Case Report

Congenital defect of the posterior arch of C1: a case report

Bilal B. Butt, Paul Gagnet, Rakesh Patel, Ilyas Aleem

Department of Orthopaedic Surgery, University of Michigan, Ann Arbor, MI, USA

Correspondence to: Ilyas Aleem, MD, MS, FRCSC. Department of Orthopedic Surgery, University of Michigan, 1500 East Medical Center Drive, 2912 Taubman Center, SPC 5328, Ann Arbor, MI 48109, USA. Email: ialeem@med.umich.edu.

Abstract: Odontoid fractures are one of the most common injuries to the cervical spine in geriatric patients. Congenital C1 arch absence, however, is a very rare anomaly found in the population. We describe the first reported case of a congenital C1 posterior arch absence and C1 anterior cleft presenting with odontoid fracture. We present the case of a 58-year-old male who was found to have a comminuted type III odontoid fracture with significant angulation and displacement. CT scan demonstrated this fracture and also demonstrated congenital cleft of his left anterior arch and absence of left C1 posterior arch. Given his anatomic anomaly, we elected to perform occipitocervical fusion. The patient underwent occipito-cervical fusion to avoid iatrogenic vertebral artery injury. He was also immobilized in a halo vest given patient-specific social factors and compromised bone quality. The patient had no intra- or post-operative complications, but a prolonged hospital stay due to alcohol withdrawal. At 3-month postoperatively the patient had no neck pain and return to baseline function. This case highlights the importance of obtaining a CT scan preoperatively to not only to further characterize the fracture but also for surgical planning and recognition of anatomic anomalies as this may significantly impact the operative strategy.

Keywords: Odontoid fracture; congenital C1 anomaly; posterior arch defect; occipitocervical fusion; case report

Submitted Jul 23, 2020. Accepted for publication Mar 05, 2021.
doi: 10.21037/jss-20-628
View this article at: http://dx.doi.org/10.21037/jss-20-628

Introduction

Odontoid fractures are one of the most common geriatric cervical spine injuries, whereas congenital C1 arch absence is a rare anomaly that is often found incidentally (1). We present the first described case of a patient with unilateral absence of C1 posterior arch and an unfused anterior arch with a displaced odontoid fracture, highlighting the unique surgical challenges present in patients with congenital C1 anomalies. C1 congenital anomalies are reviewed. Treatment options for standard type III odontoid fractures are generally immobilization with a cervical collar. It is important to note, this is patient did not sustain a standard type III odontoid fracture given his fracture extension into the joint, and subsequent deformity. We present the study in accordance with the CARE reporting checklist (available at http://dx.doi.org/10.21037/jss-20-628).

Case presentation

A 58-year-old male presented to our facility as an outside transfer, after a fall 3 days prior. Computed tomography imaging demonstrated a comminuted type III odontoid fracture with 5 mm of anterior displacement and 19 degrees of anterior angulation. The fracture extended through the C2 body on the right and violated the left C1-C2 facet, creating both a sagittal and coronal deformity. Unexpectedly he was found to have congenital absence of his left C1 posterior arch as well as an anterior arch cleft (*Figures 1*,*2*). He had no neurological deficits. MRI was obtained with findings of normal spinal cord size and signal. CT angiogram was obtained and revealed normal course of the vertebral artery (*Figure 3*).

The patient's past medical history was significant for severe alcohol dependence and seizures. We discussed operative versus non-operative treatment which included

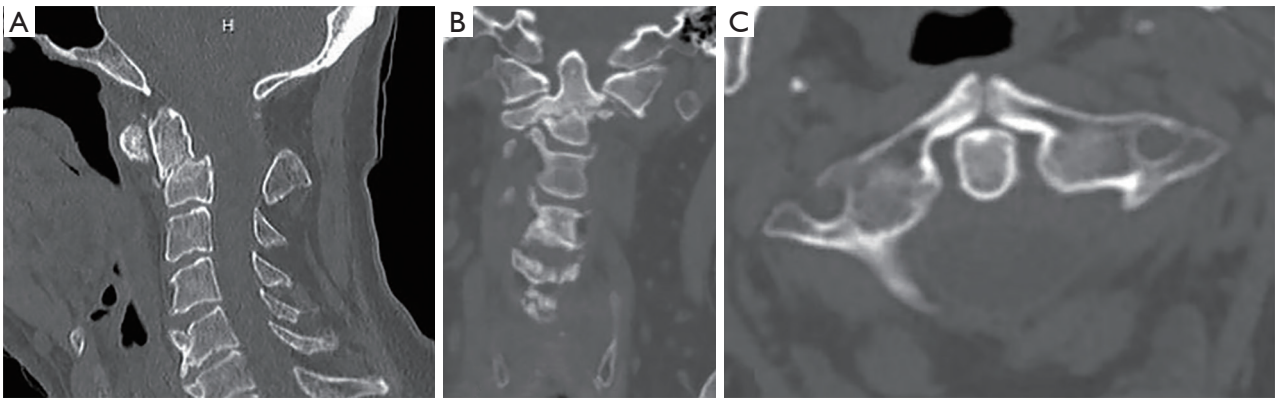

Figure 1 (A) Comminuted Type III odontoid fracture with anterior displacement in sagittal and (B) coronal plane. (C) Axial cross section with absence of left posterior arch and anterior cleft of C1.

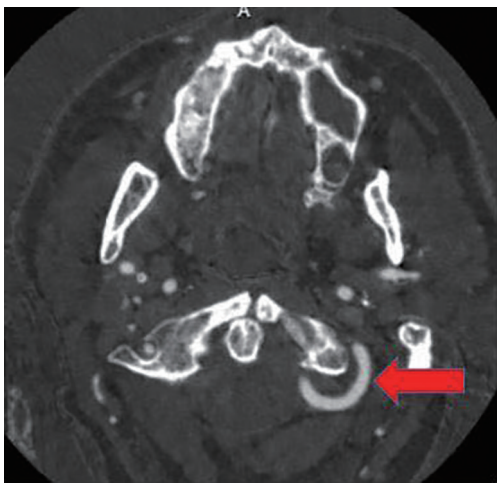

Figure 2 Congenital absence of left C1 posterior arch in 3D reconstructed image.

a halo vest application versus fusion. Due to social factors including his predisposition to falls due to alcoholism and history of non-compliance, the decision was made to undergo operative intervention in order to help mitigate further collapse and deformity. Due to congenital absence of the posterior C1 arch we felt C1 lateral mass screw fixation would risk iatrogenic vertebral artery injury. The decision was made to perform occipitocervical fusion given compromised bone quality secondary to alcoholism.

Gentle closed reduction using Mayfield traction was performed on the operating table under fluoroscopic visualization. Dissection was carried out exposing occiput to C4. Left side of C1 was not exposed given absent posterior arch to avoid iatrogenic vertebral artery injury. Occipital plate, C2 pars screws, and lateral mass screws were placed in C3-C4, given compromised bone quality. Autogenous bone graft and iliac crest strut allograft was placed from occiput to C2 and from C2-C4 and was tied in place with dissolvable suture. The patient was placed in a halo vest for further stabilization, which was self-extricated at 4 weeks postoperatively. *Figure 4* demonstrates immediate post operative radiographs. Although the patient had a prolonged hospital stay due to alcohol withdrawal, by 3-month follow up he had no neck pain and return to baseline function. He reports he was pleased with his outcome.

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient.

Discussion

Our report is the first to describe an odontoid fracture in a patient with partial absence of the C1 posterior arch. While the fracture was noted on cervical radiographs, the congenital anomalies were better appreciated on cervical CT Scan. The congenital absence of the posterior arch of C1 presented a surgical challenge in our patient due to the inability to undergo C1-C2 fusion due to a lack of fixation. We elected to proceed with occiput to C4 fusion, which does come with increased morbidity due to loss of motion at the occiput-C1 joint. In patients with spinal cord compression or irreducible odontoid fractures the treatment of choice is generally C1 laminectomy with occiput to C2 fusion (2) .

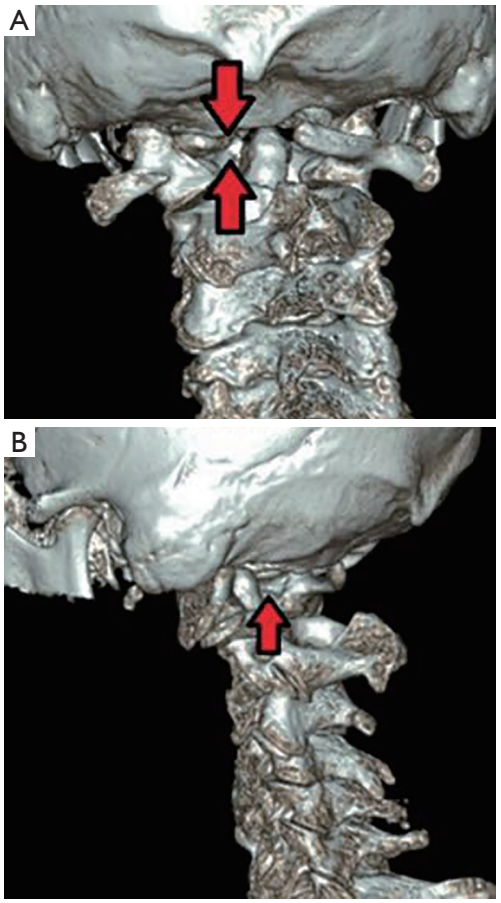

Figure 3 CT angiogram showing normal course of left vertebral artery.

C1 arch hypoplasia is due to failure of chondrogenesis (3,4), commonly associated with Klippel-Feil, Arnold Chiari, Down and Turners syndrome (5). Embryologically, the C1 vertebrae has 3 primary ossifications centers; the anterior tubercle and two lateral centers which extend forming the lateral masses and posterior arch. During the 7th week of gestations the lateral centers extend dorsally forming the posterior arch which are almost fused at birth. The posterior arches completely fuse between 3 and 5 years of age. Congenital C1 posterior arch defects are classified as: A) absent midline fusion; B) unilateral defect, C) bilateral defects, D) absence of posterior arch with presence of posterior tubercle, E) complete absence of posterior arch and tubercle (6). The overall incidence of C1 arch defects is 3.35%, with 2.6% type A, 0.54% Type B, and an anterior arch cleft only in one patient (0.09%) in a retrospective series (1).

The majority of congenital C1 anomalies are asymptomatic and found incidentally on imaging. There have been several case reports of transient paresis due to minor trauma in patients with congenital C1 posterior arch absence that had a retained posterior arch fragment (4,7). A case of a patient that remained symptomatic after minor head trauma subsequently underwent resection of the posterior tubercle which resolved the neurologic symptoms has been reported (8). Our patient, with Type B posterior arch defect and anterior arch cleft, did not have any neurologic deficits on presentation and only presented with neck pain and deformity on imaging. Given involvement

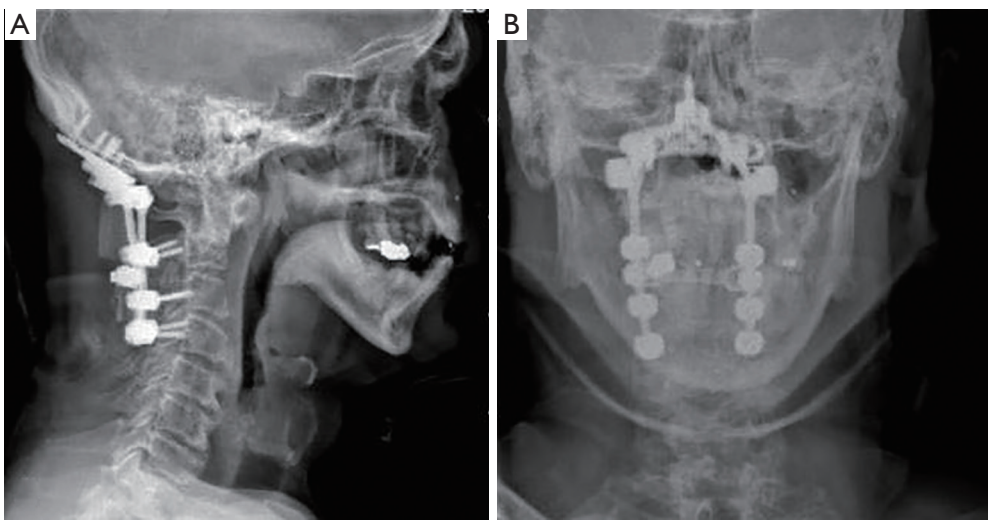

Figure 4 Immediate postoperative radiographs demonstrating occiput to C4 fusion.

of the C1-C2 joint, there was both a coronal and sagittal deformity present at the C1-C2 level. The treatment strategy here may also be applied to other C2 fractures with similar congenital anomalies.

Strengths/limitations

This case report presented, is of a scarce anomaly not frequently encountered by the spine surgeon. Having unexpectedly noted the left C1 arch anomalies on CT scan of this patient, this further emphasizes the importance of obtaining advanced pre-operative imaging in patients undergoing spinal surgery. This case also highlights that having good rapport with your patients and discussing potential risks and benefits of all interventions, though more invasive can lead to good desirable outcomes.

Conclusions

Our report is the first to describe an odontoid fracture in a patient with partial congenital absence of the C1 posterior arch, presenting a surgical challenge. This patient underwent occipitocervical fusion with good functional outcome. This report highlights the importance of obtaining a CT scan preoperatively and recognizing anatomic anomalies as this may significantly impact the operative strategy.

Acknowledgments

Funding: None.

Footnote

Reporting Checklist: The authors have completed the CARE reporting checklist. Available at http://dx.doi.org/10.21037/jss-20-628

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at http://dx.doi.org/10.21037/jss-20-628). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All figures are property of the authors, and are original. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient."

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Cite this article as: Butt BB, Gagnet P, Patel R, Aleem I. Congenital defect of the posterior arch of C1: a case report. J Spine Surg 2021;7(2):214-217. doi:10.21037/jss-20-628