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## Case Report

## Bow-hunter's syndrome due to left C7 schwannoma in a patient with bilateral absence of the posterior inferior cerebellar artery

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## 1. Introduction

Bow-hunter's syndrome (BHS), or rotational vertebral artery occlusion, is characterized by recurrent episodes of paroxysmal vertigo, nystagmus, and syncope induced by horizontal head rotation [1,2]. Typically, head rotation elicits stenosis or occlusion of the vertebral artery (VA) at the C1/C2 level, leading to compromised blood flow in the territory of the vertebro-basilar system [1–5]. As the course of the VA ranges from the subclavian artery to the craniocervical junction, stenosis or occlusion can occur anywhere in its course. The pathophysiology of BHS is not restricted to the C1/C2 level; it has been classified into atlanto-axial-, subaxial-, and mixed type BHS [1].

The anatomical structures of the posterior circulation are also thought to be associated with the onset of BHS [1,2]. Under normal conditions, when VA occlusion or stenosis occurs, the contralateral VA can contribute to the territory of the vertebro-basilar system. On the other hand, patients with poor collateralization, including a hypoplastic VA ending as posterior inferior cerebellar artery (PICA), an absent contralateral VA, or no collateral flow through the anterior circulation, can manifest vertebro-basilar insufficiency [1,2]. As the incidence of VA anomaly at the craniocervical junction is relatively high [6], a clear understanding of the course of the VA and the branching arteries of the vertebro-basilar system such as

the PICA and the anterior inferior cerebellar artery (AICA) is important.

Subaxial BHS associated with spondylotic changes of the cervical spine has been documented, however, there are few reports of BHS due to compression by a tumor [7–10]. We report a rare case of BHS in which VA compression was evident at the left C6/7 level where the VA entered the C6 transverse foramen above a C7 schwannoma. The patient was successfully treated by the posterior approach.

## 2. Case report

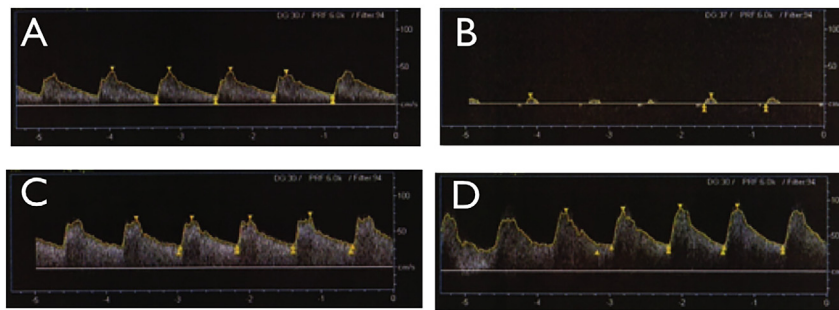
This 43-year-old woman presented with mild left arm pain, dysesthesia and vertigo. She had been in good health until 2 years ago when she first experienced gradually worsening pain at left lateral aspect of the arm and vertigo upon turning her head toward the left. Neither an orthopedic surgeon nor an otolaryngologist identified abnormalities. Her vertigo did not appear when she turned her head to the right. Duplex ultrasonography performed at a local neurosurgical clinic showed that the blood flow in the left VA was lower when her head was turned to the left than when it was in the neutral position or turned to the right (Fig. 1). BHS was suspected and she was referred to Kagoshima University Hospital for further evaluation. Her past medical history was unremarkable. Neurological examination returned almost normal findings except for pain and dysesthesia from the left shoulder to the lateral aspect of the left upper arm. Plain X-ray films (oblique view) showed dilation of the transverse foramen at the C6/7 level on the left. There was no instability on dynamic studies. Cervical magnetic resonance imaging (MRI) disclosed a heterogeneously enhanced mass at the left C6/7 transverse foramen to the paravertebral area (Fig. 2A). On brain magnetic resonance angiograms (MRA) a vertebro-basilar structure including two posterior cerebral arteries without a posterior communicating artery (PcomA), and two AICAs without an obvious PICA were observed (Fig. 2B). A 3D computed tomography angiogram (CTA) demonstrated stretching of the VA at the C6 transverse foramen (Fig. 2C). These findings indicated that the vessel was stretched between the tumor located below the vessel and the C6 transverse process above.

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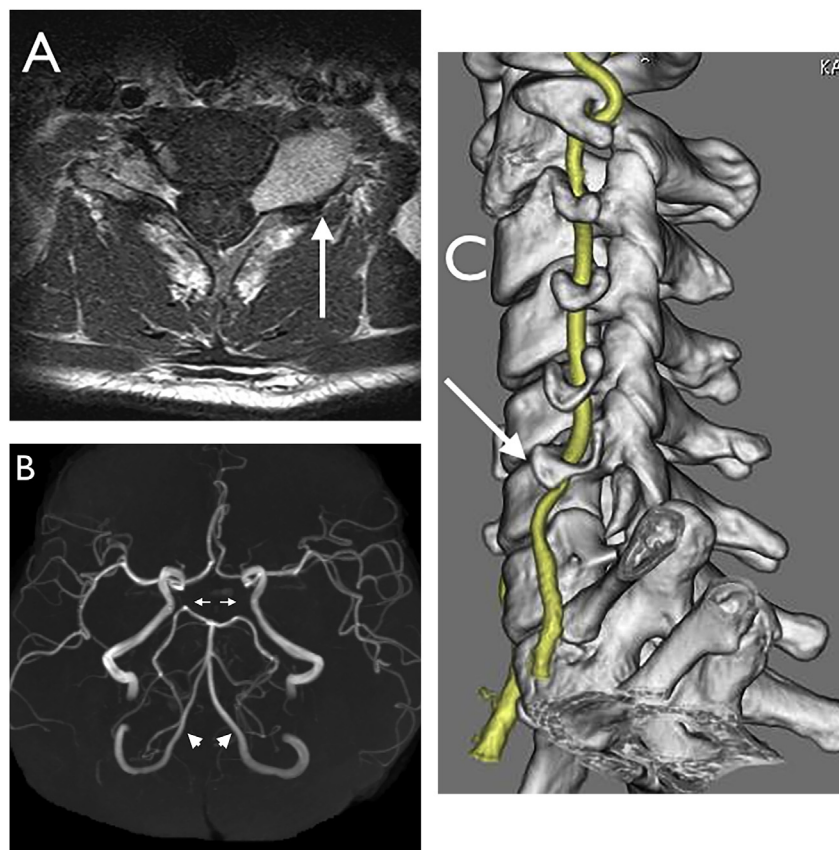
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**Fig. 1.** Duplex ultrasound images of the left vertebral artery (VA) showing adequate flow with the head in neutral position (A). Note decreased flow when the head is turned to the left (B). Duplex ultrasound images of the right VA showing adequate flow with the head in neutral position (C) and turned to the left (D).

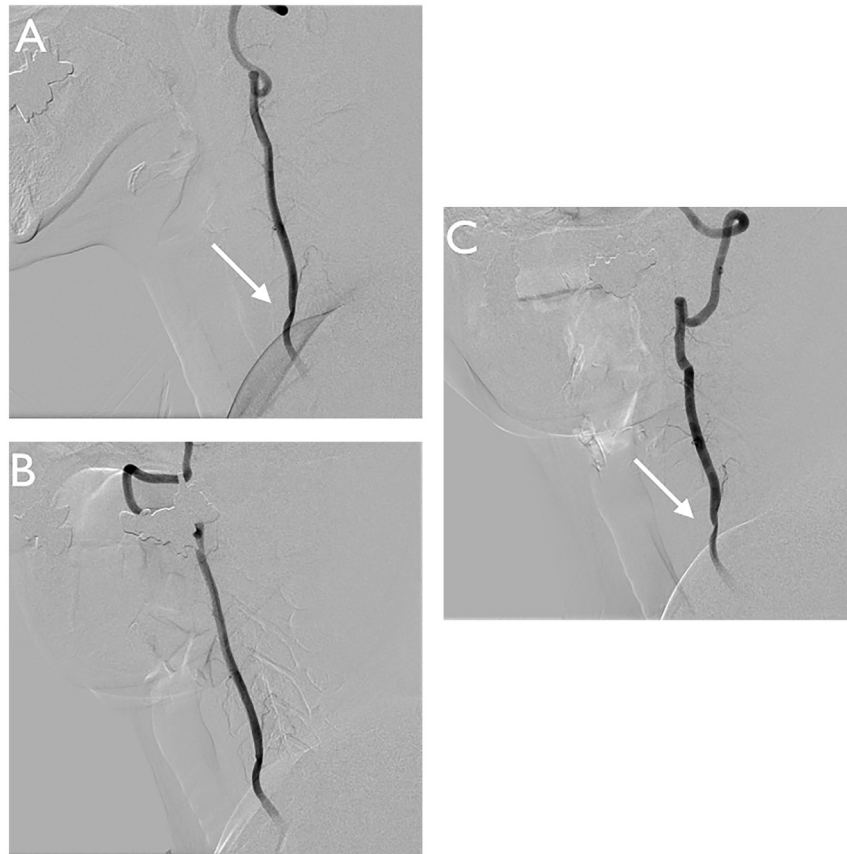


**Fig. 2.** (A) Axial Gd-enhanced, T1-weighted MR image at the C7 level showing a heterogeneously enhanced mass at the left transverse foramen (arrow). (B) Head magnetic resonance angiography disclosing a vertebro-basilar system without an obvious posterior communicating artery (arrow) and posterior inferior cerebellar artery (arrow head). (C) 3D CT angiography revealing an indentation at the C6 transverse process (arrow).

Direct angiography, performed to confirm VA compression during head rotation, revealed severe narrowing of the left VA at the left C6 transverse process when she turned her head to the left and mild stenosis when her head was in the neutral position or turned to the right (Fig. 3). During the maneuver of left head-turning she experienced vertigo. Therefore, we performed angiography to confirm the presence of stenotic changes in the left VA rather than its occlusion. No bilateral PICAs were detected; the cerebellar area was supplied by both developed AICAs (Fig. 4A and B). Each AICA was demonstrated only upon ipsilateral injection of contrast medium (Fig. 4A and B). When her head was turned to the left, the AICA on the affected left side was not observed on the contralateral VA angiogram (Fig. 4C); an ipsilateral VA angiogram showed delayed filling of the left AICA (Fig. 4D). Under a

presumptive diagnosis of schwannoma at the left C7 root (Eden type 4) [11], she underwent tumor removal via the posterior approach with facetectomy and fusion.

With the patient under general anesthesia and in the prone position with the head fixed with a Mayfield head frame, we incised the skin to expose the dorsal surface of the lamina from C5 to T2. With the aid of a navigation system we inserted a lateral mass screw at the left C5 lateral mass and a pedicle screw at the left T1 pedicle. The tumor was removed subfascially after facetectomy at the left C6/7. Doppler ultrasound confirmed pulsation in the left VA at the bottom of the tumor cavity. There was no tumor in the intradural portion. The screws were fixed with a rod and autologous bone tips from the iliac crest were inserted. The skin was closed anatomically and a subcutaneous drain was placed.



**Fig. 3.** Left vertebral artery angiogram with the head facing forward (A), turned to the right (B), and to the left (C). Note mild stenosis at the C6 transverse process (A, arrow) that is more severe when the head is turned to the left (arrow) (C).

Her postoperative course was uneventful and her rotational vertigo and pain in the left arm disappeared. 3D CTA confirmed a decrease in left VA stenosis (Fig. 5A) and MRI performed 6 months after the operation revealed almost total tumor removal (Fig. 5B).

### 3. Discussion

Underlying diseases such as atlantoaxial subluxation, rheumatoid arthritis, trauma, congenital anomaly, and tumor may elicit BHS due to vertebro-basilar insufficiency produced by the reversible obstruction or stenosis of the dominant VA at the atlantoaxial level upon head rotation [1–5]. Although a tumor is usually thought to elicit BHS [4,5], of the few reports documenting the pathophysiology of tumor-associated BHS (Table 1) [7,8,11] two were published before the MRI era. Conventional cervical X-ray followed by vertebral angiography detected a cervical tumor and VA occlusion in two earlier cases.

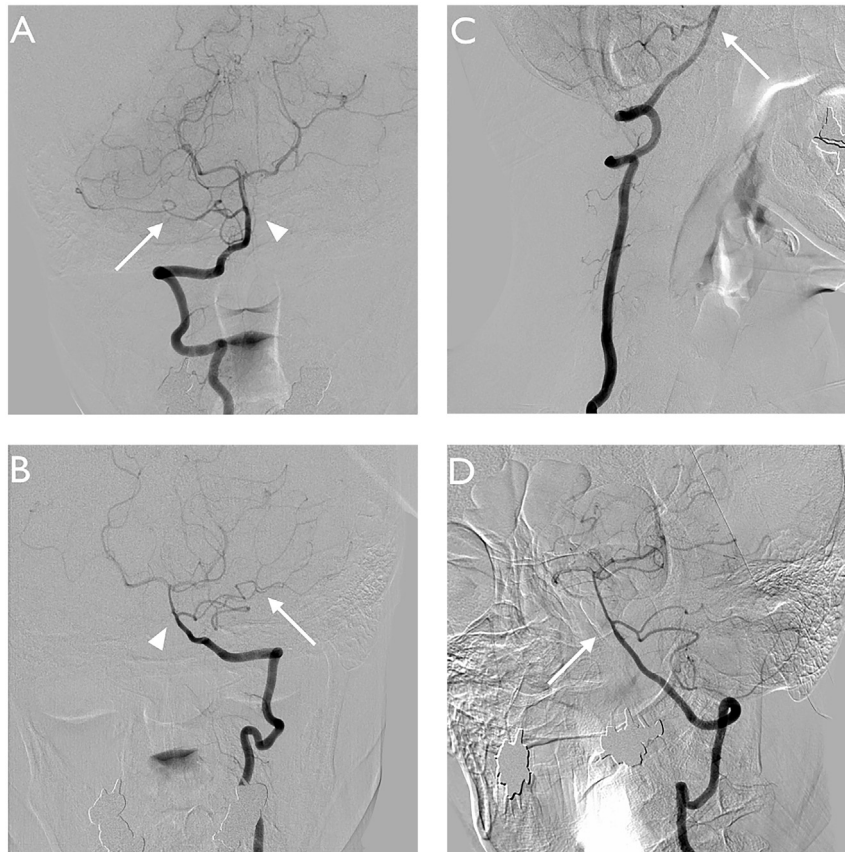
Geissinger et al. [7] reported a 51-year-old man who lost consciousness when he turned his head to the left. Plain radiographs showed enlargement of the left foramen at C3/4 and angiography revealed occlusion of the left- and patency of the right VA. He underwent tumor removal via the posterior approach. The histological diagnosis was neurofibroma. His symptoms persisted postoperatively and the authors, without angiographic confirmation, speculated that the contralateral VA was occluded upon head rotation.

George et al. [8] encountered a patient with osteochondroma arising from the left C2 transverse process that resulted in VA occlusion. Their patient suffered intermittent dizziness and

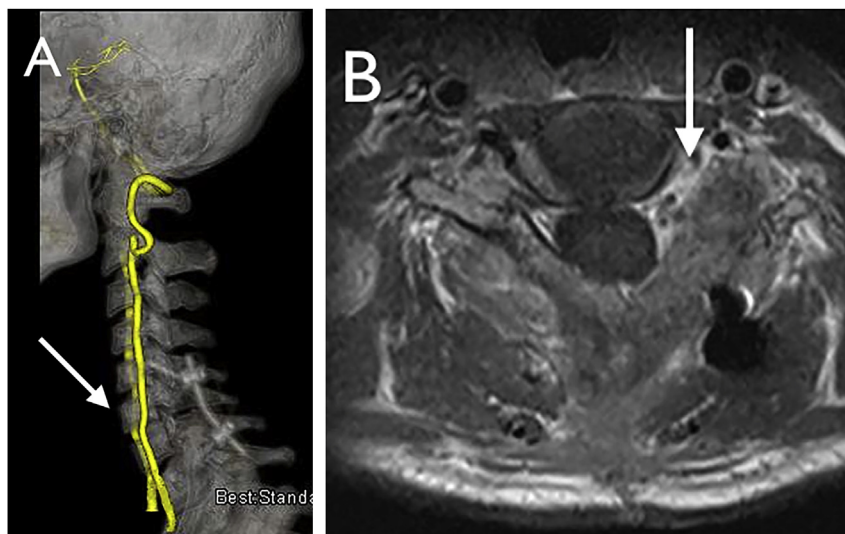
blurring of vision during head rotation. Complete surgical tumor removal was achieved via the anterolateral approach. They speculated that the cause of vertebro-basilar insufficiency was stretching of the contralateral VA at the C1–C2 level during head rotation.

Kalavakonda et al. [10] reported a rare case of a right C1 root schwannoma. Their patient suffered intermittent episodes of acute vertigo and dizziness upon head rotation to the right. Cerebral angiography showed that the right VA was occluded upon head rotation and extension. As in our case, the affected VA was not hypoplastic and did not terminate at the PICA. The contralateral VA appeared to be of the same size and was patent upon head rotation. Details on the intracranial PICA or AICA were not presented. The tumor was removed via a left extreme lateral approach; the pathological diagnosis was schwannoma. The symptoms resolved completely after the operation. The etiology at symptom onset was not discussed.

Anatomically, cervical tumors can result in stenosis or occlusion of the VA. George and Laurian [9] reported 51 patients with VA occlusion or stenosis due to an extracranial pathology. Compression was due to tumors in 34-, to osteophytes in 4-, and to fibrous bands in 5 patients; in 8 it was attributable to other factors. Symptoms of vertebro-basilar insufficiency were recorded in 13 of the 51 patients (25%). As only one of 34 patients (2.9%) who harbored tumors experienced vertigo during head rotation, stenosis or occlusion of the VA due to the tumor itself might not elicit BHS. The process leading to tumor-induced stenosis or occlusion is slowly progressive, therefore, hemodynamic changes may be well-tolerated. However, when the cerebral blood flow (CBF) in the contralateral



**Fig. 4.** Bilateral vertebral artery (VA) angiogram with the head facing forward revealing both anterior inferior cerebellar arteries (AICAs) (arrow) and absence of the posterior inferior cerebellar artery on the right (A) and left (B) VA. Note each AICA was demonstrated only upon ipsilateral injection of contrast medium (arrow head). Right VA angiogram with the head turned to the left (C) indicating the right AICA (arrow) without retrograde perfusion of the left VA. The left VA angiogram with the head turned to the left (D) shows delayed filling of the left AICA.



**Fig. 5.** (A) Postoperative axial Gd-enhanced, T1-weighted MR image at the C7 level showing total removal of the mass at the left transverse foramen (arrow). (B) 3D CT angiography revealing a dilated left vertebral artery at the C6 transverse process (arrow).

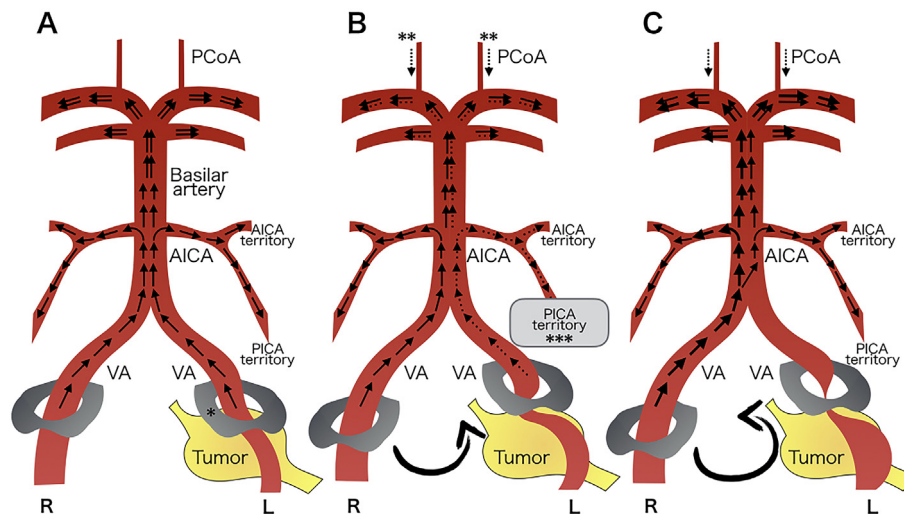
VA is disturbed, BHS may manifest. The pathophysiology in the previously-reported patients with BHS symptoms may have involved not only occlusion of the affected VA but also CBF disturbance in the contralateral VA during head rotation.

The vertebro-basilar system in our patient manifested several unique features (Fig. 6A). First, neither PcomA was observed on MRA studies. As PcomA absence is rarely found in autopsy studies, unvisualized PcomA on MRA images are thought to reflect

**Table 1**  
Literature review of bow-hunter syndrome due to tumor pathology.

Author (year)	Age/Sex	Pathology	Side/Location	Ipsilateral VA/Dominancy	Contralateral VA/Dominancy	Approach	Outcome
Geissinger (1972)	51/M	Neurofibroma	Left C3/C4	Occlusion at the C6 level/N.A.	Patent/Dominant side	Posterior approach	No change
George (1989)	33/N.A.	Osteochondroma	Left C2	Occlusion at the level of tumor/Hypoplastic	Patent/Dominant side	Anterolateral approach	Improved
Kalavakonda (2000)	52/M	Schwannoma	Left C1/C2	Patent/Normal size (Occlusion during rotation of the head to right or extension)	Patent/Normal size	Extreme lateral approach	Improved
Present study	43/F	Schwannoma	Left C6/C7	Patent/Normal size (Stenosis or occlusion during rotation of head to left)	Patent/Normal size	Posterior approach with facetectomy and fusion	Improved

N.A.; not available.



**Fig. 6.** (A) Scheme of the vertebro-basilar system of the present case showing hypoplasia of both posterior communicating arteries (PcomAs), both anterior inferior cerebellar arteries (AICAs) supplying the territory of the AICA and the posterior inferior cerebellar artery (PICA), and both vertebral arteries (VAs). The left VA runs between the tumor and the C6 transverse process (\*). (B) The scheme demonstrates “the cerebral blood flow (CBF) under the stenotic condition” in which the left VA is stretched between the tumor and the transverse process during head rotation. The estimated collateral flow through the PcomAs was low (\*\*). The left AICA is supplied by the ipsilateral VA, suggesting delayed or decreased CBF to the left PICA territory (\*\*\*). (C) Scheme demonstrating “the CBF under occlusive condition” in which the left VA is completely occluded between the tumor and the transverse process during head rotation. The left AICA is supplied by the contralateral VA because the right VA is large enough to compensate CBF in the vertebro-basilar system.

hypoplasia [12]. As collateral flow from the anterior circulation through the PcomA has been reported to depend on its size [13], we think that in our patient, the estimated collateral flow from the anterior circulation was insufficient during head rotation (Fig. 6B).

Second, both PICAs were absent and both AICAs provided posterior cerebellar circulation. The PICA usually originates from the VA intracranially as a single trunk, however, variations such as a duplicate PICA, VA termination at the PICA, extradural origins, and vessel absence has been observed. The reported incidence of bilaterally absent PICAs is 2–6.4% [14]. The symptoms of BHS vary depending on whether the AICA or PICA is the affected artery [15]. Inferior cerebellar infarction in the PICA territory is a common cause of isolated vertigo [15]. Ischemia of the labyrinth and the cochlea in the AICA territory results in vertigo, nystagmus, and auditory symptoms [15]. Our patient's chief complaint was vertigo without auditory symptoms during head rotation, indicating that the PICA—rather than the AICA territory—was primarily involved. As the territory of the PICA was more distal side from the origin of AICA (Fig. 6B), the CBF decrease or delay in the distal portion of the left AICA territory may have been attributable to stenosis or occlusion of the ipsilateral VA, resulting in our patient's transient vertigo.

Third, each AICA was demonstrated only upon ipsilateral injection of the contrast medium with the head in the neutral

position or turned to the left (Fig. 4). These observations indicate that the dominant source of supply to the ipsilateral AICA was the ipsilateral VA even under the stenotic condition (Fig. 6A and B). Angiographically, neither VA was dominant nor hypoplastic, therefore, the contralateral VA could take over the role of the affected VA in the presence of complete occlusion of the left VA (Fig. 6C). According to Stoeter et al. [16], switching of the supply to the left AICA from the left VA (Fig. 6B) to the right VA takes time (Fig. 6C). They reported delayed restoration of the antegrade flow in the VA after the resolution of stenosis in the subclavian artery in patients with subclavian steal syndrome. We speculate that dominance of the VA over the ipsilateral AICA and delayed restoration of the CBF to the left AICA was associated with the pathophysiology of our patient's BHS.

Cervical intra- and extra-foraminal schwannomas have been operated via posterior or anterolateral approaches, or a combination thereof [17–19]. When the tumor location is unilateral in the spinal canal and paravertebral space, posterior laminectomy and facetectomy has been suggested [19]. In our patient, the tumor was located mainly at the intervertebral foramen with extension to the paravertebral area; this compressed the VA anteriorly. We chose the posterior approach with facetectomy followed by unilateral screw fixation. This approach had two advantages. We were able to visualize the tumor; after facetectomy, we could differentiate

between the foraminal and the extradural tumor portion through a posterior corridor and we could determine whether there was an intradural component. The second advantage was avoidance of manipulation of the tortuous, stretched VA. On anterolateral views, the vessel was located in front of the tumor and its careful gross total removal from the posterior side avoided iatrogenic injury of the VA.

The necessity for fixation can be debated. Gu et al. [17] successfully operated 35 patients with dumbbell tumors using the posterior approach with facetectomy. They did not apply fixation and follow-up studies revealed no instability. On the other hand, Ando et al. [20] emphasized that unilateral posterior fixation is mandatory after tumor removal with facetectomy. Zdeblick et al. [21] observed significant segmental hypermobility after greater than 50% facet resection; they suggested that resection of the facet should be less than 50% to preserve the biomechanical function of the cervical vertebrae and to avoid the risk of cervical instability. Our patient manifested no postoperative instability and she suffered no associated symptoms. We are following her closely to monitor the appearance of radiological problems and clinical symptoms.

In conclusion, we reported a rare case of BHS in which compression of the VA was evident at the left C6 level where the VA entered the C6 transverse foramen above a C7 schwannoma. Our patient was successfully treated by the posterior approach. The causes of rotational VA occlusion vary and ours is the first documentation of a schwannoma at the left C7 leading to BHS. We think that our patient's BHS during head rotation was attributable to mechanical compression of the left VA by the left C6 transverse process and the C7 schwannoma, leading to a decrease in the CBF in the left AICA territory and to vertebro-basilar insufficiency.

#### Consent section

Written informed consent for publication was obtained from the patient.

#### Conflict of interest

The authors declare that they have no conflict of interest.

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