Case Report

Management of a complicated childhood Os odontoideum: A new incidence in Saudi Arabia

Fahd Nasser AlQahtani

Department of Radiology, Faculty of Medicine, Al-Baha University, Al-Baha, Kingdom of Saudi Arabia

Correspondence to: Fahd Nasser AlQahtani, E-mail: fnqatt@gmail.com

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ABSTRACT

Os odontoideum is defined as an odontoid ossicle process with smooth circumferential cortical margins that have no osseous continuity with the body of C2. The origins of Os odontoideum have much debate for both acquired and congenital causes. The incidence of Os odontoideum is unclear and difficult to investigate because symptoms not always occur. 11-year-old girl patient was presented with abnormal gait, abnormal speech, weakness of all limbs. All of these findings appeared for 3 years, and the presenting case was quite well before that time. The case was mentally normal. Physical examination revealed features of upper motor neuron deficit in the form of spasticity, hyperreflexia, and clonus. These signs were more marked in lower than the upper limbs. Vital signs were within normal limits. Palpation revealed midline tenderness all over the spine most marked down C1 to C2 with no step-offs renowned with intact cranial nerves. No evidence of sensory or motor deficits was noticed on the neurologic examination. Otherwise, no abnormalities were detected. Radiographic workup was done in the form of plain X-ray and magnetic resonance image (MRI). The computed tomography revealed Os odontoideum, as well as prominent ventricles. MRI revealed an evidence of the craniovertebral junction anomaly with most likely Os odontoideum as well as the backward displacement of the C2 vertebral body. Despite Os odontoideum is an infrequent lesion, its importance comes from associated injuries related to the cervical spine and vertebral artery injuries and should be considered in all age groups, more frequently in childhood, especially when the presenting symptoms are of upper cervical spine compression and its sequelae. Early diagnosis and proper management will prevent such complications to occur.

KEY WORDS: Os Odontoideum; Flexion/Extension; Cervical Spine; Congenital Anomalies; Myelomalacia

INTRODUCTION

Os odontoideum is considered as an anatomic variant of the odontoid process of C2 and must be put in differential diagnosis with the type 2 odontoid fracture and from the persistent ossiculum terminale. Despite it was formerly considered to be a congenital defect owing to a failure of unification between the centers of ossification of the dens with the body of C2, it may essentially represent as a hidden fracture along the dens growth plate at age ≤6 years. Associated instability and chronic symptoms are often developed.[1]

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CASE REPORT

A 11-year-old girl was presented with abnormal gait, abnormal speech, and weakness of all limbs. All of these findings appeared for 3 years, and the presented case was quite well before that time. The case was mentally normal. Physical examination revealed that upper motor neuron findings were identified in the form of spasticity, hyperreflexia, and clonus. Hyperreflexia and spasticity were present all over the upper

and lower limbs but more marked in lower than upper limbs. Vital signs were all within normal limits. Palpation revealed midline tenderness all over the spine most marked down C1 to C2 with no step-offs renowned with intact cranial nerves. No evidence of sensory or motor deficits was noticed on her neurologic examination. Otherwise, no abnormalities were detected.

A non-contrast cervical spine computed tomography (CT) scan was obtained for additional evaluation of the spinal tenderness. Coronal and sagittal reformat CT of the upper cervical spine shows Os odontoideum and backward dislodgment of the C2 vertebral body [Figure 1a-c].

One of the differential diagnoses which are an acute subluxation with associated cord injury cannot be excluded at all on CT alone, and a magnetic resonance image (MRI) of the cervical spine was highly recommended. MRI examination of the brain with I/V contrast using different pulse sequences in different planes revealed:

> *Evidence of the craniovertebral junction anomaly with most likely Os odontoideum as well as backward displacement of the C2 vertebral body which is seen significantly compressing the cervicomedullary junction in addition to the upper cervical cord with evidence of focal myelomalacia; the cervicomedullary junction and the upper cervical cord appear significantly compressed with intramedullary bright T2 signal intensity,and there was no evidence of post contrast enhancement. Mild supra- and infratentorial changes are noted possibly as a consequence of severe compromise of the cervicomedullary junction. No mass lesion or otherwise abnormal signal intensity or pathologic enhancement of the brain parenchyma, no restricted diffusion. Cavum septum pellucidum and cavum vergae are noted as a normal variant. Rather prominent cortical sulci yet with no extra-axial collection. Normal sellar region as well as the cerebellum and brain stem. The radiologist concluded that craniovertebral junction anomaly with most likely Os odontoideum and backward displacement of the C2 vertebral body with significant foramen magnum compromise as well as cervicomedullary junction and upper cervical cord focal myelomalacia with mild supra and infratentorial hydrocephalic changes like due to the impedance of cerebrospinal fluid drainage at the craniovertebral junction as described [Figure 2a and b].*

The patient was reevaluated again by neurosurgery consultant, and several flexion-extension radiographs were done and revealed instability which is more marked with flexion, between the articulations of C1 and the C2 Os odontoideum [Figure 3a and b]. Surgical fixation for atlantoaxial instability was done, and the patient has now been under follow-up.

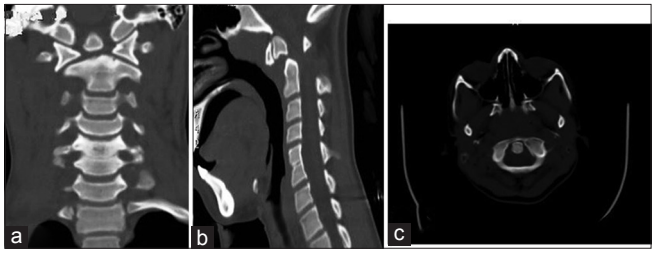

Figure 1: (a and b) Coronal and sagittal reformat computed tomography (CT) of upper cervical spine showing Os odontoideum and backward displacement of the C2 vertebral body. (C) Bone window axial view of C1 and C2 CT scan showing narrowing of the space between C1 and C2

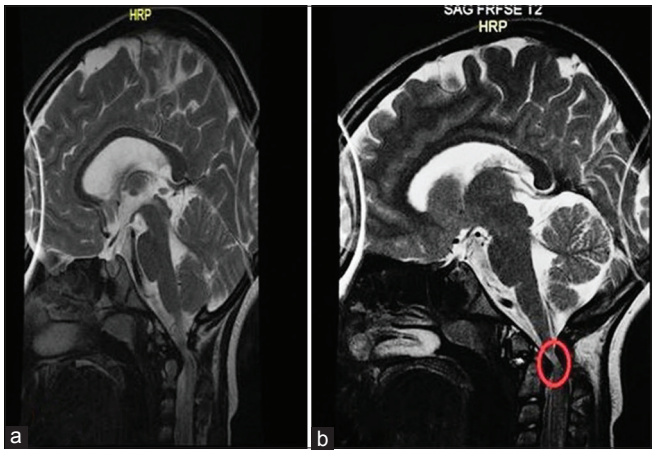

Figure 2: (a) Sagittal T2 magnetic resonance image (MRI) showing evidence of craniovertebral junction anomaly. (b) Sagittal T2 MRI showing significantly compressing the cervicomedullary junction and the upper cervical cord with evidence of focal myelomalacia (circle)

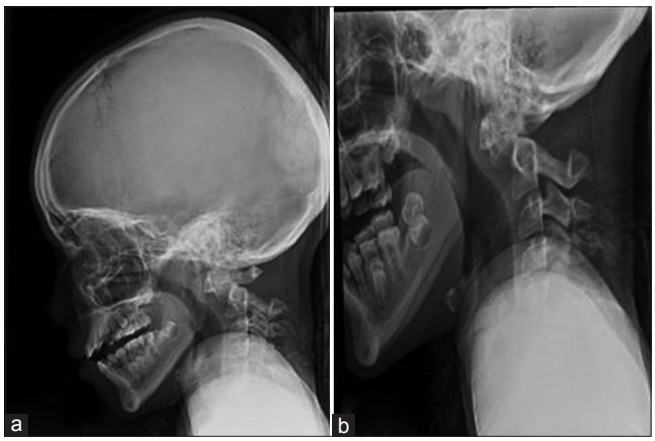

Figure 3: (a and b) Flexion and extension of the lateral upper cervical spine revealed instability which is more marked with flexion, between the articulation of C1 and C2 Os odontoideum

DISCUSSION

The historical background of Os odontoideum revealed that the first case has been illustrated as disentanglement of the odontoid process in 1863. In 1886, the term Os odontoideum for this situation was applied.[2] Os odontoideum is infrequent,

but the accurate occurrence is unknown. It may be presented and diagnosed at any age. Despite unknown etiology, it is recorded in association with many syndromes as Down and Morquio[3,4] and multiple epiphyseal dysplasias. The symptoms may include neck pain, torticollis and headache. Symptoms related to neural and vascular compression may be present.[5]

The effect of compression on the spinal cord or even decrease blood flow may lead to hypermobility of C1 and C2 in a patient who complains from cervical instability and appearance of neurological disorders.[6,7] The degree of neurologic disorders varies from an episode of widespread paresis to myelopathy and may be ended by complete spinal cord affection.[8] Ataxia and weakness frequently dominate over sensory changes. Development of infarction in both the cerebellum and brainstem associated with convulsions may occur later.[9,10]

Primarily, Os odontoideum was recognized as a congenital collapse of the union of the dens to the rest of the axis.[11,12] Nowadays, it appears that the combination of den' secondary ossification center with the odontoid base has not occurred.[13]

The plain radiography plays the first rule for the evaluation of Os odontoideum which includes flexion-extension lateral plain and open mouth anterior-posterior films. Regarding these plain views, the presence of ossicle of a smooth regular cortex separated from axis base by a wide fissure is a characteristic feature of Os odontoideum.[14-16] A 1-mm, sagittal CT scan permits a more inclusive demonstration of the atlantoaxial articulation.

MRI may distinguish any pathological alteration within the cord as myelomalacia if there is an amplified signal on T2-weighted MRI sequences in the substance of the cord.[17-20] An altered T1 signal may denote the presence of necrosis, hemorrhage within the cord, and characteristically expect a severe neurological outcome. A more than one imaging modality has been suggested to determine the nature and degree of abnormal movement in cases of Os odontoideum; for example, cineradiography, MRI, and dynamic flexion-extension have been powerfully recommended due to their capability to identify the relation of the Os odontoideum to the adjacent bony rudiments.[21-23]

There are two considerations for the treatment and management of a patient with Os odontoideum, of these, observation through clinical and radiological assessment and stabilization by surgical procedure. Stabilization is recommended in spinal instability, neurologic involvement, and intractable pain. Most authors are in agreement with some parameters if present "in flexion-extension plain film" the surgery is indicated: Sagittal plane rotational angle >20°, posterior atlanto-dens interval <13 mm, instability index >40%, and C1-C2 translation >5 mm.

The outcomes and prognostic information of Os odontoideum patients are obtained from some case series and reports.[24,25] In a study done by Klimo *et al*. who examined 77 Os odontoideum patient treated by surgical stabilization,[3] they found that fusion was attained at nearly 4.8 months for all patients and marked improvement in neck pain in 90% of cases. In addition, 50% showed improvement in their spasticity and complete resolution of myelopathy in 39% in myelopathic patients.

In the study of 35 Os odontoideum patients done by fielding, 27 were radiographically unstable. 2 months after immobilization, a solid fusion was developed opposite to 3 months in the adult. The eight stable cases were treated nonoperatively and showed an improvement at last setting of follow-up. Regarding symptomatic cases, 66% of patients subjected to the study had mechanical pain only which relieved completely after surgical union.[21]

The treatment with rigid instrumentation in 21 patients with myelopathy revealed improvement in Nurick scale of 2.3 before surgery to 0.7.[26] Zhang *et al*. revealed that patients with atlantoaxial instability are of great risk for acute spinal cord injury with exposure to minor trauma and stated that fixation plus fusion must be carried out to keep away from this injuries.[27]

CONCLUSION

The causes of Os odontoideum are controversial and can go from embryological or traumatic causes to even vascular causes. The incidence of Os odontoideum is uncertain and hard to investigate because symptoms are not always obvious. For accurate diagnosis of Os odontoideum, a correlation of history/clinical examination with the radiological investigations must be done. The flexion-extension lateral plain and open mouth anterior-posterior films play the core for radiograhic diagnosis of Os odontoideum and should be put in the priority of radiographic investigations in such cases.

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