

Dystopic Os Odontoideum: A Rare Cause of Atlantoaxial Instability

¹Joe M Das, ²K Krishnakumar, ³Suresh Nair

ABSTRACT

Os odontoideum is a relatively common developmental anomaly of the craniovertebral junction (CVJ), which is usually an incidental finding on spine imaging. But the dystopic variety of os odontoideum is uncommon and can be baleful. This anomaly can be a cause of atlantoaxial instability requiring surgical fixation. But the diagnosis of this variety requires considerable knowledge of anatomy of the CVJ as this unfamiliar entity needs to be differentiated from other CVJ anomalies like persistent os terminale, accessory ossicle of anterior arch of atlas, assimilation of atlas, and, most importantly, from orthotopic variety of os odontoideum. Here, we present a 34-year-old lady who presented with gradual-onset weakness of right side of the body. A computed tomography scan of the cervical spine revealed dystopic os odontoideum.

Keywords: Cervical atlas, Congenital abnormalities, Joint instability, Odontoid, Spine.

How to cite this article: Das JM, Krishnakumar K, Nair S. Dystopic Os Odontoideum: A Rare Cause of Atlantoaxial Instability. *J Spinal Surg* 2017;4(1):38-39.

Source of support: Nil

Conflict of interest: None

INTRODUCTION

A 34-year-old lady presented with gradual-onset weakness of right side of the body and tingling sensation in right upper limb for the past 1 month. Motor system examination revealed that she had mild elbow flexion weakness and hand grip weakness bilaterally, more on the right side. Plantar reflex was bilaterally extensor and all the deep tendon reflexes were exaggerated. Sensory system examination was normal. A computed tomogram (CT) scan (Fig. 1) of the cervical spine was performed, which showed dystopic os odontoideum (DOO) with posterior tilting of dens and hypertrophy of anterior arch of atlas.

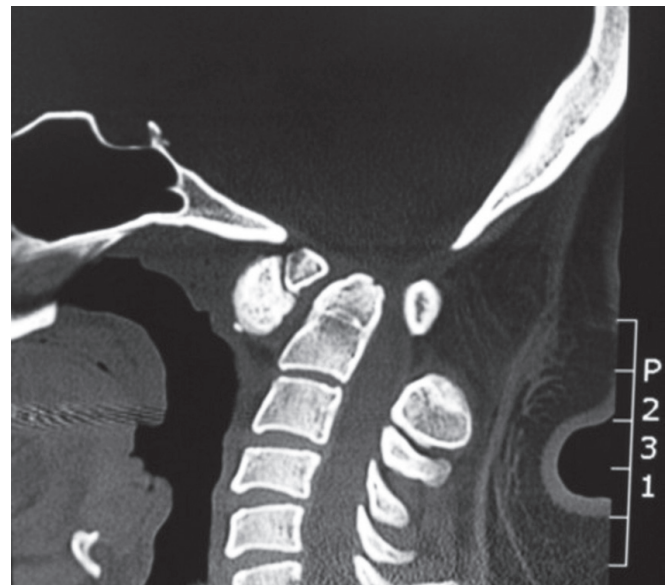


Fig. 1: Midsagittal plain CT scan of cervical spine showing dystopic os odontoideum

Os odontoideum refers to an independent ossicle located rostral to the axis bone in the position of odontoid process, which is separated from a hypoplastic dens by a variable distance and can mimic a type II dens fracture. There are two types of os odontoideum: Orthotopic and dystopic.

In the orthotopic variety (Fig. 2A), the ossicle lies in the location of the normal dens and moves with the axis body and the ventral arch of the atlas. This type is often associated with an intact cruciate ligament.

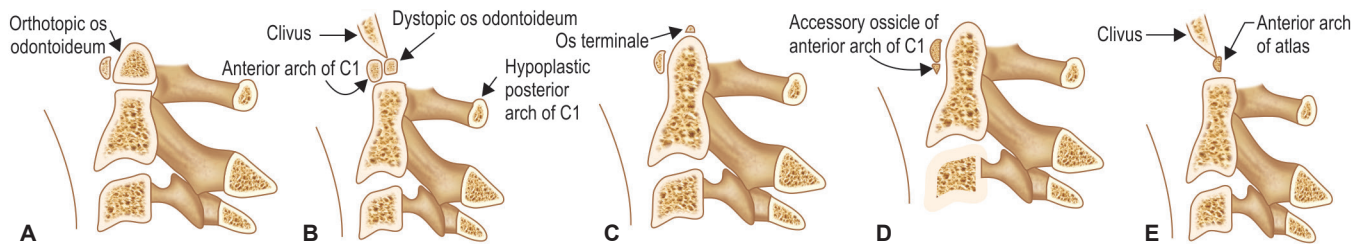
In DOO (Fig. 2B), the os is located near the basion and is often fused to the clivus. The ventral arch of the atlas is hypertrophied, and the dorsal arch is hypoplastic. As named by von Torklus and Prescher, DOO (os avis) has a greater likelihood of causing neurologic compromise than the orthotopic variety. This may occur because of dorsal compromise of the spinal cord by the ventrally located dorsal arch of the atlas during flexion; and ventral compromise by the odontoid ossicle. The DOO is thought to be congenital in origin.¹

The treatment of os avis depends on the other anomalies. Pure instability can be treated with C1-C2 posterior fusion. An absent posterior C1 arch or an occipitalized atlas would necessitate inclusion of the occiput into the fusion. Concomitant neural compression

¹Fellow, ²Additional Professor, ³Professor

¹⁻³Department of Neurosurgery, Sree Chitra Tirunal Institute for Medical Sciences and Technology, Thiruvananthapuram Kerala, India

Corresponding Author: Joe M Das, Fellow, Department of Neurosurgery, Sree Chitra Tirunal Institute for Medical Sciences and Technology, Thiruvananthapuram, Kerala, India, Phone: +919447092342, e-mail: drjoemdas@gmail.com



Figs 2A to E: Diagrammatic representation of midsagittal CT imaging of cervical spine showing: (A) Orthotopic os odontoideum; (B) dystopic os odontoideum; (C) persistent os terminale; (D) accessory ossicle of anterior arch of atlas; and (E) assimilation of atlas

caused by basilar impression or invagination of the opisthion may require simultaneous decompression.²

A persistent os terminale (Fig. 2C) refers to failure of fusion of the secondary ossification center along the superior margin of dens and can mimic a type I dens fracture.³

Accessory ossicle of anterior arch of atlas (Fig. 2D) is a normal anatomical variant which needs to be differentiated from the calcific tendinitis of longus colli or stylohyoid.

Assimilation of atlas (Fig. 2E) refers to the congenital fusion of atlas to occiput. Three fusion zones have been described by Gholve et al.⁴ Zone I involves anterior atlantal arch, zone II involves lateral masses of atlas, and zone III involves posterior atlantal arch fusion.

REFERENCES

1. La Bagnara, M.; Sure, DR.; Smith, JS.; Shaffrey, CI. Anatomy and physiology of congenital spinal lesions. In: Steinmetz, MP.; Benzel, EC., editors. *Benzel's spine surgery: techniques, complication avoidance, and management*. 4th ed. Philadelphia: Elsevier; 2017. p. 143-144.
2. Pang, D. Embryology of the cranio-vertebral junction and congenital malformations in the region. In: Banerji, D.; Pauranik, A., editors. *Progress in clinical neurosciences*. Vol. 25. New Delhi: Byword Books; 2011. p. 44.
3. O'Brien WT Sr, Shen P, Lee P. The dens: normal development, developmental variants and anomalies, and traumatic injuries. *J Clin Imaging Sci* 2015 Jun;5:38.
4. Gholve PA, Hosalkar HS, Ricchetti ET, Pollock AN, Dormans JP, Drummond DS. Occipitalization of the atlas in children. Morphologic classification, associations, and clinical relevance. *J Bone Joint Surg Am* 2007 Mar;89(3):571-578.