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Review

Treatment of os odontoideum in a patient with spastic quadriplegic cerebral palsy

Yusuf T. Akpolat [a], Ben Fegale [b], Wayne K. Cheng [a,*]

[a] Department of Orthopaedic Surgery, Loma Linda University, East Campus Ambulatory Services Building, Suite 213, 11406 Loma Linda Drive, Loma Linda, CA 92354, USA
[b] School of Medicine, Loma Linda University, Loma Linda, CA, USA

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ABSTRACT

Severe atlantoaxial instability due to os odontoideum in a patient with spastic cerebral palsy has not been well described. There is no consensus on treatment, particularly with regard to conservative or surgical options. Our patient was a 9-year-old girl with spastic cerebral palsy and unstable os odontoideum as an incidental finding. During the waiting period for elective surgical treatment, the patient developed respiratory compromise. Surgery was performed to reduce the subluxation and for C1–C2 arthrodesis and the girl regained baseline respiratory function. A CT scan was obtained 1 year after the initial surgery and revealed adequate maintenance of reduction and patency of the spinal canal. This patient highlights the fact that unstable os odontoideum can cause mortality due to respiratory distress in patients with spastic cerebral palsy. This is an important factor in deciding treatment options for cerebral palsy patients with low functional demand. We review the relevant literature.

1. Introduction

Os odontoideum (OO), first described in 1886 by Giacomini, is a cause of atlantoaxial instability in pediatric patients [1]. The weight of the current evidence points to traumatic etiologies in the majority of patients [2]. Limited reports of OO in patients with cerebral palsy have been found in the literature and patients with spastic quadriplegic cerebral palsy present a special challenge to medical professionals. This patient population is physically fragile and has very limited function [3], yet, the preservation of their life remains the goal of the physician, as well as the family.

2. Methods

2.1. History

A 9-year-old girl with profound cognitive developmental delay and spastic quadriplegic cerebral palsy was referred to the clinic due to an incidental finding of OO by radiography. The mother denied any history of trauma and stated that the girl's neurological function had been static.

2.2. Initial examination

The girl was able to breathe unassisted and a general physical examination revealed that she was severely delayed and nonverbal. She was unaware of her environment and her gaze was mildly disconjugate with esotropia of the right eye. We were able to elicit a startle response. She had no observable purposeful motor activity below the neck. The severity of her motor disability was assessed with the gross motor function classification system and scored as a five [4].

2.3. Initial radiologic studies

Radiography revealed severe anterior displacement of C1 and the tip of C2 in front of the base of the odontoid (Fig. 1). CT scans revealed complete anterior translation of the C1 arch and the upper portion of the odontoid in front of the base of C2 (Fig. 2). MRI further revealed marked narrowing of the underlying thecal sac measuring 3.5 mm in midline anterior-posterior diameter and impingement of the underlying cervicomedullary junction (Fig. 3).

2.4. Preoperative course

Treatment options were discussed with the parent and due to the severe compromise of the cervicomedullary junction, an elective surgical option was chosen. The girl then presented to the

* Corresponding author. Tel.: +1 909 558 6444; fax: +1 909 558 6118.
E-mail address: md4spine@yahoo.com (W.K. Cheng).

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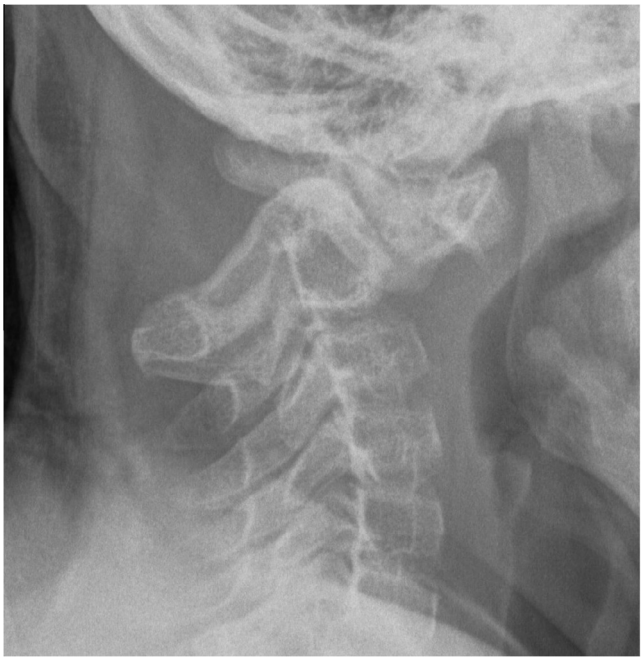

Fig. 1. Spinal radiograph shows severe anterior displacement of C1 and the tip of C2 in front of the base of the odontoid.

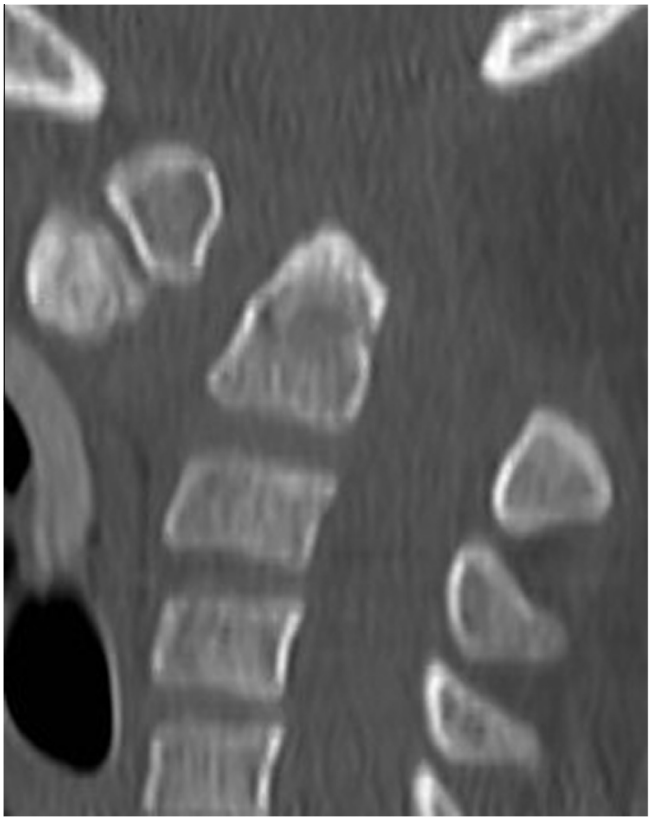

Fig. 2. Spinal sagittal CT scan shows complete anterior translation of the C1 arch and upper portion of the odontoid in front of the base of C2.

emergency room prior to her elective surgery date with abnormal breathing for 2 days prior. Her blood gas analysis revealed respiratory acidosis with a pH of 7.15 and carbon dioxide partial pressure of 104, with breathing on an inspired oxygen of 40% at a slow respiratory rate, between 8 to 10, and oxygen saturation of 88%; she had to be intubated. A chest radiograph and laboratory findings did not suggest an infectious etiology. After a thorough discussion with the pediatric intensive care unit team, it was decided that her hypoventilation was due to a central neurogenic cause, therefore, surgery was recommended.

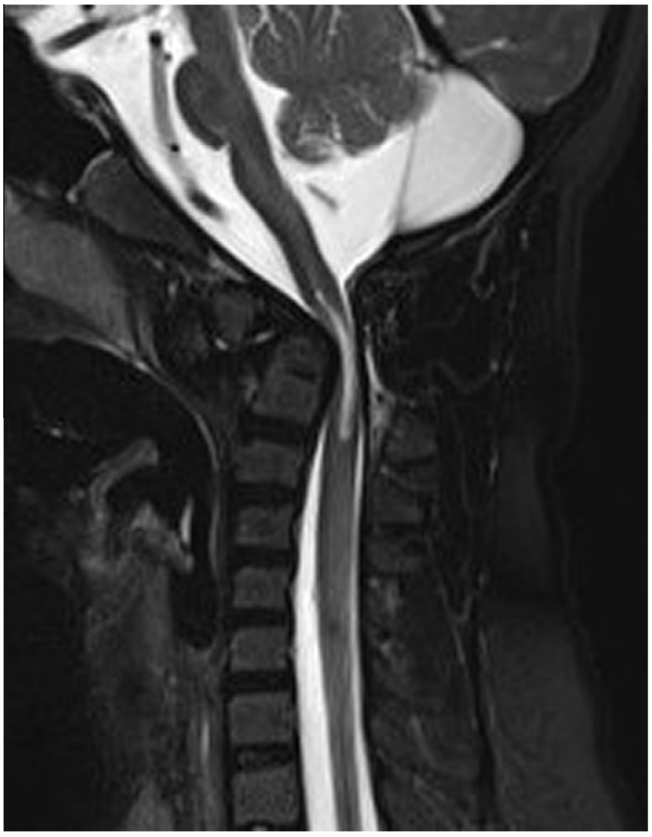

Fig. 3. Sagittal spinal T2-weighted MRI reveals marked narrowing of the underlying thecal sac and impingement of the cervicomedullary junction by severe anterior displacement of C1 and the tip of C2 in front of the base of the odontoid.

A halo was placed in a standard fashion and traction was applied. The girl's body weight was 24.5 kg (54 pounds). Serial weights were added and an adequate reduction was achieved with 15 pounds of traction within approximately 24 hours (Fig. 4).

2.5. Surgery

The girl was taken to the operating room and placed in the prone position on a Jackson spine table (Mizuho OSI, Union City, CA, USA). Traction was maintained at 15 pounds to achieve adequate reduction. A midline exposure was made from the occiput to C3. C1 lateral mass screws were placed using the technique described by Harms and Melcher [5], and C2 translaminar screws were placed using the technique described by Wright [6]. Complete reduction was achieved with distraction and hyperextension at C1 and C2. The rod was contoured and secured in a standard fashion. Fresh frozen allograft (30 cm^3) was placed over the decorticated fusion bed and a crosslink was used. Postoperative CT scans were then obtained (Fig. 5–7).

2.6. Postoperative course

The girl's postoperative course was uneventful and she was able to be weaned and extubated on postoperative day 1. Her progress has been followed for 32 months to date. CT scans obtained 1 year