

Case 2082: Os odontoideum: CT and MR findings

Subspecialty: Neuroradiology

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Patient:

Age: 38 year(s)

Sex: female

Clinical Summary

Occipital pain syndrome after minor cervical trauma.

Clinical History and Imaging Procedures

A patient with a history of recent minor cervical trauma and occipital pain, underwent CT examination of the craniocervical junction (CVJ). The examination showed the presence of a separate ossicle apart from a hypoplastic dens. An MR examination was performed in order to exclude any possible associated abnormalities. The presence of an "os odontoideum" was confirmed. No cervical cord lesions were present. An associated slight herniation of the cerebellar tonsils below the foramen magnum without syrinx was also found.

Discussion

A wide spectrum of acquired, congenital or developmental abnormalities occur at the craniocervical junction (CVJ) because of the complex developmental anatomy and the complex transition between the spinal cord and the brainstem. There are five types of odontoid anomalies: os odontoideum, ossiculum terminale, hypoplasia of the odontoid, aplasia of the odontoid base and aplasia of the odontoid. All five anomalies may lead to atlantoaxial instability and may cause identical neurological symptoms. Only radiographic findings are distinctive. The term "os odontoideum" was introduced by Giacomini in 1886 and refers to an independent bone found cranial to the axis, in the place of the dens. Plain films, CT and MR demonstrate an ossicle with rounded, cortical borders that is separated by a variable gap from the odontoid process. It is usually located in the position of the normal odontoid tip at the base of the occiput in the region of the foramen magnum. It may be fused to the basiocciput. Furthermore CT and MR may show the presence of granulation tissue owing to repeated subluxations. Many radiological indexes of stability have been proposed, however it has been shown that the excursions between flexion and extension do not reflect the true extent of instability. For this reason the value of radiological indexes is still debated. The pathogenesis of os odontoideum has been variously explained on an embryological, traumatic, or vascular basis. Some authors have recently suggested that the os odontoideum is the product of excessive movement at the time of ossification of the cartilaginous dens. Os odontoideum may be associated with Down's syndrome, Morquio's syndrome and spondyloepiphyseal dysplasia. An association has been shown between os odontoideum and congenital cervical (C5-C6) vertebrae fusion and basilar impression. Minor neck trauma is frequently associated with the onset of symptoms, which may vary from neck or occipital pain to compressive myelopathy. Decompression and/or stabilisation of the CVJ represents the therapy of choice for symptomatic patients. If atlantoaxial arthrodesis is not possible, occipitocervical fusion should also be considered in the operative management. Acute trauma may dramatically worsen symptoms, leading to urgent surgical procedures. When os odontoideum is found incidentally, close clinical and MR follow-up is recommended.

Final Diagnosis

Os odontoideum

Figure 1: CT of the craniocervical junction

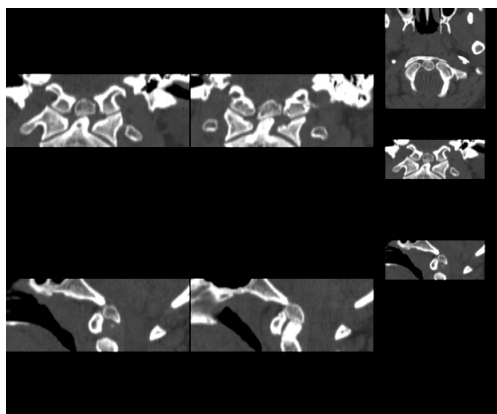


Figure 1a

This multi-planar CT reconstruction shows the presence of an ossicle, located in the position of the normal odontoid tip, with rounded, smooth cortical borders separated by a small gap from the odontoid process. No fractures are seen.

Figure 2: Sagittal T2-weighted image of the cervical spine



Figure 2a

The sagittal T2-weighted image confirms the presence of the os odontoideum. No cervical cord lesions are visible. A slight herniation of the cerebellar tonsils below the foramen magnum is present. Please note a small bright area just below the os odontoideum probably due to the presence of granular tissue.

Figure 3: Sagittal T1-weighted image of the cervical spine

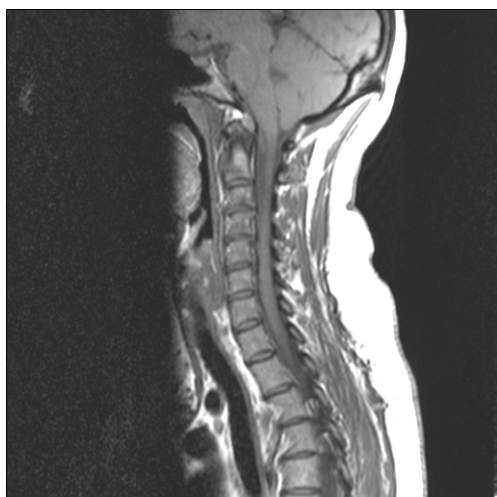


Figure 3a

The sagittal T1-weighted image confirms the radiological findings described in Figure 2.

MeSH:

[A02.835.232.834.151.383] Axis

[A02.835.232.834.151.383.668] Odontoid Process

The toothlike process on the upper surface of the axis, which articulates with the atlas above.

[C10.500.680.291] Arnold-Chiari Malformation

A group of congenital malformations involving the brainstem, cerebellum, upper spinal cord, and surrounding bony structures. Type II is the most common, and features compression of the medulla and cerebellar tonsils into the upper cervical spinal canal and an associated MENINGOMYELOCELE. Type I features similar, but less severe malformations and is without an associated meningocele. Type III has the features of type II with an additional herniation of the

entire cerebellum through the bony defect involving the foramen magnum, forming an ENCEPHALOCELE. Type IV is a form a cerebellar hypoplasia. Clinical manifestations of types I-III include TORTICOLLIS; opisthotonus; HEADACHE; VERTIGO; VOCAL CORD PARALYSIS; APNEA; NYSTAGMUS; swallowing difficulties; and ATAXIA. (From Menkes, Textbook of Child Neurology, 5th ed, p261; Davis, Textbook of Neuropathology, 2nd ed, pp236-46)

References:

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