

## Case Report

# Cervical Spondyloptosis in a Patient With Os Odontoideum: A Case Report



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## ABSTRACT

A 30-year-old man presented to our clinic with progressive neck pain and dysphagia. He had been operated on for an atlantoaxial instability (os odontoideum) 7 years ago. Imaging studies revealed cervical spondyloptosis of C5-C6 and C7-T1; the neurologic examination was intact. It was decided to correct the deformity through a circumferential approach. Thus C5, C6, and C7 corpectomy, alongside anterior column reconstruction using titanium expandable cage, reinforced by posterior spinal instrumentation from occipital bone to T3 vertebra were performed; his symptoms resolved completely following an uneventful surgery.

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## 1. Introduction

**S**pondyloptosis is the extreme degree of spondylolisthesis. The prevalence of cervical spondylolisthesis (CS) approximates 5% in the general population. CS frequently occurs in the mid-cervical spine and is associated with instability and dynamic spinal canal stenosis [1]. Congenital cervical spondyloptosis has been reported in less than 60 patients [2]. Os odontoideum (OO) as a congenital anomaly of the second cervical vertebra is associated with several syndromes and skeletal dysplasia [3]. The occurrence of high-grade CS (spondyloptosis) in a patient with OO has not yet been described. We report a case of cervical spondyloptosis in a patient with OO.

## 2. Case Presentation

A 30-year-old man was admitted to our clinic because of neck pain and dysphagia in the past several months. He had a history of occipitocervical fusion 7 years ago due to an atlantoaxial instability (OO) (Figure 1). He presented with symptoms of headache, neck pain, repeated falling, and gait instability before the index surgery. Meanwhile, on physical examination, he showed hyperreflexia and positive Hoffman signs. He had finally undergone posterior fusion from occipital bone to C4 plus C1 decompression using occipital plate and lateral mass screws for the cervical spine (Synthese Spine, West Chester, PA). His symptoms resolved and normal neurologic examination was restored. Then, he was absent for the follow-up for several years and later showed up with the aforementioned symptoms.

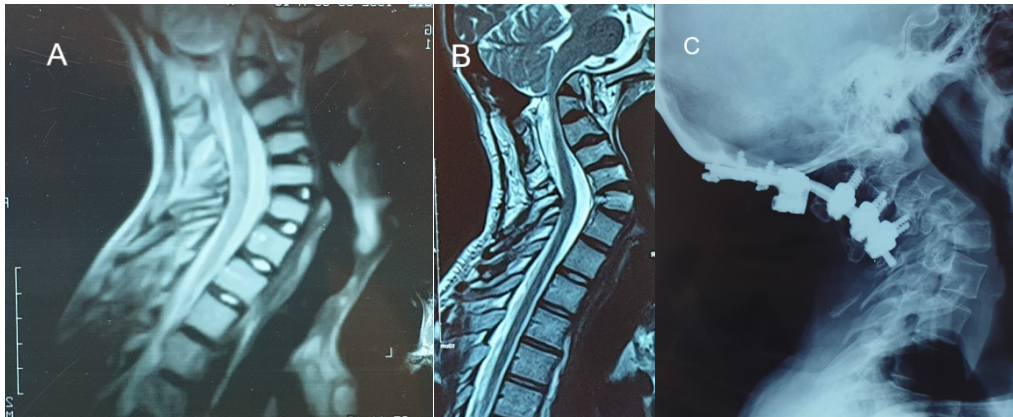
Physical examination demonstrated neither hyperreflexia nor positive Hoffman signs, and other neurologic tests, including sensation, muscle force, and gait stability (Tandem Romberg Test) proved normal. In cervical magnetic resonance imaging (MRI) and radiography, a simultaneous high-grade anterolisthesis of C7-T1 and retrolisthesis of C5-C6 (spondyloptosis) were observed (Figure 2). Retrospectively, we reviewed the MRI and the radiography of the index surgery and realized that C5-C6 mild retrolisthesis and C6 vertebra hypoplasia already existed which had progressed over the years and gave rise to the chain of spondylolisthesis. C6 hypoplasia and OO might have been related to a spectrum of formation failure in the cervical spine. Considering the progressive nature of high-grade spondylolisthesis, we decided to perform a 360° fusion. Firstly, we proceeded with the anterior approach. The patient was supine and the head was fixed in the Mayfield tongs with SSEP

monitoring. Through a standard anterolateral cervical spine approach, the skin was incised along the anterior border of the right sternocleidomastoid (SCM) muscle and blunt dissection in the interval between the carotid sheath and trachea-esophageal space was developed. Complete corpectomy of C5, C6, and C7 was performed, and then using a dynamic titanium expandable cage (Synthese Spine, West Chester, PA), the anterior column was reconstructed. The patient turned prone and through a posterior midline incision, the occipital bone to T3 was exposed bilaterally. Through a combination of the lateral mass and pedicle screws with rods (Synthese Spine, West Chester, PA), the instrumentation from the occipital bone to the T3 vertebra was carried out (Figure 3). The patient was advised to be on a cervical collar for 3 months postoperatively; he made an uneventful full recovery. 12 months after the surgery, the patient's dysphagia and neck pain were resolved completely; and the neurologic examination remained intact.

## 3. Discussion

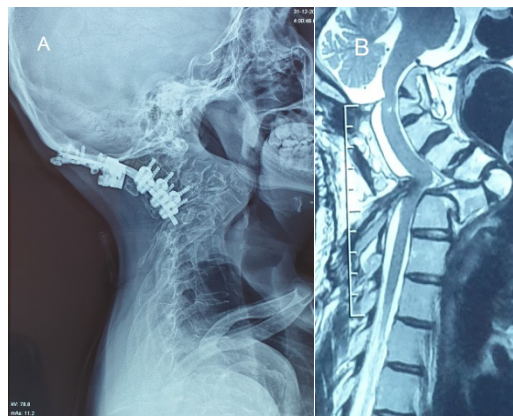
CS is probably underreported and has not received the same attention as lumbar spondylolisthesis, despite possibly being as common as the latter [4]. Isolated traumatic spondyloptosis in the cervical spine has been reported sporadically [5]. However, Bhojraj et al. [6] reported spondyloptosis at C7-T1 with late onset of cord compression symptoms in an 8-year-old girl. This is regarded as the first reported case of cervical spondyloptosis. CS develops from childhood, as Fedorchuk et al. [7] revealed the presence of a CS greater than 2 mm in 21% of the pediatric population (excluding C2-C3 pseudo-subluxation). The likelihood of CS increases with advancing age and mostly occurs in the upper cervical vertebrae that are more mobile.

The cervical spondyloptosis in ongoing pathological processes, such as neurofibromatosis type 1 [8] and the Larsen syndrome has been elucidated [9]. OO, a circumferentially corticated ossicle separated from the body of C2, is a major cause of atlantoaxial instability (atlantoaxial distance of > 5 mm or limited space available for the spinal cord <13 mm on maximum flexion or extension radiographs or MRI, irrespective of age). If instability is present, surgical stabilization is needed. The etiology of OO remains a subject of debate. Studies have supported both the congenital and traumatic origins; however, its association with Down syndrome, Morquio disease, Klippel-Feil syndrome, multiple epiphyseal dysplasias, achondroplasia, and Larsen syndrome, advocates the congenital hypothesis compared to being traumatic [3].



**Figure 1.** Cervical MRI

A: Cervical MRI at the age of 18 years (preoperational stage); B: Cervical MRI at the age of 23 years (preoperational stage); C: Radiography after occipitocervical fusion.



**Figure 2.** Radiography and MRI of the cervical spondyloptosis at the age of 30 years.

To the best of our knowledge, this is the first report of a cervical spondyloptosis in a patient with OO. The patient demonstrated C5-C6 retrolisthesis and C7-T1 anterolisthesis (spondyloptosis) which is not common for spondylolisthesis compared to C3-C4 or C4-C5 [10]. Furthermore, dysplasia of C6 vertebra which was retrospectively confirmed on MRI plus no history of trauma raises the suspicion of a congenital etiology in our patient. We believe that the cervical spondyloptosis observed in our case is strongly related to the pathophysiol-

ogy of OO development, defective bone and ligament formation, which led to the dysplasia of vertebra and incompetent joint and facet capsule; this would comprise the physiologic load and stress, and eventually gave rise to the progressive deformity of the cervical spine spondyloptosis. Hence, we favor the congenital hypothesis as the leading etiology for OO.



**Figure 3.** Radiography of occipito\_T3 vertebra fusion and plus anterior column reconstruction.

## Ethical Considerations

### Compliance with ethical guidelines

An informed consent letter was obtained from the patients who participated in this study, and they consented to the submission of this case report to the journal and publication of all the images.

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### Authors' contributions

Conceptualization and Methodology: Khodakaram Ras-tegar; Ras-tegar; data collection, data analysis, and writing-original draft: Hasan Ghandhari and Farzam Mokarami; Writing-review & editing: Ebrahim Ameri; Drafting the manuscript and data collection and literature review: Farzam Mokarami.

### Conflict of interest

The authors declared no conflict of interest.

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