Asian Journal of Neurosurgery
Vol. 8, Issue 3, July-September 2013

CASE REPORT

Atlanto-axial dislocation associated with anomalous single vertebral artery and agenesis of unilateral internal carotid artery

Anant Mehrotra, Jayesh Sardara Chunnilal, Kuntal Kanti Das, Arun Srivastava, Raj Kumar
Department of Neurosurgery, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow, Uttar Pradesh, India

ABSTRACT

We report a case of an anomalous single midline vertebral artery (VA) in a case of atlanto-axial dislocation (AAD). The left VA coursed in the midline at craniovertebral junction as there was no right VA. The left internal carotid artery was also not formed. This was a case report of a 34-year-old male patient who presented with features of high cervical myelopathy. On evaluation, he had fixed AAD. There was inadvertent intra-operative VA injury, which was sealed to control the brisk bleeding to require stenting ultimately. Patient did not survive and expired after 5 days due to brain edema secondary to compromised anomalous intracranial circulation.

Key words: Atlanto-axial dislocation, congenital anomalies, internal carotid artery, vertebral artery

Introduction

Craniovertebral junction (CVJ) is a complex area and associated congenital lesions make the understanding of the local anatomy more difficult. Vertebral artery anatomy is also likely to be distorted in these congenital anomalies. Vertebral artery (VA) injury is more likely to be encountered while doing posterior approaches. Internal carotid artery (ICA) anomalies especially agenesis are rarely associated with VA anomalies. The anomalies of ICA make the treatment of VA injury more precarious.

Case Report

We report a case of 34-year-old male who presented with history of progressively increasing difficulty in walking and progressively increasing spastic quadriparesis for 7 years. He had pain and paresthesia in his right upper limb for the past 2 years. On examination, the tone was increased in all four limbs with power 4+/5 and hand grips of 60%. Bilateral planters were extensor. Posterior columns were impaired bilaterally. He had a short neck with low hair line with restricted rotational neck movement. Computed tomography (CT) showed atlanto-axial dislocation (AAD) with atlanto-dental interval (ADI) of 8 mm in both flexion and extension with partially occipitalized atlas and basilar invagination (BI) with basal angle of approximately 148° and tip of odontoid crossing the Wackenheim clival canal line. Magnetic resonance imaging of the CVJ showed significant compression of the cord at the CVJ [Figure 1]. Transoral decompression and posterior fusion was planned in view of fixed AAD (in view of no change of ADI in flexion and extension) and BI. C1 anterior arch was drilled and tip of the dens was drilled and excised and dura was partially visualized but was not opened. So as to have an adequate decompression, bony drilling was further carried out laterally and downward. While drilling on the left side there was brisk bleeding which was controlled with muscle, glue, gelfoam and floseal. In view of suspected injury to the VA, posterior fusion was deferred and traction was continued. Intra-arterial digital subtraction angiography was planned for diagnosis of suspected VA injury and necessary therapeutic intervention. However, before shifting for digital subtraction angiography (DSA) there was rebleeding from oral wound to require re exploration and hemostasis. Patient was shifted to DSA room. DSA showed absent left ICA and the right VA. The left middle cerebral artery (MCA) was filling through the left VA and the left anterior cerebral artery (ACA) was filling from the right ICA. The left VA was taking an anomalous course with the V3 segment curving medially and reaching up to the midline just behind the odontoid process. The VA was injured in its extradural part as it was injured behind the odontoid

Address for correspondence:
Dr. Raj Kumar, Department of Neurosurgery, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Rae Bareli Road, Lucknow - 226 014, Uttar Pradesh, India.
E-mail: rajkumar1959@gmail.com

Access this article online

Quick Response Code:  
Website: www.asianjns.org  
DOI: 10.4103/1793-5482.121692

Asian Journal of Neurosurgery
Vol. 8, Issue 3, July-September 2013
Mehrotra, et al.: Cranial vascular anomalies associated with atlanto-axial dislocation

Figure 1: (a and b) Computed tomography and magnetic resonance imaging showing atlanto-axial dislocation with basilar invagination with cord compression

Figure 2: (a and b) Left vertebral artery injection and site of injury; (b) right VA injection with non-visualization of right VA; (c) non-visualization of left internal carotid artery on common carotid artery injection; (d) left VA supplying left middle cerebral artery; (e) right ICA supplying bilateral anterior cerebral artery; (f) patent left VA after stenting

and there was no sub-arachnoid blood on CT. The site of injury was visualized and the stent was placed across the rent [Figure 2]. Next day the pack was removed and no bleeding was noticed. The sensorium of the patient did not improve. CT showed diffuse brain edema and anti-edema measures were initiated, but on the 5th post-operative day the patient expired.

Discussion

A normal VA is divided into four segments: The first segment originates from the subclavian artery and enters the C6 transverse process, the second courses from C6 to C2 transverse process, the third from C2 to the foramen magnum and the fourth segment enters the cranium to attend the basilar artery. At the CVJ, the anatomy of the 3rd segment of the VA becomes especially important. The VA third segment (V3) enters the transverse foramen of Cl, where it runs in a vertical, slightly anterior direction. Just above the transverse foramen of Cl, the artery changes direction dorsally and runs further in the sagittal plane. The artery then changes its direction again and runs transversely above the posterior arch of Cl. The course of the V3 is divided into three portions:
A vertical portion between the transverse processes of C2 and C1; a horizontal portion (V3 h) in the groove of the posterior arch of the atlas; and an oblique portion where it leaves this groove and travels up to the dura mater. In a study on cadavers and on healthy individuals, Cengiz et al. found that the mean distance of the medial tip of the VA to the midpoint of the posterior tubercle of the atlas was 1.67 cm (in cadavers in both right and left VA) and 1.76 cm (right VA in healthy individuals) and 1.72 cm (left VA in healthy individuals). In a similar study by Cacciola et al., the distance between the medial most tip of the VA and the midline was 14.3 mm to 19.7 mm. In our case, the VA was taking an anomalous course in the sense that the VA reached the midline and thus the distance between the midline and the medial most tip of the VA was nearly zero and the VA was lying just behind the odontoid predisposing it to injury while performing transoral decompression.

Another anomaly in our case was the associated absence of left ICA with left MCA territory supplied by left VA and left ACA territory supplied by right ICA. Agenesis of the ICA is a rare entity and might be present without any symptoms. Agenesis of ICA has been reported to be associated with increased incidence of aneurysms, transsphenoidal encephalocoele and rete mirabilis in the cranial base, agenesis of corpus callosum, arachnoid cyst, trigeminal neuralgia, megadolichobasilar anomaly and olivoponto-cerebellar atrophy. The agenesis of ICA has not been associated unilateral agenesis of VA and AAD and to the best of our knowledge this is the first reported case of association of the three conditions (agenaesthetic unilateral ICA, contralateral VA with AAD). Unilateral ICA agenesis need not be treated as such but it might complicate treatment of other associated intracranial. This anomaly reduced the treatment options as occlusion of the only VA (left) was ruled out and only stent could have been done to maintain the patency and to control the bleeding as well.

Pre-operative computed tomography angiography (CTA) is not done routinely in cases of AAD. Whether CTA should be done in all cases of congenital CVJ anomaly needs to be studied more extensively. Even if CTA is done, CTA images of the VA at the CVJ with occipitalization of the atlas should be cautiously analyzed before surgical procedures.

**Conclusion**

VA can have an anomalous course especially in cases of congenital CVJ anomaly. The anomalous VA might even be associated with anterior circulation anomalies and might be the only source of blood supply to a part of the anterior circulation. Agenesis of ICA is a rare occurrence and its association with AAD in along with VA anomaly has not been reported earlier. Pre-operative CT angiography might be helpful in recognizing these rare variations but whether CTA needs to be done in all cases of CVJ anomalies needs to be determined.

**References**


**How to cite this article:** Mehrotra A, Chunnilal JS, Das KK, Srivastava A, Kumar R. Atlanto-axial dislocation associated with anomalous single vertebral artery and agenesis of unilateral internal carotid artery. Asian J Neurosurg 2013;8:173-5.

**Source of Support:** Nil, **Conflict of Interest:** None declared.