

Os Odontoideum: A Case Report

Os Odontoideum: Olgu Sunumu

ABSTRACT

Several congenital anomalies of the odontoid process have been described in the literature. Malformations of the dens include aplasia, hypoplasia, duplication, condylicus tertius, os terminale (os avis) and os odontoideum. Os odontoideum is the most common anomaly of the odontoid. However, there is insufficient evidence to support treatment standards and guidelines for patients with os odontoideum in the literature. In this article a patient with os odontoideum who underwent two separate operations due to spinal cord compression during cervical motion is presented and the surgical treatment modalities are discussed.

KEY WORDS: Craniovertebral junction anomaly, occipitocervical instability, os odontoideum

ÖZ

Literatürde odontoid proçesin bir çok farklı konjenital anomalisi tarif edilmiştir. Dens malformasyonları aplazi, hipoplazi, duplikasyon, kondilikus tertius, os terminale (os avis) ve os odontoideumu içerir. Os odontoideum odontoidin en sık görülen anomalisidir. Bununla birlikte, standart bir tedavi yöntemi ya da tedavide izlenecek yolu önermek için yeterli veri yoktur. Bu makalede, boyun hareketleri esnasında omurilik basısına neden olan os odontoideum nedeniyle iki kez opere edilen bir hasta sunularak, cerrahi tedavi yöntemleri ilişkili literatür eşliğinde tartışılmıştır.

ANAHTAR SÖZCÜKLER: Kraniovertebral bileşke anomalisi, oksipitoservikal instabilite, os odontoideum

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INTRODUCTION

The craniovertebral junction is one of the most common sites for malformations (22). Several congenital anomalies of the odontoid process have been described in the literature (9, 10, 13, 16). Malformations of the dens include aplasia, hypoplasia, duplication, condylicus tertius, os terminale (os avis), and os odontoideum. Os odontoideum is the most common anomaly of the odontoid (2). An os odontoideum is defined as a radiolucent oval or round ossicle with a smooth dense border of bone separate from a rudimentary odontoid process. If the ossicle is located where the odontoid tip would normally be, it is said to be orthotopic; if the ossicle is located near the base of the occiput in the region of the foramen magnum, it is termed dystopic (23). Os odontoideum may mimic a Type 1 or Type 2 odontoid fracture. The etiology of os odontoideum has been variably attributed to an embryologic, traumatic or vascular basis (6, 5, 8, 15). The atlas and the odontoid process originate from the first cervical sclerotome, whereas the body, lateral masses, and posterior arch of axis arise entirely from the second cervical sclerotome (22). The odontoid process is now thought to originate as an addition to, or a projection from, the anterior part of atlas. It separates between the sixth and the seventh weeks of gestation and then moves caudally to join the body of axis (12, 17). Although the etiology of os odontoideum remains debated in the literature, it does not play an important role in its diagnoses or subsequent management (1). There is insufficient evidence to support treatment standards and guidelines for patients with os odontoideum in the literature (11). In this article a patient with os odontoideum who underwent two separate operations due to spinal cord compression during cervical motion is presented and the surgical treatment modalities are discussed.

CASE REPORT

A 19-year-old female patient was suffering from pain in her neck and shoulders. The pain had started 3 years ago, and there had been a remarkable increase in the last 6 months. Her neurological examination revealed loss of muscle power in both arms (+4/5), hyperactivity of all deep tendon reflexes and bilateral positive Babinski sign. There was no pathology in direct cervical x-ray films. Although there was no sign of spinal canal

compression, cervical MRI revealed an os odontoideum with myelomalasia in the spinal cord at the C1-2 level and syringomyelia at the T3 level (Figure 1). Dynamic (flexion and extension) lateral cervical x-ray films and MRI were performed. The distance between anterior edges of atlas and C2 had increased to 14 mm on lateral X-ray films. The anterior dislocation of os odontoideum with atlas and the compression of the spinal canal by the posterior arcus of atlas resulting in myelomalasia of the spinal cord were observed at sagittal T2-weighted flexion MRI (Figure 2). Anterior odontoid screw fixation was performed on an elective basis taking into consideration that os odontoideum dislocates with atlas (Figure 3). In the control examination 3 months after the operation, she was still suffering from pain and her dynamic MRIs showed that compression of the spinal cord persisted (Figure 4). Thus the patient was operated on again and the posterior arcus of atlas was resected. Occiputo-cervical plate-screw fixation and autologous bone graft fusion was also performed (Figure 5). In the control examination 3 months after the second operation, the patient was completely free from pain and her MRI showed total decompression of the C1-2 (Figure 6).



Figure 1: Preoperative T2-weighted sagittal cervical MRI of the patient.



Figure 2: Preoperative T2-weighted sagittal cervical MRI of the patient in flexion position.



Figure 4: Sagittal T2-weighted cervical MRI three months after the operation.

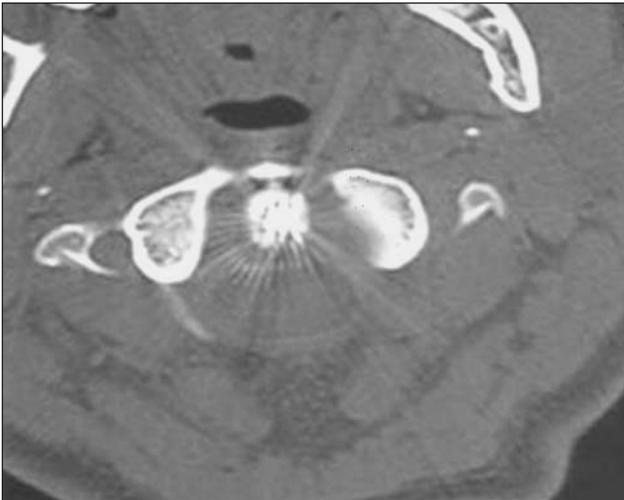


Figure 3: Early postoperative computerized tomography scan shows odontoid screw fixation.



Figure 5: Lateral cervical roentgenogram of the patient after occipitocervical fixation.



Figure 6: Sagittal T2-weighted cervical MRI shows decompression of the spinal cord three months after the second operation.

DISCUSSION

The natural history of untreated os odontoideum comprises a wide spectrum. There are many examples of both asymptomatic and symptomatic patients with known os odontoideum who have never been treated and who have had no reported new problems during follow-up over many years (19). Conversely, examples of sudden spinal cord injury in association with os odontoideum after minor trauma have also been reported (14). The

predictive factors for deterioration, particularly in the asymptomatic patient, have not been identified (11). Indications for surgical stabilization include the simple existence of an os odontoideum, os odontoideum in association with occipitocervical pain alone, and/or os odontoideum in association with neurological deficit (8, 19). Other factors that may assist in determining the need for stabilization and/or decompression include C1-C2 instability, associated deformity and spinal cord compression (11).

The aims of surgery in a patient with os odontoideum are stabilization and/or neural canal decompression when necessary as in other spinal pathologies. The preferred surgical stabilization method in the literature is posterior C1-C2 arthrodesis. Two larger series by Fielding et al. (8) and later by Spierings and Braakman (19) in the early 1980s described both operative and nonoperative management strategies for these patients. Fielding et al. (8) described 35 patients with os odontoideum, 27 with radiographic evidence of instability and 8 without instability. They performed successful C1-C2 posterior arthrodesis to 26 of 27 patients with instability and 8 patients with no evidence of C1-C2 instability were managed nonoperatively without complication. Spierings and Braakman (19) reported a series of 37 patients with os odontoideum. Of the 37 patients, 17 underwent surgical treatment while the remaining 20 were not operated for analysis of the natural history of os odontoideum. Of the 17 patients who underwent surgery, one patient had neurological worsening and two died. Of the 20 patients who were managed conservatively, 15 had no myelopathy and none of them developed myelopathy at follow-up. Neurological worsening was not observed at follow-up in any of the other 5 patients with myelopathy. The authors conclude that patients with os odontoideum without C1-C2 instability can be managed without surgical stabilization and fusion with good results.

More recent series reported in the literature provide better descriptions of the operative procedures and postoperative immobilization techniques used for patients with os odontoideum (3, 4, 6, 7, 14, 19, 20). Posterior wiring and fusion techniques supplemented with postoperative halo immobilization provided successful fusion in 40 to 100% of cases reported (4, 6, 19). Atlantoaxial transarticular screw fixation and fusion seem to have

merit in the treatment of C1-C2 instability in association with os odontoideum and to obviate the need for postoperative halo immobilization. In cases with neural compression in association with os odontoideum reduction of deformity, C1-C2 or occipitocervical fusion and internal fixation are advised (18). On the other hand, in cases with irreducible deformities supplemented dorsal or ventral decompression will be necessary (6, 18, 20). Inability to anatomically reduce the fracture and transverse atlantal ligament disruption is an absolute contraindication for direct internal fixation of the odontoid process (21). If anatomic reduction cannot be achieved, posterior C1-C2 arthrodesis should be considered (21). In our case, simultaneous motion of atlas and os odontoideum on preoperative dynamic MR images was clear evidence of an intact transverse atlantal ligament. Thus, anterior odontoid screw fixation was performed for preventing simultaneous motion of the atlas and os odontoideum. Nevertheless, C1 arcus resection with a second operation was needed after determination of persistent spinal cord compression on control dynamic MR images three months after operation.

CONCLUSION

Patients who have no neurological deficit and have no instability at C1-C2 on flexion/extension studies can be managed without operative intervention. Cases with myelopathy in association with instability are the most suitable candidates for surgery. C1-C2 transarticular screw fixation and fusion seem to be the best surgical choice in patients who have instability and reducible deformity alone. In patients with irreducible deformity, C1-C2 or occipitocervical fusion after ventral or dorsal decompression and internal fixation are advised. Odontoid screw fixation is not advised in this pathology since it is a chronic process.

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