Spastic quadraparesis following a relatively minor injury - the ‘os odontoideum’

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Abstract
We present a case of an adult male who sustained severe spinal injury following relatively minor injury. Plain film, computed tomography (CT) scans and magnetic resonance (MR) images aided the diagnosis of os odontoideum with C1-2 subluxation and high cervical spinal cord compression. Incidental detection of an os odontoideum should not be considered as a variant anomaly but should be referred for appropriate spinal evaluation and surveillance.

Introduction
Os odontoideum is an ossicle with smooth circumferential margins representing the odontoid process or dens. It lacks continuity with the body of C2 and its importance lies in the fact that trauma may compromise a previously asymptomatic atlanto-axial joint. We present a case of an adult male who sustained severe spinal injury following relatively minor injury. We believe that an os odontoideum should not be considered as a variant anomaly, and because such patients are vulnerable to severe spinal cord injury, they should be referred for spinal evaluation and surveillance.

Case report
A 22-year-old man sustained an injury to his neck when he was pushed into the back of a pick-up truck. He reports that when his head hit a metal plate in the truck, he felt dizzy followed by a sensation of pins and needles in his arms and legs. He was subsequently unable to move his arms and legs at all. His childhood had been uneventful, and he had worked as a bricklayer’s assistant.

He was admitted with a diagnosis of spastic quadraparesis. He has recovered to some degree, being able to...
walk unaided for short distances and has regained some power in both hands. Plain film, computed tomography (CT) scan and magnetic resonance (MR) images are presented in Figs 1 - 5. A diagnosis of os odontoideum with C1-2 subluxation and high cervical spinal cord compression was made.

Discussion

Os odontoideum (Greek 'tooth like') is a rare anomaly of the second cervical vertebra which was first described from postmortem studies by Giacomini in 1886. By definition, an os odontoideum is a small accessory ossicle with smooth, well-corticated borders that is separated from the base of a shortened odontoid process. The size and shape of the ossicle can vary. The atlanto-axial joint exhibits the greatest mobility in the vertebral column and is also the least stable. Os odontoideum frequently leads to atlanto-axial instability with spinal-cord compression. Atlanto-axial instability can arise from multiple causes including inflammatory arthritides, trauma, infection, laxity of the transverse ligament, e.g. trisomy 21 and other congenital abnormalities. Plain cervical spinal radiographs are usually sufficient to obtain a diagnosis.

Os odontoideum has been classified into two anatomic types, orthotopic and dystopic. Orthotopic describes an ossicle that moves with the anterior arch of C1, whereas dystopic defines an ossicle that is functionally fused to the basion. Minor trauma can produce symptoms when the dystopic os is freed and subluxes anterior to the arch of C1.

The gap between the os odontoideum and the odontoid process extends to above the superior articular facet of the atlanto-axial joint. This is in contrast to an acute fracture or non-union of the odontoid in which the gap is narrow and irregular and extends into the body of the axis below the level of the superior facet.
Os odontoideum is associated with hypoplasia of the posterior arch as well as anterior arch hypertrophy, compensating for the increased stress placed on the latter. It occurs commonly in trisomy 21, Klippel-Feil and other cranio-cervical malformations.

The aetiology of os odontoideum remains unsettled but this does not play a major role in its diagnosis or management. The proponents who feel that it is a congenital abnormality suggest that the mechanism involves the failure of the os terminale to fuse, or failure of the main part of the odontoid process to fuse with the body of the axis. Fielding et al. attributed os odontoideum to a disunited fracture by presenting cases with a history of trauma and previously normal radiographs. They propose that subsequent to an odontoid fracture, the proximal portion is pulled by the contraction of the alar ligament and non-union is facilitated by vascular interruption. In our case, a second ossicle inferior to the anterior arch is uncommon and probably points to a traumatic origin.

The literature supports the ability of cervical plain films to establish the diagnosis of os odontoideum but their sensitivity and specificity as well as the need for additional confirmatory studies is still unreported. Flexion and extension plain films have been used to depict the degree of C1 and C2 instability. Anterior subluxation of the os in relation to the body of C2, as was the case in this patient, predominates over posterior subluxation. CT scans have been used to better define the bony anatomy of the os odontoideum and the odontoid process. The role of MRI has been to evaluate the spinal cord and to provide additional anatomical information. Both CT and MRI assist in operative planning of unstable os odontoideum.

The degree of C1-2 instability does not seem to correlate with the neurological status in patients with os odontoideum and a sagittal spinal canal diameter on plain films of 13 mm or less is strongly associated with myelopathy. Asymptomatic patients with os odontoideum, with or without C1-2 instability may be managed with clinical and radiological surveillance. Those with symptoms and/or neurological signs and C1-2 instability may be managed with posterior C1-2 internal fixation and fusion.

**Conclusion**

Os odontoideum is a potentially mechanically unstable condition which is associated with trisomy 21 and other congenital conditions. Minor trauma may result in severe spinal cord injury. Cervical plain films are usually diagnostic. Both CT and MRI assist in operative planning. The incidental detection of an os odontoideum should not be considered as a variant anomaly but should be referred for appropriate spinal evaluation and surveillance. This may have special relevance in children whose sporting activities may have to be restricted.