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Atlantoaxial Instability with Persistent Second Intersegmental Artery

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Abstract

Understanding the anatomy of the vertebral artery is essential while manipulating the craniovertebral joint during surgery. Its anomalous course in congenital atlantoaxial dislocation makes it more vulnerable to injury. Preoperative dedicated computed tomography (CT) angiography helps identify the artery's position and plan for surgical procedure. A 13-year-boy presented with neck pain and spastic quadriparesis for 1 year. Radiological imaging of the craniovertebral junction revealed atlantoaxial instability with basilar invagination. His CT angiography of neck and brain vessels revealed an anomalous course of the vertebral artery due to a persistent second intersegment artery. He underwent posterior atlantoaxial fixation after mobilization of the vertebral artery. His clinical condition significantly improved after surgery. We report a case of management of an atlanto axial dislocation with persistent second intersegment artery and describe the role of vertebral artery mobilization during surgery.

Keywords
- atlantoaxial instability
- vertebral artery
- CT angiography
- 3D CT
- Os odontoideum

Introduction

The craniovertebral junction (CVJ) is a complex area of multiple joints, bones, ligaments, and vessels. Vertebral arteries (VAs) are the principal vessels closely placed to the important bony and neural structures.[1] Because of the complex nature of CVJ, it is prone to various congenital and acquired pathologies and often requires surgical correction. While dealing with these pathologies, a complete understanding of anatomy is crucial. The VA is the critical structure concerned during the surgery around CVJ. Congenital CVJ anomalies are often associated with the anomalous course of VAs, including high-riding VA, fenestrated VA, persistent first intersegment arteries, and persistent second intersegmental arteries.[2] Persistent second intersegmental artery associated with CVJ anomalies like atlantoaxial dislocation (AAD) is uncommon. We report a case of management of an AAD with persistent second intersegment artery and describe the role of VA mobilization during surgery.

Case Report

A 13-year-old boy presented with neck pain followed by progressive spastic quadriparesis and imbalance while walking for 1 year. He was unable to do his day-to-day activities without assistance. His general examination revealed a short neck, low hairline, right-sided torticollis, cleft lip, and absent uvula. A neurological examination revealed modified Ashworth Grade 3 hypertonia in all four limbs. His power in all four limbs was IV/V (according to the Medical Research Council grading. His deep tendon reflexes in both upper limbs and lower limbs were exaggerated. Babinski signs were positive, and posterior column involvement was on both sides.

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Magnetic resonance imaging (MRI) of CVJ showed significant anterior compression of the cervicomedullary junction due to sharp bony indentation on sagittal T1-and T2-weighted images and on axial T2-weighted images with widened atlantoaxial interval (►Fig. 1A, B). Also, the hyperintense signal was noted in the cervicomedullary junction on sagittal T2-weighted images due to chronic compression. Computed tomography (CT) scan of CVJ on a sagittal view (►Fig. 1C) and coronal view (►Fig. 1D) revealed irregular cortical bone fragment at the posterior aspect of the anterior arch of the atlas suggestive of Os odontoideum. The Wackenheim's line was seen transecting the residual odontoid process representative of basilar invagination (BI). CT scan of CVJ also revealed fixed AAD, abnormal configuration of left C1-C2 joints, and absence of posterior arch of atlas and its spinous process (►Fig. 2A–E). Three-dimensional (3D) CT scan CVJ also showed similar findings in the atlas and atretic left C2 foramina transversarium (►Fig. 3A, B). 3D CT angiography CVJ revealed an abnormal course of the left VA, which after exiting the C3 neural foramina had subisthmic course passing beneath the C2 lamina and not through its neural foramen (►Fig. 3C, D). This was suggestive of the persistence second intersegmental artery on the left side. In view of the congenital nature of AAD and associated bony and vascular anomalies, we also prepared a 3D bony model using CT and CT angiography images. 3D bone model showed bony abnormalities and an abnormal course of the left VA (►Fig. 3E). It helped us to plan the surgical procedure, including mobilization of the left VA and planning screw trajectories.

The patient underwent posterior C1-C2 fixation under general anesthesia. He was given a prone position with his neck in slight extension. The skull traction was applied using Gardner-wells tong with a 5-kg weight. Skin and facia were cut open by a midline vertical incision from the inion to C3 spinous process. Subperiosteal dissection was done, and the C1-C2 complex was identified. An anomalous course of the left VA was identified and confirmed with the vascular Doppler. The artery was meticulously dissected and mobilized medially (►Fig. 4A) (►Video 1). Subsequently, the C1-C2 joint was exposed and opened bilaterally, and end plates were removed and filled with bone pieces. Also, by gentle manipulation, C1-C2 joints were distracted, and distraction was maintained by impacting approximate-sized titanium spacers filled with bone grafts. Bilateral C1 lateral mass and C2 pars screws were inserted and connected with rods. A particular precaution was taken while inserting the C2 pars screw on the left side as the medially mobilized VA was close to the entry point (►Fig. 4B). Both C1-C2 screws were inserted safely, preserving the bilateral VA. Occiput and C2 lamina were drilled out and covered with bone graft harvested from the posterior superior iliac crest, and the wound closed in layers.

The postoperative course was uneventful, and his neurological status slowly improved. A postoperative MRI and CT scan of CVJ images revealed correction of BI and significant decompression of the CV junction (►Fig. 5A–F).

Discussion

Over the past two decades, CVJ surgeries have undergone a significant revolution. This is applied even in irreducible AAD. Traditional surgeries like transoral odontoidectomy with posterior fixation are less followed nowadays due to complications like the transoral spread of wound infection.[3,4] Though various procedures are described in the literature, presently method of choice for this pathology is the posterior fixation of the atlas and axis using lateral

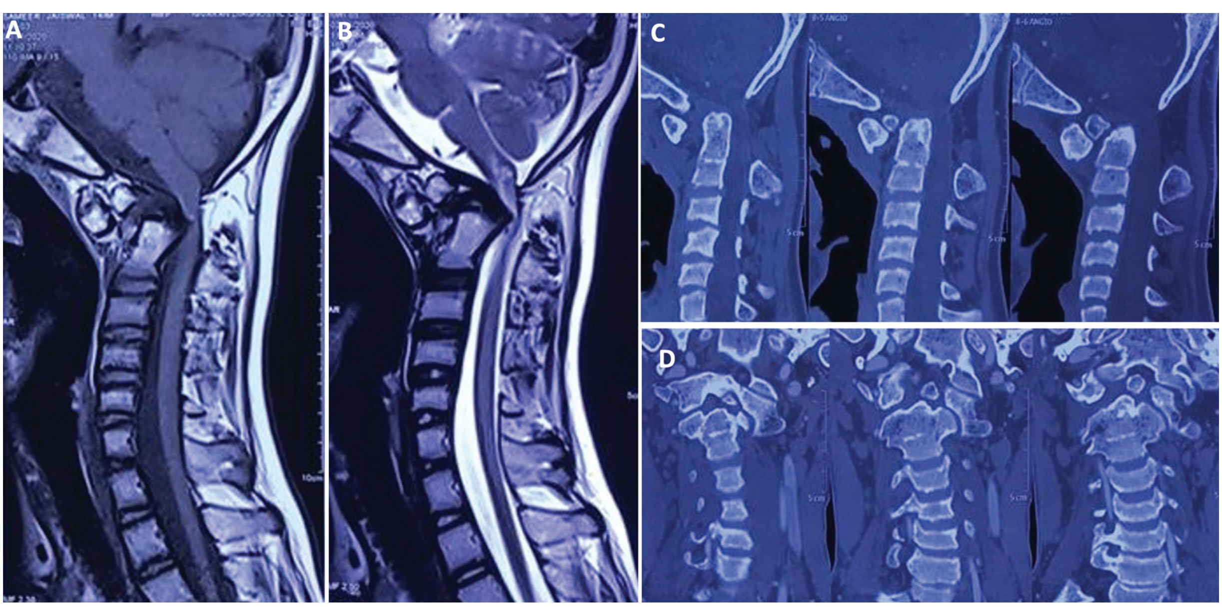

Fig. 1 Magnetic resonance imaging (MRI) of craniovertebral junction (CVJ) showing significant anterior compression of the cervicomedullary junction due to sharp bony indentation on sagittal T1 (**A**) and T2 (**B**)-weighted images. Also, hyperintense signals noted at the cervicomedullary junction on sagittal T2-weighted images due to chronic compression. Computed tomography scan of CVJ showing irregular, well cortical bone fragment at the posterior aspect of the anterior arch of atlas suggestive of Os odontoideum on sagittal view (**C**) and coronal view (**D**).

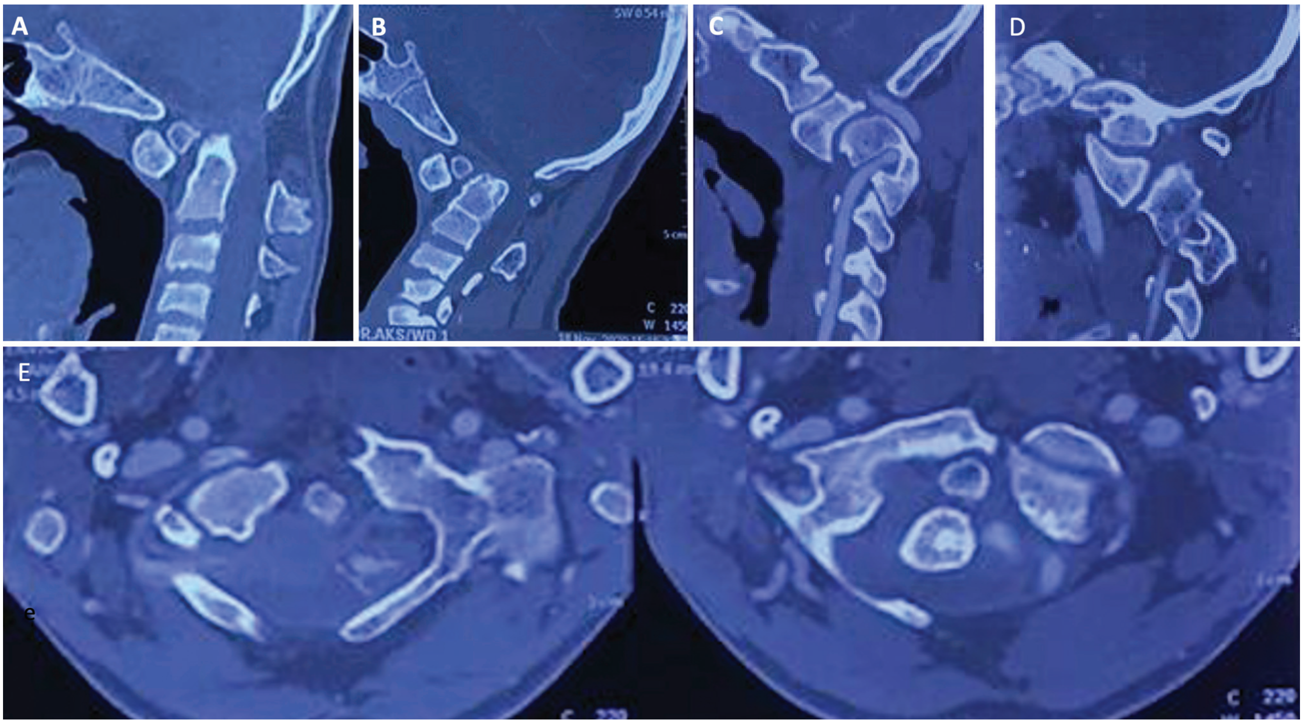

Fig. 2 Computed tomography (CT) scan craniovertebral junction (CVJ) sagittal view in flexion (**A**) and extension (**B**) images revealing fixed atlantoaxial dislocation. CT scan CVJ sagittal view in neutral position showing abnormal configuration of left C1-C2 joint (**C**) and right C1-C2 joint (**D**) as well as axial images showing Os odontoideum, absent of posterior arch of atlas and its spinous process (**E**).

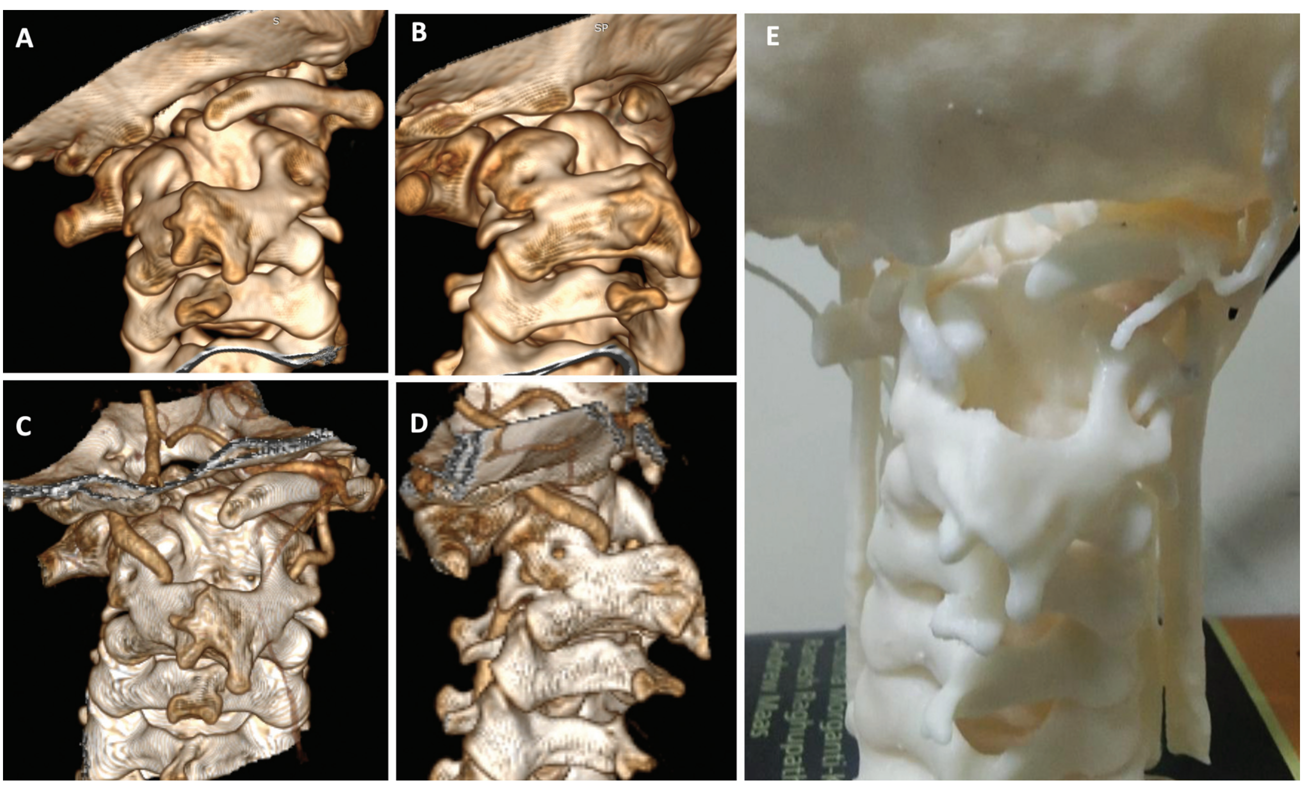

Fig. 3 Three-dimensional computed tomography (CT) scan craniovertebral junction (CVJ) images showing absent posterior arch of atlas on left side, absent C1 spinous process, and atretic left C2 foramina transversarium (**A, B**). Three-dimensional CT angiography CVJ showing abnormal course of left vertebral artery; after exiting C3 neural foramina it had subisthmic course passing beneath the C2 lamina and not through its neural foramen (**C, D**). Three-dimensional bone model showing bony abnormalities and abnormal course of left vertebral artery (**E**).

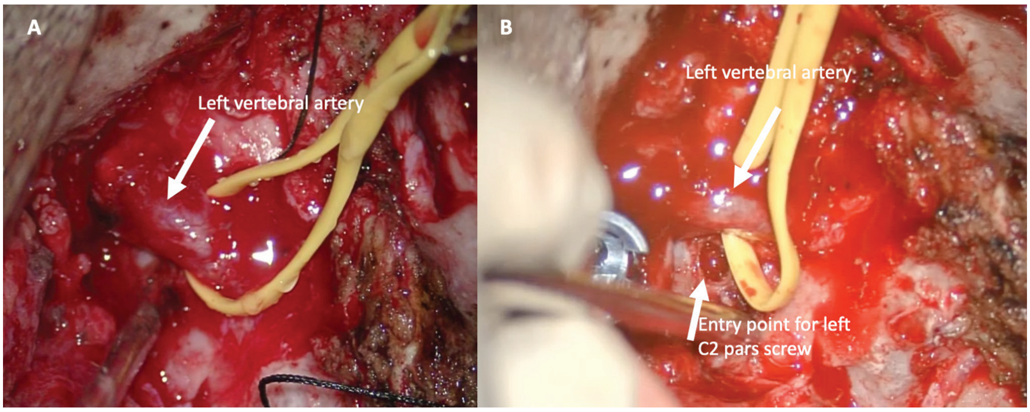

Fig. 4 Intraoperative images; arrow showing medially mobilized left vertebral artery (**A**) and arrow relation of C2 pars screw entry point with mobilized left vertebral artery (**B**).

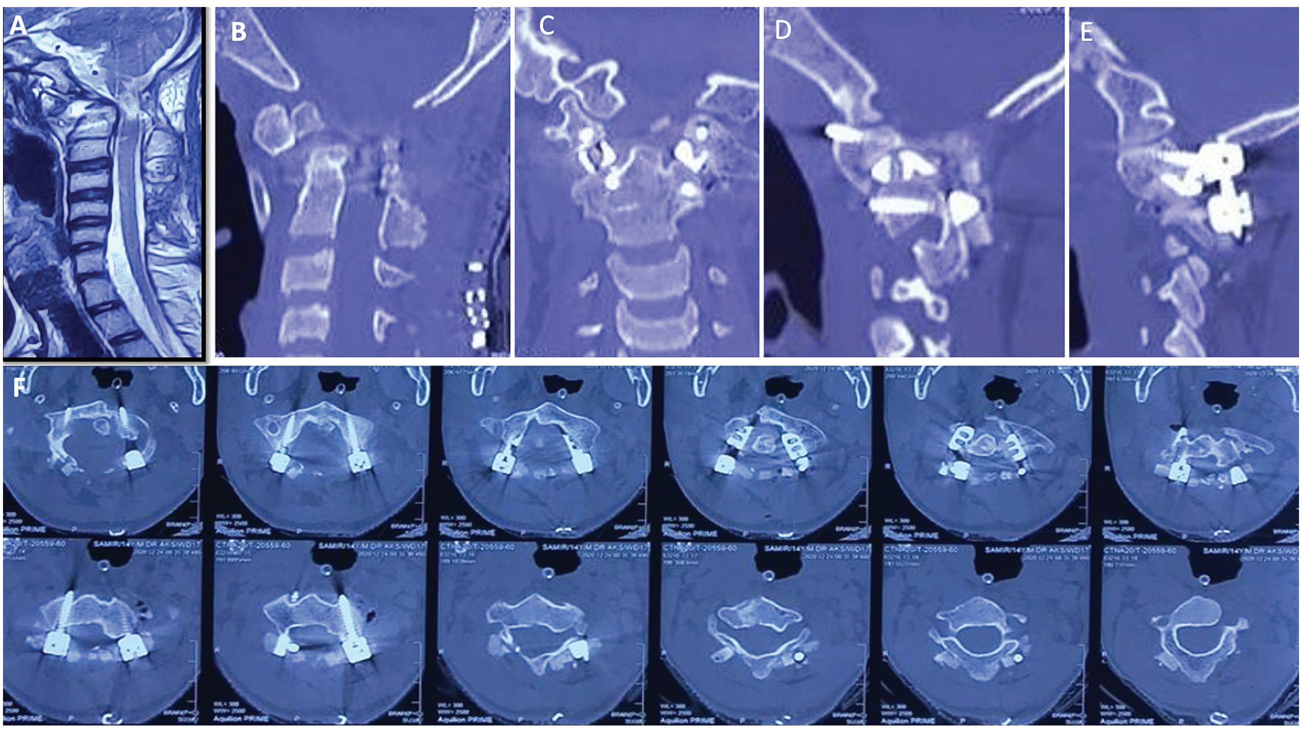

Fig. 5 Postoperative magnetic resonance imaging scan of craniovertebral junction (CVJ) T2 weighted sagittal view (**A**) and computed tomography scan of CVJ showing reduction in basilar invagination (sagittal view, **B** and coronal view, **C**) and atlantoaxial dislocation sagittal views (left C1-C2 joint, **D** and right C1-C2 joint, **E**) with spacers and screws in situ on sagittal images and on axial images (**D–F**).

masses and pars screws, respectively.[5] It is the shortest possible fixation and maintains mobility at the craniovertebral joint. The surgery aims to decompress neural structures to achieve acceptable biomechanical alignment and function by fusing C1-C2, thereby improving the region's local hemodynamics.[2] However, lateral mass screws and pars screw placement is technically demanding and requires expertise.

VAs are the main arteries of the posterior circulations and supply blood to the brain and spinal cord. Various anatomical variations exist in the development of these arteries, including asymmetry, irregular and insufficient development; these include duplication and fenestration of VA,[6,7] the variable origin of either one or both the arteries,[8] and high riding VA.[9] Prior understanding of these variations is very much essential to prevent intraoperative catastrophic events.

The VA is the most common artery injured during atlantoaxial fixation. It becomes even more susceptible to injury when the course is uncommon. VA injury is a rare complication of CVJ surgery but is devastating when it occurs. The reported incidence of VA injury during spinal surgery ranges from 0.1 to 0.5% in large series.[10] VA injury may occur in approximately 4.1% of patients during surgery at the CVJ.[11]

VA injury during surgery may lead to catastrophic bleeding, and the patient may develop a postoperative neurological deficit. To prevent vascular insult, preoperative evaluation of this region is imperative. CT scan CVJ with the 3D reconstruction of bone and CT angiography are the most important investigative tools before planning safe CVJ surgery. Also, the color-coded 3D model plays a vital role in better understanding bony and vascular abnormalities in relation to each other.[12]

Embryologically, the VA develops from a long anastomotic channel that interconnects the seven cervical intersegmental arteries.[13] The other intersegmental arteries involute during their development, and vertical channels persist to form the normal VA. Depending on the abnormalities in involution, various VA anomalies may occur. A persistent first intersegmental artery is one in which the VA, after exiting the C2 foramen transversarium, passes below the C1 posterior arch instead of its normal course through the C1 transverse foramen.[14] The VA anomaly described in our case is entirely different from the previously reported ones. After exiting the corresponding C3 transverse foramen, the VA on the left side passed medially and below the C2 isthmus in proximity to pars interarticularis. The left-sided second intersegmental artery (between C2 and C3) might have persisted, giving rise to this anomaly. The transverse foramina of C2 on the left side was also atretic. Correctly identifying this anomaly in CT angiography has its management implications. Here, the VA hugs the thin pars interarticularis of C2. A screw violating the C2 inferior wall will likely injure the VA. Therefore, placing a transarticular or pars screw may not be a good choice in these cases, as a bony breach inferiorly could be disastrous. In such cases, skeletonizing the VA, V3 segment, and putting the screws under vision is advisable. This way the chance of injuring the VA is minimal. A neuronavigation-guided screw placement may further minimize the risk. The other option for such a VA anomaly would be inserting translaminar screws, albeit with slightly inferior biomechanical stability. In the presence of anomalous VA, occipital–cervical fusion is an alternative. However, this hampers the neck movements adversely, which adds to the morbidity caused to the patient and may have delayed complications.[15]

Conclusion

Congenital craniovertebral anomalies are often associated with VA anomalies. 3D CT scans and CT angiographic images help in the early identification of these anomalies. Preoperative knowledge about persistent second intersegment artery, early identification, and intraoperative mobilization of the VA facilitate placement of C1-C2 intra-articular screws with good clinical outcome.

Video 1

Online content including video sequences viewable at: https://www.thieme-connect.com/products/ejournals/html/10.1055/s-0043-1776050.

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Conflict of Interest
None declared.

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