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CASE REPORT

Vertebral artery dissection caused by atlantoaxial dislocation: a case report and review of literature

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Abstract

Background Atlantoaxial dislocation (AAD) is the most common craniovertebral junction malformation (CVJm) which are anomalies of the bones and soft tissues surrounding the foramen magnum. It usually leads to neurologic abnormalities because of instability of this mobile area. But vertebral artery dissection (VAD) caused by AAD is uncommon.

Case report We report a 15-year-old boy who presented with acute onset of bilateral VAD leading to posterior circulation ischemic stroke (PCIS). Computed tomography angiography (CTA) indicated dissection and occlusion of bilateral intracranial vertebral arteries and AAD with os odontoideum. After antithrombotic treatment for 3 months, the patient got complete revascularization and received posterior C1-C2 fusion.

Discussion There have only been tens of cases of PCIS caused by CVJm. We reviewed these relevant literatures and suggested that more attention should be paid to vascular impairment for patients with CVJm.

Keywords Atlantoaxial dislocation (AAD) · Vertebral artery dissection (VAD) · Posterior circulation ischemic stroke (PCIS)

Introduction

Cervicocerebral artery dissections account for nearly 2% of all ischemic strokes [1]. Among them, vertebral artery dissection (VAD) is an uncommon but important cause of posterior circulation ischemic stroke (PCIS) in young patients. VAD can be caused by neck trauma; however, it may be painless and usually occurs without a clear history of trauma [2]. The incidence of spontaneous vertebral artery dissection is estimated at 1–1.5 per 100,000 per year [3]. Atlantoaxial dislocation (AAD) is the most common craniovertebral junction malformation (CVJm) which are the anomalies of the bones and soft tissues surrounding the foramen magnum and often lead to instability of this mobile area [4]. It is assumed that the absence of stability induced by AAD is a risk factor of VAD [5]. However, VAD caused by AAD has been reported rarely. Here, we report a case of bilateral VAD caused by AAD with os odontoideum (OS) resulting in PCIS.

Case report

The patient was a 15-year-old boy who presented with a sudden vertigo, unsteady gait, tinnitus of the right ear, nausea, and brief unconsciousness without any recent head or neck trauma. Magnetic resonance imaging (MRI) showed infarcts in the right cerebellum and the splenium of the corpus callosum (Fig. 1). One hundred milligrams of aspirin per day was prescribed by the local hospital for 11 days. His symptoms gradually improved and then the patient stopped taking aspirin. Unfortunately, symptoms reappeared with neck stiffness 2 days later. Then, the patient was referred to Department of Neurology of Xuanwu Hospital.

Neurological examination revealed ataxia of right extremities. Vision test revealed bilateral disturbance of abduction and horizontal nystagmus. Cervical vascular ultrasound showed false lumens on V3 segments of bilateral vertebral arteries (VAs) suggesting VAD. Three-dimensional computed tomography angiography (3D-CTA) showed occlusion of bilateral intracranial VAs and basilar artery (BA). In addition, 3D-CTA also revealed AAD and OS (Fig. 2). Then, the patient received whole cerebral digital subtraction angiography (DSA) which indicated proximal BA

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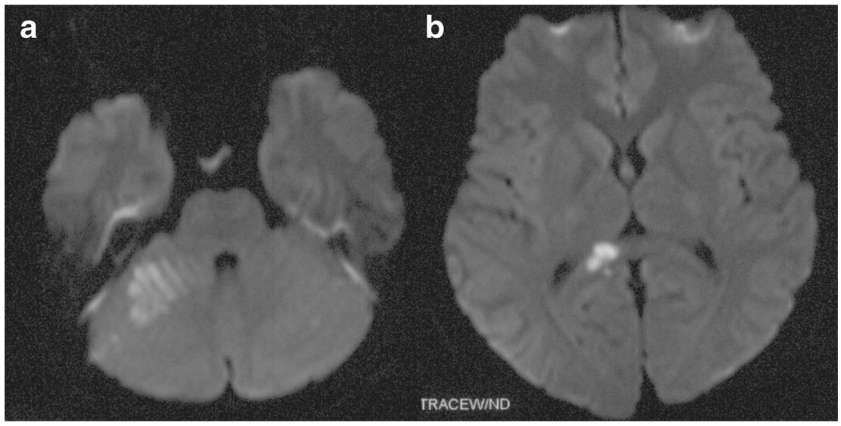

Fig. 1 **a** Hypertensive signal in T1 imaging of MRI suggesting infarct of the right cerebellum; **b** infarct of the splenium of the corpus callosum

was occluded and the right posterior inferior cerebellar artery (PICA) was not visualized (Fig. 3). He was treated with 150 mg warfarin per day sustaining the international normalized ratio between 2 and 3 since admission. After oral anticoagulation treatment as an outpatient for 3 months, the patient got favorable clinical improvement and another cervical CTA revealed complete revascularization. Then, the patient received posterior C1-C2 fusion surgery performed by Department of Neurosurgery of Xuanwu Hospital. He had a smooth postoperative recovery and was discharged from the hospital on the 5th day after operation with intact neurological function.

Discussion

This case showed a boy who presented with PCIS as the first manifestation of AAD which caused VAD. AAD is the most common type of CVJm which usually causes neurologic abnormalities by direct compression of the spinal cord or obstruction of the cerebrospinal fluid pathways. It is not common that CVJm leads to VAD, even rarer to PCIS. There have been only tens of cases of PCIS caused by CVJm reported in literature, most of which are single cases. Kulkarni et al. reported a case series [6] which included seven Indian patients diagnosed with PCIS

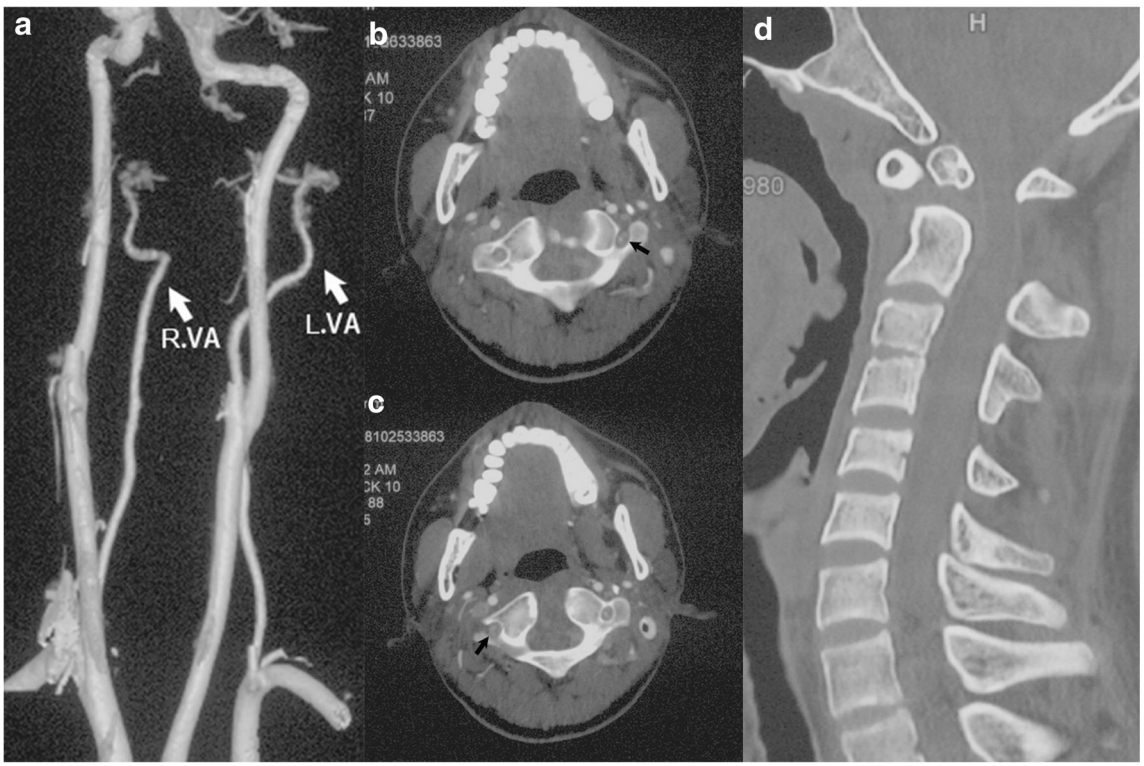

Fig. 2 **a** Cervical three-dimensional computed tomography angiography (3D-CTA) showed occlusion of the bilateral vertebral arteries (white arrows). **b** Axial view showed the left vertebral artery intimal flap (black arrow) secondary to vertebral artery dissection. **c** Axial view showed a narrow eccentric lumen of the right vertebral artery (black arrow). **d** Sagittal view showed atlantoaxial dislocation and os odontoideum

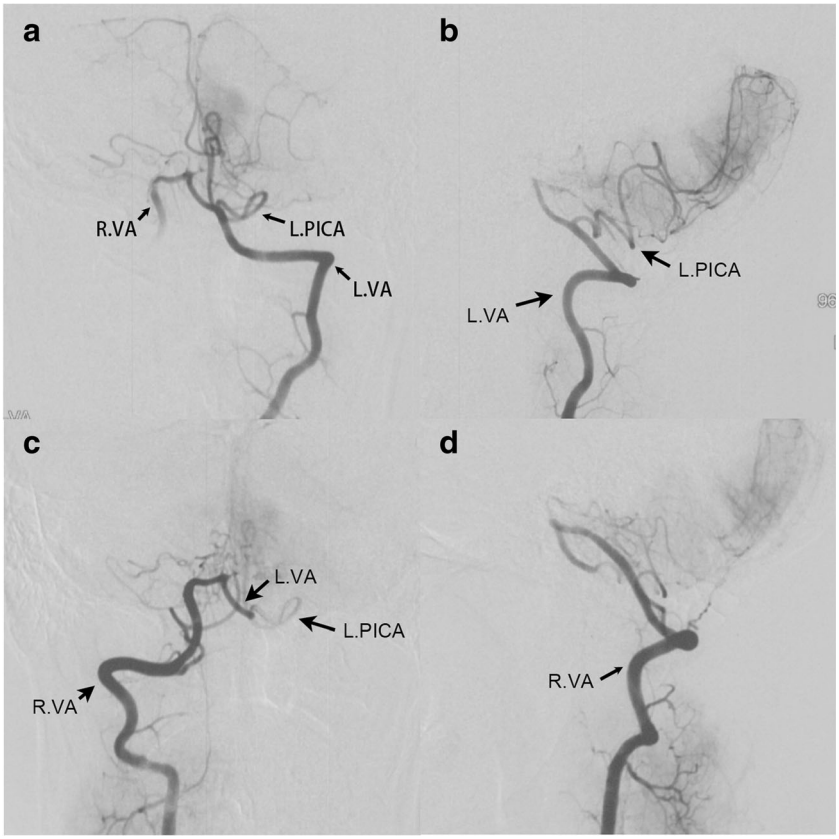

Fig. 3 **a**/**b** DSA of the left vertebral artery (L.VA) showed occlusion of the basilar artery and reflux of the right vertebral artery (R.VA). The left posterior inferior cerebellum artery (L.PICA) is visible. **c**/**d** DSA of the right vertebral artery showed occlusion of the basilar artery and non-visualization of the right PICA

caused by CVJm. Among the cases reported so far, the ratio of male to female was approximately 4.5:1, and a majority of them are pediatric or young adult patients. Patients were usually admitted to the hospital because of PCIS as the first manifestation without previous CVJm diagnosis.

The exact mechanism of VAD caused by CVJm is still unknown. It is generally thought that repeated motion, kinking, and stretching of a hypercurved VA which coursed in unstable occipitoatlantoaxial complex would trigger intimal tears that serve as dissection and thrombus formation [5]. Neck or head trauma are significant risk factors for VAD with CVJm but only happened in some patients. Zotter et al. reported a boy with basilar invagination who developed symptoms of cerebellar stroke following tracheal intubation which caused neck hypertension during an appendectomy under general anesthesia [7]. It reminds us more attention is needed on protection of the neck and head for the CVJm patients, not only in daily life but also during pertaining iatrogenic procedures.

Like the majority of patients with VAD caused by AAD, the dissection of our patient occurred at the third segment (V3) of VA which starts as the artery exits the foramen transversarium of C2 and ends at the point where the artery penetrates the dura at the foramen magnum. The high prevalence of VAD of V3 segment may result from the higher mobility of bone structures surrounding the VAs compared with the relatively fixed foraminal segment (V2) and intracranial segment (V4). In our case, DSA showed shortened and straighter course of the left VA. This phenomenon was described as "stretched loop sign" by Sawlani et al. and predisposed to thrombosis and occlusion of the VA [8].

It is generally recommended that antithrombotic therapy should last for 3–6 months for VAD. Our patient's initial antiplatelet therapy was insufficient which only lasted for 11 days and might be a potential cause for the subsequence ischemic stroke. Surgery for correcting malformations is also important to prevent stroke recurrence for patients with VAD caused by AAD. To our knowledge, the optimal timing to perform surgery remains unknown. Complete revascularization of VAD usually occurs within the first 6 months after the onset of symptoms. Arauz et al. found the estimated rate of complete revascularization was 45.9% at 3 months, and 62.3% at 6 months [9]. To relieve the impact on VAs by perioperative interruption of antithrombotic drugs, we performed surgery for our patient after revascularization when the endomembrane was supposed to be stable [5]. Warfarin was stopped 5 days before surgery and resumed 24 h after surgery, according to ACCP 9th ed. guidelines [10]. The majority of patients with VAD caused by AAD have a favorable outcome on stroke recurrence and neurologic function following surgery within 1–3 years [6, 11].

Conclusion

AAD rarely leads to VAD which is an important cause of PCIS for young and middle-aged patients. A majority of patients presented PCIS as the first manifestation without being diagnosed with AAD or CVJm. The instability of CVJ structures may predispose them to VAD and more attention is needed on vascular impairment with CVJm patients.

Compliance with ethical standards

Conflict of interest statement On behalf of all authors, the corresponding author states that there is no conflict of interest.

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