Case Report

Cervical myelopathy involving os odontoideum with retro-odontoid cyst and atlanto-axial instability: A case report

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ABSTRACT

Os odontoideum is an unusual anatomic variant of the dens of C2 defined as an independent osseous element separated from the axis; the etiology is a topic of debate, with investigative studies supporting congenital and traumatic origins, clinical manifestations vary from asymptomatic forms, underlying C1-C2 instability to compression of the spinal cord or verteobasilar ischemia. We report a case of a patient with a history of minor trauma 5 years ago, she suffered from neck pain. The clinical examination was normal. Radiological examination including X-ray, CT, and MRI showed cervical myelopathy involving os odontoideum with C1-C2 instability and compressive retro-odontoid cyst. Imaging has an important role in the management of os odontoideum, from diagnosis to therapy.

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Introduction

The definition of os odontoideum is common throughout the literature; an osseous mass that is non-continuous with the body of C2 [1,5,6], first described in Giacomini in 1886 [13,1,7]. The etiology of os odontoideum remains debated in the literature with evidence for both traumatic and congenital theories [1,5,6]. The etiology, however, does not play an important role in diagnosis or subsequent management [5].

Patients with os odontoideum can be asymptomatic or present with a spectrum of neurological deficits [1]. Diagnosis usually can be made with X-ray, MRI, and CT can assess spinal cord integrity and C1-C2 instability [1,6].

Asymptomatic cases without evidence of radiologic instability are typically managed with regular clinical and radiologic examination [2,3]. The existence of atlanto-axial insta-

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We report a case of a 29-year-old woman, who had a history of minor trauma 5 years ago in a car accident, and she was not explored because of being asymptomatic, without any radiological investigation.

She suffered from neck pain 2 years ago. Clinical examination was normal without neurological deficits. Radiological examination including X-ray (Fig. 1); CT (Fig. 2) and MRI (Fig. 3) showed os odontoideum with retro-odontoid compressive cyst and C1-C2 instability engendering cervical myelopathy.
For being asymptomatic without neurological manifestations or deficit, after consultation with a multidisciplinary health team including; neurosurgeons, neurologists, and radiologists, and thorough discussion with the patient, the decision was to adopt close surveillance with periodic neurological and radiological examination and to operate subsequently to guarantee the stability.

Discussion

The etiology of os odontoideum is controversial and different theories exist.

Congenital hypothesis: The den is formed by the fusion of the apex and the body of dens, the apex originating from the pro atlas sclerotome and the body originating from the first cervical sclerotome [1,4]. The axis results from the fusion of the dens and the body of the axis originating from the second cervical sclerotome. The apex of dens, the body of dens, and the body of the axis are arranged in an up to down manner to form upper and lower synchondroses among them and the failure of ossification and calcification of lower synchondrosis result in os odontoideum [1,4]. Os odontoideum is also associated with congenital malformations, such as Klippel–Feil syndrome, Down syndrome, Marquio syndrome, or neurofibromatosis, which further strengthens the congenital theory [1,11]. This condition is also reported in identical twins supporting the congenital theory [4].

Acquired hypothesis: The deficiency in arterial blood supply and the rareness of trabecular bone in the base of the dens, this make it vulnerable to injuries caused by minor trauma or repeated micro-trauma, the apex of dens separates from the base of the dens, and then os odontoideum is formed by subsequent contraction of the Alar ligament - connecting the occipital bone and the tip of the dens - and cephalic displacement of the broken ends of the dens [1,12].

Still, some other theories suggest the combination of traumatic and congenital origins [4].

Regardless of the etiology, the clinical management of os odontoideum remains the same [1,6].

Clinical manifestations vary from asymptomatic forms, underlying atlanto-axial instability in the early stage to compression of the spinal cord [1,6]. With the increase the age, cervical myelopathy is aggravated, atlanto-axial stability was decreased by cervical trauma, symptoms such as neck pain, torticollis, numbness, weakness, and activity limitation appeared, then paralysis or death can occur in serious conditions [1,6,5,7]. C1–C2 instability can lead to stenosis with resultant myelopathy secondary to vascular compromise, the tension of the cord, or bony compression. Vertebro-basilar ischemia can occur due to either kinking or thrombosis of the vertebral arteries [6]. In very rare cases retro odontoide cyst due to C1–C2 instability is the origin of spinal compression [2].

Mouth-open cervical anterior-posterior radiograph show os odontoideum with a smooth intact cortex separated from the body of the axis. Flexion-extension lateral radiographs showed atlanto-axial instability and spinal canal stenosis [14]. CT images show clearly the morphology and location of the os odontoideum; atlas and axis, CT with multiplanar reconstructions can show the adjacent structures and also be helpful to identify other malformations of craniovertebral junction [1]. MRI has superior soft-tissue resolution and obvious advantage in showing spinal cord compression and signal changes in the spinal cord [1].

The C1–C2 instability with normal vertebral morphology usually was evaluated by measuring the atlas-odontoid interval (A–OI); it can be defined as A–OI more than 3 mm in adults and 4 mm in children, respectively [1,9]. Spinal stenosis was defined as the effective sagittal canal diameter of less than 13 mm, with spinal cord compression [1,10].

In this case, A–OI measured 6 mm and sagittal canal diameter measured 9 mm.
Our case represents a radio clinical discordance; the patient likely had pain neck without any neurological deficits; however, MRI showed cervical myelopathy involving C1–C2 instability.

For asymptomatic cases, conservative management with clinical and radiological surveillance may be the option [1,3,6]. Patients with neurological symptoms and C1–C2 instability are generally treated with posterior fixation and fusion [1,7]. Medical imaging can be used to show favorable anatomy, such as the length and direction of the pedicles for surgical intervention, help to determine the trajectory for screw placement, and improve the accuracy of screw placement [1,6,7,8].

Conclusion

Os odontoideum is defined by a smooth, well corticated osicle located superior to and separated from the dens. Treatment depends on the existence of radiological evidence of C1–C2 instability with spinal compression or neurological deficits, in this case, posterior fixation and fusion are recommended. A multimodality imaging approach including X-ray, CT, and MRI had an important role in the management of os odontoideum, from diagnosis to therapy.

Patient consent

Informed consent was obtained from all individual participants included in the study.

REFERENCES

[1] Wang Q, Dong S, Wang F. Os odontoideum: diagnosis and role of imaging. SurgRadiolAnat 2020;42:155–60. doi:10.1007/s00276-019-02351-3.

[2] Pape S, Laforge R, Latrobe C, Leroux J, Roussignol X, Ould-Slimane M. Cervical myelopathy involving os odontoideum and retro-odontoid cyst treated with Harms C1–C2 arthrodesis. Orthopaed Traumatol Surg Res 2016;102(6).

[3] Laghmari Mehdi, et al. Os odontoideum: à propos d’un cas et revue de la littérature. Pan African Med J 2012;13:44. doi:10.11604/pamj.2012.13.44.1930.

[4] Tang X, Tan M, Yi P, et al. Atlantoaxial dislocation and os odontoideum in two identical twins: perspectives on etiology. Eur Spine J 2018;27:259–63. doi:10.1007/s00586-017-5116-5.

[5] OsOdontoideum. Neurosurgery 2002;50(Suppl. 3):S148–55. doi:10.1097/01.brs.0000214935.70868.1c.

[6] Hedequist Daniel J, Andrew Z. OsOdontoideum in children. J Am Acad Orthop Surg 2020;28(3):e100–7. doi:10.5435/JAAOS-D-18-00637.

[7] Wu X, Wood KB, Gao Y, Li S, Wang J, Ge T. Surgical strategies for the treatment of osodontoideum with atlantoaxial dislocation. J Neuroureg Spine SPI 2018;28(2):131–9. Retrieved November 7, 2021, from https://thejns.org/spine/view/journals/j-neurosurg-spine/28/2/article-p131.xml.

[8] Rozzelle Curtis J, Aarabi Bizhan, Dhall Sanjay S, Gelb Daniel E, Hurlbert RJ, et al. OsOdontoideum. Neurosurgery 2013;72(Suppl. 3):159–69. doi:10.1227/NEU.0b013e318276ee69.

[9] Johal J, Loukas M, Fishah C, et al. Bergmann’s osicle (ossiculumterminalis): a brief review and differentiation from other findings of the odontoid process. Childs NervSyst 2016;32:1603–6. doi:10.1007/s00381-016-3199-7.

[10] Spierings EL, Braakman R. The management of osdontoideum. Analysis of 37 cases. J Bone JtSurg Br 1982;64:422–8. doi:10.1097/01.brs.0000214935.70868.1c.

[11] Nguyen JC, Pollock AN. Osodontoideum. PediatrEmerg Care 2015;31:225–7. doi:10.1097/PEC.0000000000000411.

[12] Dias RP, Buchanan CR, Thomas N, et al. Osodontoideum in Wolcott-Rallison syndrome: a case series of 4 patients. Orhanpet J Rare Dis 2016;1:1–5. doi:10.1186/s1302 3-016-0397-z.

[13] Giacomin Ci. Sull’esistenza dell’ “osdontoideum” nell’uomo. Gior Accad MedTorino 1886;49:24–5.

[14] Zhang Z, Wang H, Chao L. acute traumatic cervical cord injury in pediatric patients with osodontoideum: a series of 6 patients. World Neurosurg 2014;83 1180.e1–1180.e6. doi:10.1016/j.wneu.2014.12.036.