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Hemifacial microsomia (oculo-auriculo-vertebral spectrum) in an individual from the Teramo Sant'Anna archaeological site (7th–12th centuries of the Common Era, Italy)

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ABSTRACT

Background: This study is based in an analysis of the skeletal remains of an adult male from the Teramo Sant'Anna archaeological site (7th–12th centuries of the Common Era, Teramo, Italy).
Results and discussion: The individual shows distinct abnormalities that principally involve asymmetric hypoplasia and dysmorphogenesis of the facial skeleton. The combination of these findings and the absence of abnormalities of the spine strongly suggest diagnosis of the congenital malformation known as hemifacial microsomia. This very heterogeneous syndrome affects primarily aural, ocular, oral and mandibular development. Despite the lack of clinical information and the absence of soft tissue, it was possible to perform a differential diagnosis for this palaeopathological case. Mastication was probably altered considering that the mandible is extremely asymmetric and lacks true condyles. The temporomandibular joints are present, but the right one is hypoplastic and abnormal in shape. There is evidence of bilateral dislocation, and the facial muscles are hypertrophic.
Conclusions: This case represents an important contribution to the palaeopathological literature because this is an uncommon condition that has not been widely documented in ancient skeletal remains.

1. Introduction

Facial asymmetry is common in humans, which is seen as a disproportionality or imbalance between the right and left sides. Indeed, a mild degree of asymmetry is normal and acceptable in the average face (Chia, Naini & Gill, 2008; Jones & Tier, 2012). However, when the disparity is significant, this can cause both functional and aesthetic problems through a series of factors that can affect the underlying skeletal structure and/or soft tissue. The aetiology includes: (i) congenital disorders that originate prenatally; (ii) acquired disorders that result from injury or disease; and (iii) developmental deformities that arise during growth without any conspicuous aetiology (Cheong & Lo, 2011).

Oculo-auriculo-vertebral spectrum (OAVS) is a congenital malformation complex that is characterised by a wide spectrum of symptoms and phenotypes that can vary in severity from case to case. However, such abnormalities tend to principally involve asymmetric hypoplasia and dysmorphogenesis of the facial skeleton, the external and middle ears, the facial soft tissues, and/or the spine. In 20% of individuals with OAVS, there is marked facial asymmetry, although some degree of asymmetry is evident in 65% of cases (Cohen, Rollnick & Kaye, 1989; Gorlin, Cohen & Hennekam, 2001). In the literature there is considerable discrepancy in the reported frequency of OAVS, ranging from 1 per 5,642 individuals (Grabb, 1965) to 1 per 45,000 individuals (Morrison, Mulholland, Craig & Nevin, 1992).

Hemifacial microsomia (HFM) is one of the more common forms of this OAVS complex, and it primarily affects aural, oral and mandibular development. HFM can vary from mild to severe, and in many cases, involvement is limited to one side of the face, although bilateral involvement can also occur with more severe expression on one side (Gorlin et al., 2001). Goldenhar syndrome is considered as a variant of HFM, and it is characterised additionally by vertebral anomalies and epibulbar dermoids (Gorlin et al., 2001).

Thus, OAVS encompasses both HFM and Goldenhar syndrome. Goldenhar syndrome might also be a more complicated form of OAVS, while HFM might represent a milder form. Thus, an individual with just a small or abnormal ear and no other problems, might be at the very mildest end of this spectrum (Cohen et al., 1989).

Although OAVS is a relatively common modern clinical finding, few cases have been reported in the palaeopathological literature. The

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published cases have been exclusively based on palaeoepidemiological studies (Castro, Moreno & Aspillaga, 1997), morphological examinations of skeletal remains (Ginestà, Díaz & Ollich, 2013; Nagar & Arensburg, 2000; Panzer, Cohen, Esch, Nerlich & Zink, 2008), or ceramic culture (Pachajoa, Rodríguez & Isaza, 2010). It appears that the relative lack of cases reported to date might be due to several factors, such as the little attention that has been paid to this condition in recent years.

The purpose of this paper is to describe a probable case of OAVS in an individual recovered from the Teramo Sant'Anna archaeological site (7th-12th centuries of the Common Era (CE); Teramo, Italy). Considering the lack of similar evidence from archaeological skeletal remains worldwide, this case is also extremely valuable to the understanding of the incidence of this type of congenital malformation among past populations.

2. Material and methods

2.1. Archaeological context

In 1980, the *Soprintendenza Archeologica dell'Abruzzo* undertook a series of annual excavation campaigns under Sant'Anna Square, in the historic centre of Teramo (Fig. 1). During the period from 1980 to the summer of 1990, the remains of a Medieval Cathedral were discovered, which according to historic documents existed between the centuries 6th-14th CE, next to a Roman *domus*. Dozens of graves were also found, which contained mainly well-preserved skeletal remains. There were also hundreds of human and animal bones distributed sporadically at various stratigraphic levels throughout the excavated area (Capasso, Di Muzio, Di Tota & Spoletini, 1990). The Medieval necropolis extended all around the perimeter of the Cathedral, so that part of it also impinged on the ruins of the ancient Roman *domus*. Radiocarbon dating demonstrated an outward spreading use of the necropolis with respect to the perimeter of the Cathedral (Calderoni & Petrone, 1990); indeed, the graves closest to the perimeter walls of the Cathedral are the oldest (600–685 CE), compared to the more distant ones, located on the Roman *domus* (1000–1160 CE) (Capasso et al., 1990).

2.2. Preservation and completeness

The skeletal remains considered here were classified as individual T.D-I.31. The cranium was almost complete (i.e., missing only a fragment of the occipital bone), with a well-preserved mandible. The upper body elements were optimally preserved despite the fragmentation of the ribs and left scapula. The lower body elements were also well preserved. The only missing bones were the majority of the right and left tarsals, carpals, metatarsals, metacarpals and phalanges. The skeletal remains are housed at the University Museum of Chieti ('G. d'Annunzio' University of Chieti–Pescara, Chieti, Italy).

Fig. 1. Location of Teramo, in the region of Abruzzo, Italy.