Clinical Reasoning: A 37-Year-Old Man Presenting With Intermittent Dizziness and Brief Choreiform Movements

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Abstract
In this report, we describe the case of a 37-year-old man presenting with intermittent episodes of dizziness and choreiform movements. MRI of the head showed an acute infarction in the left cerebellar hemisphere and multiple chronic infarctions in the cerebellum and pons. Vessel imaging also showed dynamic stenoses in the posterior circulation. This case provides a framework for evaluating young patients with intermittent dizziness and recurrent strokes and highlights the need to combine anatomical angiographies or hemodynamic studies with head rotational behavior.
Key words

Recurrent posterior circulation infarction; Stroke in young adults; Vertebral artery variation; Dynamic stenosis

Section 1

A 37-year-old man presented with episodes of dizziness, and loss of coordination for one week, and acute choreiform movements of his left arm for one day.

Neurological examination disclosed normal mental status and cranial nerves. Motor system examination revealed choreiform movements of his left upper extremity, uncoordinated finger-to-nose and heel-knee-shin movements on the left side. Muscle strength, tone, deep tendon reflexes, sensation and the Romberg test were normal. All were normal on the right side except for a positive Babinski sign.

He had no hypertension, diabetes, or hyperlipidemia, and denied smoking, drinking, staying up late or toxic exposures.

Questions for consideration:

1. Where do you localize the patient's symptoms and clinical signs?
2. What is your differential diagnosis at this stage?

Section 2

The symptom of dizziness originates from damage to the vestibulocerebellar system.

Given the deficits in coordination on the left side, we localized an acute lesion in the
left cerebellar hemisphere. The right Babinski sign indicates an impairment in the left corticospinal tract which may be chronic. Left choreiform movements suggest injury of right basal ganglia, involving the putamen, caudate or thalamus. Together, arteries of the posterior circulation including left vertebral artery (VA) and/or its branches, and right posterior cerebral artery (PCA), are probably involved.

The initial concern is an acute vascular event such as vertebrobasilar insufficiency or posterior circulation stroke. Even rarer, beauty parlor stroke syndrome (BPSS) and patent foramen ovale (PFO) should also be considered. BPSS is referred to as a stroke caused by the acute arterial dissection brought on by extreme backward head positions, such as those maintained during hair washing. The patient denied the experience of having his head bent backward prior to the onset of dizziness. PFO is a small hole between the upper chambers of the heart, through which a blood clot could pass from the right atrium directly into the left atrium, and thereupon into systemic circulation toward the brain, causing dizziness or cryptogenic stroke.

For the differential diagnosis of transitory choreiform movements, common etiologies include vascular, metabolic, epileptic or autoimmune diseases. For example, limb-shaking transient ischemic attack, presenting as a paroxysmal involuntary hyperkinetic movement associated with carotid occlusion; chorea associated with non-ketotic hyperglycemia; focal motor seizures presenting as jerking, transitory limb movements; Leucine-rich glioma-inactivated 1 antibody encephalitis accompanied by involuntary muscle jerk in arm and face, could be easily confused with each other. Laboratory and neuroimaging tests are of great value to differentiate these conditions.

Laboratory examinations, including blood parameters, electrolytes, glucose, D-dimer (0.19 μ g/mL; normal < 0.5 μ g/mL), LDL (2.95 mmol/L; normal < 3.36 mmol/L),
were within normal ranges. Neither transcranial doppler (TCD) bubble test nor right heart contrast echocardiography detected PFO.

Brain MRI showed both acute infarction in the left cerebellar hemisphere and chronic infarctions in the cerebellum and pons bilaterally (Figure 1, A and B). Head CTA showed a patent but hypoplastic left VA, and the stenosis in the P1 segment of the right PCA and in P1 and P2 segments of the left PCA (Figure 1C). Digital subtraction angiography (DSA) demonstrated bilateral fetal-type PCAs (not shown). Notably, the chorea in the left arm only lasted for 2 days, after which normal arm function was recovered. Brain MRI did not reveal any acute infarction in basal ganglia, therefore we speculated that the reversible choreiform movements were induced by a transient ischemia, most likely in the right thalamus where the blood supply was restricted due to the insufficiency in the P1 segment of the right PCA (Figure 1C). Intriguingly, we did not find any evidence of complete occlusion or dissection in the vertebrobasilar arteries, so where do the cerebellar strokes come from?

Questions for consideration:

1. Is there any useful historical information on this patient?
2. What further investigations would you order to figure out the etiology?

Section 3

In light of multiple chronic cerebellar infarcts, the patient’s medical history was reviewed. In particular, although treated with aspirin 100 mg/d, the patient had experienced two other episodes of strokes within the past two years. Notably, he had occasionally had intermittent neck pains and vertiginous symptoms with extreme head
turning to the left also for two years, while the symptoms could be relieved immediately by returning head to the neutral position. It occurred to us that the cause for the recurrent strokes was unusual.

Intriguingly, neck CTA showed left VA originating from aortic arch (Figure 1D), rather than the left subclavian artery. Notably, the left VA was thinner than the right VA (Figure 1, E and F), and abnormally dilated at the V2 segment due to the compression by a bony spur of the transverse process at C5 before entering C4 transverse foramen (Figure 1D). Without evidence of persistent VA stenosis or dissection, we suspected the cerebellar strokes were caused by a dynamic or reversible vessel stenosis in the V2 segment of VA around C5 level.

Questions for consideration:

1. What is your final diagnosis?
2. How does patient’s aberrant VA relate to recurrent strokes?

Section 4

The clinical constellation of intermittent dizziness and recurrent strokes caused by a dynamic or reversible vessel occlusion in a young patient made us think of bow hunter’s syndrome (BHS). BHS is a rare yet important etiology of posterior circulation stroke, characterized by recurrent attacks associated with head rotations\(^1\).

To verify the possibility of BHS, the patient further underwent dynamic SPECT. When his head was in the neutral position, SPECT revealed adequate blood flow in the cerebellar hemispheres bilaterally (Figure 2, A and B), indicating patent vertebral
vessels. In contrast, blood flow was decreased in the left cerebellar hemisphere when the patient turning head 45° to the left (Figure 2, D and E), evidence for the presence of a dynamic stenosis in the left VA. We also carried out a TCD test associated with head rotation. Compared with the normal blood flow in the neutral head position (Figure 2C), the blood flow of the left VA demonstrated a higher flow resistance during leftward head movements (Figure 2F), suggestive of the diagnosis of BHS. Note that no abnormalities were detected in the rightward head rotational SPECT or TCD tests (not shown). The reconstructed CTA further revealed a dynamic stenosis in the left VA at C5 in response to various head rotations to the left. Particularly, the degree of stenosis change was likely to be associated with the extent of head rotation (Figure 2, G–I). Similar results were observed in DSA (Figure 2, J and K), thus confirming the diagnosis of BHS.

Discussion

Nondominant VA involvement, as in our case is rare in BHS. We assume bilateral fetal-type PCAs limited collateral blood flow from the anterior cerebral circulation, thus aggravating the ischemia induced by the temporary stenosis of left VA when the patient turned head leftwards. In previous case reports where symptomatic BHS was caused by nondominant VA, none possessed a variant anatomical origin and course of VA like ours.2,3 BHS caused by nondominant VA compression seldom exhibits severe symptoms. Indeed, our patient only suffered from slight dizziness during the rotational tests.

The origin of left VA from aortic arch is seen in 4.4% (153/3460) of cases4. The variant vessel which usually enters the cervical vertebrae transverse foramina at a
level higher than C6 entered C4 transverse foramen instead in our case, a phenomenon observed in less than 20% (29/153). A configuration in which the left VA branches of the aorta is considered likely to be at higher risk of dissection, due to a larger shear stress compared with a VA of subclavian artery origin. In our case, VA dissection was not detected in both high-resolution MRI and DSA, but vessel dilation and a dynamic stenosis did occur at the C5 level. We speculate that the left VA of aortic origin might receive direct cardiac pulsatile flows, thereby gradually the vessel became vulnerable and dilated.

Although DSA is the gold standard diagnostic technique, noninvasive SPECT and TCD tests are simple and easily accessible for the initial screening of BHS. There is no unified guideline for the treatment of BHS and choices of treatment include conservative management, endovascular procedures and surgery. A study of 19 BHS patients under conservative treatment showed that none of them developed stroke in 37.5-months’ follow-up, indicating conservative treatments might be safe as a first-line therapy. Whereas some studies argue that surgical treatment is effective and can be associated with a higher favorable prognostic outcome. In general, this disease carries a good prognosis if treated properly. Presented with the options, our patient opted for conservative management including anticoagulation therapy (Rivaroxaban 10 mg/d) and avoiding excessive left flexion of the neck using a cervical collar. No recurrence of stroke has occurred in one-year’s follow-up so far.

Taken together, we report a unique case of BHS, presenting with brief choreiform movements due to the stenosis in the right PCA, and simultaneously with intermittent dizziness associated with recurrent cerebellar strokes which were caused by dynamic stenoses of the nondominant left VA. The combination of noninvasive neuroimaging and hemodynamic investigations, combined with head rotational behaviors, is highly...
recommend for the diagnosis of BHS.

References


**Appendix: Authors**

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Diffusion-weighted imaging showing diffusion restriction in the left cerebellar hemisphere suggestive of acute infarction (A, red arrow-head). T2 weighted image showing multiple areas of hyperintensity in bilateral cerebellar hemispheres and pons suggestive of chronic ischemic changes (B, white arrow-heads). Head CTA revealing a patent but hypoplastic left VA (C, leftward arrow) and dominant right VA in comparison. Note the stenosis in the P1 segment of the right posterior cerebral artery and in P1 and P2 segments of the left posterior cerebral artery (C, downward arrows). Neck CTA showing that the left VA aberrantly originates from the aortic arch between left carotid artery and left subclavian artery, and dilates at the C5 cervical level (D, white arrow) then enters the C4 transverse foramen, rather than the typical C6 level. The axial planes of CTA showing asymmetrical VAs in either V1 segment.
(E, white arrow) or V2 segment (F, white arrow). Yellow lines in the right panels indicate the corresponding transverse levels.
In the neutral head position (A), SPECT demonstrates adequate blood flow in the left cerebellar hemisphere (B, dashed frame); TCD displays a normal spectrum with blunt peaks and gentle slopes (C, white arrow) and the pulsatility index (PI) is 0.94. When the patient turns his head 45° to the left (D), the blood flow in left cerebellar hemisphere is decreased (E, dashed frame); the peaks of TCD spectrum become sharp (F, white arrow) and PI is increased to 1.64, indicating a higher flow resistance. In the neutral head position shown in the reconstructed CTA (G), there is no occlusion of
the left VA except for a vessel dilation at the C5 cervical vertebral level. During
leftward head rotation to approximately 45° (H) or 90° (I), a clear stenosis could be
seen along the dilation of the left VA at the transverse process of C5 cervical vertebral
level. The area framed in red are enlarged and reconstructed in the insets, and the
narrowing sites are indicated by white arrows. The results of the CTA shown in G and
I, are confirmed by reconstructed DSA performed in the neutral head position (J,
black arrow) and 90° head rotations leftwards (K, black arrow), respectively.
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