CASE REPORT

Congenital anomaly of the posterior arch of the atlas: a rare risk factor for posterior circulation stroke

Zhi-yuan Ouyang,¹ Min-jian Qiu,¹ Zhe Zhao,² Xiao-bing Wu,³ Lu-sha Tong¹

¹The Second Affiliated Hospital of Zhejiang University, School of Medicine, Hangzhou, China

²Department of Orthopedics, Beijing Tsinghua Changgung Hospital, Tsinghua University, Beijing, China

³The Second People Hospital of Quzhou, Quzhou, China

Correspondence to

Dr Lu-sha Tong, The Second Affiliated Hospital of Zhejiang University, School of Medicine, Hangzhou, China; rosa00tong@163.com

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SUMMARY

We report an extremely rare case involving a posterior arch anomaly of the atlas, causing a vertebral artery dissection (VAD) induced posterior circulation stroke. A 16-year-old girl was admitted to hospital because of new onset dizziness. VAD related multiple infarction in the posterior circulation was revealed. The congenital posterior arch anomaly of the atlas, along with instability of the atlantoaxial joint, were discovered accidentally during follow-up. This is the first case of ischemic stroke related to atlantal posterior arch aplasia and atlantoaxial instability. Although rare, it reminds us that investigation of the craniocervical junction should be considered when stroke occurs in young patients.

congenital anomaly of the atlantoaxial joint has been reported rarely. Moreover, malformation of the posterior arch of the atlas is even rarer and is usually assumed to be benign.^{4 5}

CASE PRESENTATION

A 16-year-old female dancer complained of persistent dizziness and ataxia for a week. Nausea and vomiting were also present, with left-sided numbness and weakness. Her past medical history was unremarkable. Diffusion weighted imaging demonstrated newly occurring multiple infarctions in the right thalamus and cerebellum (figure 1A, B). DSA revealed dilation and an intraluminal filling deficit in the V3 segment of the right vertebral artery (figure 1C). VAD induced multiple thromboembolic infarction was considered, and dual antiplatelet therapy was administered.

OUTCOME AND FOLLOW-UP

Four months later, the main symptoms were significantly relieved and repeat DSA was performed, showing a dissecting aneurysm formed in the V3 segment of the right vertebral artery (figure 1D). One year after onset, despite complete recovery, repeated CT angiography showed an even larger aneurysm, and a congenital posterior arch anomaly

BACKGROUND

Vertebral artery dissection (VAD) is reported to have an estimated incidence of approximately 1.87 per 100 000, and more than half of VAD cases emerge at the V2–V3 segments.^{1–3} Risk factors for VAD are trauma, autoimmune diseases, infections, abrupt movements of the head, sporting activities, manual treatment of the neck, metabolic bone diseases, and congenital anomalies.^{2 3} Among these risk factors, traumatic atlantoaxial dislocation is assumed to be the most common cause while

Figure 1 (A) Diffusion weighted imaging (DWI) showing multiple ischemic lesions in the right thalamus and (B) right hemisphere of the cerebellum. (C) DSA demonstrating a filling defect and dilation in the V3 segment of the right vertebral artery (arrow) and (D) a newly formed dissecting aneurysm in the same site after 4 months (arrow). (E, F) Axial and reconstructed CT of the atlantoaxial joint, highlighting the absence of the posterior arch of the atlas with atlantoaxial dislocation (red arrow) (E) and deviation of the odontoid peg from the middle line (F). (G, H) Severe atlantoaxial instability on flexion/extension X-rays of the cervical spine. (I, J) Coronal and reconstructed three-dimensional CT showing the dissecting aneurysm (black arrow).

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of the atlas was discovered (figure 1E–J). Flexion/extension X-rays of the cervical spine (figure 1G, H) were performed and severe atlantoaxial instability was confirmed. Surgery fixing of the atlantoaxial luxation was advised but was rejected by the patient.

DISCUSSION

The vertebral artery has four parts. The second part (intraforaminal) is more or less fixed to the cervical vertebra and the third part (V3) is mobile with loops; both are highly susceptible to VAD.^{3–6} A variety of risk factors have been reported, including history of mechanical trauma, labor or delivery, recent infection, and rheumatoid diseases.^{3–5 7} It is rare to see VAD related to non-traumatic atlantoaxial dislocation, and there are only very limited reports of malformation of the posterior arch of the atlas.^{4 5} The neurological symptoms associated with atlantal posterior arch defects have been mostly described as chronic neck pain or quadriplegia in the setting of a minor trauma to the head or neck.^{5 8} To the best of our knowledge, this is the first case of ischemic stroke caused by VAD induced embolic infarction, related to atlantal posterior arch aplasia and atlantoaxial dislocation.

Previous reports have suggested that the increased risk of vertebrobasilar territory ischemia is mainly attributed to instability between the odontoid peg and the anterior arch of the atlas.⁹ It is noted that craniovertebral junction anomalies are often multiple, coexisting, and include atlantoaxial dislocation.⁴ Regarding our patient, coexistence of atlantoaxial dislocation

and deformity of the posterior arch of the atlas were observed during follow-up. Using reconstructed CT angiography, we were able to disclose the skew of the odontoid peg (instead of rotation in healthy patients). In this way, we assumed the abnormal twist and retraction of the vertebral veins would make them more susceptible to dissection. Given the congenital anomaly at the atlantoaxial junction, although there was no history of trauma, the profile of a dancer may have added to the risk of atlantoaxial instability, and exaggerated the dissecting aneurysm, which contributed to the posterior circulation stroke.

Contributors Z-YO and M-JQ visited and followed-up the patient. Z-YO, M-JQ, X-BW, and L-ST collected and analyzed the data. ZZ provided the diagnosis and suggested this patient. L-ST conducted the study. All authors contributed towards the study design and drafting of the article.

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Learning points

- We have reported a rare case in the setting of a craniovertebral junction anomaly, demonstrating ischemic stroke rather than spinal compression.
- In young patients with posterior circulation stroke, investigation of the craniovertebral junction should not be overlooked.
- Dancing may be a high risk profession related to artery dissection with atlantoaxial instability.

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