaxillary bone. Both areas present lower body temperature what is essential to *Mycobacterium leprae* survival and multiplication (Rendal et al. 1973).

Surprisingly, LO was not reported since Danielsen’s publication, i.e. in the last four decades. Hitherto, no clinical cases are described and its etiological and epidemiological significance are poorly understood.

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Material

St. Jørgen’s leprosarium cemetery (Fig. 1) was in use between the 13th-16th/17th centuries (Arentoft 1999). The human remains exhumed are housed at the ADBOU (Anthropological Database of the Odense University), at Danish University.

Skeleton 572 was exhumed during the 1980/1 archeological campaign.

This individual died between 13-19 years old (Matos 2009). The skull present 24 teeth, 21 in the alveoli, and the facial bones are well preserved.

Thus, many questions are still waiting for an answer:

Is LO pathognomonic of leprosy? If so, does it indicate a very early onset of clinical leprosy in childhood (before 6 years old)?

Our finding reinforces the need of research regarding the nosologic and epidemiologic significance of LO. This will benefit from further clinical evidence, both from old medical literature and leprosy patients from endemic countries. Additionally, systematic search of this condition in skeleton from leprosaria and regular cemeteries is recommended.

References


