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SHORT REPORT

A Case of *Os odontoideum* in the Palaeopathological Record

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ABSTRACT *Os odontoideum* is an uncommon abnormality of the second cervical vertebra, the aetiology of which is contentious. Whether congenital or acquired, the condition often results in atlantoaxial instability and subsequent compression of cervical cord or vertebrobasilar vessels. The bioarchaeological study of a medieval sample from the Christian necropolis of Cacela Velha (Algarve, southern Portugal) provided the opportunity to describe one of the first cases of this rare anomaly ever reported in the palaeopathological record. Copyright © 2007 John Wiley & Sons, Ltd.

Key words: Os odontoideum; palaeopathology; medieval; Cacela Velha; Portugal

Introduction

Cacela Velha is a fortified village located in the Algarve, southwestern Portugal. The foundation of Cacela Velha, by the Phoenicians, goes back to the year 804 BC (Garcia, 2002). Under Roman rule the village was known as *Hicetele Cacetala* and became an important fortress, protecting the ancient bay of Montegordo. Throughout the Islamic dominance of the Iberian Peninsula, Cacela Velha exerted a significant role in *Gharb al-Andalus* (the name the Arabs gave to the southwestern region of the Iberian Peninsula) as a commercial and religious centre (Cavaco, 1984; Sanchez, 2000).

The warrior monks of the military order of Santiago, commanded by Dom Paio Peres, subjugated Cacela Velha in 1239 (Brandão, 1632). The Islamic village was destroyed. In its place Christians built a small church with an adjacent necropolis. This sepulchral space was active during the 13th and 14th centuries (Garcia, 2002).

This paper considers the presence of *os odontoideum*, a rare abnormality of the axis vertebra, in an adult female individual from the Christian necropolis of Cacela Velha.

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Material and methods

In 1998 and 2001, during extensive archaeological excavations by the 'Historical Patrimony Centre of Cacela', 74 individuals were exhumed from the Christian necropolis of Cacela Velha. The bones are in good condition and the skeletons are almost completely intact, both because they were buried in the sand, which is an excellent preservation medium, and because excavation was conducted extremely carefully (Garcia, 2002).

The presence of *os odontoideum* was recognised through macroscopic evaluation of an adult female skeleton. Observation was aided by a magnification lens and plain radiographic analysis.

Os odontoideum

The axis (second cervical vertebra) is the pivot upon which the atlas – but not the head – turns

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(Wynsberghe *et al.*, 1995). Embryologically, the axis is formed from the proatlas, the C1 sclerotome and the C2 sclerotome (Dai *et al.*, 2000). It ossifies from five prime centres of ossification – from both halves of the neural arch, from the true centrum of the axis, and from each half of the body of the dens (Scheuer & Black, 2000). The true centrum of the axis begins ossification from a solitary centre between the fourth and fifth months of intra-uterine life (Wollin, 1963). At about the same time, two laterally placed ossification centres emerge in the odontoid process, which rapidly unite so that the intradental synchodrosis fuses, undoubtedly by the time of birth and perhaps as early as 7 or 8 months *in utero* (Ogden, 1984). An intradental sulcus is found on the posterior surface of the dens and persists until 3 or 4 years of age (Scheuer & Black, 2000). The dentocentral junctions mingle between 4 and 6 years of age. All lines of fusion will generally disappear by 9–10 years, even though a diminutive horizontal fissure may remain for quite some time (Fullenlove, 1954). It is thought that this may be a place of latent weakness if there is trauma in this region (Scheuer & Black, 2000). At approximately 2 years of age, *ossiculum terminale* appears. This nodule increases in size and ultimately fuses with the apex of the dens at the age of 12 (Todd & D'Errico, 1926).

As a rare abnormality of the second cervical vertebrae, *os odontoideum* has been infrequently acknowledged since Giacomini's original description in 1886 (Dai *et al.*, 2000). *Os odontoideum* is discernible as a separate round or oval ossicle situated faintly cephalad to, or in the normal position of, the odontoid process. There is a transverse gap between the ossicle and the body of the axis (Clements *et al.*, 1995; Fukuda *et al.*, 2003). The cranial portion of the dens fragment continues to obtain a blood supply from the apical arcade. The caudal segment becomes avascular and resorbs, leaving the characteristic rounded ossicle. Below the ossicle, there is a shortened odontoid process protruding from the vertebral body of the axis above the atlantoaxial facet joint space. The size of the *os odontoideum* may vary, but it is typically smaller than the normal dens (Teng *et al.*, 1989).

The aetiology of *os odontoideum* remains indefinite, but both acquired and congenital mechanisms have been suggested. In the allegation of congenital aetiology for *os odontoideum* (Greenberg, 1968), two theories have been advanced: failure of the odontoid process to fuse with the body of the axis, and failure of the odontoid apex to fuse with the main portion of the odontoid process (Fukuda *et al.*, 2003). Today, this last failure is known as persistent *ossiculum terminale* (Barnes, 1994; Scheuer & Black, 2000). *Os odontoideum* has been distinguished to have a round or oval shape and a smooth margin. These findings uphold the congenital mechanism (Dai *et al.*, 2000). The congenital origin for *os odontoideum* is also supported by some authors who emphasise its association with syndromes such as trisomy 21, Klippel-Feil syndrome and multiple epiphyseal dysplasia (Sherk & Dawoud, 1981; Forlin *et al.*, 1992). Furthermore, familial *os odontoideum* has been reported (Kirlew *et al.*, 1993).

In contrast, Fielding *et al.* (1980) attributed *os odontoideum* to an unrecognised rupture in the region of the base of the odontoid. They postulated that subsequent to a fracture of the odontoid process, a contraction of the alar ligaments exerts a distraction force that pulls the portion away from the base, leading to a disruption of the blood supply to the odontoid process which induces non-union. Ricciardi *et al.* (1976), Ellis & Kaan (1993) and Galli *et al.* (2001) described cases of an acquired *os odontoideum* subsequent to trauma, and considered that *os odontoideum* was due to disruption of blood supply secondary to a purely ligamentous injury. Schuler *et al.* (1991) elegantly described the development of an *os odontoideum* subsequent to trauma in a child. Besides that, unlike most congenital malformations, *os odontoideum* tends to occur as an isolated entity without other regional anomalies.

Os odontoideum may be clinically silent, but when the odontoid process is disrupted the atlas will be displaced along with the odontoid process, leading to atlantoaxial unsteadiness. The reserve space in the atlantoaxial articulation is relatively greater, so patients sometimes have no complaints or neurological deficits although atlantoaxial instability occurs. Nevertheless, if atlantal displacement exceeds the limit of reserve space, clinical symptoms will emerge (Dai *et al.*,

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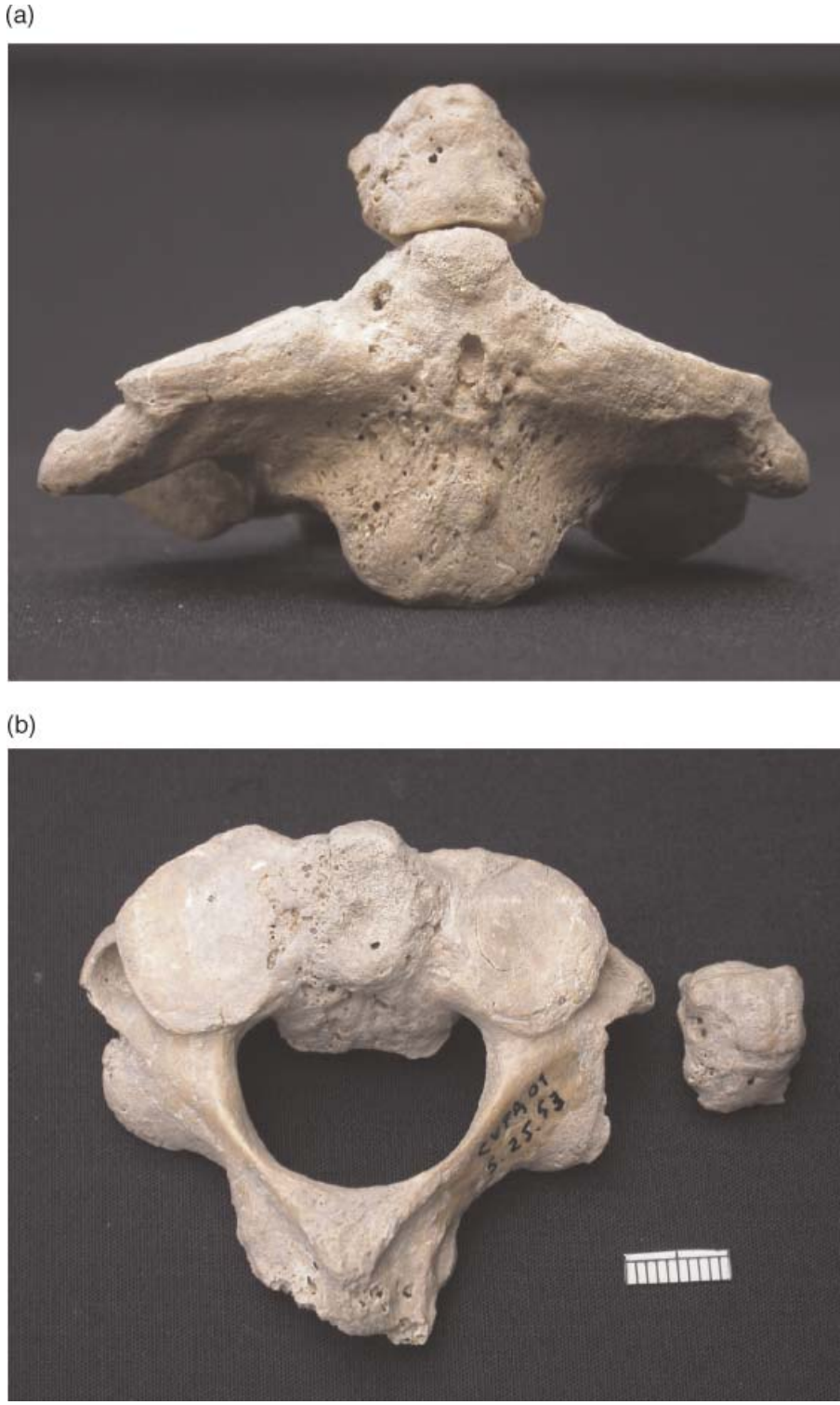

Figure 1. The axis vertebra of the study skeleton: the dens of the axis is transversely separated from the body. (a) Anterior view. (b) Superior view. This figure is available in colour online at www.interscience.wiley.com/journal/oa.

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Figure 2. Detailed view of the ossicle. This figure is available in colour online at www.interscience.wiley.com/journal/oa.

2000). Symptoms reported by patients are intricate, and comprise cervical pain, increased or decreased range of cervical movement, hand weakness, nausea and vertigo, among many others (Clements *et al.*, 1995; Galli *et al.*, 2001).

The pathological skeleton

The present study refers to a female individual of middle age (30–50 years). The skeleton is in very good condition and almost complete. This individual's height was about 160 cm (Mendonça, 2000). Incipient vertebral osteoarthritis was reported in the thoracic and lumbar column. In the inferior plate of T6 to T12 and the superior plate of L2 and L4, Schmorl's nodes were noted. From T1 to L3 laminal spurs were recorded. No other pathologies were registered in this skeleton.

In this individual the dens of the axis was transversely separated from the body (Figure 1a, b). The separate ossicle was placed in the same position as the dens and displays a round shape with a smooth margin at the base. The ossicle measured 13 × 13 mm (Figure 2). Underneath the ossicle there was a protrusion from the vertebral body of the axis, higher than the atlantoaxial facet joint space. This protuberance

Figure 3. Close view of the axis showing the protrusion from the vertebral body of the axis beneath the ossicle, higher than the atlantoaxial facet joint space. This figure is available in colour online at www.interscience.wiley.com/journal/oa.

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exhibited a distinct sclerotic reaction, which has promoted new bone formation with a fine-grained appearance (Figure 3). The atlas facet for dens displayed significant modification. This articular surface showed an irregular, abnormal conformation, with no signs of osteoarthrosis.

Discussion

The anomaly observed in the axis vertebra of an adult female skeleton exhumed from the Christian necropolis of Cacela Velha probably relates to an abnormality known as *os odontoideum*. The odontoid process is clearly separated from the body of the axis, presenting a round configuration and a smooth margin base. This configuration suggests a congenital origin (Dai *et al.*, 2000). Below the ossicle, there is a shortened odontoid process protruding from the vertebral body of the axis above the atlantoaxial facet joint space, showing sclerotic reaction and new bone formation. As such, this particular case could also be the result of a dental fracture. A minor traumatic event may result in unnoticed bony or ligamentous injury. The blood supply of the dens enters superiorly via the accessory, alar and apical ligaments, and inferiorly through the body of the axis. As a result, non-union occurs in as many as 62% of odontoid peg fractures. Trauma might compromise its feeble blood supply, resulting in either non-union of a fracture or arrested ossification and failure of fusion (Fielding *et al.*, 1980; Clements *et al.*, 1995).

The lesion observed could be attributed to another nosological possibility: *ossiculum terminale persistens*. However, persistent *ossiculum terminale* is differentiated from *os odontoideum* by its ossicle, which is much smaller. Also, in the former condition the apex of the dens is 'v'-shaped (Scheuer & Black, 2000), which is clearly not the case here. The *ossiculum terminale* typically attaches to the anterior aspect of the atlas, but it may append to the anterior edge of the foramen magnum or remain a detached entity (Barnes, 1994).

Conclusion

The bioarchaeological study of a medieval skeletal sample from Cacela Velha (Algarve, southern Portugal) offered the oppurtunity to depict one of the first cases of an abnormality of the second cervical vertebra, *os odontoideum*, ever reported in the palaeopathological record.

The rarity of this anomaly surely causes difficulties for its positive identification in archaeological settings. However, the unremitting presence of an experienced anthropologist in the field and careful excavation of all the small bones of the skeleton will certainly bring new cases of *os odontoideum* to light.

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