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An 18-year-old man with fenestrated vertebral arteries, recurrent stroke and successful angiographic coiling

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Abstract

Fenestration of vertebral arteries has been reported in association with thromboembolic brain infarctions. However, few cases have been reported in which recurrent infarction occurred in spite of adequate anticoagulation. We report a young man with fenestrated vertebral arteries and stroke who failed to respond to standard anticoagulation therapy but did well with angiographic coil obliteration of an abnormal vertebral segment. An 18-year-old left-handed man presented with acute onset of dizziness and headache. No trauma or other stroke risk factors were identified. Left cerebellar infarction was seen on CT, but the cause could not be identified by brain and neck MRI, MRA, or CTA. Bilateral fenestrated vertebral arteries were identified with conventional angiography. Although the patient recovered fully and was treated with anticoagulation, he suffered a recurrent stroke 1 month later involving the right cerebellum while he was on a therapeutic dose of warfarin. Repeat arteriography showed a spontaneous dissection within one of the fenestrated vertebral segments. Since receiving angiographic coil obliteration of the pathologic segment, he has been free of all symptoms. We conclude that the patient sustained recurrent thromboembolic events in his posterior circulation due to spontaneous dissection within a fenestrated vertebral artery segment. Conventional angiography and emergent interventional embolization were essential to his diagnostic evaluation and therapeutic intervention.

Keywords

Cerebellar infarction; Thromboembolic stroke; Posterior circulation; Fenestrated vertebral artery; Interventional radiology

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1. Introduction

The vertebral arteries may be redundant because of two anomalies: duplication and fenestration. Fenestration differs from duplication in that the artery has a single origin and then separates into two parallel arteries along the length of its course [1]. Fenestrations of the extracranial course of the vertebral arteries are rare and are usually noted incidentally during angiographic studies or post mortem examinations [2]. Some authors have suggested that fenestration of vertebral arteries is a congenital abnormality without pathological significance [2]. Arterial dissection and thromboembolism can arise within fenestrated vessels, and shearing forces induced by vigorous head movement may be responsible [3]. Typically, these dissections have presented with subarachnoid hemorrhage [3,4]. Aneurysm formation can also occur [5]. Anticoagulation has been suggested when redundant vertebral arteries are implicated as the cause of embolically mediated neurological events [6]. However, anticoagulation is not 100% effective, and surgical or endovascular obliteration of the redundant lumen offers the possibility of definitive treatment [3,6]. Stenting or transcatheter occlusion of the fenestrated vessels could be effective in select cases. We report a patient with bilateral vertebral artery fenestration who had irregularities in one lumen, which most likely resulted in recurrent stroke despite adequate anticoagulation. The patient subsequently underwent endovascular obliteration of the abnormal segment and is now symptom-free.

2. Case report

This 18-year-old left-handed man experienced the sudden onset of “the worst headache” of his life while driving. The pain was panencephalic and associated with intermittently blurred vision, photophobia, nausea and vomiting. He took tramadol and acetaminophen but became progressively obtunded. Head CT showed a left cerebellar hypodensity, and brain MRI demonstrated acute ischemia in the same distribution (Fig. 1). The CSF was unremarkable. MRA of the brain and CTA of the neck were normal, except that the vertebral arteries at the level of C1 were not well visualized. A conventional vertebral angiogram was then performed, which revealed bilateral vertebral artery fenestration at C1, with slow flow and irregularity in one lumen on the left (Fig. 2). Based upon these findings, anticoagulation with low molecular weight heparin was administered, followed by warfarin within 2 weeks of the event. A hypercoagulability panel was notable for a borderline-positive dilute Russell Viper Venom Time (DRVVT) and an elevated lipoprotein (a) of 63 mg/dl.

The patient did well for just over a month, when he had the acute onset of headache and right neck pain while driving. At home, his mother noticed slurred speech and a lack of coordination. He was taken to the emergency room where a head CT showed a subtle right cerebellar hypodensity in addition to the old left cerebellar infarct. International normalized ratio (INR) was 3.8. MRI showed a new ischemic infarct in the right cerebellar hemisphere and medial left occipital lobe (Fig. 3). Dysarthria worsened, and he was transferred to University of Colorado Hospital (UCH). On admission to UCH, the patient’s speech was dysarthric, but there were no other neurological abnormalities. He was treated with heparin and remained neurologically stable throughout his 16-day hospitalization. On the 7th hospital day, diagnostic angiography showed bilateral fenestration of the vertebral arteries and aneurysmal dilation, with dissection in the same fenestrated left vertebral segment that had slow flow on the previous arteriogram (Fig. 4). On the next day, he underwent catheter-directed coil obliteration of the vessel with the aneurysmal dilation (Fig. 5). There were no complications. He was then maintained on heparin until a transition to warfarin was completed, and he was discharged with no neurologic deficits. Subsequent evaluation as an outpatient disclosed that he remained normal neurologically at nine months of follow-up. Further hematologic evaluation concluded that he had no hypercoagulable disorder.

3. Discussion

This case demonstrates that fenestration of vertebral arteries with subsequent strokes can, when there is an anatomic abnormality in a fenestrated segment, fail medical management with anticoagulation and require endovascular intervention. Previous reports have demonstrated the efficacy of coiling in similar situations [3], but none have shown the efficacy of this approach when medical management has failed in fenestrated arteries. In addition, these other cases presented with subarachnoid hemorrhage and not dissection with embolic stroke [3,4]. Further, few descriptions in the literature have demonstrated, both clinically and radiographically, the progression of such a dissection in a fenestrated artery. Our patient had bilateral vertebral artery fenestration and progressive spontaneous dissection in one segment, which likely resulted in recurrent stroke despite adequate anticoagulation. Previous case reports have demonstrated bilateral fenestrations of the vertebral artery with trigeminal neuralgia, but never stroke [7]. Our patient subsequently underwent endovascular obliteration of the diseased segment and is now free of all symptoms. Similar cases with duplicated vertebral arteries and thrombosis have failed medical therapies and required surgical intervention [6], and in some, death has occurred [8]. This sequence of events has never been demonstrated in a fenestrated vertebral artery. Cases of subarachnoid hemorrhage and dissecting aneurysm of a fenestrated vertebral artery have responded to immediate endovascular therapy [3,4].

Our case raises the question of whether endovascular obliteration should be considered as the primary treatment when an anatomic abnormality is detected in a fenestrated segment, or whether medical therapy should always be the primary treatment. In this case, we felt that it would have been premature to offer invasive treatment at the time of the first stroke, given the inherent risks and the lack of firm evidence in the literature to favor this approach. In addition, the patient had several initial indicators that he might have a hypercoagulable state that would warrant a medical trial of an anticoagulant such as warfarin; this diagnosis, however, was eventually excluded by detailed subsequent hematologic evaluation. Thus, we conclude that medical therapy is still indicated as the initial approach in such cases. However, in view of the slow flow in one left vertebral fenestrated segment seen on the first arteriogram, timely follow-up arteriography prior to the second stroke would likely have shown progression of the dissection, allowing consideration of endovascular obliteration of the segment before recurrent neurological symptoms developed. Therefore, our second conclusion is that patients with fenestrated vertebral arteries and strokes should have sequential imaging following their initial event. Detailed studies evaluating the efficacy of anticoagulation compared with endovascular surgery in this setting are clearly needed. At present, patients such as ours with stroke and anomalous neurovascular anatomy warrant thorough case-by-case review using combined neurologic, neurointerventional, and neurosurgical expertise to consider whether either fenestrated segment harbors a potentially emboligenic defect.

This case also demonstrates the utility of conventional angiography in strokes of uncertain etiology, particularly in younger patients. Neither conventional MRA nor CTA identified the fenestrated vertebral anomaly during the evaluation of the first stroke. Had these studies been relied upon solely, the patient would have been at substantial risk of morbidity and even mortality. Previous studies have demonstrated the limitations of MRA and CTA in imaging the posterior circulation [9,10]. Our patient illustrates that conventional angiogram remains the gold standard for visualizing the posterior circulation, because both CTA and MRA failed to identify the fenestrated vessels.

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Fig. 1.
Initial T2 MRI showing acute left cerebellar infarct (diffusion weighted imaging was also positive).

Fig. 2.

(a) Initial right vertebral angiogram-AP. There is a fenestration of the right vertebral artery at the level of C1 (extending from the black arrow to the white arrow). (b, c) Initial left vertebral arteriogram-AP (b) and lateral (c). There is a fenestration of the left vertebral artery at the level of C1 (extending from the black arrow to the white arrow). There was slower flow through the medial limb. There is minimal deformity of this limb at the time of this arteriogram.

Fig. 3.
Diffusion-weighted MRI at readmission, showing new right cerebellar and medial left occipital lobe infarcts.

Fig. 4. Second left vertebral arteriogram after right cerebellar stroke. There is now a fusiform dissecting aneurysm in the medial limb of the fenestrated left vertebral artery. There is also a small filling defect at the basilar tip consistent with a small embolus.

Fig. 5.
After coil occlusion of the medial limb of the left vertebral artery fenestration. There is complete cessation of flow through the dissecting aneurysm with minimal retrograde flow into the distal portion of the embolized medial limb.