

## Os Odontoideum: A Rare Case Report

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### Abstract

*Os odontoideum is very rare and its exact frequency is unknown. Most of cases are incidently detected and others are diagnosed when patient becomes symptomatic.*

*We report on a 24-year-old married female presenting with neck pain radiating to both upper and lower limb more in right upper limb since 15 years. Radiological findings demonstrated an os odontoideum and osseous continuity between the occiput and an ossicle, termed an "occipitalized os odontoideum." The occipitalized ossicle and atlas moved as a functional unit from the body of the axis. Eighteen months later, he remained free of symptoms and showed solid bone fusion. Our report provides useful knowledge regarding treatment of extremely rare osseous anomalies in the craniovertebral junction.*

**Keywords:** os odontoideum, treatment

### INTRODUCTION

An os odontoideum is a rare condition in the craniovertebral junction (CVJ) which can cause mild progressive myelopathy or sudden spinal cord injury even after minor trauma. It was first seen in 1863 by Giacomini and he gave it the name as os odontoideum. In this disorder the translation of atlas on the axis can compress the spinal cord or vertebral arteries results in the neurological deficit. Here, we report our experience with a patient who had myelopathy due to atlantoaxial instability secondary to os odontoideum associated with an osseous continuity between the occiput and an ossicle, which was termed an "occipitalized os odontoideum." To our knowledge, this is the first report describing an occipitalized os odontoideum.

### CASE REPORT

A 24-year-old female presented with pain in neck, motor weakness and numbness in the left upper extremity. The pain was dull aching, progressive localised to neck region with out any radiation. On examination, the patient had short neck with restricted neck

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movement. Mouth opening was normal, mallmapati score was 1. Neurological examination revealed motor weakness (grade 4/5) in his left triceps and intrinsic muscles of left hand. She had decreased sensation in left hand, intact cranial nerves and hyperreflexia in the left upper extremity with a positive Hoffmann sign. She did not present with any history of weakness in lower limbs. Knee and ankle reflexes were normal bilaterally, with negative Babinski signs. No bowel or bladder difficulties.

Plain lateral radiographs showed an os odontoideum, and atlantoaxial instability was demonstrated during flexion-extension. Reconstruction computed tomography (CT) images demonstrated osseous continuity between the occiput and an ossicle, termed as “occipitalized os odontoideum.” The occipitalized ossicle and atlas moved as a functional unit from the body of the axis. Magnetic resonance imaging (MRI) demonstrated intramedullary high signal intensity changes at the posterior arch of the atlas. High signal-intensity changes without spinal cord compression suggest the presence of focal spinal cord contusion due to instability between the cranial unit and the body of the axis. The patient was given the option of posterior spinal cord decompression with fixation or fusion of C1-C2 vertebrae. The patient was operated and kept on clinico-radiological surveillance, anti-inflammatory drugs and physiotherapy of neck. If in future, the neurological status deteriorates then we will do the operative management.

## DISCUSSION

An os odontoideum is defined as the dissociation between the body of the axis and the dens, such that a disconnected ossicle takes the place of an intact odontoid process.[2] Surgical treatments, such as posterior atlantoaxial fixation with an atlas lateral mass screw and axis pedicle screw, have been reported as the mainstay of treatment for the patients who have os odontoideum and show neurological symptoms. They have also been reported, on occasion, to be a preventive treatment for spinal cord injury in patients without neurological symptoms.[1,3,4] Although the choice of surgical procedures for patients with atlantoaxial instability depends on the pathology of the instability, the patients' individual anatomical features, and their comorbidity, pre-operative imaging in the craniovertebral junction may often show concomitant diseases or conditions, such as osseous anomalies,[5,6] vertebral artery anomalies,[7] and congenital disease.[8] Knowledge of the treatments for these coexisting states can help the surgeon to prevent intra- or post-operative complications. We reported a case of an occipitalized os odontoideum, which was an extremely rare osseous anomaly in CVJ, causing spinal cord myelopathy. **We believe that our experience provide a basis for the selection of surgical procedure in this rare condition in CVJ.**

The surgical stabilization of the os odontoideum with atlantoaxial fusion involving transarticular screw fixation or atlas lateral mass and axis pedicle fixation with polyaxial screws and rods have been described as mainstay surgical treatments. Pre-operative imaging revealed instability between the unit and the body of the axis, not atlantodental or occipitotlas instability. Two previous reports regarding surgical treatment for atlantoaxial instability with an occipitalized atlas, resembling the current case with the presence of a fused cranial unit, have been published. Jain *et al.*[9] performed occipitotlas posterior fusion for 46 patients having congenital atlantoaxial dislocation with an occipitalized atlas. After that, Goel *et al.*[10] reported eight patients with an occipitalized atlas who had a mobile and reducible atlantoaxial dislocation and underwent lateral mass plate and screw fixation instead of fixation of the much longer plates or rods necessary for occipitocervical fixation. The method could be used even in

the situation where the facet of the atlas is occipitalized. This latter report supports our consideration that the shortest fixation for the extent of instability is the ideal intervention. Use of atlantoaxial fixation, instead of occipitoaxial fixation, has the advantage that it can provide stabilization in the anteroposterior direction, while still preserving the flexion-extension motion between the cranial unit and the body of the axis. Although occipitoaxial fixation for this condition seems to be a reasonable strategy for surgical treatment, the longer fixation can spoil the flexion-extension motion and increase the mobility and loading at the inferior adjacent segment.

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