



## OS ODONTOIDEUM: A RARE CONGENITAL ANOMALY

### OS ODONTOIDEUM: NADİR BİR KONJENİTAL ANOMALİ

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#### SUMMARY:

Os odontoideum is a separate odontoid process from the body of the axis. It is the most common anomaly of the odontoid process. The etiology of os odontoideum remains controversial, although there is emerging consensus on both traumatic etiology and a congenital source. Patients with this condition can be asymptomatic or present with a wide range of neurological dysfunctions. It may cause cervical instability, atlantoaxial dislocation and myelopathy. This anomaly can mimic Type-I and II odontoid fractures. Accurate diagnosis is mandatory to prevent treatment failures. There is a role for conservative treatment of an asymptomatic incidentally found, radiologically stable, and non-compressive os odontoideum. However, surgical treatment has a definitive role in symptomatic cases.

In this study, the case is presented of a 31 year- old male patient with neck pain who was diagnosed with incidental os odontoideum. The diagnosis of acute odontoid fracture was discarded in this case as the radiological findings were of a characteristic cortex with smooth contours, and there was no history of recent trauma, sclerosis or hypertrophy of the anterior tubercle of the atlas.

**Keywords:** Os odontoideum, congenital anomaly, cervical instability

**Level of evidence:** Case report, Level IV.

#### ÖZET:

Os odontoideum odontoid sürecin aksisin gövdesinden ayrı bir kemik parça olmasıdır. Odontoid sürecin en sık rastlanan anomalisidir. Os odontoideum etiyojisi tartışmalıdır. Travmatik ve konjenital etiyojisi arasında konsensus hâlâ sağlanamamıştır. Os odontoideum hastaları asemptomatik, semptomatik ya da çok çeşitli semptomla başvurabilirler. Servikal instabilite, atlantoaksiyel dislokasyon ve myelopatiye neden olabilir. Bu anomali Tip-1 ve 2 odontoid fraktürleri taklit edebilir. Doğru olarak tanımlanması olası tedavi yanlışlıklarını önlemek için önemlidir. Asemptomatik olanlar radyolojik istikrarlı ve nöral basısı olmayan os odontoideum olguları konservatif tedavi ile tedavi edilirken, nöral bası ya da semptomatik olanlarda cerrahi tedavisinin yapılması gerekir. Bu çalışmada boyun ağrısı şikayeti ile başvuran ve ossikulun karakteristik düzgün konturlu korteksinin bulunması, yakın zamanda geçirilmiş travma öyküsünün olmaması, atlasın anterior tuberkülünün skleroz ve hipertrofisi nedeniyle akut odontoid proses fraktürü dışlanarak os odontoideum tanısı konulan 31 yaşındaki erkek hastamızı sunduk.

**Anahtar kelimeler:** Os odontoideum, ayırıcı tanı, boyun ağrısı, servikal instabilite

**Kanıt düzeyi:** Olgu sunumu, Düzey IV.

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## INTRODUCTION:

Congenital anomalies related to the odontoid process have been defined in literature as aplasia, hypoplasia, duplication, condylus tertius, os terminale (os avis) and os odontoideum. Of these, the most frequently seen is os odontoideum<sup>6</sup>. The os odontoideum is an oval or round piece of bone with regular cortical edges, located behind the atlas anterior arch, and is separate from the hypoplastic odontoid process. It has been reported under various names such as “Cleft Dens Disease”, “Incomplete Dens Disease” and “Independent Dens Fragment Disease”<sup>9</sup>. First described in literature by Giacomini in 1866, the etiology of os odontoideum is still controversial<sup>2</sup>. The embryology of the odontoid process is complex and the reasons for the controversy are that there is evidence of both congenital and acquired causes<sup>2-3,9</sup>.

Os odontoideum may cause various symptoms such as atlantoaxial instability, spinal cord compression, myelopathy, neck pain and respiratory dysfunction<sup>2</sup>. Although narrowed spinal canal seen on radiographs and atlantoaxial instability are debated as factors increasing the severity of clinical symptoms, no clear correlation has been seen between radiological findings and clinical symptoms<sup>11</sup>.

This anomaly may mimic Type-1 and Type-2 odontoid fractures. Accurate identification is important to prevent the possibility of incorrect treatment. The case is here presented of a patient who presented with complaints of neck pain at an external centre and was diagnosed with an odontoid fracture, was referred to our clinic with the indication for emergency surgery, but as a result of the examination was diagnosed with os odontoideum as an uncommon cause of neck pain.

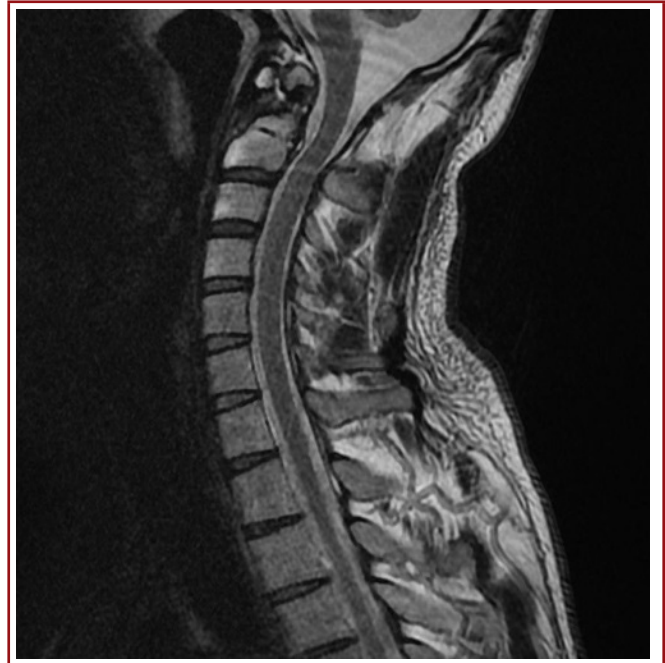
## CASE REPORT:

A 31-year old male presented at an external centre with neck pain and on the magnetic resonance imaging (MRI), there was a very severe neck fracture and so the patient was referred to our clinic with the need for urgent intervention. There was no history of trauma.

The patient reported that he had experienced sporadic neck pain. In the physical examination of the patient, there was nothing remarkable. Cervical MRI and cervical spinal CT were applied. On the cervical MRI, a regular bordered ossicle was observed in the posterior of the atlas anterior arch, separate from the hypoplastic odontoid process. There were no findings of signal change in the spinal cord, oedema in the bone or trauma to the surrounding tissues, no damage to the ligamentous structures or narrowing in the canal (Figure-1).

On examination of the cervical spine CT, a small round odontoid process with regular and uniform cortical borders was observed to have separated from the C2 odontoid process

base with a wide radiolucent gap and to be located behind the atlas anterior arch (Figures-2, 3).



**Figure-1.** Sagittal T2-weighted MRI showing the os odontoideum part separate from the main body and natural spinal cord signal intensity.



**Figure-2.** Reformatted sagittal CT imaging showing hypoplasia of the odontoid process and dystopic os odontoideum separate from the odontoid process, surrounded by well-bordered cortex.



**Figure-3.** Reformatted coronal CT imaging showing the os odontoideum part separate from the main body.

The bone was located close to the clivus (dystopic os odontoideum). There were no findings of instability on the MR and CT findings. As there was characteristic regular contoured cortex of the odontoid bone and sclerosis and hypertrophy of the atlas anterior tubercle, acute odontoid process fracture was discounted and the diagnosis was made of os odontoideum. The patient had no neurological deficit and stabilization treatment was applied with an external orthosis and follow-up.

## DISCUSSION:

Os odontoideum, which is defined as a separate piece of bone in the posterior of the C1 anterior arch is classified into 2 types of dystrophic or orthopic depending on the anatomic placement of the free, independent fragment. In the dystrophic type, the majority of the free fragment is seen to be attached in the clivus inferior third and in the less commonly seen orthopic type, the free fragment moves as in its normal location<sup>3,6,8-9</sup>. In literature it has been reported that the dystrophic type is most likely congenital<sup>8</sup>. In the current case, os odontoideum was determined as the dystrophic type.

Os odontoideum must be differentiated from a dens fracture. Hypertrophy and sclerosis of the atlas anterior tubercle are used to differentiate os odontoideum from an acute dens fracture<sup>2,9,13</sup>. Hypertrophy of the atlas anterior arch and impairment of the line to the spinolaminar complex are radiographic findings but are not specific to os odontoideum<sup>5,13</sup>.

In dens fractures, the corners and cortex are irregular. The difference in os odontoideum is that it is oval or round with regular edges, the cortex is protected and the corners are relatively sclerotic<sup>2,12</sup>. Specifically, the distance between fracture fragments is narrower and the axis extends within the corpus below the level of the superior facets of the vertebrae. On dynamic radiographs, just as the structure of fracture fragments is destroyed so the compatibility of the ends is demonstrated<sup>9</sup>. In the current case, as there was a regular contoured cortex characteristic of the odontoid process, there was no recent history of trauma and there was sclerosis and hypertrophy in the atlas anterior tubercle, acute odontoid process fracture was discounted and the patient was diagnosed with os odontoideum.

The etiology of os odontoideum is still a matter of debate. That it is seen together with anomalies such as Down's syndrome, Klippel-Feil syndrome and multiple epiphyseal dysplasia, supports that it is congenital<sup>2,7,10,13</sup>. In recent studies, it has been suggested that a reliable radiological finding of congenital os odontoideum is the joint finding known as the 'jigsaw sign' between the atlas anterior arch and the odontoid and this supports the view of congenital etiology<sup>13</sup>.

It has been reported that the impairment of dens blood flow associated with the stretching of the alar ligaments which are important in providing atlantoaxial stability, could originate from non-union in the odontoid<sup>4</sup>. It is thought that it may occur because of odontoid fracture not diagnosed in childhood or a lack of immobilisation or in some cases following bone and ligament damage associated with trauma in early childhood<sup>1</sup>. Reasons have been suggested of impaired dens blood flow after trauma and non-union in the odontoid or a pseudo joint. This is said when the odontoid does not show union<sup>13</sup>. Some references in literature have reported that the odontoid process has developed normally but could not attach to the C2 corpus because of abnormal movement<sup>9</sup>. The attachment of the odontoid process to the C2 corpus should be completed between the ages of 5-7 years<sup>13</sup>.

Whatever the reason, this situation causes the development of a small hypoplastic odontoid in the upper part of the axis corpus and the development of a separate ossicle with no continuity with the axis corpus in the posterior of the atlas anterior arch. In the current case, as there was no history of trauma in childhood, the os odontoideum was considered to be congenital.

Os odontoideum cases may be asymptomatic or symptomatic. As the spinal canal is relatively wide at the C1-2 level, symptoms are not seen in this area<sup>5</sup>. In some asymptomatic cases, the patient is not treated and continues for years with no new problems observed<sup>2</sup>. In symptomatic cases, neck and shoulder pain is most often seen. Less frequently reported symptoms are headache, torticollis, thinness and weakness. The most

serious complication of C1-C2 instability develops because of spinal cord compression or obstruction of the vertebral artery. In literature, many asymptomatic and symptomatic cases have been reported who have not been treated and have continued for years with no new problems observed during follow up.

Asymptomatic cases that are diagnosed incidentally must be well evaluated and the indication for surgery should not be given immediately<sup>9</sup>. In the current case, as there was no complaint other than pain, there was no history of trauma and no neurological deficit was determined, the patient was given detailed information about his condition, a conservative treatment method was selected and the patient was closely monitored. In patients with imaging abnormalities between C1-C2, although os odontoideum is rare, it should be kept in mind as the most frequently observed anomaly of the odontoid process. Asymptomatic and stable cases must certainly be well evaluated and it should be considered not necessary to give indications for surgery immediately.

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