Recurrent Syncopal Episodes Due to Atlanto-Axial Instability – An Unusual Presentation of Os Odontoideum

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ABSTRACT

Os odontoideum is a rare cause of atlantoaxial instability. Despite being first described in 1886, its pathogenesis is still unclear, with theories for both congenital and traumatic etiologies. We report on an atypical presentation of this pathology, in a patient who was being investigated for recurrent syncopal episodes during performance of everyday activities, and who was unexpectedly found to have an os odontoideum. The patient underwent occipitocervical fusion and had complete resolution of all her symptoms postoperatively.

CASE DESCRIPTION

A 45-year-old female was referred to the neurosurgical department after being investigated for recurrent syncopal episodes for one week's duration. She reported progressively worsening light-headedness on any bending forward which resulted in loss of consciousness for approximately 15–30 seconds. This occurred while doing daily chores and activities. There was no acute history of trauma or heavy lifting. She denied having any headaches, neck pain/stiffness, nausea/vomiting, fever or seizures. No cardiovascular symptoms including palpitations, dyspnoea or chest pain were elicited.

In her past history, she reported being in a motor vehicular accident at the age of 12, and spending several weeks in a cervical collar, but was unable to give further details on the incident. No imaging was available from that incident. The patient had no other significant medical history, was a non-smoker and had no comorbidities.

On neurological examination, her GCS was 15/15, and she appeared comfortable. Her gait was normal, with no ataxia or spasticity. There was increased tone in all her limbs, with 5/5 power demonstrated in all myotomes. She was hyper-reflexic (3+) with upgoing plantars bilaterally, but no clonus. No Hoffman's sign was elicited. Cranial nerve examination was normal. The patient experienced significant light-headedness during flexion of the neck, which resolved once the head was returned to a neutral position. She had neither neck tenderness nor a positive Spurling's sign.

Both non-contrast CT and MRI scans of the brain were performed, with neither revealing any intracranial abnormalities. However, the lower cuts showed upper cervical spinal cord compression. Controlled flexion/extension X-rays, CT and MRI of the cervical spine were then ordered. There was an abnormality of the odontoid process, and a significantly widened atlanto-dental interval of 8mm (normal < 2.5mm in adult females). The anterior arch of C1 had an ossicle fused to it. There was no evidence of an acute fracture. Flexion extension X-rays showed significant motion of the ossicle-C1 complex in relation to the dens (Figure 1). There was evidence of myelomalacia of the cervical cord on T2-weighted MRI sequences (Figure 2). Notably, the left vertebral artery was hypoplastic. This was in keeping with a diagnosis of os odontoideum, and likely causing vertebra-basilar ischemia. Investigations for rheumatoid arthritis were performed, with both the erythrocyte sedimentation rate (5mm/hr) and rheumatoid factor (8 IU/mL) being within normal reference ranges for the patient's age.

The patient was counselled about the need for surgical stabilisation of the upper cervical spine, and consented for posterior C1-2 fusion, with possible occipito-cervical fusion if necessary. Neuromonitoring was utilized. A posterior midline incision from the inion to C4 was made, and subperiosteal dissection of the musculature performed. Exposure of C1 lateral masses and C2 pedicles was done. The C1 lateral masses were not amenable to safe screw placement, and the decision made for occipito-cervical fusion. Lateral mass screws were placed in C3, and an occipital plate and rod construct created, with final occiput-C3 fusion (Figure 2). Intraoperative X-rays revealed good position of the hardware, and standard layered closure was done. There was no vertebral artery injury.

Postoperatively, the patient had no further light-headedness or syncopal episodes on bending forward. She was discharged to the outpatient clinic. On 3-month follow-up, she remained symptom free, and had resumed full activity and, at one year, had no recurrence of syncope.

DISCUSSION

Introduction

Since the first description of os odontoideum (OO) by Giacomini in 1886, it remains a rarely encountered pathology.[1] There is separation of a portion of the odontoid process from the remainder of the axis, leading to a well-circumscribed proximal ossicle. This ossicle may either be in its normal position (orthotopic) or in any other position (dystopic).[2] This includes the foramen magnum region, fusion to clivus or anterior arch of C1.[3]

Etiology

While its pathogenesis still remains unclear, two major theories are proposed. Firstly, congenital lack of ossification and fusion at the boundary between the basal dens and body of C2 has been described by Pang et al.[4] This congenital theory is supported by the fact that there is an increased incidence in families, twins and Down's syndrome.[5] The second major theory is that of a post-traumatic etiology, with a chronic non-united fracture of the dens due to avascular necrosis and osseous remodeling.[2,3,6]

Clinical Presentation

This can be widely varied, including occipito-cervical neuralgia, neck pain, pyramidal signs, lower cranial nerve deficits, vertebra-basilar ischemia or even totally asymptomatic. One study of 279 patients managed over the course of 10 years showed pyramidal signs and extremity weakness to be the most common signs, which is in contrast to previous studies that described neck pain to be the primary complaint.[2] Symptoms can be vague and subtle, and of a chronic nature, but precipitated by mild trauma.[5]

Figure 1: Top row – Dynamic (flexion/extension) X-rays showing ossicle-C1 complex motion in relation to the dens. Bottom row – Axial CT showing ossicle – C1 anterior arch complex, and sagittal CT displaying widened atlanto-dental interval of > 8mm.

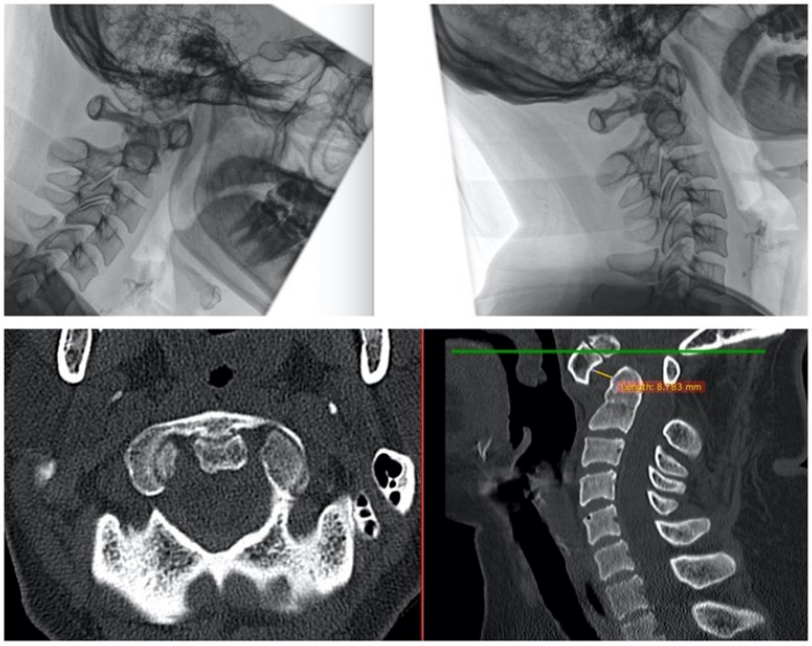
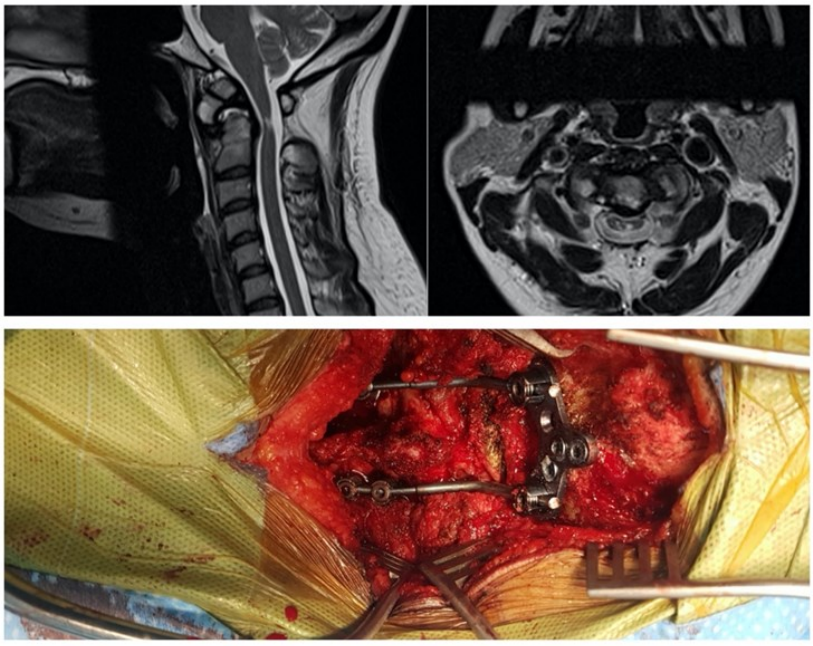


Figure 2: Top – T2 MRI sequence showing myelomalacia of upper cervical cord (bright spot posterior to dens. Bottom – final intraoperative occipito-cervical construct.



Investigations

Plain X-rays can be difficult to interpret, but dynamic flexion-extension views usually reveal the movement of the ossicle—C1 anterior arch complex in relation to C2. CT scans give more precise anatomic definition and diagnosis of OO as well as the type present (orthotopic vs dystopic). It also allows measurement of the atlanto-dental interval to be taken, and thus assessment of atlanto-axial instability. MRI gives information on any compromise of the spinal cord, and can show vertebral artery anomalies, which are critical in operative planning.

Treatment

Stability of the C1-2 complex requires an intact odontoid process, transverse and alar ligaments.[3] Therefore, OO is inherently unstable. Surgical management is the standard of care and involves posterior fusion of the C1-2 complex. This reduces motion and the subsequent risks of upper cervical cord injury, including sudden death, stroke, worsening myelopathy, paraplegia, etc. Surgical options are transarticular C1-2 fixation (modified Magerl), C1 lateral mass – C2 pedicle (Goel-Harms) and occiput—C2 pedicle screw (Abumi).[2,7] Based on individual anatomy and intraoperative findings, the fusion may be extended to include subaxial lateral mass screws, as in our patient (occiput-C3).

Outcomes

Goel et al, in a series of 190 surgically treated cases over the course of 20 years, showed excellent clinical and surgical outcome, particularly with the Goel-Harms technique. No patient in their series needed reoperation, and there was resolution of symptoms clinically.[5] There will be a significant loss (50%) of rotation of the neck with C1-2 fusion, and patients may experience difficulty in adjusting to this. Inclusion of further subaxial and occipital segments further reduces the range of cervical motion and patients should be appropriately counseled prior to surgery.

CONCLUSION

Os odontoideum remains a rarely encountered pathology, with incredible clinical variability in its symptomology. Its true pathogenesis is still not confirmed, but there is evidence for both congenital and traumatic etiologies. By definition, it is an unstable atlanto-axial pathology, and surgical fusion should be offered to all symptomatic patients. At this time, only level 3 evidence exists to guide its management.

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