Recent Review Article on Os Odontoideum

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Abstract

Introduction: Os-odontoideum is a rare condition, described radiographically and clinically as a congenital anomaly of the second cervical vertebra (axis), it a smooth, independent ossicle of variable size and shape separated from the base of a shortened odontoid process by an obvious gap, with no osseous connection to the body of C2.

Material and methods: This study reviewed the literature on OO to evaluate its etiology, the clinical presentations, differential diagnosis, imaging modalities and outcomes in the management of asymptomatic and symptomatic cases of os odontoideum. PubMed, EMBASE, Google Scholar and Cochrane key articles were searched.

Discussion: Considering etiology traumatic hypothesis is favoured over congenital hypothesis as per recent literature on OO. Clinical presentation varies from asymptomatic to mild neck pain to severe myelopathy and neurodeficit. Various C1-C2 instrumentation and fusion techniques like wiring, trans articular screw and laminar screws have been described with good success rates.

Conclusion: Os odontoideum is a rare condition with limited existing literature. Considering significant risks involved if conservative management is opted like severe neurodeficit to sudden death on trivial trauma and the recent improvement of imaging tools helping to understand the pathology of the disease, surgery can be indicated even in an incidentally detected os odontoideum. Although for asymptomatic stable patients' individual case-by-case approach can be considered depending on factors such as age, activity level, comorbidities, syndromic association and radiographic findings.

Keywords: Os-odontoideum, Surgical management, Etiology, Atlanto-axial instability, Complications

Introduction

Os-odontoideum was first described by Giacomini in 1886 [1]. Bevan in 1863 and Cunningham in 1886 also described OO on the basis of anatomical and autopsy studies [2]. The word os odontoideum is derived from Latin language meaning os (bone) and odontoideum (tooth-like). It is a rare condition, described radiographically and clinically as a congenital anomaly of the second cervical vertebra (axis), it a smooth, independent ossicle of variable size and shape separated from the base of a shortened odontoid process by an obvious gap, with no osseous connection to the body of C2. It is classified variously depending on anatomy, stability, etiology and symptomatology. It is classified anatomically into 2 types, dystopic and orthotopic, by Fielding et al in 1980 [3, 4]. In dystopic type OO is fused to basion while in orthotopic OO lies in the normal position on the odontoid process and moves with the atlas anterior arch. It is classified on the basis of the extent of excursion of the atlas from the axis on dynamic imaging as stable or unstable. It is classified etiologically as congenital and post-traumatic. OO has been classified as asymptomatic or symptomatic depending on clinical presentation and an increasing risk of cervico-medullary compression [5, 6, 7]. This study tries to review and address the various controversies regarding etiology and management options of os odontoideum.

Materials and Methods

We undertook a review of the literature on OO to evaluate its etiology, the clinical presentations, differential diagnosis, imaging modalities and outcomes in the management of asymptomatic and symptomatic cases of os odontoideum. PubMed, EMBASE, Google Scholar and Cochrane key articles were searched. Key words like 'Os-odontoideum', 'surgical management', 'etiology', 'Atlanto-axial instability' 'complications' were used. Additional articles were

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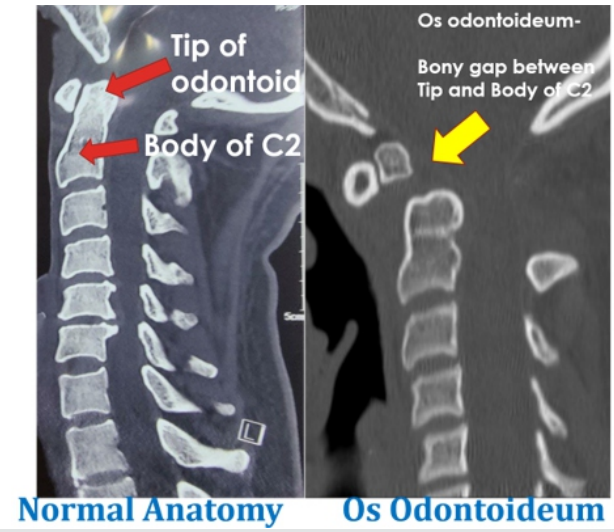

Figure 1: Normal anatomy and Os Odontoideum

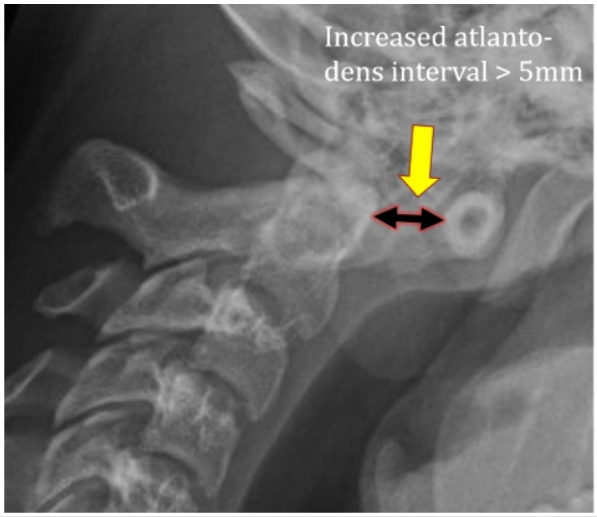

Figure 2: Unstable Os Odontoideum with increased Atlanto-Dens interval >5mm

identified by checking the references manually. Articles were reviewed by two independently reviewers.

Discussion

Pathophysiology:

The etiology of the OO has always remained controversial since its description in 19th century. The aetiologies proposed are congenital or traumatic. Although the current evidence from literature is in more support of traumatic over the congenital hypothesis. The authors supporting of the congenital hypothesis describe OO results from a failure of fusion of the dens with the body of the axis during embryonic development or failure of fusion of the secondary ossification center at the apex with its main part of the dens or the failure of proper caudal migration of the dens during development. The congenital OO etiology is further supported by its presence in the identical twins, siblings and association of OO with many congenital syndromes like Down's syndrome, Morquio's disease, achondroplasia, Klippel-Feil syndrome, Larson syndrome and other associated anomalies at the CVJ with no significant history of trauma. The congenital hypothesis was debated as the neurocentral synchondrosis is located below the level of the superior articulating facet, whereas the gap in OO is frequently located above the plane of the superior articulating facet [7, 8, 9].

Fielding and Griffin in 1980s proposed the proposed the acquired/post traumatic/vascular hypothesis, describing formation of OO due to an unrecognized fracture to the odontoid after trivial fall in childhood with the subsequent contraction of the apical and alar ligaments, the distraction of the fractured fragment, and then reducing or detaching the blood supply and giving rise to the OO [3, 4].

Clinical Presentation

OO is more commonly seen among males and in the second and third decades of life. The clinical presentation in OO patients is highly variable ranging from incidental finding in asymptomatic patients, local symptoms like neck pain, restriction of neck movements, shoulder pain, torticollis, and occipital headaches, also cervical myelopathic symptoms and signs like weakness, paresthesia, ataxia, urinary complains and symptoms related to vertebra-basilar ischemia [10].

Differential Diagnosis of OS

On the basis of clinical presentation differential diagnosis of OO can vary from cervical spondylosis, mechanical neck pain, and degenerative disc disease or atlantoaxial subluxation as seen in rheumatoid arthritis. It could also be confused with acute fracture of dens. Although OO could be ruled out on the history itself by absence of a significant history of fall or trauma and depending on radiologically characteristic like the smooth surface of the ossicle and the below body of C2, and also the presence sclerosis and hypertrophy of the anterior tubercle of the atlas [7, 8].

Imaging and indicators of instability (Figure 1)

OO can be diagnosed on plain radiographs with the anteroposterior, lateral neutral and dynamic views (flexion and extension) and open mouth view. In addition to plain lateral radiographs, CT of the CVJ to understand bony anatomy and abnormalities, CT angiography to study course of vertebral artery, and MRI of the CVJ to look for cord compression and soft tissue compression and anomalies. Recently use of kinematic MRI to diagnose OO was advised by Hughes et al [11]. Recent article by Goyal et al described criteria for atlantoaxial instability.

Criteria for AAI [12, 13]

- Atlanto-dens interval of more than 5 mm (Figure 2)
- Overriding of the anterior arch of the atlas over the odontoid
- Space available for the cord (SAC) of less than 13 mm (Figure 3)
- Violation of the Steel's rule of thirds (one-third cord, one-third odontoid, and one-third safe space) (Figure 4)
- Translation of the tip of the odontoid of more than 4 mm of the basion

Figure 3: Unstable Os Odontoideum with cord signals with reduced available space for the cord[SAC]

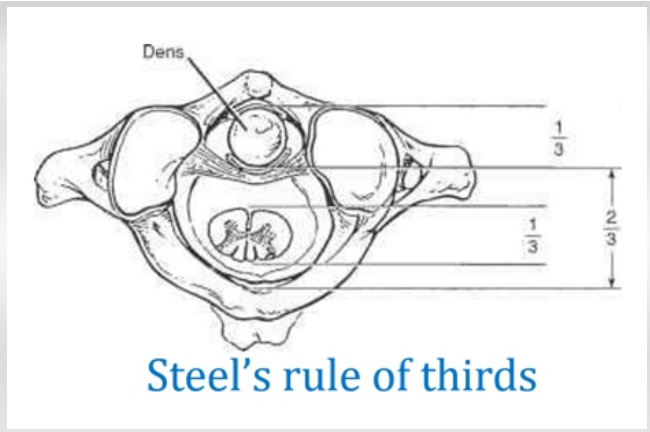

Figure 4: Steel's Rule of thirds

Management

The American Association of Neurological Surgeons and the Congress of Neurological Surgeons (AANS/CNS) provided evidence-based clinical recommendations for the treatment of os odontoideum in 2001 [14, 15].

- No clinical evidence was found supporting the recommendation of operative treatment standards or guidelines for os odontoideum.
- For asymptomatic patients, clinical and radiographic surveillance may be the appropriate management.
- Patients with neurological symptoms/signs and C1–2 instability are generally managed with posterior fixation and fusion.
- Other operative measures may be warranted in cases of irreducible cervicomedullary compression and/or occipitocervical instability.
- The paucity of high quality studies was noted

The evidence based clinical studies recommend surgical management is in symptomatic OO patients (e.g. cervical myelopathy). Although, controversy still exists in the management of asymptomatic patients.

In 1982, Spierings and Braakman [16] noted high mortality in OO patients who were treated with surgical management, so authors suggested conservative treatment in more than 60% of their cases, including among those who were asymptomatic.

In 2000, Dai et al [6]. suggested that five asymptomatic cases of OO which were managed conservatively remained stable at follow-up. In 2008, Klimo et al [8, 9] contradictorily believed that stressful events can easily cause spinal cord injury. Klimo et al recommended surgical intervention in OO, regardless of the clinical presentation. Literature has suggested in asymptomatic stable OO patients even a trivial trauma could lead to significant neurological deficit or death [17, 18, 19].

Surgical techniques:

Various surgical techniques has been advised like

- Occiput-cervical fusion- rarely used as it reduces range of motion
- C1 – C2 wiring
- C1 C2 trans articular screw
- C1 lateral mass- C2 pars screw
- C1 lateral mass- C2 lamina screw
- C1 – C2 wiring

Gallie et al [20, 21] in 1939 described the C1-2 wiring technique elaborately. In this technique wires are passed under the lamina of C1 arch and hooked to spinous process of C2. The tricortical iliac crest bone graft is compressed between C1-C2 to achieve fusion. Recently in 2002, Brockmeyer et al used cables instead of wires for C1-C2 stabilization. Inability to correct rotatory forces, non-union and gradual loss of reduction are major drawbacks of this technique. Recently few authors described use of hooks at C1 posterior arch to improve stability and strength of construct.

- C1 C2 trans articular screw instrumentation

Magerl et al [22] in 1987 demonstrated C1-C2 trans articular screw technique. It provides biomechanically strong and stable construct in comparison with C1-C2 wiring. Although long learning curve, technically demanding precision and accuracy, risk of neurovascular injuries like high riding vertebral artery injury, this technique showed promising results over the years [23].

- C1 lateral mass and C2 pars screw instrumentation

Goel and Laheri [24] in 1994 described C1 lateral mass and C2 pars screw with plate instrumentation with good results. Harms et al [25] in 2001 modified this technique with use of polyaxial screws and rod system. This technique showed relatively easy learning curve and good clinical and radiological outcomes. It is one of the safest, reproducible, most accepted and widely used procedure in the current literature.

- C1 lateral mass- C2 lamina screw

In certain cases, there is presence of high riding vertebral artery increasing risks of its injury. In such scenario use of C2 laminar screws is well described with good outcome.

Limitations

This study has few limitations. As this study is not a systematic review, the evidence about each debatable issue is inherently

insufficient. However, this study will provide the current concept about etiology and current concepts in management of os odontoideum. Also this study could not suggest specific guidelines because of the paucity of highly qualified, relevant studies.

Conclusion

Os odontoideum is a rare condition with limited existing literature. The etiology is debatable, but recent literature favours acquired/post traumatic/vascular hypothesis. OO could present with wide manifestations of symptoms, precise clinical and radiological diagnosis could help in OO treatment and prevent likely devastating complications. Considering significant risks involved if conservative management is opted like severe neurodeficit to sudden death on trivial trauma and the improvement of imaging tools helping to understand the pathology of the disease, morphology of the nearby vital structure along with the relative safety of the current surgical procedures, surgery can be indicated even in an incidentally detected os odontoideum. Although for asymptomatic stable patients' individual case-by-case approach can be considered depending on factors such as age, activity level, comorbidities, syndromic association and radiographic findings.

Declaration of patient consent : The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his/her consent for his/her images and other clinical information to be reported in the Journal. The patient understands that his/her name and initials will not be published, and due efforts will be made to conceal his/her identity, but anonymity cannot be guaranteed.

Conflict of Interest: None; **Source of Support:** None

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