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Case Report

Bow hunter's syndrome in a patient with vertebral artery atresia, an arcuate foramen, and unilateral deafness: a case report

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ABSTRACT

Bow hunter's syndrome (BHS) is a rare cause of vertebrobasilar insufficiency that occurs when the vertebral artery (VA) is occluded on rotation of the head and neck. This dynamic occlusion of the VA can occur anywhere along its course after it arises from the subclavian artery. Although most cases are associated with compression by osteophytes, cervical spondylosis, or lateral disc herniation, BHS has a highly variable clinical course that depends on the patient's specific anatomy. Therefore, it may be important for clinicians to be aware of anatomical variants that predispose individuals to BHS. Here, we report on a patient with BHS who was found to have two uncommon anatomical anomalies: an atretic right VA and a left-sided arcuate foramen.

Introduction

Structures within the posterior cranial fossa, including the brainstem and cerebellum, are predominantly vascularized by the vertebral arteries (VAs). Ischemic events involving this posterior circulation account for 20% of all strokes and can result in devastating paralysis and even death [1]. Transient or incomplete ischemia within this vascular territory can cause vertebrobasilar insufficiency (VBI), which is characterized by symptoms such as vertigo, nystagmus, and syncope [2]. Rarely, VBI can be transiently induced by rotation of the head and neck [3]. This condition is known as Bow hunter's syndrome (BHS) or rotational VA occlusion. BHS was first formally described in 1978 by B. F. Sorensen in a patient who suffered

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Wallenberg syndrome after archery practice [4]. Although this initial case report described a patient who suffered permanent deficits after a rotation-induced occlusive event, the term BHS is now broadly used to describe a reversible condition.

The mechanism underlying typical BHS can be conceptualized as a "two-hit" model. The "first-hit" may be congenital or acquired, most often one VA is severely hypoplastic or stenotic. The "second-hit" occurs when the contralateral (and now dominant) VA is occluded by a bony-overgrowth, fibrous band, or herniated disc on rotation of the head [5,6]. Atlantoaxial compression of the dominant VA by a cervical spine osteophyte has been shown to be the most common cause of BHS [7]. However, this model is limited in describing all cases. The VA can be compressed anywhere along its course into the posterior cranial fossa. There are even case-reports documenting BHS after the occlusion of the non-dominant VA [8]. As a result, the clinical presentation and radiographic findings in patients with BHS are highly variable [3]. A more thorough understanding of both normal and abnormal VA anatomy may be helpful for clinicians while evaluating patients who present with symptoms of VBI. Here, we present a case of BHS in a patient with alterations in both the course and structure of the VAs.

Case report

A 59-year-old Caucasian female presented to her pain management physician for a preprocedural evaluation. She was scheduled to undergo a cervical facet rhizotomy, but on questioning the patient reported dizziness when she rotated her head to the right. These episodes lasted only a few seconds and were associated with lightheadedness and nausea. All symptoms resolved completely on returning her head to the neutral position. The patient reported that these symptoms had started within the past year, but they did not significantly affect her daily life. The dizziness could not be reproduced with any other head or neck movements. There was no recent history of illness, vomiting, tinnitus, changes in vision, postural instability, or syncopal episodes. Her medical history was significant for chronic bilateral neck and shoulder pain of 13 years and right-sided congenital sensorineural deafness. Her hearing loss was diagnosed at around age 3 years without apparent cause. She was not aware of any significant childhood illnesses, head trauma, exposure to ototoxic drugs, or a family history of hearing loss.

As a result of the patient's report of rotational dizziness, a magnetic resonance angiogram was ordered to rule out any major vascular occlusion or dissection. Axial time-of-flight noncontrast magnetic resonance angiogram demonstrated normal right and left internal carotid arteries. The left VA was of normal course and caliber. There was reduced flow through the right VA, which was suspicious for a VA dissection (Fig. 1). The patient was promptly sent to the emergency department for confirmatory computed tomography angiography. These images demonstrated a dominant left VA, which was the sole supply to the basilar artery. The V1-V4 segments of the right VA were markedly hypoplastic, but patent into the posterior cranial fossa. The V4 segment terminated as the right posterior inferior cerebellar artery and did not contribute toward the basilar artery (Fig. 2). On the left side of C1, a bony bridge can be seen extending from the lateral mass, over the groove for the left VA, and nearly connecting to the posterior arch (Fig. 3). There was no further evidence of dissection or infarction, so the patient was discharged from the emergency department and received the facet rhizotomy several days later.

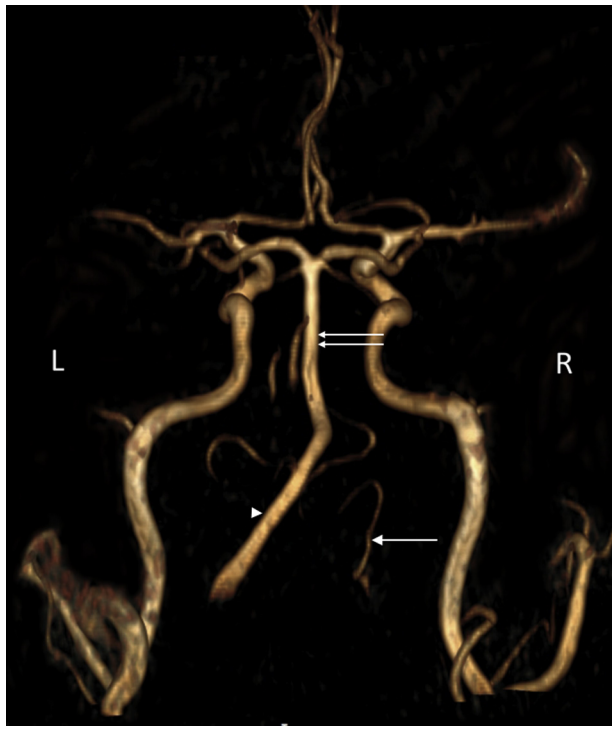

Fig. 1 – Posterior view of 3D time-of-flight magnetic resonance angiogram (MRA) rendering. The right vertebral artery is decreased in caliber (arrow). The left vertebral artery (arrowhead) and basilar artery (double arrows) are both of normal course and caliber.

Discussion

This case highlights several interesting variations in the course and structure of the VA. Upward of 69% of healthy adults are thought to have some degree of size discrepancy between the right and left VAs, with the left being larger than the right in most cases [9–11]. Vertebral artery hypoplasia (VAH) is a condition that occurs when one VA is significantly smaller than the other, but still connects to the basilar artery system [12]. The diameter cut-off used to define VAH varies depending on the study, but generally range from <2 to <3 mm [13]. VAH has been reported in anywhere from 2.34%-28.9% of healthy adults, with the right VA more frequently affected than the left [14–16]. Although most cases of VAH are asymptomatic, numerous studies have identified it as an independent risk factor for regional hypoperfusion and posterior circulation infarction [13,17,18].

The computed tomography angiography findings in our patient revealed a hypoplastic VA that terminated as the posterior inferior cerebellar artery instead of the basilar artery, which meets criteria for the diagnosis of vertebral artery

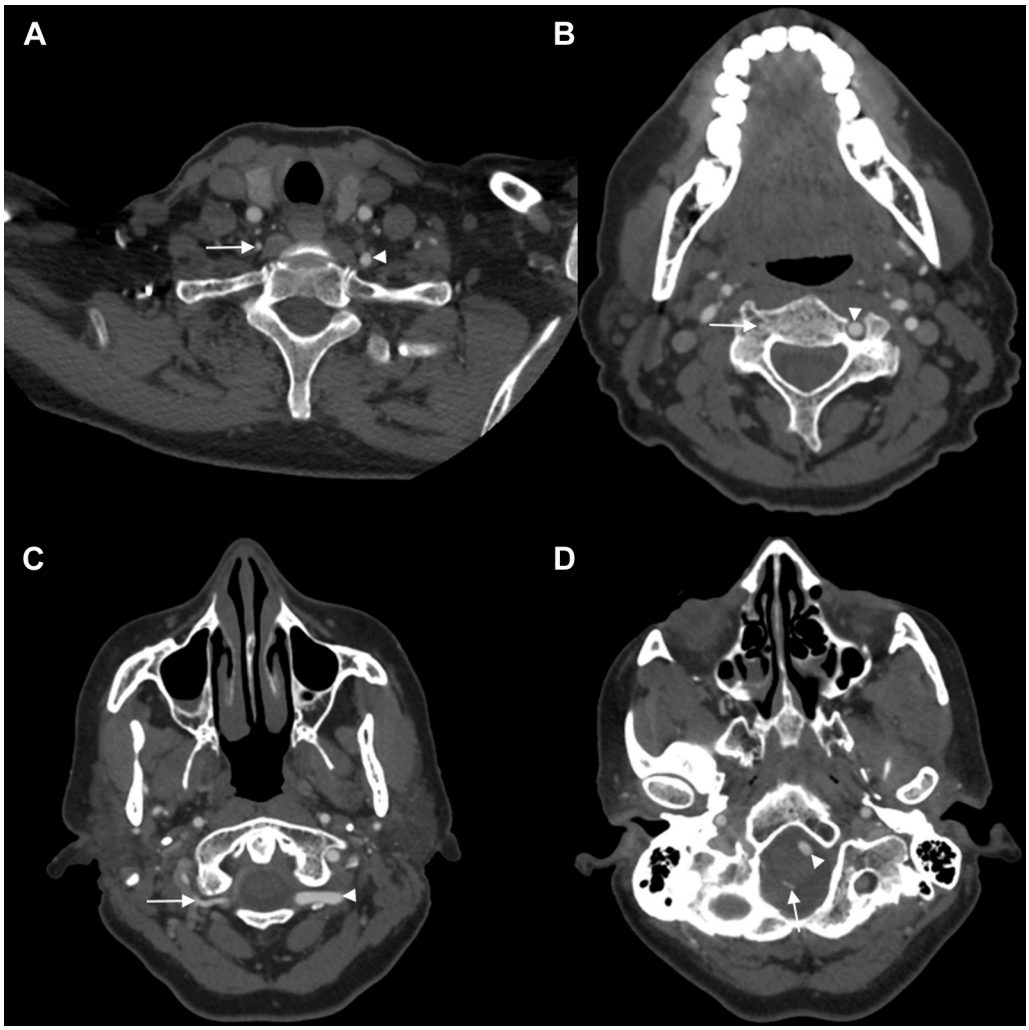

Fig. 2 – Axial section images of computed tomography angiography (CTA). Notice the marked size difference between the right vertebral artery (arrow) and left vertebral artery (arrowhead). (A) V1 segments of the vertebral arteries after they branch off the subclavian arteries at the level of the first thoracic vertebrae (T1). (B) V2 segments of the vertebral arteries as they ascend the transverse cervical foramina of C3. (C) V3 segments of the vertebral arteries as they emerge from the transverse cervical foramina and wrap around the posterior arch of C1. (D) V4 segments of the vertebral arteries within the posterior cranial fossa. The right vertebral artery has terminated as the posterior inferior cerebellar artery (arrow), and the left vertebral artery (arrowhead) continues to ascend anterior to the medulla to form the basilar artery.

atresia (VAA) [12]. VAA is thought to be relatively rare; one study estimated its prevalence at around 3.4% in the healthy population [15]. Currently, only around 30 case-reports specifically document VAA in the literature [19]. However, this is likely an underestimation of the actual prevalence of VAA. Numerous other studies document VAA as an incidental finding or in association with another condition [3,20].

Although under-reported, VAA is a clinically significant variant. Recent investigations have raised concerns about the safety of cervical spine manipulation in patients with VAA [19]. VA dissections have been reported after manipulation of the cervical spine [21,22]. Such an event could be especially devastating in these patients given the lack of collateral flow to the brainstem. Premanipulative provocative tests may help clinicians identify patients with poor collateral flow through the vertebrobasilar system [23]. Furthermore, it is important that radiologists be aware of VAA as it can be easily misdiagnosed as a VA dissection. A broader awareness of this anomaly may improve time-to-diagnosis and enable future researchers to establish a more accurate estimate of the actual incidence of VAA.

In our patient, we believe the right-sided VAA to be the "first hit" that predisposed her to BHS. A recent meta-analysis similarly identified VAA as a contributing factor in at least 9 of 126 patients with BHS [3]. On the left- side, the presence of a bony bridge extending over the groove for the VA on C1 likely served as the "second-hit" for BHS. This anatomical variant is most commonly known as an arcuate foramen, but has been given several different monikers in the literature including Kimmerle's anomaly and ponticulus posticus [24]. Arcuate foramina have been estimated to occur in around 16.7% of the general population and are thought to form as a result of abnormal ossification of the posterior atlantooccipital membrane [25,26]. Although generally asymptomatic, the

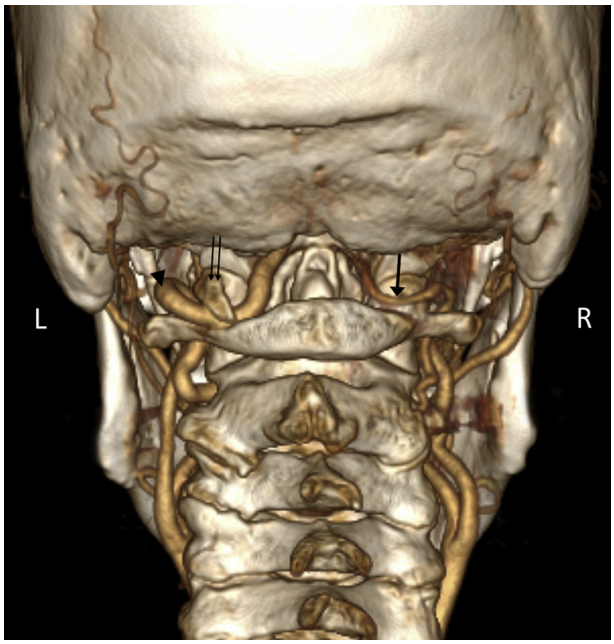

Fig. 3 – Posterior view of 3D-CTA rendering. The dominant left vertebral artery (arrowhead) can be seen passing underneath a bony bridge that spans between the lateral mass and posterior arch of C1 (double arrows). The atretic right vertebral artery (arrow) can be seen as it wraps around the normal posterior arch of C1.

presence of an arcuate foramen has been associated with migraine, chronic neck pain, and VA dissection [27–29].

Cadaveric studies have demonstrated a close adherence between the VA and the overlying bony bridge which tethers the VA onto the posterior arch of C1 [30]. In the present case, we hypothesize that the left-sided arcuate foramen limited the normal movement of our patient's dominant left VA such that during right-ward rotation of the head, it was stretched to the point of occlusion. Because of the lack of collateral flow through the right VA, there may have been a transient ischemic event in the cerebellar and labyrinthine circulation which induced the symptoms of BHS [31].

Surgical decompression or fusion procedures have been used with some success to treat cases of BHS [3]. As our patient's symptoms were not debilitating, she elected to simply avoid provocative head movements and has been generally asymptomatic during follow-up. The question that now remains is: could her right sided congenital sensorineural hearing loss be associated with her atretic right VA? Hearing loss in young children may be congenital or acquired [32]. Both categories encompass a myriad of conditions that cause deafness including syndromic and nonsyndromic genetic disorders, bony anomalies, infections, drug exposure, and prematurity [33–35]. As the details of our patient's early childhood medical history are relatively unknown, it remains unclear as to the exact etiology of her deafness. Disruption of blood flow to the inner ear is recognized as a cause of deafness, particularly in cases of sudden sensorineural hearing loss [36,37]. Several studies have attempted to correlate sudden hearing loss with other vascular anomalies of the circle of Willis with mixed results [38,39]. To our knowledge, there is no documented association between VA anomalies and hearing disturbances. Further investigation of VA anatomy in adults and children with idiopathic sensorineural hearing loss would be required to establish that the cooccurrence of these two findings in our patient are anything beyond coincidental.

This case illustrates how variations in the course and structure of the VA can predispose an individual to BHS. Clinicians evaluating patients with rotation-induced dizziness should be aware that these anomalies may alter their presentation and influence the clinical decision-making process. Vascular imaging that includes the symptomatic position could be especially illustrative in cases such as this. Furthermore, we present a novel association between VAA and unilateral deafness. Although we cannot establish causality between these two findings, their concordance in this patient may help direct future research and clinical evaluations of patients with unexplained unilateral congenital deafness.

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