A Subtype of Bow Hunter's Syndrome Requiring Specific Method for Detection: A Case of Recurrent Posterior Circulation Embolism due to “Hidden Bow Hunter's Syndrome”

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Objective: A condition that presents with recurrent embolism due to “hidden bow hunter's syndrome,” a subtype of bow hunter's syndrome with a different pathogenic mechanism, is reported.

Case Presentation: The patient was a 78-year-old male who exhibited recurrent embolic stroke of the posterior circulation territory resistant to medical treatment. DSA showed occlusion of the right vertebral artery (VA), but dynamic left vertebral arteriography (VAG) presented no change in blood flow. Since indirect signs of occlusion and recanalization of the right VA were retrospectively obtained, dynamic right VAG was performed again on another day, which revealed that the occluded right VA in the neutral neck position recanalized when the neck was rotated to the left. Suspecting that thrombi formed during occlusion scattered with recanalization, we performed embolization of the parent artery in the distal right VA for the prevention of recurrence.

Conclusion: This pathological condition should be considered as a differential diagnosis if unexplained ischemia of the posterior circulation is accompanied by unilateral VA occlusion.

Keywords ▶ bow hunter's syndrome, vertebral artery stump syndrome, cryptogenic stroke, vertebrobasilar insufficiency, rotational vertebral artery compression

Introduction

Bow hunter's syndrome (BHS) is a disorder that causes disturbance of the posterior circulation due to stenosis or occlusion of the vertebral artery (VA) associated with movements such as rotation of the cervical spine.^{1,2)}

Although it is a rare condition, it must be remembered as a differential diagnosis in the evaluation of the cause of ischemia of the posterior circulation territory.

In this report, we present a case that exhibited recurrent embolism of the posterior circulation due to “hidden BHS” caused by a different pathogenic mechanism from BHS.

Case Presentation

The patient was a 78-year-old male exhibiting recurrent episodes of dizziness and unsteady gait. He had histories of hypertension, cervical spondylosis, surgery for stomach cancer, and surgery for prostate cancer.

The patient began to note recurrent narrowing of the visual field and episodes of dizziness and unsteady gait from about 1 month before the visit. One day, he became unable to rise and was transported to our hospital by an ambulance. Findings on arrival: Mild motor ataxia of the left upper and lower extremities was noted. Diffusion-weighted MRI

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Fig. 1 On the initial head MRI, DWI showed high-intensity areas in the left PICA and left SCA territories (**B**). FLAIR imaging showed a high-intensity area in the left PCA territory in addition to the above regions (**A**). At recurrence, MRI showed new DWI high-intensity areas in the bilateral SCA and left PCA territories (**C**). DWI: diffusion-weighted MRI; PCA: posterior cerebellar artery; PICA: posterior inferior cerebellar artery; SCA: superior cerebellar artery

(DWI) of the head showed high-intensity areas in the left posterior inferior cerebellar artery (PICA) and left superior cerebellar artery (SCA) territories. FLAIR imaging revealed high-signal areas in the posterior cerebellar artery (PCA) territory in addition to the above regions (**Fig. 1A** and **1B**). Basi-parallel anatomical scanning (BPAS) MRA showed no hypoplasia in the bilateral Vas, but, on MRA, the right VA was not delineated from the origin, and the left VA was narrowed in the cervical region (**Fig. 2**). Blood biochemistry tests or clotting tests showed no particular abnormality. Electrocardiogram (ECG) was negative for arrhythmia.

Course after admission: The subacute lesions that were hyperintense on FLAIR imaging were consistent with the symptoms observed from 1 month before the visit, and the lesions that were hyperintense on DWI were considered to have occurred on the day of the visit. The condition was judged to be serial episodes of embolic stroke in the posterior circulation territory. The pathogenic mechanism was considered likely to be angiogenic embolism due to stenosis of the left cervical VA or embolism associated

with atherosclerotic occlusion of the right VA. The patient was admitted on the day of the visit and was treated with medication by continuous intravenous infusion of argatroban, intravenous injection of edaravone, and oral administration of aspirin at 100 mg. However, the patient newly exhibited dysarthria and exacerbation of right motor ataxia on the 4th hospital day. MRI showed new DWI high-intensity areas in the bilateral SCA territories and the left PCA territory (**Fig. 1C**). No marked change was noted in MRA findings. To determine the source of emboli, trans-thoracic echocardiography, cervical ultrasonography, Holter ECG, and MRI plaque imaging of the aortic arch to the carotid arteries were performed, but no clear source of emboli was detected.

Angiography: DSA was performed for the first time on the 6th hospital day (**Fig. 3**). While left vertebral arteriography (VAG) showed stenosis at the C6/7 level, it was mild. The distal right VA was delineated via the VA union with stagnation of the contrast medium. We also performed dynamic left VAG in consideration of the possibility of BHS, but the change in the stenosis rate associated with

Fig. 2 On BPAS, the right VA was not hypoplastic. On MRA, however, the right VA was not visualized from the origin (**A** and **B**) double arrows, and the left VA was stenosed in the cervical region (**C**) arrowhead. BPAS: basi-parallel anatomical scanning; VA: vertebral artery

Fig. 3 On subclavian artery angiography (**A** and **B**), the right VA was slightly visualized at the origin but was occluded in the distal portion (**A**) arrow. The left VA showed stenosis at the C6/7 level, but it was mild (**B**) arrowhead. Dynamic angiography showed slight changes in the stenosis rate with neck rotation but no marked change in blood flow (**E** and **F**) arrowhead. On left VAG, the distal part of the right VA was retrogradely visualized and was stagnated (**C** and **D**) double arrows, but, on dynamic angiography, the distal part of the right VA disappeared during left rotation of the neck (**E** and **F**) double arrows, and laminar flow was noted in the BA. The anterior spinal artery was visualized from the proximal side of the union of the left VA (**C**). BA: basilar artery; VA: vertebral artery; VAG: vertebral arteriography

Fig. 4 On dynamic right VAG, the right VA was occluded during right rotation of the neck and in the neutral position (**A** and **B**) but was patent during left rotation (**C**). In addition, angiography during the patent period revealed marked stenosis of the right VA at the C6/7 level and a radiolucent image suspected to be a thrombus distally to the stenosed area (**D**) arrowhead. Cone beam CT showed stenosis of the right VA due to compression by a bony spur of the Luschka's joint at C6/7 (**E** and **F**) arrow. VA: vertebral artery; VAG: vertebral arteriography

neck rotation was slight, and no clear change in blood flow was noted. The right VA was slightly visualized at the origin but was occluded in the distal part. We had not performed dynamic right VAG. After the examinations, we retrospectively evaluated dynamic left VA angiograms and confirmed disappearance of delineation of the right VA via the VA union during left rotation of the neck and laminar flow in the basilar artery (BA). We judged this to be an indirect finding suggesting patency of the right VA associated with left rotation of the neck. Then, we attempted to confirm recanalization of the right VA by dynamic cervical ultrasonography, but the attempt failed due to poor delineation of the VA. We, therefore, performed second DSA and dynamic right VAG on the 10th hospital day (**Fig. 4**). A catheter was inserted to the origin of the right VA, and the contrast medium was injected slowly by hand without applying pressure. While the right VA was occluded in the neutral head position, it was recanalized during left rotation and anterior flexion. Also, angiography during recanalization showed marked stenosis of the right VA at the C6/7 level and a radiolucent image suggestive of a thrombus distal to the stenosed area. Cone beam CT by similar hand injection showed stenosis of the right VA due to compression by a bony spur of the Luschka's joint at C6/7. Neck imaging was performed in about 30° and 60° rotation, and recanalization of the right VA was observed only at 60°.

Treatment plan: From the above imaging findings, the pathogenic mechanism in this patient was judged to be embolism caused by scattering of thrombi formed in the right VA associated with its occlusion and recanalization depending on the neck position. We evaluated medical treatment including anticoagulant therapy as well as decompression by removal of the bony spur, anterior fixation of the cervical spine, and occlusion of the parent artery in the distal part of the right VA as therapeutic options for the prevention of recurrence and selected occlusion of the parent artery for the safety and reliability. Until endovascular treatment, neck rotation was restricted with a cervical collar, and argatroban was administered continuously. From 1 week before endovascular treatment, antiplatelet therapy was increased to aspirin at 100 mg plus clopidogrel at 75 mg. **Endovascular treatment:** On the 29th hospital day, endovascular treatment was performed under intravascular local anesthesia. A 6 Fr sheath was inserted via the right femoral artery. A 6 Fr guiding catheter was placed in the left VA. A PX SLIM 90° (Penumbra Inc., Alameda, CA, USA) was inserted and placed in the right VA near the craniocervical junction via the VA union. A Penumbra coil Complex ExtraSoft 2 mm × 4 cm (Penumbra Inc.) was inserted, and a cage was formed in the horizontal segment, followed by insertion of the same coils while reducing the size. Sufficient embolization could be achieved according

Fig. 5 Preprocedural right VAG (A). A microcatheter was inserted from the left VA and placed in the right VA near the craniocervical junction via the VA union (B). Coil embolization was performed in the horizontal segment (C). On postprocedural left VAG, reflux to the right VA was reduced (D). Right VAG showed termination of the antegrade blood flow after the bifurcation of a muscle branch on the proximal side of the coil mass (E). VA: vertebral artery; VAG: vertebral arteriography

to images after insertion of six coils (21 cm). Left VAG after the procedure showed reduced reflux to the right VA. Right VAG showed interruption of antegrade delineation on the proximal side of the coil mass. No intraprocedural symptoms or change in vital signs were noted (**Fig. 5**).

Postprocedural course: No apparent periprocedural complication was noted except that MRI showed asymptomatic microinfarctions in the right cerebellar hemisphere. After endovascular treatment, no recurrence of embolic stroke was observed, and antiplatelet regimen was reduced to aspirin at 100 mg alone. On the 56th hospital day, the patient was transferred to a rehabilitation hospital with an modified Rankin Scale (mRS) score of 2. The patient remained free of recurrence 9 months after endovascular treatment.

Discussion

BHS and “hidden BHS”

BHS is a pathological condition in which the posterior circulation is impaired by stenosis or occlusion of the VA associated with rotation or flexion of the cervical spine, causing transient ischemic attacks and embolic stroke.^{1,2)}

It used to be considered to occur frequently at the atlantoaxial level, but a recent review of the literature suggested that the percentage of BHS involving lower cervical vertebral levels is also high.²⁾ In BHS, the VA is patent in the neutral position but is stenosed or occluded due to bony compression during movements. Compression often occurs when the neck is rotated to the opposite side of the lesion. In the present case, the VA was occluded due to bony compression in the neutral position but was recanalized during neck rotation to the unaffected side unlike usual cases. In this patient, thrombi are considered to have formed due to congestion of blood at a site in the VA occluded in the neutral neck position and have scattered to peripheries with restoration of the blood flow due to relief of occlusion with neck movements (**Fig. 6**). Since conditions similar to our case have not been reported to our knowledge, we define this pathological condition as “hidden BHS.” This condition, in which the VA on the affected side is occluded in the neutral neck position, cannot be detected by common blood vessel examinations and remains undetected unless angiography is performed with suspicion of recanalization of the occluded VA with neck movements. It is considered a very rare condition, but, if unexplained embolic stroke of the posterior

Fig. 6 In BHS, the VA is patent when the neck is in the neutral position but is stenosed or occluded under bony compression during neck movements (**A**). In hidden BHS, the VA is occluded due to bony compression in the neutral neck position but is recanalized when the neck is rotated to the intact side, allowing thrombi formed in the VA to embolize the distal area (**B**). BHS: Bow hunter's syndrome; VA: vertebral artery

circulation is accompanied by occlusion of the unilateral VA, examinations with this condition in mind are necessary.

Methods to diagnose hidden BHS

Based on the present case, we evaluated findings that suggest hidden BHS. They include 1) recurrent embolic stroke localized in the posterior circulation territory without a clear source of emboli as pathological features, and imaging findings such as 2) occlusion of the unilateral VA and no hypoplasia or aplasia on BPAS MRI, 3) partial visualization of the origin and distal portion of the occluded VA on MRA or DSA, 4) disappearance of reflux to the

contralateral VA and appearance of laminar flow in the BA despite no decrease in blood flow of the ipsilateral VA on dynamic DSA of the patent VA, and 5) bony spurs and instability on radiography or plain CT of the cervical spine. 4) may be overlooked on DSA aimed to demonstrate BHS. On dynamic imaging, if the unilateral VA is occluded in the neutral neck position; usually, the patent VA must be assumed to be the affected side, and attention must be paid to the patent VA for direct confirmation of its temporary occlusion. At this time, however, images should be examined with attention also to 4), which is an indirect sign of temporary recanalization of the occluded VA.

In our patient, dynamic angiography was performed by low-velocity and low-pressure hand injection from the affected side with cone beam CT to establish the diagnosis. By this method, however, there is a risk of distal scattering of thrombi from the site of occlusion. Particularly, when 4) described above is observed, dynamic VAG of the occluded side, which involves the risk of distal embolism, may be omitted, because the possibility of hidden BHS is considered high. In addition, modalities such as MRA, cervical ultrasonography, and transcranial ultrasonography have been reported to be useful for the diagnosis of BHS,³⁻⁵⁾ and implementation of these mildly invasive examinations should be considered as they may demonstrate recanalization of the occluded VA. In our patient, we performed cervical ultrasonography with changes in head position but could not diagnose the condition due to difficulty in visualization of the VA. We did not perform MRA or transcranial ultrasonography with changes in head position.

Treatments for hidden BHS

Although BHS has been treated primarily by surgical procedures such as decompression and fixation of the cervical spine⁶⁻⁸⁾ and conservative approaches such as anticoagulant therapy and neck immobilization⁹⁾ or occasionally by stent placement¹⁰⁻¹²⁾ and parent artery embolization,¹³⁾ no standard treatment has been established. Basically, these options should also be evaluated as treatments for hidden BHS, but the possibility of recurrence remains after surgical and conservative treatments. In our patient, who was asymptomatic when the VA was occluded, prevention of scattering of thrombi by occlusion of the parent artery was considered optimal from the viewpoints of the safety of the procedure and certainty of prevention of recurrence. In endovascular treatment, the affected VA was embolized extradurally distal to the site of stenosis across the VA union from the intact VA. It is necessary to check the presence of perforating branches and the anterior spinal artery in advance to prevent obliterative complications. It may be difficult to approach the lesion from the intact side due to a sharp angle of bifurcation of the VA union and narrowing of the contralateral VA, but embolization on the proximal side of the stenosed area of the affected VA should not be performed because of the possibility of persistence of blood flow via the anastomoses from vessels such as the deep carotid artery, ascending carotid artery, and external carotid artery.

On the other hand, blood flow of the VA may be impaired bilaterally in BHS. If occlusion of the parent artery has

been performed for the affected VA, future treatment options may be restricted in the event of blood flow disturbance also in the contralateral VA. Accordingly, treatment must be selected after careful individual evaluation.

Embolic stroke of the posterior circulation territory

Cardiogenic embolism and atherosclerosis of major vessels are common causes of embolic stroke in the posterior circulation territory. However, if infarction recurs in the posterior circulation territory alone, angiogenic embolism due to atheroma or stenosis/occlusion of the origin of the vertebrobasilar system and arterial dissection is possible as differential diagnosis.¹⁴⁻¹⁶⁾ Particularly, embolism from stenosis/occlusion of the VA origin is attracting attention as VA stump syndrome.¹⁶⁾ Regular search for the source of emboli is carried out, but it is often impossible to identify the cause. Dynamic angiography is usually omitted unless there are findings that warrant a suspicion of BHS. If, on the other hand, the unilateral VA is not visualized on MRA, and the vascular contour observed by BPAS MRI is inconsistent, atheromatous occlusion and arterial dissection are often suspected, but there is also the possibility of reversible occlusion due to compression as in our patient. If the source of emboli cannot be determined in a patient with recurrent embolic stroke localized in the posterior circulation territory accompanied by occlusion of a unilateral VA, it is worth suspecting hidden BHS and performing dynamic angiography. The conditions of hidden BHS may be "hidden" in patients in whom the cause of the disorder has not been identified. Accumulation of cases is awaited to clarify the frequency of hidden BHS in infarctions of the posterior circulation territory.

Conclusion

Diagnostic approaches in consideration of hidden BHS should be evaluated if unexplained embolic stroke in the posterior circulation territory is accompanied by unilateral VA occlusion. Embolization of the parent artery may be useful for the prevention of recurrence of this condition.

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Disclosure Statement

Neither the first author nor any of the coauthors have any conflicts of interest to disclose regarding this paper.

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