

Imaging Findings of Chronic Subluxation of the Os Odontoideum and Cervical Myelopathy in a Child with Beare-Stevenson Cutis Gyrata Syndrome after Surgery to the Head and Neck

Chow Wei Too,¹MBBS, Phua Hwee Tang,¹MBBS, M Med (Diag Radiol), FRCR

Abstract

Introduction: Although uncommon, fractures of the os odontoideum are known to occur in children under 7 years old, following acute trauma. **Clinical Picture:** We report a case of chronic subluxation of the os odontoideum resulting in cervical myelopathy in a child with Beare-Stevenson cutis gyrata syndrome after surgery to the head and neck. **Treatment and Outcome:** The patient was initially put in a Halo vest, following which occipital cervical fusion was performed. **Conclusion:** Subluxations and fractures at the odontoid synchondrosis are rare but should be anticipated in young children with risk factors for instability of the cervical spine.

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Key words: Myelopathy, Os odontoideum, Subluxation, Synchondrosis

Introduction

Fractures of the odontoid typically involve the synchondrosis between the odontoid process and body of the axis in the paediatric age group. However, to our knowledge, a chronic subluxation of the os odontoideum resulting in cervical myelopathy has yet to be described in a child with the Beare-Stevenson cutis gyrata syndrome.

Case Report

The patient is a known case of Beare-Stevenson cutis gyrata syndrome. Beare-Stevenson cutis gyrata syndrome is a rare autosomal dominant disorder and consists of cutis gyrate (corrugated skin furrows), acanthosis nigricans, skin tags, craniofacial anomalies (particularly craniosynostosis and ear defects), anogenital anomalies and a prominent umbilical stump.¹ Chiari 1 malformation has been described in a child with such a syndrome.² Mutations in the FGFR2 gene, which encodes for the fibroblast growth factor receptor 2, has been described in some patients with Beare-Stevenson cutis gyrate syndrome and is thought to be the cause of premature calvarial fusion.³

The patient was delivered via lower section caesarean section for antenatally detected craniosynostosis. She had a ventriculoperitoneal shunt insertion on day 2 of life for congenital hydrocephalus. On day 13 of life, she

had a permanent tracheostomy performed for choanal atresia and has since remained on home bilevel positive pressure (BIPAP) ventilation. At 2 months of life, she had her first posterior cranial fossa decompression for Chiari 1 malformation. She would have a total of 3 posterior cranial fossa decompressions (including C1 laminectomy) by the time she presented with her current problem. Other procedures performed include: Fronto-occipital advancement and Le Fort III osteotomy with anterior cranium remodelling for turribrachyencephaly at 6 months and 2 years of age, respectively; laposcopic fundoplication and button gastrostomy for severe gastro-esophageal reflux at 2 years of age. She is also on long-term antibiotics for right grade 3 vesicoureteric reflux.

She presented at 6 years and 2 months of age with increasing respiratory distress, requiring increased day time BIPAP requirements. She also had cyanotic episodes which required bagging through the tracheostomy. During the initial ward stay, her blood pressure was found to be elevated. Magnetic resonance imaging (MRI) of the brain was performed to look for a central cause for the hypertension after a CT brain scan revealed no ventriculomegaly.

Imaging Findings

MRI showed kinking of the cervicomedullary junction

¹ Department of Diagnostic Imaging, KK Women's and Children's Hospital, Singapore

Address for Correspondence: Dr Tang Phua Hwee, Department of Diagnostic Imaging, KK Women's and Children's Hospital, 100 Bukit Timah Road, Singapore 229899.

Email: phuahwee@yahoo.com

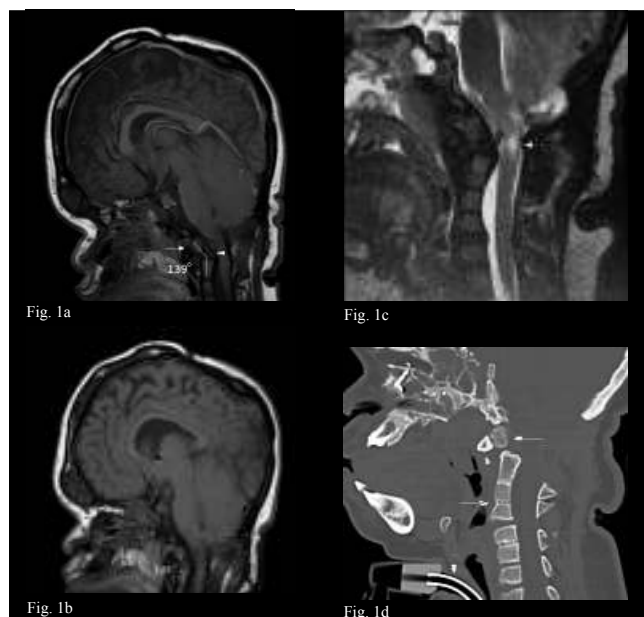


Fig. 1. Imaging at presentation at 6 years and 2 months of age. Sagittal T1 image of the head (Fig. 1a) shows anterior subluxation of the anteriorly angulated os odontoideum (arrow) at the synchondrosis (arrowhead) which improved with head repositioning (Fig. 1b). Sagittal T2 image of the upper cervical spine (Fig. 1c) shows high T2 signal in the cervicomedullary junction (broken arrow) due to myelopathy. CT of the cervical spine (Fig. 1d) shows an anteriorly angulated subluxed os odontoideum (double arrows). Note congenital fusion of the anterior and posterior bony elements of C2 and C3 (line arrow) and the airway maintained by a tracheostomy (double broken arrows).

and subluxation of the os odontoideum at the synchondrosis. The subluxation improved after head repositioning but the cervicomedullary kink persisted. There was associated high T2 signal in the cervicomedullary junction, which was attributed to myelopathy. The C1 lamina was deficient, as a result of C1 laminectomy. Review of previous imaging showed gradual subluxation of the odontoid synchondrosis since the MRI done at 5 years 3 months of age.

Treatment and Outcome

The patient was initially put in a Halo vest, following which occipital cervical fusion was performed as a definitive treatment. Blood pressure improved following these interventions.

Discussion

Although rare, fractures through the synchondrosis of the odontoid process have been described after trauma and is limited to children under 7 years of age where the synchondrosis has not fused.^{4,5} If there is associated displacement of the odontoid peg, the displacement is anterior in more than 90% of the cases.⁶

The fulcrum/pivot point in a child is located higher in the cervical spine compared to adults. The synchondrosis is the weakest area in this region as the head and C1 move together

as a unit with respect to C2 during trauma.⁴ Biomechanical analysis has demonstrated the shearing forces encountered by the odontoid synchondrosis at the point of maximal head flexion during trauma.⁵ Other factors that predispose to odontoid synchondrosis injury in the young child would be the under-developed neck and paravertebral muscles, physiological ligamentous laxity and horizontal orientation of the facet joints. Cord injury associated with such fractures occurs at the cervicothoracic junction⁶ and is attributed to the stretching of the cord.

Os odontoideum was formerly thought to be due to a congenital failure of fusion of the odontoid dens to the axis, now believed to be due to a fracture of the odontoid synchondrosis before its fusion at age 7. Ossiculum terminale, on the other hand, is thought to be due to a failure of the secondary ossification of the dens to fuse with the base of the odontoid. A persistent ossiculum terminale is much smaller than an os odontoideum, located at the tip of the dens and not associated with significant instability.⁷

Kyphotic deformity of the cervical spine post-laminectomy has been described, with risk factors being: multi-level laminectomy,⁸ laminectomy of C2 or lower, operations complicated by local wound infection resulting in weakness of the paraspinal muscles.⁹ These spinal deformities are more likely the younger the patient, probably due to the increased elasticity of the ligaments in children. It is thought however that laminectomies involving the occiput, C1 and C2 are unlikely to result in spinal deformities because of the unique structure of these vertebral bodies and their ligaments. The facet joints are situated more anteriorly at C1/2 and thus unlikely to be disrupted during laminectomy unlike the lower levels.

As C1 laminectomy in combination with posterior fossa craniotomy in children has not been shown to lead to cervical subluxation⁹ and there is no history of trauma in our patient, we believe that the chronic subluxation of the os odontoideum in our patient was due to a combination of 2 factors. The patient's large head relative to her neck as well as weakened posterior cervical muscles and ligaments from the multiple surgeries may have resulted in increased flexion force at C1/2 level which the weakened posterior ligamentous complex (ligamentum flavum, interspinous and supraspinous ligaments, posterior paraspinal muscles) could not counteract. The focal myelopathy seen at the cervicomedullary junction is probably related to chronic repetitive impingement given its location.

Management of subluxation of the os odontoideum depends on whether it can be reduced with repositioning or cervical traction. If such measures are successful, they should be carried out promptly to stabilise the spine. If these measures are unsuccessful, decompressive surgery and subsequent fixation is required.¹⁰

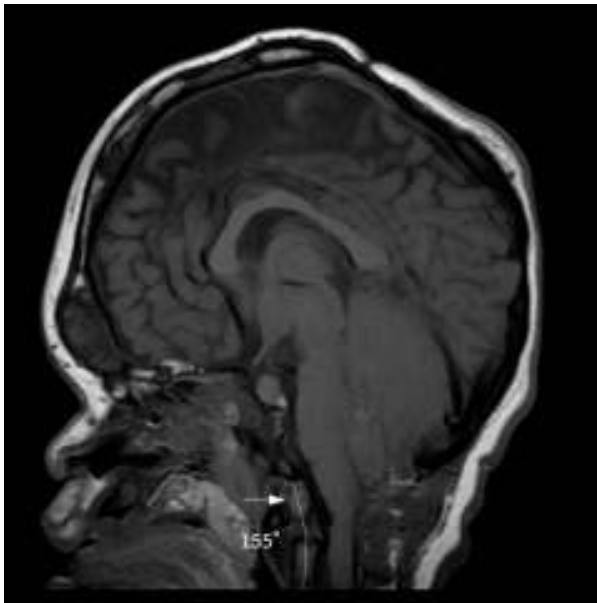


Fig. 2. Earlier MRI performed at 5 years and 3 months of age shows a slightly anteriorly angulated and minimally anteriorly subluxed os odontoideum (arrow).

Conclusion

Subluxations and fractures at the odontoid synchondrosis are rare but should be anticipated in young children with risk factors for instability of the cervical spine. This is the first report of a non-traumatic chronic subluxation of the os odontoideum resulting in cervical myelopathy in a child

with Beare-Stevenson cutis gyrata syndrome after surgery to the head and neck.

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