Incidental os odontoideum: current management strategies

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Os odontoideum was first described in the late 1880s and still remains a mystery in many respects. The genesis of os odontoideum is thought to be prior bone injury to the odontoid, but a developmental cause probably also exists. The spectrum of presentation is striking and ranges from patients who are asymptomatic or have only neck pain to those with acute quadriplegia, chronic myelopathy, or even sudden death. By definition, the presence of an os odontoideum renders the C1–2 region unstable, even under physiological loads in some patients. The consequences of this instability are exemplified by numerous cases in the literature in which a patient with os odontoideum has suffered a spinal cord injury after minor trauma. Although there is little debate that patients with os odontoideum and clinical or radiographic evidence of neurological injury or spinal cord compression should undergo surgery, the dispute continues regarding the care of asymptomatic patients whose os odontoideum is discovered incidentally. The authors' clinical experience leads them to believe that certain subgroups of asymptomatic patients should be strongly considered for surgery. These subgroups include those who are young, have anatomy favorable for surgical intervention, and show evidence of instability on flexion-extension cervical spine x-rays. This recommendation is bolstered by the fact that surgical fusion of the C1–2 region has evolved greatly and can now be done with considerable safety and success. When atlantoaxial instrumentation is used, fusion rates for os odontoideum should approach 100%.

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Key Words • incidental os odontoideum • atlantoaxial instability • atlantoaxial fusion • screw fixation

OS ODONTOIDEUM is an anatomical abnormality in which the tip of the odontoid process lacks continuity with the body of C-2. It appears as a smooth-margined, apical osseous segment separated from the base of the odontoid process by an obvious gap. Os odontoideum may be discovered as part of a workup for neck pain and/or neurological symptoms, but it is also often found incidentally. Most spine surgeons agree that patients with signs or symptoms of neurological dysfunction should undergo stabilization, but the role for surgical stabilization in asymptomatic patients and those with neck pain alone remains controversial.

In an article in the “Guidelines for the Management of Acute Cervical Spine and Spinal Cord Injuries” supplement published in 2002 jointly by the American Association of Neurological Surgeons and the Congress of Neurological Surgeons, the authors reviewed the available literature on os odontoideum and found no Class I or Class II data to generate treatment standards or guidelines.1 The following management option was given for patients with incidental os odontoideum based on the available Class III data: “Patients with os odontoideum, either with or without C1–2 instability, who have neither symptoms nor neurological signs may be managed with clinical and radiographic surveillance.” Even so, the authors acknowledged that patients with C1–2 instability are at risk for future spinal cord damage and that surgical stabilization and fusion of C1–2 is “meritorious.” In fact, the literature regarding the management of os odontoideum remains limited to Class III data. Here we present the case of a patient with an incidentally discovered os odontoideum and review the embryological aspects and relevant upper cervical spinal anatomy and literature. We conclude by presenting a management strategy that we believe is rational given our experience, along with data from the available literature.

Illustrative Case

Clinical Presentation

This 14-year-old girl was admitted to the hospital after having possible seizures, intermittent headaches, and dizziness. She underwent MR imaging of her brain, which showed a mild Chiari I malformation with approximately 6 mm of tonsillar herniation, but also a dysplastic odontoid. On her cervical spine MR images there was no spinal cord signal change or syringohydromyelia, and the VA anatomy was normal (Fig. 1A). A CT scan confirmed the presence of an os odontoideum. The os fragment was

Abbreviation used in this paper: VA = vertebral artery.
fused to the posterior aspect of C-1, consistent with a dystopic os odontoideum (Fig. 1B and C). There was no abnormal atlantoaxial motion on flexion, but with extension, the patient had posterior subluxation of 9–10 mm (posterior instability, Fig. 1D and E). We concluded that although her os odontoideum was discovered incidentally, surgical fusion was indicated because she showed evidence of instability.

**Treatment and Postoperative Follow-Up**

After approximately 1 month in a collar, the patient underwent a posterior C1–2 fusion performed using C-1 lateral mass screws coupled to C-2 pars interarticularis screws (Fig. 1F and G). Posterior iliac crest autograft was fashioned and secured between C-1 and C-2. We elected not to perform a posterior fossa decompression for her mild Chiari malformation because we believed it was asymptomatic. A CT scan obtained 4 months after surgery showed a solid fusion.

**Embryological Development and Pathogenesis**

During normal embryological development, the odontoid process is derived from somites 4 (fourth occipital sclerotome, “C0”); 5 (first cervical sclerotome, “C1”); and 6 (second cervical sclerotome, “C2”). The body of the odontoid begins as part of the centrum of the first cervical vertebra, but becomes separated from the atlas to fuse with the remainder of the axis between the 6th and 7th weeks of gestation. The apex of the odontoid is derived from the fourth occipital sclerotome, commonly known as the proatlas. At birth, there is an epiphyseal growth plate called the neurocentral synchondrosis that separates the body of the axis (C-2) from the dens (C-1). This synchondrosis is not located at the base of the dens, but is well below the level of the superior articulating facet of the axis and into its body. The blood supply to the odontoid is also unique. The VA provides both an anterior and a posterior ascending artery, but the neurocentral synchondrosis prevents further rostral supply by these arteries. Therefore, the odontoid is supplied by a descending supply superiorly called the apical arcade.

The cause of os odontoideum remains the subject of debate. It was once believed to be a congenital lesion caused by a failure of fusion between the first and second sclerotomes (that is, across the neurocentral synchondrosis). A familial form of the condition and os odontoideum found in identical twins have been reported in patients with no preceding history of trauma, supporting the congenital hypothesis; however, this theory has been questioned because, as previously stated, the neurocentral synchondrosis is located below the level of the superior articulating facet, whereas the gap in os odontoideum is frequently located above the plane of the superior articulating facet. Nonetheless, some authors still believe that a congenital mechanism is plausible. Currarino proposed that os odontoideum represents an abnormal complete or partial embryonic segmentation of the midportion of the odontoid between chondrification segments X and Y.

Most physicians now believe that the ossicle re-
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represents a posttraumatic phenomenon resulting from a nonhealed odontoid synchondrosis fracture. In essence, the odontoid fragment fails to remodel and unite with the body of C-2. This scenario is supported by the observation that many patients with os odontoideum have a history of remote trauma, particularly in early childhood. The most commonly cited theory was proposed by Fielding et al., who suggested that with a fracture or disruption through the neurocentral synchondrosis, the alar ligaments that attach to the apex of the odontoid may gradually distract the fragment away from the base. The apex and base of the odontoid continue to have adequate perfusion, but the midportion suffers from lack of blood supply and thus contributes to poor healing.

The “traumatic cause” hypothesis is supported by case reports of patients with a previously documented intact C-2 who later were found to have os odontoideum after remote trauma. Schuler et al. reported on a 2-year-old patient who fell out of her crib and complained of neck pain; her initial cervical x-ray was normal. After continued neck pain, repeat cervical x-rays were obtained 13 months after her injury, which demonstrated os odontoideum with atlantoaxial instability. A recent case report by Zygourakis et al. supports the delayed formation of an os odontoideum through a combination of trauma and vascular compromise. Their patient was a 2-year-old girl who sustained a C1–2 ligamentous injury but no bone injury. Four years later an os odontoideum was found, and 10 years after trauma the child had developed mild atlantoaxial hypermobility that continued to be managed nonoperatively. It is likely that os odontoideum most commonly arises as a result of remote trauma, but some cases are also probably due to faulty embryogenesis. Os odontoideum needs to be differentiated from a persistent ossiculum terminale. The latter represents congenital nonunion of the odontoid body from a terminal ossicle located above the transverse atlantal ligament. Thus, atlantoaxial instability is usually not associated with an ossiculum terminale.

Presentation and Imaging Features

Rowland et al. categorized symptoms of os odontoideum into 4 groups: Group 1, local symptoms only (for example, neck pain, headaches) and no signs of myelopathy; Group 2, posttraumatic transient myelopathy; Group 3, persistent myelopathy; and Group 4, cerebral symptoms suggestive of posterior circulation ischemia. Patients with symptoms in Group 2 classically have transient complete tetraplegia immediately after a traumatic event, which is often minor in nature. Those with symptoms in Group 3 have chronic, often progressive myelopathy because of repeated trauma to the spinal cord as a result of excessive motion of the ring of C-1 and the ossicle. Patients with posterior circulation symptoms are quite rare, but have been reported. Asymptomatic patients, by definition, have incidentally discovered os odontoideum. This group comprised 15% of patients in our recent series.

Os odontoideum is a rare radiographic diagnosis. Two types of os odontoideum have been defined based on the position of the dens tip: orthotopic and dystopic. In the orthotopic type, the dens is in an anatomical position. In dystopic os odontoideum, the dens tip is in any other position. Most commonly, the fragment is located near the foramen magnum, where it may fuse with the clivus, or it may be fixed to the anterior ring of the atlas. Plain cervical x-rays are often sufficient to suggest the diagnosis of an os odontoideum, but a high-resolution multiplanar CT scan is necessary to provide the detailed anatomy of the os odontoideum and any associated cervical or skull base anomalies for surgical planning. Dynamic flexion and extension lateral radiographs may be obtained to determine whether there is any abnormal atlantoaxial motion and, if so, in which direction is the subluxation: anterior, posterior, or both.

In our series, a subset of 60 patients with os odontoideum who underwent dynamic imaging had predominantly anterior instability (70%), with almost equal posterior instability (10%) or combined anterior/posterior instability (13%). No motion was seen in 7% of the patients. Although many spine surgeons believe that a patient with normal mentation can undergo volitional flexion and extension imaging without risking neurological injury, particularly in those who are without symptoms, these studies should be performed with caution in those who have had recent neurological injury or chronic myelopathy. An MR imaging study should also be done to evaluate for any intrinsic spinal cord signal change at the level of os odontoideum that would indicate prior or continuing spinal cord trauma. Rarely, synovial cysts may develop as a result of the abnormal motion, and these are best evaluated using MR imaging. Careful evaluation of the regional vascular anatomy is necessary because anomalies have been reported in association with an os odontoideum, particularly the presence of an anomalous VA or persistent fetal anterior-posterior circulation connection.

Biomechanics and Stability

The stability of the C1–2 joint is determined by the integrity of the odontoid process, the transverse ligament, and to a lesser extent, the alar ligaments. Thus, the biomechanical principles of C1–2 stability are inherently violated with os odontoideum. With an incompetent odontoid process, the spinal cord may be compressed posteriorly during flexion by the posterior ring of C-1 or on extension by posterior translation of the os itself (Fig. 2). In the absence of an intact odontoid process, we believe that the existing stabilizing structures—the thinner vertical portion of the cruciate ligament (an extension of the posterior longitudinal ligament), the thin anterior longitudinal ligament, and the lax capsular ligaments—are insufficient to provide the necessary spinal stability and protection of the spinal cord. An os odontoideum is biomechanically analogous and in some cases difficult to distinguish from a chronic nonunited Type 2 odontoid fracture. Although it is well accepted among most spine surgeons that Type 2 fractures, whether acute or chronic but not healed, are unstable and require treatment, there continues to be a significant difference of opinion among surgeons regarding the treatment of an incidental os odontoideum.
Descriptions of the natural history of os odontoideum are very limited and dated. Several reports have presented evidence of patients who were treated conservatively without further incident. Fielding et al. reported a series of 35 patients, 8 of whom had no radiographic evidence of C1–2 instability and underwent conservative management. Each of them remained without symptoms at follow-up evaluation. Similarly, all 5 of the asymptomatic patients with os odontoideum in whom the condition was managed without surgical intervention by Dai et al. remained stable at follow-up. Spierings and Braakman analyzed 37 patients with os odontoideum, in 20 of whom the condition was managed conservatively. The authors found that patients with a minimal sagittal diameter (defined as the distance between the posterior border of the body of C-2 and the posterior atlantal arch on flexion) of < 13 mm had the greatest risk of developing permanent or progressive cord signs. This suggests that incidental os odontoideum may be managed without surgical intervention unless the minimum sagittal diameter is < 13 mm or there is progressive neurological decline. Although these studies suggest that in some patients with incidental os odontoideum the condition can be safely managed conservatively, in sum the studies provide less than definitive evidence as to the natural history and risks associated with untreated os odontoideum.

Sudden death and significant neurological morbidity with minor trauma have both been reported as the initial presentation of patients with a previously undiagnosed os odontoideum. Choit et al. reported on 2 children who each had an os odontoideum that had not been appreciated on initial imaging and that caused subsequent morbidity. One of the children sustained a severe high spinal cord injury after a minor accident. We previously presented 3 case examples of patients in whom an os odontoideum was initially diagnosed but who never received treatment and who subsequently developed a spinal cord injury. More recently, Zhang et al. reported on 10 patients with atlantoaxial instability from os odontoideum, including 3 who were asymptomatic, who later suffered a spinal cord injury. The mechanism was a fall in 6 patients, a minor motor vehicle accident in 3, and assault in 1. Based on this experience, the authors advocated fusion and instrumentation for all patients with radiographically unstable os odontoideum, whether they were symptomatic or not.

White and Panjabi defined clinical instability as “the loss of the ability of the spine under physiological loads to maintain its pattern of displacement so that there is no initial or additional neurological deficit, no major deformity, and no incapacitating pain.” For some patients with os odontoideum, everyday physiological loads are well tolerated and supported by the accessory structures described previously. Given the numerous reports of patients suffering neurological injury from minor trauma, however, we believe that the ability to tolerate physiological loads or minor trauma cannot be assumed, because the potential loss of that gamble could be catastrophic.

For some patients, particularly young children and teenagers, it is difficult or even impossible to limit day-to-day activity to “physiological” loading, without severely restricting their activity and thus negatively impacting the quality of their life. This includes the normal day-to-day episodes of ground-level falls in toddlers or, for example, falls from playground equipment in older children. For teenagers, giving up contact or collision sports may be relatively easy, but the pushing and shoving that goes on in school hallways, as another example, may not be so easy to avoid. Conversely, it is relatively easy for most young adults with os odontoideum to modify their activities to reduce the risk of spinal cord injury. Their greatest risk obviously lies in the possibility of a motor vehicle accident, which is a risk shared by almost everyone.

**Fig. 2.** Diagrams depicting an os odontoideum in neutral (A), flexed (B), and extended (C) positions. With an os odontoideum (A), abnormal motion may occur anteriorly, posteriorly, or both. Flexion (B) can cause translation of the C-1 ring and ossicle complex to the point that it may impinge on the dorsal aspect of the cord. On extension (C), the anterior ring with the ossicle can strike the ventral aspect of the cord. Reproduced with permission from Klimo Jr, P, Kan P, Rao G, et al. Os odontoideum: presentation, diagnosis, and treatment in a series of 78 patients. Clinical article. J Neurosurg Spine 9:332–342, 2008.
Incidental os odontoideum is why we choose to use the patient’s age as a surgical selection criterion for an incidentally discovered os odontoideum; we believe that young patients are more likely to need protection than older ones.

Management of Incidental Os Odontoideum

We believe that all patients with neurological symptoms or significant neck pain due to os odontoideum should undergo surgical stabilization. Patients with incidentally discovered os odontoideum should be considered for surgery on a case-by-case basis. Those patients who show evidence of radiographic instability at the atlantoaxial level, who are relatively young (<20 years old), and who have bone anatomy favorable for screw fixation should be strongly considered for surgery. Asymptomatic patients who do not meet the criteria given above may be monitored with serial radiographs and clinical evaluations to detect whether instability or significant symptoms are developing. Arvin et al.3 presented similar recommendations, arguing that patients with a stable os odontoideum without evidence of spinal cord compression should be evaluated annually with dynamic plain x-rays, repeat MR imaging in 5 years, and avoidance of all contact sports.

Surgery

Excellent clinical results and high rates of fusion (>90%) for both adults and children who have undergone instrumentation of the C1–2 level have been demonstrated in numerous series.12,14,15,22,28,35 Surgical techniques and fusion technology have evolved tremendously over the last several decades. Screw fixation techniques have largely replaced the stand-alone graft/wiring procedures such as the Gallie and Brooks techniques of the past, leading to excellent results.13,16 Placement of transarticular C1–2 screws was the main method of screw fixation of the C1–2 joint, but this technique involved a risk of injuring the VA.13 The surgeon now has available other types of screws, including C-1 lateral mass screws, C-2 pedicle or pars screws, C-1 hooks, and C-2 laminar screws, to minimize the chance of injuring the VA. Consequently, the use of transarticular screws has become less popular. Hardware can now be placed with minimal risk of neurological or vascular injury. Although harvested autograft, typically from the patient’s posterior iliac crest, is still the main grafting substrate, there is now evidence that allograft may work just as well, thus eliminating donor site-related issues, particularly pain.16 Although there is a loss of approximately 40%–50% of rotation with fusion of C1–2, we have found that older adults seem to adapt well to this loss of motion. Younger adults and children often have little, if any, limitation to motion after atlantoaxial fusion, because other spinal levels assume more rotatory function. It is our opinion that the loss of rotation after a C1–2 fusion seems to be a small price to pay for securing spinal stability and avoiding potentially catastrophic cord compression.

Conclusions

The incomplete odontoid process found in os odontoideum results in weakness of the C1–2 joint. It is potentially dangerous to assume that the remaining supporting structures are strong enough to sustain physiological loads and forces from even minor trauma. Patients with an incidentally discovered os odontoideum should be considered carefully for surgery, taking into account their age, activity level, and radiographic findings, including evidence of atlantoaxial instability and anatomy favorable for surgical instrumentation. Surgery can be done safely, with minimal risk of complications, and with an excellent chance of achieving optimal clinical results.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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