

Case Report

Bow Hunter's Syndrome Caused by Atlo-Occipital Malformation: Dynamic Color Doppler Sonography Diagnosis

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Keywords

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- Atlanto-occipital assimilation
- Rotational vertebrobasilar insufficiency

Abstract

Bow hunter's syndrome (BHS) is a rare symptomatic vertebrobasilar insufficiency in which vertebral artery (VA) is mechanically occluded during head rotation. Various pathologic conditions have been reported as causes of bow hunter's stroke. To our knowledge, however, just two cases associated with complex cranio-cervical junction (CCJ) malformation with atlanto-occipital assimilation were reported. We present an unusual case of rotational occlusion of VA in a 12-year-old male with bow hunter's syndrome manifesting as repeated vertebrobasilar ischemic attacks induced by left head rotation during sport practice. Blood flow changes were dynamically detected with Color Doppler Sonography. Computer Tomography revealed the complex CCJ malformation. For accurate diagnosis and treatment of BHS, neuroimaging with cervical rotation is mandatory.

ABBREVIATIONS

BHS: Bow Hunter's Syndrome; VA: Vertebral Artery; CCJ: Cranio-Cervical Junction; MRA: Magnetic Resonance Angiography; CT: Computer Tomography; CDS: Color Doppler Sonography

INTRODUCTION

Bow hunter's syndrome (BHS), also known as rotational vertebral artery syndrome, is rare cause of symptomatic vertebrobasilar insufficiency that arises from mechanical compression of the vertebral artery (VA) at the atlanto-axial level during head rotation. BHS is usually demonstrated in adults and related symptoms are due to CCJ malformation associated with vascular comorbidity [1,2]. The term was coined by Sorensen in 1978, reporting a patient becoming symptomatic during archery [3]. The underlying pathology is dynamic stenosis or compression of the VA by abnormal bony structures with neck rotation or extension in many cases, such as osteophyte, disc herniation, cervical spondylosis, tendinous bands or tumours [4]. On the basis of the site of compression, BHS can be divided into i) atlantoaxial BHS, ii) subaxial BHS, and iii) mixed type. The left VA is involved in a significantly ($p < 0.0001$) higher number of BHS patients than the right one [4]. Due to the rarity of this pathology, there are no guidelines for its diagnosis and treatment [1]. However, it has been reported that neuroimaging with cervical rotation is mandatory for diagnosis and treatment of BHS. We present the

unusual case of a young patient with rotational occlusion of the right VA associated with a complex cranio-cervical junction (CCJ) malformation without any vascular pathology. To our knowledge, this is the third case of association between BHS and complex CCJ malformation such as atlanto-occipital assimilation.

CASE PRESENTATION

A 12-year-old male presented at our Emergency Department with a history of vertigo and drop attacks during tennis practice. In particular, hyperextension and right head rotation (smash tennis position) were able to immediately produce dizziness, vertigo and fall without loss of consciousness. Symptoms always resolved completely within few seconds when he returned his head to the neutral position and in supine position, without any neurological consequence. A physical and neurological examination was normal. He previously had played football without any neurologic symptoms.

Considering his young age, Color Doppler Sonography (CDS) and Magnetic Resonance Angiography (MRA) were performed as first-line imaging. Dynamic CDS examination, both in clinostatic and orthostatic position and during head rotation, showed occlusive flow pattern in the right VA when the patient turned his head 45° right and during hyperextension (Figure 1). Blood flow velocity quickly increased and symptoms disappeared when head rotation was released into the neutral position. Moreover,

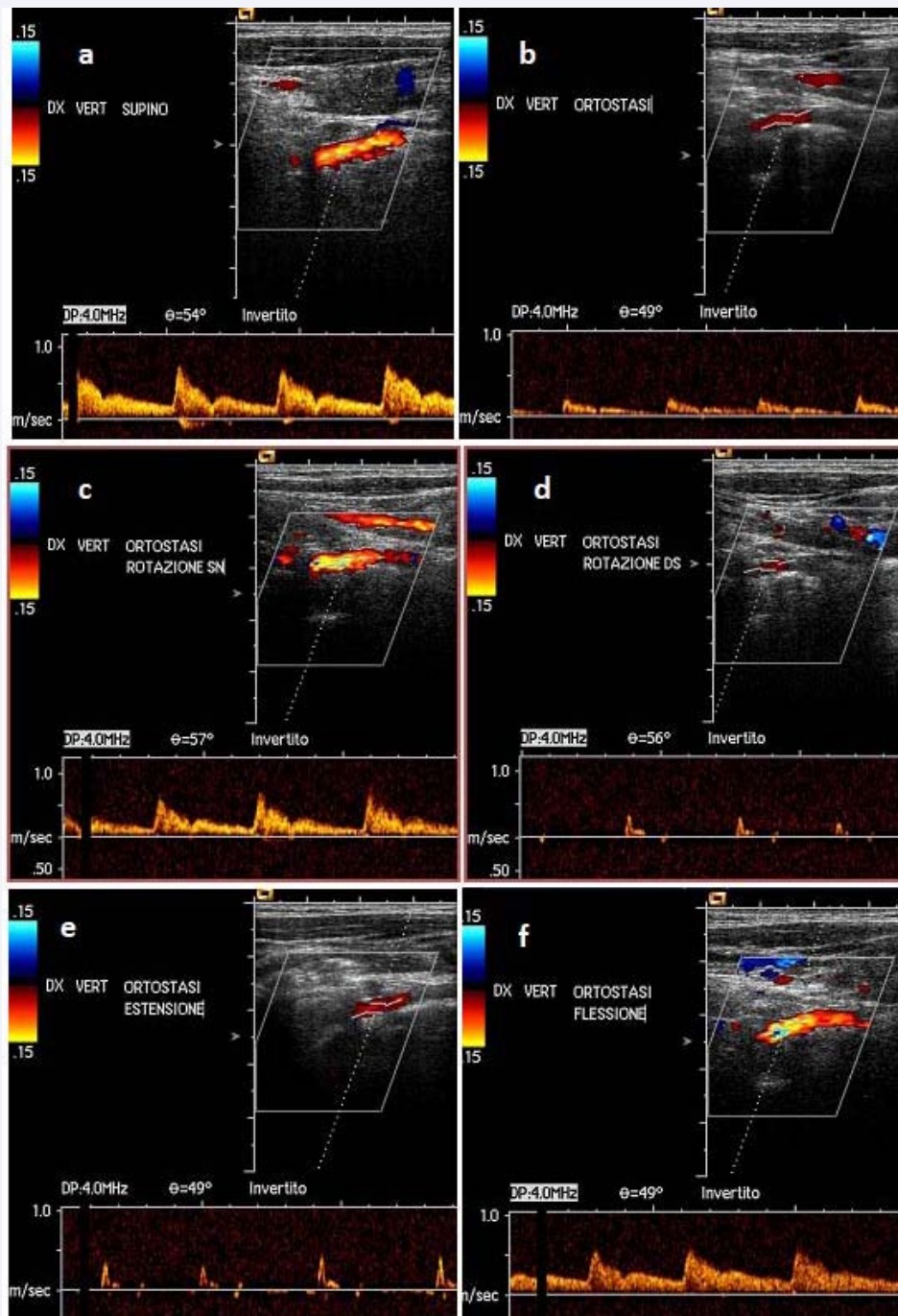


Figure 1 Dynamic CDS examination. Right VA flow changes from clinostatic to orthostatic position during neutral head position (a,b). The exam showed the different flow patterns in the right VA when the patient turned his head 45° left (c) and right (d), and during hyperextension (e) and flexion (f). Note the right VA Doppler wave is reduced in orthostatic position (b), during right rotation combined with orthostatic position (d), and during head extension in orthostatic position (e).

supine position was enough to avoid clinical evidence of vertebral vertebrobasilar insufficiency.

The patient was examined by using a 1.5-T MR unit (Siemens Avanto, Erlangen, Germany). DWI and T1/T2-weighted imaging images did not reveal cerebral ischemic changes due to thromboembolic events. However, irregular caliber and signal abnormality of the distal extra-cranial segment of the right VA

(V3 tract), suspicious for dysplasia, were found. MR angiography demonstrated the tortuosity of the right VA at C1 level, with segmental stenosis and abnormal course, and raised suspicion for cranio-cervical junction (CCJ) malformation (Figure 2).

Therefore, a Computed Tomography (CT) of the cranio-cervical region was obtained: a complex CCJ malformation was evident. The study showed anterior arch of the atlas rachischisis,

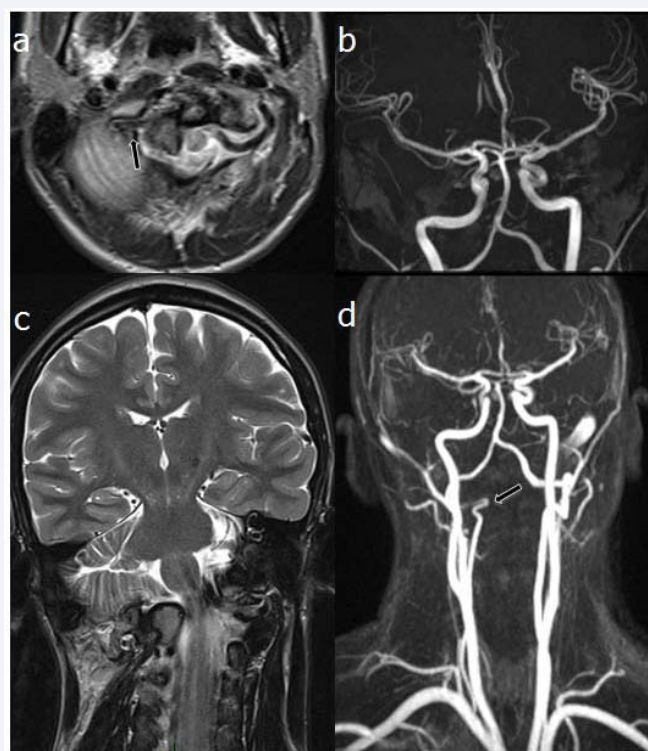


Figure 2 Axial T2-weighted image shows the asymmetry between the right and the left vertebral artery (arrow) (a). Coronal T2-weighted image reveals the abnormal head position (c). MRA confirms the involvement of right vertebral artery in the CCJ malformation (b;d). Note V3 tract of right VA: reduced caliber and tortuosity at C1 level (arrow) (d). None intracranial vascular abnormalities are demonstrated (b).

incomplete right atlanto-occipital assimilation, atlanto-axial fusion, odontoid process partial rotation and basilar invagination. Incomplete C3-C4 body and C6-C1 posterior arch rachischisis was associated. Transverse foramina of C1-C2 were asymmetrical, with narrowing of the right one (Figure 3).

Asymmetric location and vessel wall irregularity of V3 tract of right VA were confirmed by digital angiography. MRA and DSA did not demonstrate any other vascular abnormalities of intra and extracranial vessels, allowing ruling out fibro muscular dysplasia or vasculitis or Moya patterns, which are possible reasons of childhood transient ischemic attack (TIA). Moreover, clinical and laboratory evaluation excluded systemic diseases such as autoimmune vasculitis or hematologic disorders. Considering the age, we decided on a conservative treatment. After patient received the instruction to restrict head rotation to the right and hyperextension, no neurological symptom has occurred.

DISCUSSION

Extracranial narrowing or occlusion of the VA resulting from head-turning may occur a) at the first segment of the VA originating from the subclavian artery and entering into the transverse C6 foramen, b) at the second segment of the VA passing through the C5 to C2 transverse foramina, and c) at the atlantoaxial level [5]. BHS refers to symptomatic vertebrobasilar insufficiency caused by mechanical occlusion or compression of the VA at the atlantoaxial or subaxial level during neck and head rotation. It was first described by Sorensen [3] in 1978, in a patient who developed a posterior circulation infarction during

archery practice; accordingly, the term 'bow hunter's syndrome' was coined. The affected VA is usually contralateral to the side of neck rotation [6]. A recent review of the literature demonstrated that BHS is rare but associated with pathognomonic and serious presentation [7]. The condition may present with dizziness, nausea, dysarthria, dysphagia, transient blurring of vision in one or both eye fields, gait disturbance, headaches, and other sensorimotor findings such as tinnitus, hearing loss, syncope, or drop attacks [8]. Symptoms are correlated with rate of inflow of vertebral-basilar system.

Various pathologic conditions have been reported as causes of BHS. It has been observed in association with many physical activities, such as archery [3] or basketball/handball [9], with cervical spondylosis [10], massive facet hypertrophy at the C1-2 level [11], chiropractic manipulation [12], compression by cervical sympathetic chain [13], erosive rheumatoid arthritis of C1-C2 [14], and tendinous bands.

Recently, Duan et al. [4], distinguished primary from acquired BHS, which is due to the complications of cervical spondylosis and surgery or to injury. BHS can also be categorized into pediatric BHS, juvenile BHS and adult BHS, based on patient age at final diagnosis. Pediatric patients may present with congenital anomalies such as bony malformation at the atlantoaxial level or a suboccipital bony protuberance impinging on the VA [4,15]. To our knowledge, however, very few papers reported BHS associated with complex CCJ malformation [5,16], and our case is the first pediatric BHS with complex CCJ malformation in literature.

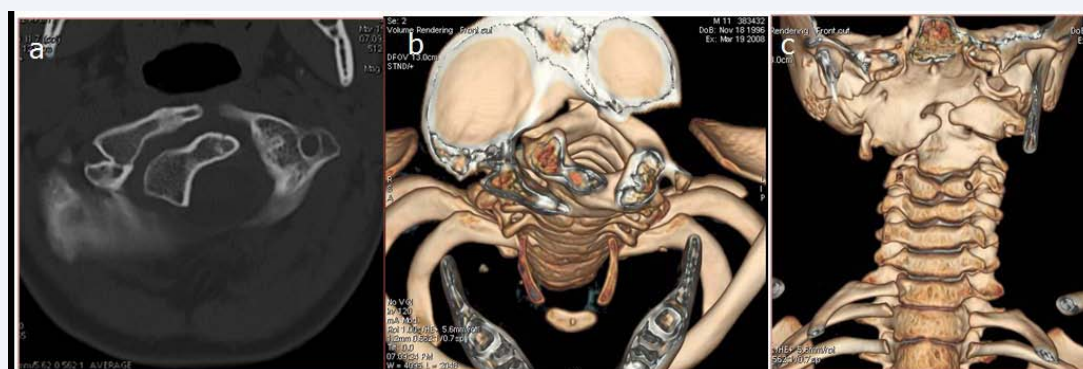


Figure 3 CT scan reveals the complex CCJ malformation (a). Reconstructed coronal CT scan with 3D volume rendering (b,c) reveals incomplete assimilation of the right lateral C1 masses and the occipital condyles. Right atlanto-occipital assimilation, incomplete fusion of the hypoplastic occipital condyles and C1 posterior arch rachischisis. Eccentric positioning of the odontoid process at the C-1 level is demonstrated.

We found an incomplete right atlanto-occipital assimilation, such as C1 anterior arch rachischisis, atlanto-axial fusion, odontoid process rotation with basilar invagination, and incomplete C3-C4 body and C6-C1 posterior arch rachischisis. Atlas assimilation is known to be related to serious vascular and neurologic problems, like vertebra-basilar insufficiency [17-19]. In particular, Sakai et al. [5], presented a case of BHS associated with atlanto-occipital assimilation; they affirmed that 3D-CTA with his head rotated clearly showed the VA occluded between the axis and atlas. CT was crucial in the detection and characterization of the CCJ malformation. CTA with 3D reconstructions can give very precise information about compressing elements and the topographical relationships between the vascular anatomy and the surrounding structures in the same image [4].

In our case, right head rotation produced blood flow reduction and consequent reversible vertebrobasilar insufficiency. No cerebral ischemic insults due to thromboembolic events were demonstrated. Only a segmental V3 irregularity was demonstrated by MRA and DSA, leading to the hypothesis of vessel wall damage by repetitive shear stress due to CCJ malformation instead of fibromuscular dysplasia. Conservative management, surgery and endovascular therapy are the three main treatment methods for BHS. However, for accurate diagnosis and treatment of BHS, neuroimaging with cervical rotation is mandatory [4].

Imaging approaches, such as CDS, CT and angiography, as well as MR/MRA, are widely used in the diagnosis and evaluation of this syndrome [4]. Dynamic angiography has been considered the gold standard of diagnosis [7]. In our case, the vertebrobasilar insufficiency was clinically evident only when the orthostatic position and the hyperextension/right rotation of head together occurred; however, hemodynamic changes, even without symptoms, were present also during right rotation alone. These asymptomatic hemodynamic changes were demonstrated during a dynamic CDS examination in supine position. Dynamic CDS exam may reveal feeble physiological hemodynamic alterations. On the contrary, in dynamic angiography these feeble hemodynamic changes may be masqueraded because the manual selective vertebral artery contrast medium injection could alters the pathophysiological pattern. Thus, dynamic angiography is mandatory when a surgical approach is proposed, but in our case

non-invasive technique, such as CDS, MRA and CTA, allowed to achieve the correct diagnosis. DSA was useful to depict the entire cerebral vascular tree, and therefore to rule out other possible reasons of childhood TIA, such as moyamoya syndrome and fibro muscular dysplasia.

CDS is able to demonstrate the hemodynamic alterations without interfere with arterial inflow. Particularly, CDS has been increasingly employed as a non invasive, inexpensive, and safe technique for measuring cerebrovascular function, especially in young patients [4]. VA hemodynamic changes can be found by CDS in patients with BHS when the patient slowly turns his head [20,21]. Therefore, CDS can be a significant alternative and non-invasive procedure to screen for those patients with signs and symptoms suspicious of this syndrome: its CDS reversible hemodynamic pattern is pathognomonic, and cannot be confused with other pathologies such as post-traumatic dissection and / or occlusion, as well as fibro muscular dysplasia and vasculitis.

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