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# A Rare Cause of Myelopathy: Os Odontoideum

## *Nadir Bir Miyelopati Nedeni: Os Odontoideum*

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**Anahtar Kelimeler:** Kraniyoservikal bileşke anomalileri, os odontoideum, miyelopati

**Dear Editor,**

A 60-year-old female patient was admitted with difficulty in walking. We learned that her symptoms started with head and neck pain 6 years ago, and that difficulty in walking was added to her symptoms over time. In the last few years, she started to have upper extremity weakness and atrophy, and we learned that she could not mobilize herself for the past three months. Her medical history and family history were normal. At the initial neurologic examination, she was conscious, oriented and cooperative, and cranial nerves were intact. Proximal dominant quadriparesis syndrome was present. Deep tendon reflexes were increased in the upper extremities, hyperactive in the lower extremities, and Hoffman's and Tromner's signs were bilaterally positive. Bilateral Achilles and patellar clonus were present, she had bilateral extensor plantar reflex, her superficial tactile sensation was normal, and cerebellar tests were impaired due to paresis. There was bilateral atrophy of the forearm, interosseous, thenar, hypothenar, and peroneal muscle groups with no fasciculation. We found that the patient was able to move with broad-based and ataxic gait by unilateral support on a flat surface. Laboratory examinations revealed normal complete blood count and routine blood biochemistry. There was no significant pathologic finding in the collagen tissue examinations performed from blood. The venereal disease research laboratory test (VDRL): 1/4 and Treponema pallidum hemagglutination assay (TPHA): 1/640 were positive in the infection panel. With these results, the patient was referred to the infectious diseases clinic. Considering syphilis, 24 million units/day of crystallized penicillin and lumbar puncture (LP) were suggested. In the CSF examination, the protein was found to be high, close to the upper limit (52.6 mg/dL). Lactate was 2.02 and glucose was 75 mg/dL (simultaneous blood glucose: 103 mg/dL), and direct examination and cerebrospinal fluid (CSF) cytology were normal. Depot penicillin treatment was initiated and outpatient clinic follow-up was suggested twenty days later because the VDRL and TPHA in the CSF were negative for neurosyphilis. In somatosensory evoked potential studies, bilateral responses with delayed latencies were obtained in the upper extremities, but no response was obtained in the lower extremities. Nerve conduction studies were normal in the electroneuromyography examination. In a needle electromyography (EMG) examination, chronic neurogenic involvement and motor unit potential loss were detected. No denervation and fasciculation were observed. Bilateral renal cysts were detected in thoraco-abdominal computed tomography. A carotid and vertebral artery Doppler examination revealed increased resistance in the vertebral artery. Cerebral and whole- spinal magnetic resonance imaging examination revealed a prominent posterior angulation of the odontoid in the craniocervical junction, spinal cord compression, and myelomalacia (Figure 1). After considering all these findings, the patient was diagnosed as having os odontoideum. The neurosurgery clinic recommended surgery but the patient did not consent.

Os odontoideum is a rare craniocervical junction anomaly with a smooth cortical edge, located behind the anterior arc of the atlas and separated from the main body of the odontoid process (1). It was first described by Giacomini in 1866 (2,3). Odontoid process anomalies may occur due to acquired and genetic reasons. Acquired lesions often occur with posttraumatic

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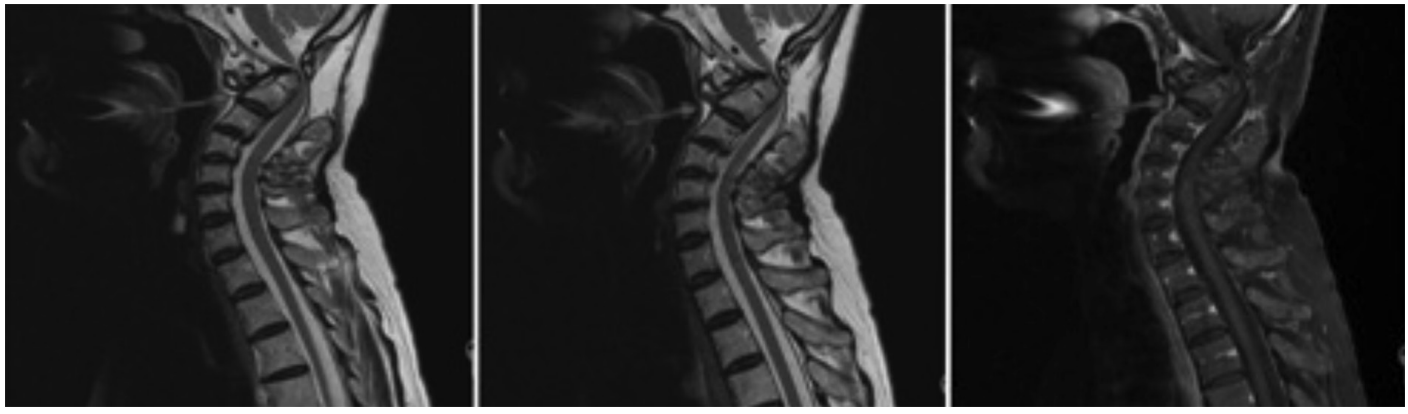

**Figure 1.** The cervical magnetic resonance imaging examination revealed a prominent posterior angulation of the odontoid, spinal cord compression and myelomalacia in the craniocervical junction.

causes. Congenital anomalies of the odontoid process include aplasia, hypoplasia, duplication, condylus tertius, persistent os terminale, and os odontoideum (2). The most common is os odontoideum (1,2). The fact that it is reported together with many developmental anomalies in the literature supports congenital causes (1,2,3,4). Studies have reported an association with *Hox* and *Pax-1* genes (4). Some patients are asymptomatic, and it is incidentally detected. Symptomatic patients present with atlantoaxial instability and myelopathy (1,2). Surgical treatment is the first option in symptomatic patients and provides very good results in experienced hands (5). This article was written to draw attention to os odontoideum, a rare cause of myelopathy.

**Ethics**

**Informed Consent:** Consent form was filled out by a participant.

**Peer-review:** Internally peer-reviewed.

**Authorship Contributions**

Surgical and Medical Practices: M.B., Z.S.Ş., M.D., Concept: M.B., Z.S.Ş., M.D., Design: M.B., M.D., Data Collection or Processing: M.B., Z.S.Ş., Analysis or Interpretation: M.D., Literature Search: M.B., Z.S.Ş., Writing: M.B.

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## References

1. Crockard H, Stevens M. Craniovertebral junction anomalies in inherited disorder: Part of the syndrome or caused by the disorder? Eur J Pediatr 1995;154:504-512.
2. Komut E, Kültür T. Os Odontoideuma Bağlı Miyelopati. KÜ Tıp Fak Derg 2015;17:41-45.
3. Schuler TC, Kurz L, Thompson E, Hensinger RN. Natural history of os odontoideum. J Paediatr Orthop 1991;11:222-225.
4. Özeren E, Şimşek S. Kraniyoservikal bileşkenin gelişimi ve anomalileri. Türk Nöroşir Derg 2015;25:104-109.
5. Le Pape S, Gauthe R, Latrobe C, Leroux J, Roussignol X, Ould-Slimane M. Cervical myelopathy involving os odontoideum and retro-odontoid cyst treated with Harms C1-C2 arthrodesis. Orthop Traumatol Surg Res 2016;102:817-820.