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Case Report

Absent posterior element of axis vertebra with myelopathy treated with C2, C3, and C4 arthrodesis: A case report and review of literature

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ABSTRACT

A congenital abnormality of the posterior element of the axis (C2) vertebra is extremely unusual, with just a few occurrences documented in the literature. The majority of individuals have no symptoms and are discovered by chance on plain radiography for neck discomfort, radiculopathy, or trauma. We describe a case of a 13-year-old boy who was born without the posterior element of the C2 vertebrae, resulting in C2-C3 spinal instability and compressive myelopathy.

Keywords: Absent C2 posterior arch, Congenital anomaly of C2, Craniovertebral junction anomaly, Computed tomography craniovertebral junction angiography, 3D reconstruction, Atlantoaxial instability

INTRODUCTION

Congenital anomaly of the posterior element of the axis (C2) vertebra is an uncommon condition, occurring in 0.15% of the general population.[1] The axis vertebra has a complex development. C2 vertebrae grow in three stages from the second spinal sclerotome. Pre-cartilage, chondrification, and ossification are the three stages of cartilage formation. Chondrification of the posterior arch begins at the pedicle in the 6th week of development and continues till the midline in the 4th month. There are four primary ossification centers: Two for the neural arches, one for the body, and one for the odontoid process. It is also characterized by four secondary ossification centers – one for the body, one each for transverse processes, and one for the tip of the spinous process. Failure of these ossification centers during development leads to congenital anomalies of the axis vertebra. The lack of C2 vertebral posterior elements is caused by dysregulated gene expression, improper tissue contact, or cellular migration and proliferation during development. Most of the patients are asymptomatic but some may present with cervical myelopathy or simple neck pain. We here present a case of a 13-year-old male with congenital absence of the posterior element of C2 vertebrae with C2-C3 spinal instability with compressive myelopathy.

CASE REPORT

A 13-year-old boy patient complained of increasingly worsening walking problems with the left foot drop during the previous year. There was no history of trauma. There was no history of neck pain. Mild tenderness over the back of the neck at C2-C3 levels was seen. His neurological examination revealed weakness (Medical Research Council's grading, MRC grade – 4/5) of the left lower limbs with the left foot drop. Bilateral Lasegue tests were negative. Deep tendon reflexes across major joints were brisk. Bilateral plantar reflex was extensor. Bilateral Hoffmann signs were positive. Bilateral ankle clonus and patellar clonus were present and ill sustained. No involvement of bladder and bowel was seen. Sensory modalities were intact. The breath-holding time was 42 s. The modified Japanese Orthopaedic Association score for the patient was 16/18.

Plain radiographs, magnetic resonance imaging (MRI), and computed tomography (CT) craniovertebral junction (CVJ) with cervical spine revealed absent posterior arch of C2 vertebra. In ultrasound abdomen, MRI of the lumbar spine with the thoracic spine was done to rule out any other congenital anomaly. Electromyography showed normal findings. MRI CVJ showed major compression at the C2-C3 vertebral level causing myelopathic changes in the cord which was correlated to the clinical complaints and examination of the patient [Figure 1a]. The absent posterior elements of the axis vertebra can be seen clearly on CT scan axial views [Figure 1b]. CT CVJ revealed listhesis (Meyerding Grade I) at the C2-C3 vertebra with the absent posterior arch of the C2 vertebra. There was no evidence of C2 and C3 vertebral

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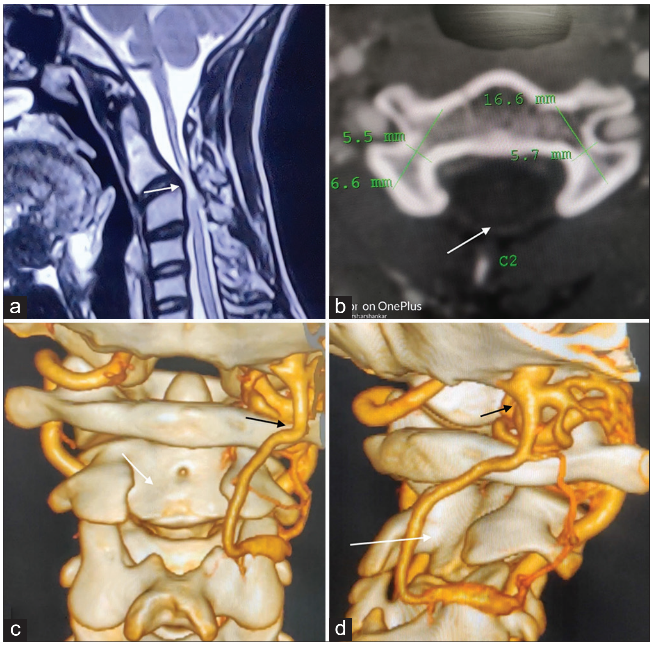

Figure 1: (a) Pre-operative MRI cervical spine showing compressive myelopathy at the craniovertebral junction with (b) pre-operative computed tomography (CT) cervical spine showing absent axis (C2) vertebral posterior elements shown with white arrows. (c) CT craniovertebral junction with angiography of neck vessels with 3D reconstruction showing absent posterior C2 elements (shown with white arrows). (d) An aberrant bunch of vessels (shown with black arrows) on the right C1-C2 vertebral junction with origin from the right vertebral artery at C3 vertebra and ending at C1-C2 vascular bunch.

fracture on the radiograph. The CT angiography of the neck vessels with 3D reconstruction with special emphasis on vertebral artery showed an aberrant bunch of vessels on the right C1-C2 vertebral junction with origin from the right vertebral artery at C3 vertebral level and ending at C1-C2 vertebral vascular bunch [Figure 1c and d]. The left vertebral artery had a normal course.

The patient was taken for surgery. Pre-operative positioning was as per standard guidelines. Fluoroscopic guidance was used for surgery. The C2-C3 vertebral joint was partially reduced intraoperatively. Bilateral C2 vertebra pedicle and C3 and C4 vertebra lateral mass were fixed with titanium screws and rods system [Figure 2a-d]. Onlay autologous bone grafts were placed between C2, C3, and C4 vertebral lamina. The post-operative CT CVJ revealed an adequate decrease in C2-C3 displaced joints as well as an increase in effective canal diameter at the C3 body level [Figure 2e and 2f]. The patient had significant improvement in his symptoms and was discharged with a hard cervical collar for 3 months. On a 6-month follow-up, the patient was significantly relieved of the symptoms. Informed consent was taken from the patient to use his information and imaging for this paper.

DISCUSSION

The development of the axis (C2) vertebra is complicated. The cartilaginous arch ossifies by 3–4 years of age.[2] The posterior elements develop in the pre-cartilage stage and failure at this stage leads to absent posterior elements and facets which, in turn, cause instability. O'Rahilly *et al*. in their study of nine embryos found that by Stage 23 of embryogenesis (8 post-ovulatory weeks), the neural arches of the C2 vertebra extended laterally from the centrum and formed a complete or incomplete foramen transversarium. During this stage, neural arches produce the pedicle, articular facets, and a portion of our lamina.[3]

The complete absence of bony and cartilaginous components of posterior elements signifies early defects during embryogenesis.[3] Senoglu *et al*. in their study on 1354 CT scans and cadavers found 3.35% of the absent posterior arch of the atlas.

Most of the patients are asymptomatic. It is commonly an incidental finding on plain radiograph for neck pain or radiculopathy or trauma. Instability can cause headaches, neck discomfort, paresis, and other symptoms. The C3 vertebra's hypertrophied spinous process provides a compensatory mechanism for supporting the interspinous ligament and paraspinal muscles, which are typically linked to the axis vertebra's spinous process.[4] Symptoms and neurological deficit are likely to be due to subtle instability-related microtrauma rather than spinal cord compression.[5]

The anomaly can be diagnosed on a plain radiograph but confirmed on a CT scan. MRI can be used to rule out other potential diagnoses. Even when there is no radiological indication of instability, direct physical assessment of joints and bone can detect instability.[5] CT angiography with the 3D reconstruction of the neck vessels provides a detailed course of bilateral vertebral arteries around the CVJ. This helps in planning surgery and avoiding complications. We, at our institute, routinely do a pre-operative CT angiography of neck vessels in patients with CVJ anomalies planned for surgery.

A critical objective of therapy is to stabilize the area rather than decompress it.[6] Sharif *et al*. in their study concluded that asymptomatic patient needs conservative treatment with rest, analgesia, wearing a collar, muscle relaxant, and physical therapy. Surgery, on the other hand, is the best option for a patient with an unstable condition or symptomatic spinal cord compression.[7] Surgery is the preferred therapy for unstable situations, painful lesions, cervical spine injuries, intractable long-term neck discomfort, and modest neurologic deficits caused by neck trauma.[1,8] We chose to operate on our patient as he had compressive symptoms and C2-C3 vertebral instability.

In the past, C2-C3 vertebral fusion was used but now C1-C2 vertebral fusion is more popular for the absent

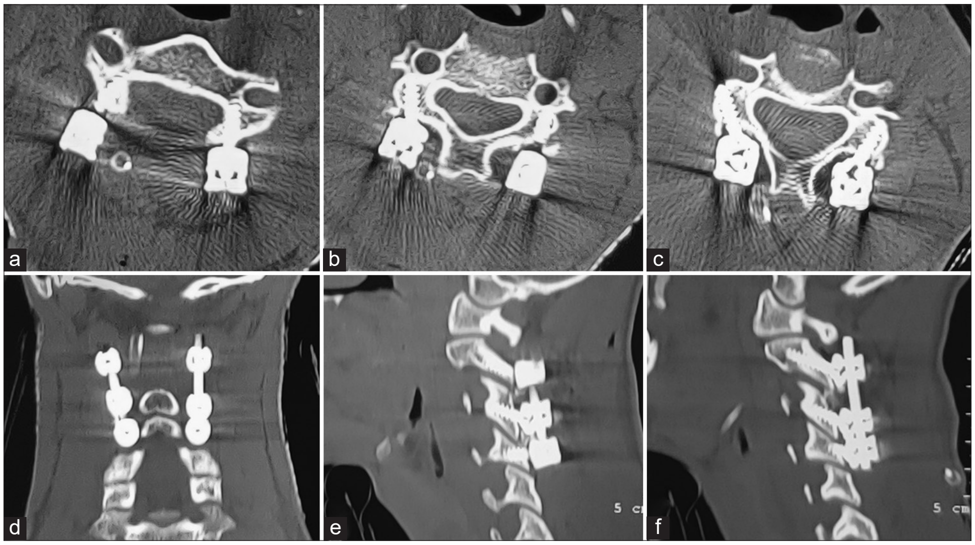

Figure 2: Post-operative CT craniovertebral junction showing axial views at (a) C2, (b) C3, and (c) C4 vertebra with (d) coronal view. Post-operative CT craniovertebral junction showing sagittal views at the (e) right pedicles and (f) left pedicles with increased effective canal diameter can be seen at the level of the C3 vertebral body.

posterior arch of C2 vertebra.[9] Trivedi *et al*. recommend the use of occiput-C3 vertebra fusion for absent C2 vertebra posterior elements seeing the good outcome in their patient. Kataria *et al*. used C1 vertebra lateral mass and C3 vertebra pedicle screw and rod fusion. We here performed a bilateral C2 vertebra pedicle and C3 and C4 vertebra lateral mass fixation with titanium screws and rods system with onlay autologous bone graft placed between C2, C3, and C4 lamina. Although the long-term outcome of this construct is still to be evaluated in coming times, in theory, it proves to be better construct than occipit-C3 fixation as the majority of the neck movements occur at C1-C2 joints. Our post-operative results were satisfactory with optimal reduction of C2-C3 vertebral instability and clinical improvement in our patient.

CONCLUSION

The absent posterior element of the axis vertebra is a very rare congenital anomaly. Most of the patients are asymptomatic. Diagnosis is made incidentally on a plain radiograph for mild neck pain or trauma. CT angiography with the 3D reconstruction of neck vessels should be routinely done in patients planned for surgery. This helps in avoiding complications. Surgery is the treatment of choice in unstable lesions or symptomatic patients whereas conservative management is advised for those who are asymptomatic. Very few cases have been described in the past with mostly good post-operative results. As a result, recommending a unified guideline in the treatment of absent posterior elements of the C2 vertebra is still debatable and requires additional research.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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