Case Report

Case of Atlantoaxial Dislocation with Assimilated C1, Absent Posterior C2, Butterfly C3, and Fused Subaxial Cervical Spine: Management Dilemma with Multiple Segmentation and Formation Defects

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Segmentation defects are often seen with congenital atlantoaxial dislocation (AAD) though an associated absence of posterior arch of C2 and butterfly C3 is rare. Apart from rarity, the combination of formation and segmentation defects adds to the management dilemma. We report a case of AAD with assimilated atlas, absent C2 posterior arch, C3 butterfly vertebra with floating posterior elements, and fused C4–C6. The child was managed by C1–C2 fusion alone with immediate symptomatic improvement. The presence of formation defects such as adjacent butterfly vertebra and absent posterior elements does not alter the management of AAD. Fusing the C1–C2 joints appears to be a balanced approach.

Keywords: Absent posterior C2, atlantoaxial dislocation, butterfly vertebra

INTRODUCTION

Segmentation defects of cervical spine are often seen in cases of congenital atlantoaxial dislocation (CAAD). The most common associated anomalies are assimilated atlas and C2–C3 fusion. Such dislocations were often treated by fusing occipital squama to C2–C3.[1] Rarely, C2 posterior elements may be absent and fusion for atlantoaxial dislocation (AAD) in these cases necessitates inclusion of lower segments, thereby restricting neck movements further.[1–4] The presence of adjacent level butterfly body complicates it further. With the focus shifting to fusing C1 and C2 joints rather than posterior elements, inclusion of lower segments can be avoided even in such cases. The presence of C3 butterfly vertebra is incidental and may not add to the instability.

We report a case of CAAD with assimilated atlas, absence of posterior arch of axis, butterfly C3 vertebrae, floating C3 posterior arch, and fused subaxial cervical spine combined with anomalous course of vertebral arteries (VAs). The AAD was treated by C1–C2 fusion and the rationale for such a treatment is discussed.

CASE REPORT

A 14-year-old boy presented with neck pain, progressive neck tilt, and spastic quadripareisis of 1 year duration. He had difficulty in swallowing and was bed ridden for 4 months. Examination revealed low hairline, torticollis, and severe spasticity with grade 3/5 power. The modified Japanese Orthopaedic Association score was 9/17 and breath hold time was 8 s.

Dynamic X-ray of cervical spine revealed AAD, absent spinous process of axis, and fused lower cervical spine. Computed tomography (CT) with three-dimensional reconstruction done showed occipitalization of atlas. Posterior arch of axis and transverse foramen were absent, C3 body was split into halves (butterfly vertebrae), and the posterior elements of C3 were not connected to the bifid body (floating). They were connected to the C4 facet alone. C4–C6 vertebral bodies were fused. C1–C2 joints were oblique [Figure 1].[5] CT angiography revealed anomalous course of vertebral artery.[6] VAs were skirting along lateral surface of C2 body and turned posteromedially just short of superior facets of C2.

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The patient was put on Crutchfield’s tong traction starting with 7%–8% of body weight increased over a period of 24 h. Lateral cervical radiographs showed distraction with partial reduction. The patient was operated through midline posterior incision. Inferior surface of C1 arch traced and dissected to identify C1–C2 joints. Bilateral C2 root ganglia were transected to obtain a panoramic view of the C1–C2 joints. The anomalous VA was addressed appropriately, by dissecting and safeguarding it.\(^6\) C1–C2 joints were opened and drilled comprehensively to make them flat in sagittal and coronal plane.\(^7\) Spacers were inserted with bony chips taken from rib graft. C1 screws were inserted into lateral masses and C2 screws were directly inserted into C2 body just below C2 facets. Further manipulation of C1–C2 joints were manipulated using rod holder and loosely fastened rod to realign facets.\(^8\) The arteries were protected while drilling the C1–C2 joints and inserting the screws.

Postoperatively, the patient improved neurologically with immediate relief from spasticity. Immediate postoperative scan revealed good reduction of the dislocation and was discharged after a week [Figure 2]. In the 2nd postoperative week, the patient came back with high-grade fever and tachypnea. Although his improved neurological status was maintained, the chest X-ray showed right lower lung showing pneumonic patch [Figure 3]. Repeat CT scan was obtained showing no construct failure. Unfortunately, the patient succumbed to Type 1 respiratory failure.

**DISCUSSION**

The absence of posterior elements of axis is rare. The posterior arch of axis develops from the second spinal sclerotome. The defect in the posterior elements of the axis may be caused by the failure of the extension of the chondrification centers in the posterior arch, or by the failure of the ossification process. Formation of the posterior arch goes through stages of precartilage, chondrification, and ossification.\(^9\) At 8\(^{th}\) week of embryogenesis, the neural arches of the axis extend laterally from the centrum and also form a complete or incomplete foramen transversarium.\(^10\) The neural arches proceed directly backward to a variable extent to form the pedicle, articular process, and the lamina. A defect in various stages leads to variety of anomalies. Defective migration in early embryogenesis leads to complete absence of posterior arch or bifid or floating arch. Defective ossification leads to cartilaginous arch. Most cases with absent posterior arch of cervical spine in radiography have cartilaginous arch rather complete absence which was noted intraoperatively.\(^2-4\) Complete absence of the posterior elements of axis including absence of transverse foramen and inferior lateral masses as in our case indicates the failure of dorsal migration of cells from the second spinal sclerotome.

Anomalies of vertebral body can be defects pertaining to formation (partial or complete) or those related to failure of segmentation. There are three ossification centers responsible for completeness of a vertebra. A central one for body and two lateral centers for neural arches. The failure of the lateral centers to join with the central one possibly leads to floating arch. The intercalation of central ossification center or a sagittal cleft caused by the persistence of the notochord gives rise to a butterfly vertebra.\(^10\)
Abnormal PAX1 expression leads to failure of segmentation giving rise to fused vertebrae. A combination of formation defects and segmentation defects can coexist. However, the combination of absent C2 posterior arch, with C3 butterfly body and floating C3 posterior elements along with occipitalized C1 and C4–C6 fusion, is rare.

The management of CAAD would remain C1–C2 fusion despite associated other complex anomalies. In the past, posterior elements of C2–C3 formed an integral part of the construct. Currently, the focus is on fusing the C1–C2 joints and absence of C2 posterior arch does not preclude this. In addition, the normal weight bearing shifts from laterally placed O-C1–C2 joints to midline vertebral bodies in subaxial cervical spine. The posterior elements of cervical spine have little role in weight bearing. In cases of butterfly vertebra, the two parts of body were connected by cartilage and prevent splaying. This possibly transmits the weight efficiently to the normal vertebral bodies below. Most often, the butterfly vertebra are asymptomatic and do not require treatment. This is well illustrated in a reported case of Larsen syndrome with AAD, where the C3–C4 bodies were hypoplastic with kyphotic deformity. The hypoplastic C3–C4 vertebrae were not treated and the patient improved with treatment of AAD alone.

These cases helped us rationalize our management to a limited C1–C2 fusion.