Os odontoideum was first described in the 19th century by Giacomini, who found that it is associated with craniovertebral junction instability; it is an abnormality of the cervical axis characterized by an independent ossicle, which might be variable in size.

“Os odontoideum” is a Latin term for “a tooth-like bone”. Fielding et al classified os odontoideum into dystopic and orthotopic, based on the relationship of the odontoid process to the axis and the clivus. The ossicle is identified as orthotopic when it is located at its normal position posterior to the anterior arch of C1 and dystopic when it is located in a displaced position and usually fused with the basion. The small odontoid process is separated from the ossicle which is usually lying cranial to the superior facets of C2.

The synchondrosis that separates the dens and the base of C2 usually fuses at the age of 3 years. The ossiculum terminale, which is the tip of the odontoid process, starts appearing on plain radiographs at the same age comprising the secondary ossification center and fuses with the dens at the age of 12 years.

The debate on the etiology of os odontoideum remains yet unsettled. It was believed in the past to be a congenital anomaly due to failure of fusion. The hypothesis was supported by the finding of a familial form of the condition in identical twins with no traumatic history. However, most scholars nowadays support the hypothesis of a posttraumatic phenomenon representing the condition due to a previous odontoid synchondrosis fracture that did not achieve union.

Os odontoideum is commonly seen in males in their second and third decades. Patients with this condition may be asymptomatic or symptomatic of a wide range of neurological deficits and vascular insults.

Descriptions of the presentation, optimal management, and surgical interventions for os odontoideum are limited and controversial.

The aim of this presentation is to report a dystopic ossicle and to emphasize the widely variable natural history of this condition and to discuss the management options currently preferable.

THE CASE

A fifty-seven-year-old female presented with progressive cervical and sub-occipital pain radiating to both shoulders and upper arms for the past year. The pain was associated with progressive gait imbalance and dizziness. There was no history of trauma.

Examination revealed spastic gait, severe midline tenderness of the neck, painful cervical range of motion, positive Lhermitte’s sign, bilateral upper and lower limb hyperreflexia with clonus, positive Babinski sign, positive Hoffman’s test, and inverted brachioradialis reflex. No motor weakness was found.
Plain radiographs of the cervical spine along with flexion and extension views revealed the presence of os odontoideum (dystopic type), see figure 1 (A-D). Further imaging revealed significant thinning of the spinal cord (2 mm) and high signal within the cord consistent with myelomalacia, see figure 2 (A-C).

Intraoperative, the patient underwent oral fiberoptic intubation and was positioned prone by logrolling with the head positioned on Mayfield headrest. Under fluoroscopic guidance, closed reduction of C1-C2 achieved. Subsequently, a midline incision was made, and she underwent instrumented C1-C2 fusion according to Goel and Harms technique with posterior decortication and application of allograft, see figure 3. The patient tolerated the procedure with no perioperative complications or postoperative new neurological deficit.

The postoperative period was uneventful. The patient was kept on a hard cervical collar and started mobilization the next day. Six months postoperatively, the patient’s pain had decreased significantly, her dizziness and numbness subsided, and her gait improved with persistence of clonus and hyperreflexia.

**DISCUSSION**

Studies revealed supporting evidence that symptomatic patients with os odontoideum associated with radiological or clinical instability should undergo surgical fusion. Asymptomatic patients can be managed conservatively provided there is regular clinical and radiological follow-up. Such patients should be aware of the possibility of disease progression and neurological deterioration even with minor trauma. Therefore, strict avoidance of contact sports should be advised.

In 1968, Greenberg et al classified atlantoaxial dislocation as reducible (RAAD) or irreducible (IAAD). This classification is mandatory because the surgical strategy in the management of os odontoideum is quite different in the case of reducible relative to irreducible atlantoaxial instability. Special considerations should be taken when managing os odontoideum operatively; unlike cases of recent odontoid fractures, direct anterior screw fixation is found to be associated with significantly lower rate of fusion; therefore, contraindicated in cases of os odontoideum. Nevertheless, anatomical variations of the vertebral artery should be excluded prior to surgery by performing CT angiography or magnetic resonance arteriography.

As performed in our case, the principle of treating reducible atlantoaxial dislocation (RAAD) is reduction followed by posterior atlantoaxial osteosynthesis fixation and arthrodesis. In cases of irreducible atlantoaxial dislocation (IAAD), the treatment goal is to protect the cervicomedullary structures from further compression and to relieve the current cord compression. In such cases where the deformity is irreducible, cord decompression can be achieved through two different approaches.
strategies: resecting the compressive pathology, or by releasing C1-C2 facet joints anteriorly or posteriorly. The decompression should be followed by stable internal fixation and fusion12.

Current clinical and radiological outcomes of surgical treatment of os odontoideum indicate that Goel and Harms fusion, which was used in our case, is considered a sound option for posterior C1-C2 arthrodesis in adults and even pediatric cases of os odontoideum13,14.

The range of cervical rotation decreases by approximately 40%–50% following atlantoaxial fusion. Older adults usually adapt well to this decrease. Nevertheless, avoiding serious complications of the neural and vascular compression weighs out such outcome5.

As implemented in the reported case, the current advancements in contemporary segmental screw fixation methods improved surgical outcome regarding fusion rates and C1-C2 stability. These advancements also allowed us to avoid halo-vest immobilization which helped to improve postoperative rehabilitation15.

CONCLUSION

Os odontoideum is a cervical spine anomaly that has a variable natural history, a wide range of presentation and a controversial etiology that may necessitate future work to improve evidence. Multiple radiological modalities are necessary to guide the treatment, although plain radiographs are usually diagnostic. Surgical stabilization is the management option of choice in symptomatic patients with evidence of myelomalacia. The surgical advancements in treating os odontoideum have significantly improved the outcome and eliminated the need of postoperative brace stabilization.

REFERENCES