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Case Report

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Os odontoideum with progressive symptoms: case report

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ABSTRACT

A 53-year-old male patient was admitted to the neurosurgery outpatient clinic with neck pain. He had difficulty performing fine manual skills, had numbness in his hands, and felt as if he was stepping on a space while walking. He declared that his head occasionally fell forward involuntarily. He had no urinary or fecal incontinence or neurogenic claudication. On neurologic examination, there was no loss of muscle strength and no pathologic reflex. He was skillful in cerebellar tests. He declared no history of falls, impacts, or accidents. From the patient's hospital digital records, it was seen that he was admitted to the neurosurgery outpatient clinic with right arm and neck pain before, and surgical treatment was recommended to him due to the os odontoideum, but he refused this treatment. As the patient became symptomatic in the following period, atlantoaxial instability was found on cervical dynamic X-rays. For all these reasons, he was advised to undergo surgical treatment, but he re-refused the operation. In conclusion, it was considered that close follow-up with conservative treatment in asymptomatic os odontoideum patients may be appropriate. However, it was suggested that the possibility of AAI should be considered in patients who become symptomatic.

Keywords: Os odontoideum, atlantoaxial instability, conservative treatment, surgery

INTRODUCTION

Os odontoideum (OO) is an anatomical variant of the C2 dens, defined as an independent ossicle of variable size with smooth circumferential cortical edges separated from the axis.[1] Congenital and acquired causes are discussed in the etiology of os odontoideum and there is still no clear information about its epidemiology. Patients with os odontoideum may be asymptomatic or present with a spectrum of neurologic deficits. Conservative treatment is recommended for asymptomatic patients, while surgical treatment is recommended for symptomatic patients.[2] In this case report, a patient with os odontoideum is discussed.

CASE

A 53-year-old male patient was admitted to the neurosurgery outpatient clinic with neck pain which had been present for 4 years but had intensified in the last 1 month. He declared difficulty in performing fine manual skills such as buttoning buttons and zipper pulling, numbness in his hands, and felt as if he was stepping on a space while walking. He also declared that his head occasionally fell forward involuntarily. He had no urinary or fecal incontinence or neurogenic claudication. He had no history of falls, impacts, or accidents. On neurologic examination, it was observed that his cervical posture was impaired; his head was turned forward and slightly to the right in its natural position (torticollis). His muscle strength was normal, but his biceps and patella reflexes were hyperactive. No pathologic reflex (such as Babinsky's sign or Hoffman's sign) was detected. He was skillful in cerebellar tests.

When the patient's hospital digital records were analyzed, it was seen that the brain magnetic resonance (MR) images performed five years ago for headache showed findings os odontoideum and myelomalacia in the spinal cord at that segment. It was also seen that at that time, he was admitted

to the neurosurgery outpatient clinic with right arm and neck pain, his neurological examination findings were found normal, and the os odontoideum and myelomalacia in the spinal cord were also seen in the cervical MR images. Surgical treatment was recommended to the patient, but the patient refused the treatment and therefore he was referred to physical therapy. In addition, the patient was evaluated by the neurology department two years ago with complaints of numbness and weakness in bilateral upper extremities, dizziness, involuntary dorsiflexion of the feet from the ankle after waking up in the morning, difficulty in maintaining balance while walking in the road from time to time and feeling like falling. It was understood that myelomalacia and os odontoideum and myelomalacia in the spinal cord persisted in the cervical MR images performed at that time (Figure 1). Current cervical MR images of the patient showed that the os odontoideum was separated from the C2 vertebra corpus compartment at the skull base (dystopic os odontoideum) and migrated 9.5 mm from the skull base towards the superior (bacillary invagination). In addition, the distance between the separated os odontoideum and the distal end of the clivus was narrowed. At this level, there was anterior compression of the medulla oblongata and proximal spinal cord and increased intensity due to this effect (myelomalacia). The anterior-posterior diameter of the proximal part of the spinal canal was 5.4 mm (Figure 1).

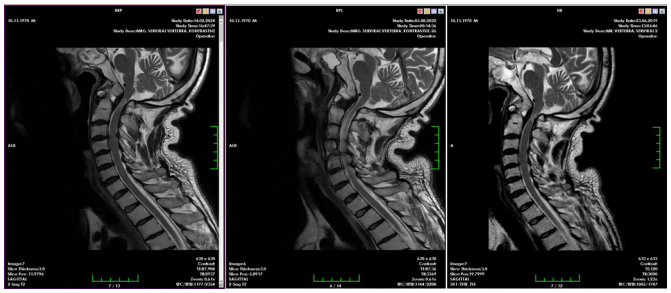

Figure 1. Cervical MR images from various dates showing os odontoideum, narrowing of the cervical canal, basilar invagination, and myelomalacia.

As the patient became symptomatic in the following period, to reveal cervical instability cervical computed tomography and cervical dynamic X-ray were performed to evaluate the bone structures and their mobility more clearly. Computed tomography showed that the dens fragment healed by adhering to the lower end of the clivus with possible apical ligament calcification after a possible odontoid dens fracture and narrowed the foramen magnum with parallel extension of the occipital bone (Figure 2).

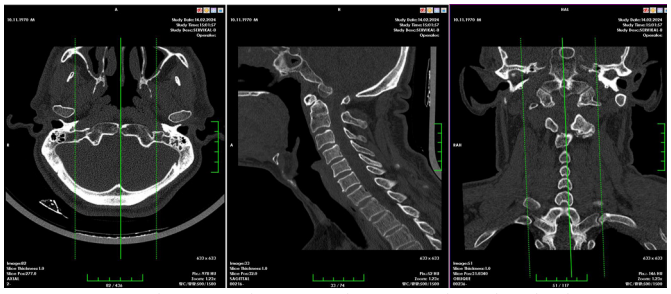

Figure 2. Cervical computed tomography images show that the dens fragment healed by adhering to the lower end of the clivus with possible apical ligament calcification after a possible odontoid dens fracture and narrowed the foramen magnum with the parallel extension of the occipital bone

When these radiologic images were compared with the old images, no additional pathologic findings were observed. However, cervical dynamic X-rays revealed that the patient had serious cervical instability (Figure 3).

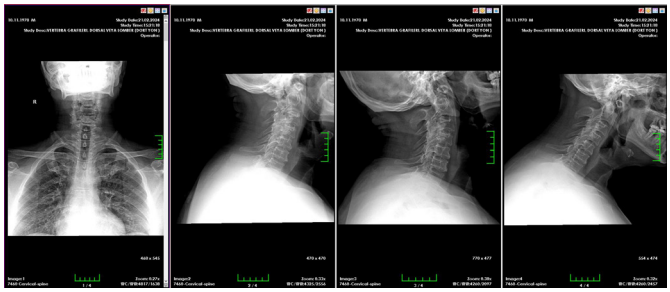

Figure 3. Cervical dynamic X-ray images show atlantoaxial instability.

For all these reasons, the patient was advised to undergo surgical treatment, but the patient refused the surgical intervention.

DISCUSSION

Today, two main theories are discussed for the formation of the os odontoideum: congenital and traumatic.[3] The congenital hypothesis argued incomplete fusion of the dens and axis vertebral bodies (segmental defects) due to developmental failure of the synchondrosis during embryonic development, non-traumatic osteonecrosis, congenital malformations (neurofibromatosis, skeletal dysplasias), autosomal dominant inheritance.[2] In the traumatic theory, it is argued that contraction of the alar ligament following an unrecognized odontoid fracture may lead to avascular necrosis and osseous remodeling contributing to ossicle formation. At the same time, deficiencies in arterial blood supply and trabecular bone at the base of the dens may predispose the dens to stress fractures caused by repeated microtrauma.[3,4] However, considering that os odontoideum is most commonly seen at the base of dens and not in synchondrosis, traumatic etiology has started to be accepted more than congenital etiology.[5] In our patient, there was no history of previous trauma or evidence of congenital malformation. However, considering the patient's age and the onset of symptoms in the last few years, it was thought that the os odontoideum may have a traumatic origin.

Common local symptoms of os odontoideum may include neck pain and stiffness, torticollis, ataxia, shoulder pain, headache, restricted neck movement, fatigue, hoarseness, respiratory dysfunction, swallowing difficulties, isolated occipital-cervical pain, upper extremity paresthesia including intermittent tingling and numbness in the upper limbs, lower extremity weakness, and gait disturbance.[6] In these patients, the os odontoideum may cause abnormal atlantoaxial instability (AAI) in both anterior and posterior directions. In these patients, compression of the vertebral artery and subsequent vascular complications may contribute to cervical myelopathy, along with spinal cord tension or bone compression. Myelopathic deficits can range from mild paresis or transient myelopathy to progressive tetraplegia, bulbar signs, and even death.[7] These patients may have central cord syndrome, hypoventilation syndrome (Ondine's

curse), Brown-Sequard syndrome, Lhermitte phenomenon, sleep apnea, lower cranial nerve dysfunction, hyperesthesia, bowel and bladder dysfunction, hypoesthesia, allodynia, hyperalgesia, vertebral artery occlusion and ischemia of the brainstem and posterior fossa structures can lead to seizures, cervical vertigo, syncope, visual disturbances, and in severe cases, sudden death. Late neurologic deterioration occurs in only 4% of patients.[2] In our patient, neurologic examination findings were normal except for a gradually increasing number of symptoms over the last few years. These symptoms included difficulty in performing fine manual skills such as buttoning buttons and pulling zippers, numbness in the hands, and the occasional sensation of stepping on a gap when walking.

The management of asymptomatic os odontoideum is controversial due to limitations in understanding its natural history. There are numerous studies in the literature showing that long-term conservative management is successful for patients with stable os odontoideum, as well as studies reporting cases of neurologic deterioration and sudden death. Non-surgical treatment methods for asymptomatic patients include serial imaging, longitudinal radiographic follow-up, and clinical observation. Immobilization consisting of a cervical collar or cervical traction may also be used.[8] It should be kept in mind that the initially stable os odontoideum may begin to develop AAI and associated symptoms. Patient education regarding the potential risks and avoidance of contact sports are also recommended.[9s] When os odontoideum was diagnosed in our patient, surgical treatment was primarily recommended to him five years ago. However, since the patient did not accept surgical treatment and neurologic examination findings were found to be normal, a conservative treatment option had to be applied. However, the patient's symptoms gradually increased in the following period and surgical treatment was recommended on his current admission again, but the patient refused this treatment again. The patient was obligatorily followed up again and the risks that might occur in the absence of surgical treatment were explained in detail again.

Sagittal spinal canal diameter <13 mm is strongly associated with myelopathy and studies report a 10% chance of developing this condition. Dystopic configurations of the os odontoideum and round morphology types are also risk factors for myelopathy and AAI. Surgical decompression and stabilization are recommended for patients with radiological indications of AAI, dynamic myelopathy, or neurological dysfunction. The most commonly used surgical treatment method is "posterior C1-C2 screw fixation and fusion". Other techniques include "sublaminar cabling of C1-C2" and "occipitocervical fusion". In addition, in recent years, "endoscopic endonasal resection techniques" have been applied more and more frequently in craniovertebral junction pathologies, including os odontoideum. It should not be forgotten that this approach should be considered as a complement, not an alternative, to the transoral-transnasal route, and that appropriate patient selection and surgical experience are important. Postoperative complications may include wound infection, cerebrospinal fluid leakage, persistent muscular neck pain, neurologic and vascular injury, anesthesia complications, pseudarthrosis, and hardware loosening.[8] A higher risk for perioperative complications has been reported in patients with unstable os odontoideum with cord compression or congenital ligamentous laxity.[10] In our patient, radiologic images showed myelomalacia findings for at least five years and the canal diameter was 5.4 mm at its narrowest point. However, since his previous symptoms were poor, neurological examination findings were found normal, and he refused the surgical intervention, he was initially treated conservatively. However, the progression of symptoms over time and dynamic radiographs performed to determine the cause of this progression revealed the development of AAI. Surgical treatment was recommended to the patient because of both the progressive symptoms increase and the presence of AAI, but, the patient did not accept the surgical treatment again.

CONCLUSION

In light of all these findings, it was argued that close and close follow-up of asymptomatic os odontoideum patients with conservative treatment may be appropriate considering the severity of complications of surgical treatments. On the other hand, it was suggested that the possibility of AAI should be considered in patients who become symptomatic.

ETHICAL DECLARATIONS

Informed Consent

The patient signed and free and informed consent form.

Referee Evaluation Process

Externally peer-reviewed.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

Financial Disclosure

The authors declared that this study has received no financial support.

Author Contributions

All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

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