Journal of Clinical Neuroscience xxx (2015) xxx–xxx

Contents lists available at ScienceDirect

Journal of Clinical Neuroscience

journal homepage: www.elsevier.com/locate/jocn

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

ARTICLE INFO

Article history:
Received 2 February 2015
Accepted 8 March 2015
Available online xxxx

Keywords:
Atlanotoaxial dislocation
C1–C2 dislocation
C1–C2 subluxation
Cerebral palsy
Craniovertebral junction
Os odontoideum
Respiratory distress

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).

http://dx.doi.org/10.1016/j.jocn.2015.03.004
0967-5868/

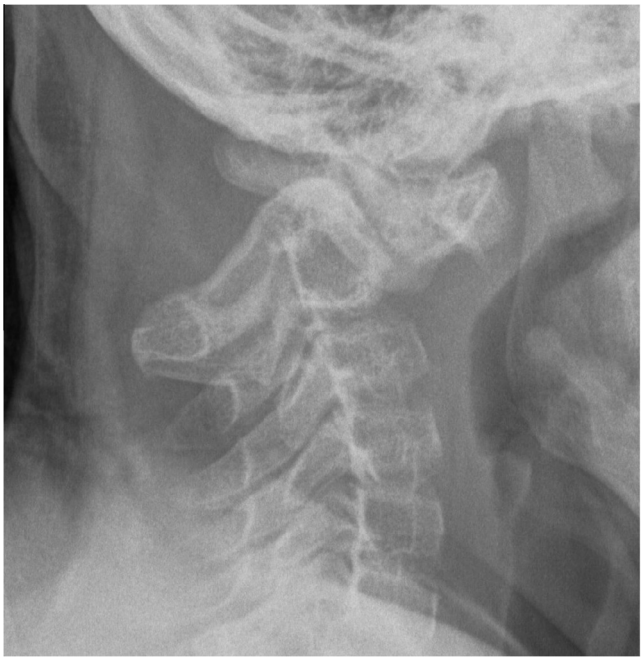

Fig. 1. Spinal radiograph shows severe anterior displacement of C1 and the tip of C2 in front of the base of the odontoid.

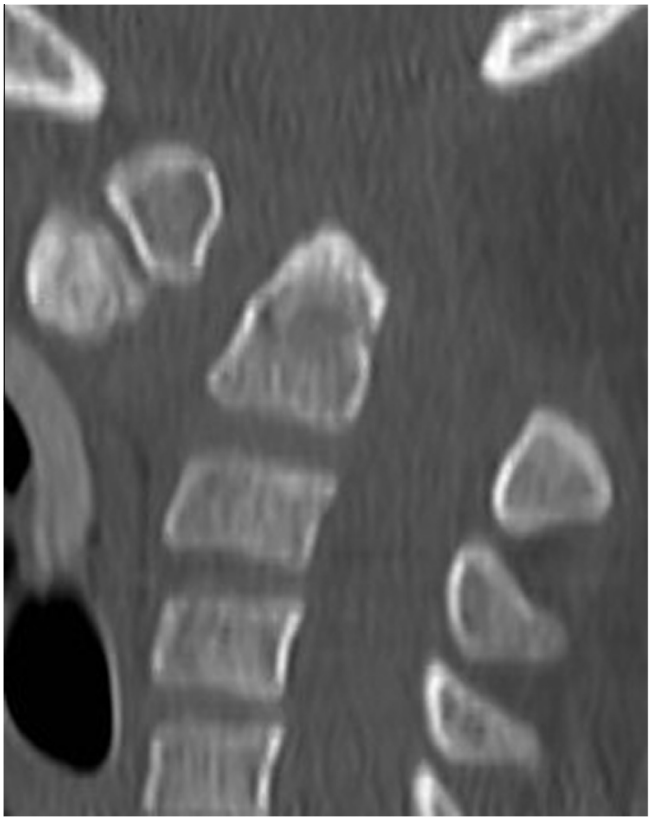

Fig. 2. Spinal sagittal CT scan shows complete anterior translation of the C1 arch and upper portion of the odontoid in front of the base of C2.

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.

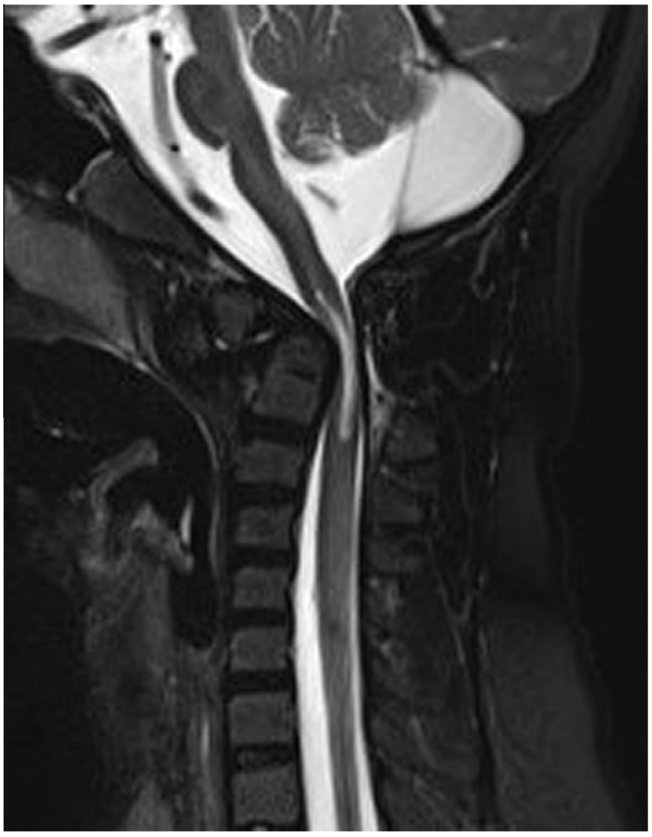

Fig. 3. Sagittal spinal T2-weighted MRI reveals marked narrowing of the underlying thecal sac and impingement of the cervicomedullary junction by severe anterior displacement of C1 and the tip of C2 in front of the base of the odontoid.

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

Please cite this article in press as: Akpolat YT et al. Treatment of os odontoideum in a patient with spastic quadriplegic cerebral palsy. J Clin Neurosci (2015), http://dx.doi.org/10.1016/j.jocn.2015.03.004

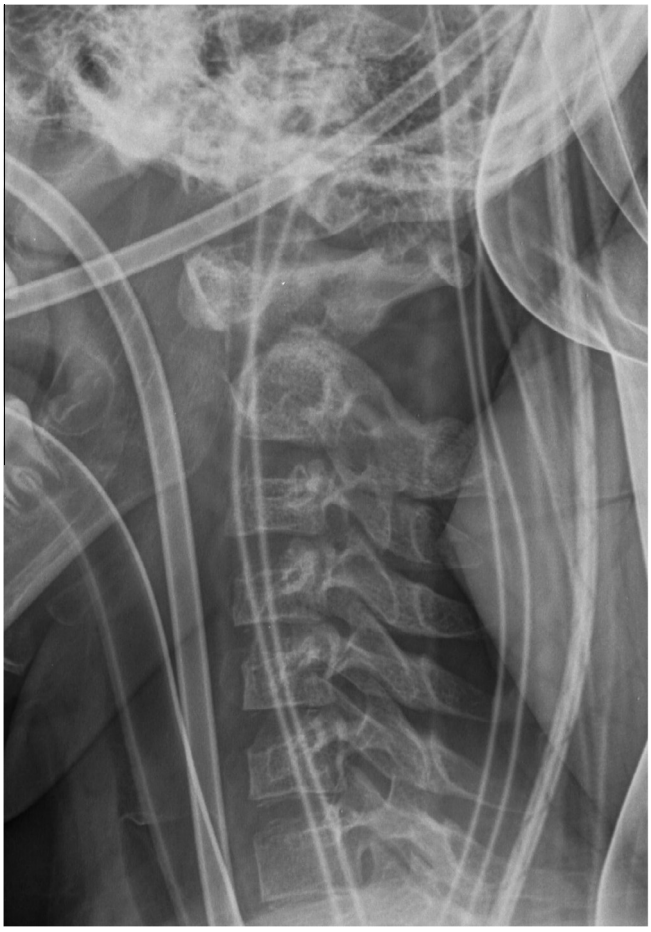

Fig. 4. Spinal radiograph of the 9-year-old spastic cerebral palsy patient with os odontoideum (weight 54 pounds) (Fig. 1–3) showing reduction with 15 pounds traction after 48 hours.

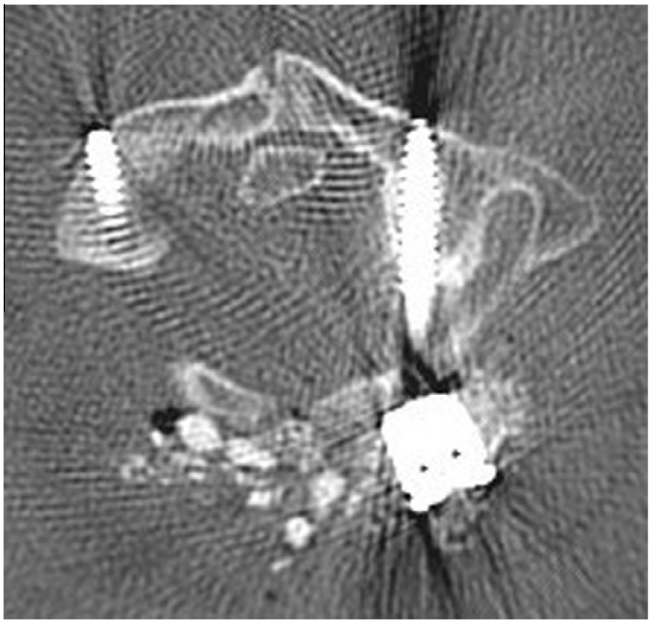

Fig. 5. Postoperative axial spinal CT scan showing the C1 bilateral lateral mass screws for treatment of severe anterior displacement of C1 and the tip of C2 in front of the base of the odontoid.

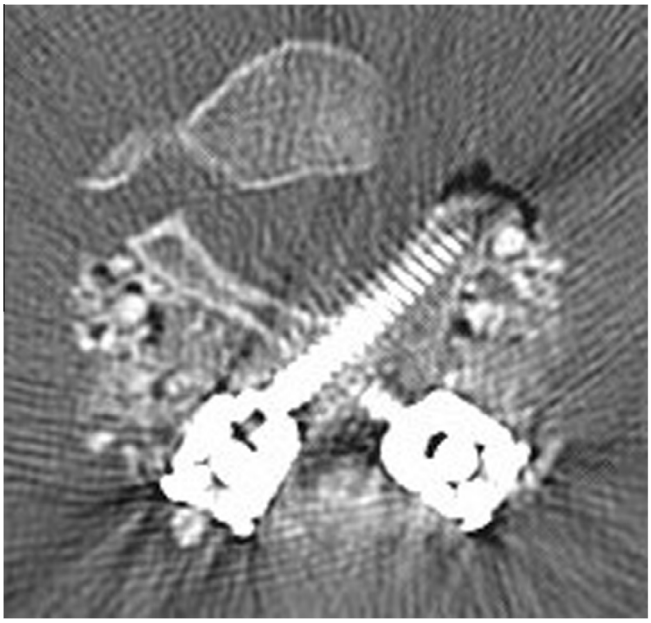

Fig. 6. Postoperative axial spinal CT scan showing the C2 translaminar screw for treatment of severe anterior displacement of C1 and the tip of C2 in front of the base of the odontoid.

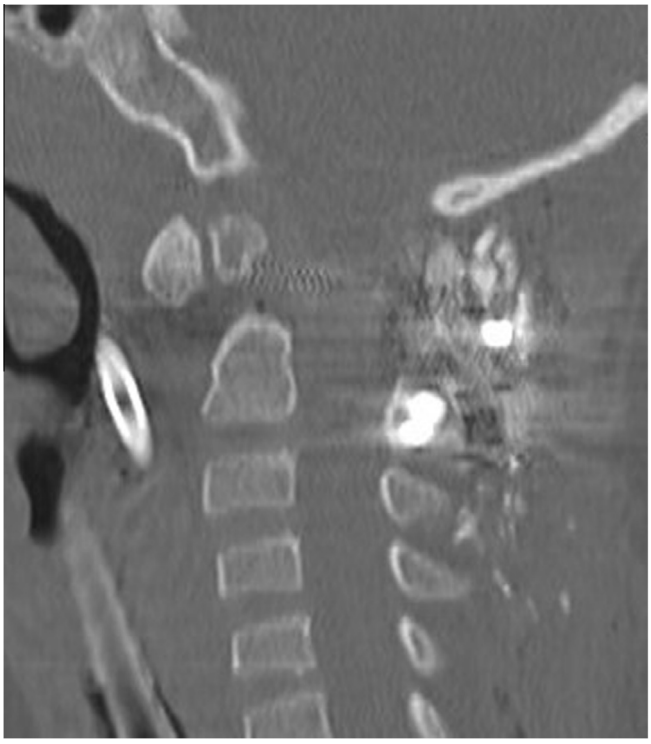

Fig. 7. Postoperative sagittal spinal CT scan showing adequate reduction and patency of the spinal canal.

after the initial surgery revealed adequate maintenance of reduction and patency of the spinal canal (Fig. 8).

3. Literature Review

The English language literature was searched from the year 1980 to present and we found five case reports on OO in cerebral

Please cite this article in press as: Akpolat YT et al. Treatment of os odontoideum in a patient with spastic quadriplegic cerebral palsy. J Clin Neurosci (2015), http://dx.doi.org/10.1016/j.jocn.2015.03.004

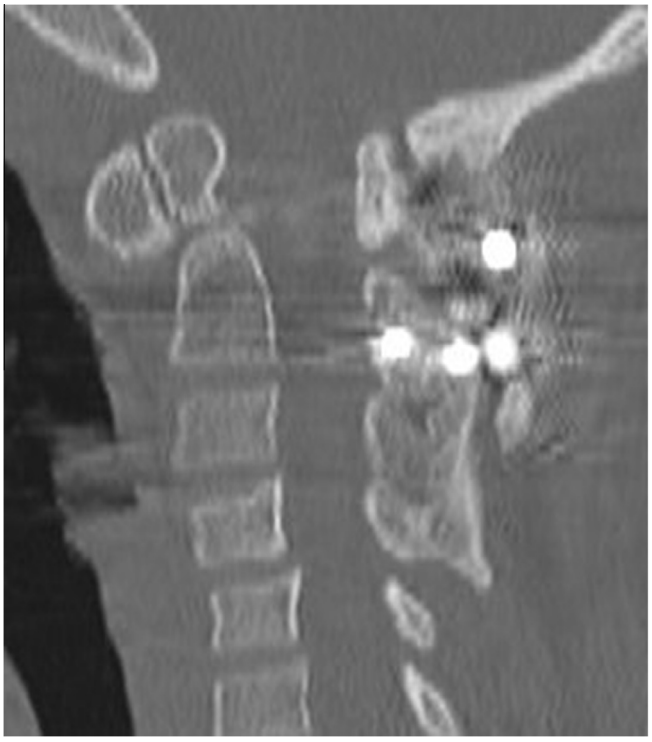

Fig. 8. CT scan 1 year postoperatively revealed adequate maintenance of reduction and patency of the spinal canal.

palsy patients (Table 1). Three patients were categorized as the spastic type, and two the dyskinetic type of cerebral palsy.

In the case series by Juhl et al. [7], two patients had OO with spastic type cerebral palsy. In this study, one patient, who had decompression alone, continued to have neurologic deterioration *versus* another patient who had both decompression and fusion and had resolution of symptoms including headache and a slight improvement in gait. The third patient was reported in the case study by Gigante et al. [8]. This patient was initially misdiagnosed as having a nonresectable cervicomedullary spinal cord tumor, but had a C1–C2 arthrodesis and neurologic improvement was achieved.

The other two reported patients with OO had the dyskinetic type of cerebral palsy and were treated nonsurgically. In the case study by Amess et al. [9], the patient had athetoid cerebral palsy with no spondylosis, congenital narrowing of the spinal canal or thickening of the ligamentum flavum. The proposed cause of OO was thought to be trauma secondary to rotational athetoid-dystonic neck movements. This patient was treated with steroids and continued to have progressive neurologic compromise. In the case study by Trabaca et al. [10], they described a dyskinetic cerebral palsy patient with an incidental finding of OO. This patient was treated nonsurgically and had stable neurologic function.

4. Discussion

The etiology of OO is greatly debated. Currently, the two predominate theories are congenital and traumatic etiologies [2,11]. Trauma is the more widely accepted theory, largely due to the work of Fielding et al. [12]. He proposed that an unrecognized fracture of the dens followed by contraction of the alar ligaments causes distraction of the fragment. The base of the dens, being more distal to the apical blood supply, then undergoes avascular necrosis [13]. This process leads to the formation of a smooth ossicle. Fielding's theory is further supported by a several case reports with a history of trauma associated with OO [14–17]. Additionally, a review of embryology does not support the congenital theory, which contends causation by a failure of fusion between the first and second spinal sclerotomes [2,11]. This failure would cause a separation below the level of the superior articular facets whereas OO is above that location.

Dystonic posturing causing excessive flexion, extension and rotation of the neck has been proposed as a mechanism for degenerative changes of the vertebrae leading to subsequent cervical radiculomyelopathy [18]. This is easily applicable to dyskinetic cerebral palsies. However, even with other types of cerebral palsy, as with the reported spastic cerebral palsy patient, it is most likely that OO is a result of unrecognized trauma. In our review, and including our patient, there are two dyskinetic cerebral palsy patients and four spastic cerebral palsy patients reported. In our patient, it is likely that trauma may have occurred but that the patient was unable to communicate her distress. Cerebral palsy is also associated with low bone mineral density for multifactorial reasons [3]. This suggests that our patient was vulnerable to fracture. Additionally, the base of the dens has a propensity to low bone mass and cortical thickness, even in normal individuals [2]. Patients with cerebral palsy of all types are frequently reported to suffer craniovertebral junction instability.

Another controversy in OO is the decision for surgical intervention. In patients with unstable OO or when neurologic changes are present, decompression and fixation is indicated [19]. There are times when stable OO is found incidentally on radiographs. In these patients, management with close follow-up for the next 5 years or preventative fusion is appropriate [2,20]. Stability is typically measured by comparing active flexion and extension radiographs [20]. There is no clear method to perform flexion and extension imaging on patients who are mentally challenged with such profound cerebral palsy and undertaking appropriate prophylactic surgery to stabilize a patient with OO has not been well established in the literature. In some patients, monitoring of stable OO leads to no neurological compromise. However, there are conflicting case reports of minor trauma necessitating emergent surgical intervention due to the changing neurological status of the patient. Looking specifically at patients with spastic cerebral palsy and OO, two prior reported patients suggest that posterior cervical fusion may lead to neurologic improvement, and one patient who had decompression alone continued to deteriorate. Our patient had resolution of her respiratory compromise immediately after surgical reduction and stabilization.

Lastly, an important aspect regarding this patient was the loss of respiratory drive as the presenting neurological deficit. Loss of respiratory drive was thought to have occurred due to cervicomedullary compression as a result of the instability [21]. In our review, Patient 4 had C1–C2 instability that was misdiagnosed as an intramedullary spinal cord tumor due to compression [8]. This is the only reported cerebral palsy and OO patient with a change in respiratory drive.

There are other case reports of OO with respiratory compromise but without cerebral palsy. In a report on OO [22], instability caused symptoms of sleep apnea and myelopathy. An MRI demonstrated atrophy of the spinal cord and intramedullary high signal intensity at the level of C1. A preoperative polysomnogram reported obstructive sleep apnea/hypopnea syndrome with a component of central sleep apnea syndrome. It was concluded that since it was the lower brain stem that was affected, apnea was due to narrowing of the airway as a result of subluxation. This conclusion can be disputed as the lower brain stem and ventral medulla are key sensors of rising CO_2 [23] and damage to these areas can lead to loss of respiratory drive. It is further stated in the case report that palsy of cranial nerves V, VII, IV, and X might have led to airway muscle collapse. This also seems unlikely as

Please cite this article in press as: Akpolat YT et al. Treatment of os odontoideum in a patient with spastic quadriplegic cerebral palsy. J Clin Neurosci (2015), http://dx.doi.org/10.1016/j.jocn.2015.03.004

Table 1
Cerebral palsy patients with os odontoideum: A review of the literature

Patient	1	2	3	4	5
Study	Juhl 1983 [7]	Juhl 1983 [7]	Amess 1998 [9]	Gigante 2011 [8]	Trabaca 2011 [10]
Age (years), Sex	33, M	6, F	12, M	8, F	12, M
CP	Spastic	Spastic	Dyskinetic	Spastic	Dyskinetic
Possible cause of OO	-	Infection	Traumatic	-	Traumatic
Neurological signs	Tetraplegia	CP-related, stable	Reduction in typical dystonic movements, difficulty swallowing, urinary and fecal incontinence	Progressive spastic quadriparesis	CP-related, stable
Years from onset	32	-	10	8	-
Intervention	Occipitocervical spondylodesis and decompression	Decompression	Steroid treatment	Posterior C1–C2 instrumented fusion with iliac crest autograft	No intervention
Results	Headache disappeared, gait slightly improved	Progression of symptoms until death at 11 years old	Muscle spasm unresponsive to drug therapy, increasing hypertonicity, flexion contractures, atrophy of the spinal cord at the level of C1–C2 (from MRI)	Improvement in tone, hyperreflexia and strength	Stable

- = not reported, CP = cerebral palsy, F = female, M = male, OO = os odontoideum.

apnea only occurring during sleep. Another case report of OO [24] reported similar loss of respiratory drive. In this report, myelopathy and shortness of breath were the presenting symptoms. MRI demonstrated similar cervicomedullary compression at the level of C1 and it was determined that this patient was suffering from central hypoventilation syndrome or Ondine's curse. Ondine's curse is characterized by periods of apnea but with preserved ability for inspiration on command. This patient's postoperative course was unique as conscious shortness of breath was relieved but sleep related periods of apnea persisted. The patient was reported to have no daytime issues with oxygenation but required mask ventilation for sleep. OO caused C1 compression and then was linked to respiratory failure due to lower brainstem and medullary dysfunction. This patient recovered from her loss of respiratory drive after decompression and posterior fixation and it is thought that her difficulties were most likely due to disruption of the respiratory pathways in the lower brainstem and not obstruction. These findings bring attention to respiratory failure as a possible fatal complication in patients with unstable OO.

Low functional demand cerebral palsy patients need urgent investigation of atlantoaxial instability when signs of decreased ventilation are present. The family's decisions regarding treatment of these patients can be difficult due to their low quality of life and the life saving measures of surgery [10]. Disease management decisions are made case-by-case in discussions with the patient (where possible) and their family.

5. Conclusion

Treatment decisions for unstable OO in patients with spastic quadriplegic cerebral palsy are difficult. Even if the finding of OO is incidental, unstable OO can cause mortality due to respiratory distress. This may serve as an important factor in decision making for both the clinician and patient's family.

Conflicts of Interest/Disclosures

The authors declare that they have no financial or other conflicts of interest in relation to this research and its publication.

References

[1] Giacomini C. Sull' esistenza dell''os odontoideum' nell' uomo. Gior Accad Med Torino 1886;49:24–8.
[2] Arvin B, Fournier-Gosselin MP, Fehlings MG. Os odontoideum: etiology and surgical management. Neurosurgery 2010;66:22–31.
[3] Henderson RC, Lark RK, Gurka MJ, et al. Bone density and metabolism in children and adolescents with moderate to severe cerebral palsy. Pediatrics 2002;110:e5.
[4] Rethlefsen SA, Ryan DD, Kay RM. Classification systems in cerebral palsy. Orthop Clin North Am 2010;41:457–67.
[5] Harms J, Melcher RP. Posterior C1–C2 fusion with polyaxial screw and rod fixation. Spine 2001;26:2467–71.
[6] Wright NM. Translaminar rigid screw fixation of the axis. Technical note. J Neurosurg Spine 2005;3:409–14.
[7] Juhl M, Seerup KK. Os odontoideum. A cause of atlanto-axial instability. Acta Orthop Scand 1983;54:113–8.
[8] Gigante PR, Feldstein NA, Anderson RC. C1–2 instability from os odontoideum mimicking intramedullary spinal cord tumor. J Neurosurg Pediatr 2011;8:363–6.
[9] Amess P, Chong WK, Kirkham FJ. Acquired spinal cord lesion associated with os odontoideum causing deterioration in dystonic cerebral palsy: case report and review of the literature. Dev Med Child Neurol 1998;40:195–8.
[10] Trabacca A, Dicuonzo F, Gennaro L, et al. Os odontoideum as a rare but possible complication in children with dyskinetic cerebral palsy: a clinical and neuroradiologic study. J Child Neurol 2011;26:1021–5.
[11] Sankar WN, Wills BP, Dormans JP, et al. Os odontoideum revisited: the case for a multifactorial etiology. Spine (Phila Pa 1976) 2006;31:979–84.
[12] Fielding JW, Hensinger RN, Hawkins RJ. Os odontoideum. J Bone Joint Surg Am 1980;62:376–83.
[13] Sakaida H, Waga S, Kojima T, et al. Os odontoideum associated with hypertrophic ossiculum terminale. Case report. J Neurosurg 2001;94:140–4.
[14] Lowry DW, Pollack IF, Clyde B, et al. Upper cervical spine fusion in the pediatric population. J Neurosurg 1997;87:671–6.
[15] Zhang Z, Zhou Y, Wang J, et al. Acute traumatic cervical cord injury in patients with os odontoideum. J Clin Neurosci 2010;17:1289–93.
[16] Giussani C, Roux FE, Guerra P, et al. Severely symptomatic craniovertebral junction abnormalities in children: long-term reliability of aggressive management. Pediatr Neurosurg 2009;45:29–36.
[17] Qureshi MA, Afzal W, Malik AS, et al. Os-odontoideum leading to atlanto-axial instability–report of surgery in four cases. J Pak Med Assoc 2008;58:640–2.
[18] Konrad C, Vollmer-Haase J, Anneken K, et al. Orthopedic and neurological complications of cervical dystonia–review of the literature. Acta Neurol Scand 2004;109:369–73.
[19] Yoshida G, Kamiya M, Yoshihara H, et al. Subaxial sagittal alignment and adjacent-segment degeneration after atlantoaxial fixation performed using C-1 lateral mass and C-2 pedicle screws or transarticular screws. J Neurosurg Spine 2010;13:443–50.
[20] Visocchi M, Di Rocco C. Os odontoideum syndrome: pathogenesis, clinical patterns and indication for surgical strategies in childhood. Adv Tech Stand Neurosurg 2014;40:273–93.
[21] Bach CM, Arbab D, Thaler M. Treatment strategies for severe C1C2 luxation due to congenital os odontoideum causing tetraplegia. Eur Spine J 2013;22:29–35.
[22] Kawaguchi Y, Iida M, Seki S, et al. Os odontoideum with cervical mylopathy due to posterior subluxation of C1 presenting sleep apnea. J Orthop Sci 2011;16:329–33.
[23] Spyer KM, Gourine AV. Chemosensory pathways in the brainstem controlling cardiorespiratory activity. Philos Trans R Soc Lond B Biol Sci 2009;364:2603–10.
[24] Campbell E, Brown J. Case report of os odontoideum causing Ondine's curse. Br J Neurosurg 2013;27:836–7.

Please cite this article in press as: Akpolat YT et al. Treatment of os odontoideum in a patient with spastic quadriplegic cerebral palsy. J Clin Neurosci (2015), http://dx.doi.org/10.1016/j.jocn.2015.03.004