Case Report

C2 segmental type of vertebral artery on the unfused side of partially occipitalized atlas

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ABSTRACT

Patients with craniovertebral junction anomalies are more likely to undergo craniovertebral junction procedures because of neurologic complications. Vertebral arteries (VAs) are more prone to injury in these cases because of an often anomalous course, with potentially disabling or grave sequel. In our patient with partially occipitalized atlas, CT angiogram revealed that 1 of the VAs passed through the bony canal in the occipitalized and/or fused bone, whereas the VA on unfused side was also unexpectedly anomalous and of C2 segmental type. To the best of our knowledge, such anomaly has never been reported in studies of patients with occipitalized atlas in the literature.

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Introduction

C2 segmental type of vertebral artery (VA) is a rare anomaly in which the VA traverses posteromedially and enters the dura between C2 and C1 after emerging from the transverse foramen of C2. The VA then moves up in the anterior two-thirds of the spinal canal to join the other VAs to form the basilar artery [1]. A higher incidence of C2 segmental type of VA has been found in people with craniovertebral junction (CVJ) anomalies, cases that are more likely to require surgery at the CVJ. As the VA may get injured during surgery or other procedures in this region because of its anomalous course with resultant grave consequences, knowledge of VA abnormalities is very important to both radiologists and surgeons in such cases. We report a case were the VA was anomalous at the unexpected side of partially occipitalized atlas.

Case report

An 11-year-old girl presented with gradually worsening restricted neck movements. As she did not respond to antispasmodics, additional imaging studies were done. X-ray of the neck revealed mild basilar invagination. As per our
institutional protocol, CT angiogram (CTA) of the neck was performed to further delineate the craniovertebral junction in this case (Fig. 1). The CTA revealed partial occipitalization of atlas with fusion of the anterior arch, lateral pedicles, and adjacent small right portion of the posterior arch of the atlas with the occiput. The maximum intensity projection image (Fig. 2) and 3D volume rendered image (Fig. 3) of the CTA of the neck showed the left vertebral artery entering into the spinal canal below the unfused arch of atlas (star) in setting of partial occipitalization of atlas.

Discussion

The C2 segmental type of VA is referred to in the literature as persistent first intersegmental artery. The reported incidence varied by the size of the sample and the methods used; incidence reported by conventional angiography was about 0.7% in disease-free people and much higher in patients with occipitalization of the atlas or Klippel–Feil syndrome [1]. Uchino et al reported a 3.2% incidence of anomalous VA at CVJ using MRA [2]. These incidence data could be exaggerated because of possible selection bias. Furthermore, many of the studies cited were retrospective and had different target populations.

During fetal development, arterial bridges called intersegmental arteries (ISAs) develop between the aorta–carotid systems and the more dorsally located longitudinal neural arterial systems (future basilar and spinal arteries). In the neck area, 7 ISAs develop; the first one occurs in the proatlas area between the atlas and the occiput. According to Padget, the ISA in the proatlas area is not numbered and was given the name “proatlan tal intersegmental artery (PIA)” [3]. The remaining 6 ISAs were numbered: the first ISA (FIA) as named is between...
C1 and C2. The sixth ISA is the future subclavian artery. The nomenclature can be confusing sometimes, and many articles have referred to the PIA as FIA [3, 4].

The cervical VA develops normally from longitudinal anastomosis of these 7 ISAs. All ISAs regress, except the last (to become subclavian artery) and the dorsal part of PIA, which becomes the horizontal part of VA above the atlas and joins the basilar artery [3, 4]. The C2 segmental type of VA results when the distal part of the FIA, instead of the PIA, persists; thus, the VA enters the spinal canal between C2 and C1, that is, below the C1 vertebra [1]. The C2 segmental type of VA is also called persistent first intersegmental artery, which can be a misnomer as the FIA is, in fact, only the persistent dorsal part of the artery, and not the complete or whole persistent artery. Accordingly, "C2 segmental type of VA" is a better nomenclature in our opinion.

Rogers et al reported a case of fatal hematoma that developed after C1–C2 cisternal puncture. At postmortem, anomalous VA was seen entering the spinal canal and subarachnoid space below C1 (persistent first intersegmental artery or C2 segmental VA), indicating the significance of missing such anomaly [5]. Our patient had partially occipitalized atlas and stabilizing surgery was being contemplated; thus, detecting any anomalous VA was critical. Unexpectedly, we detected the C2 segmental artery on the unfused side of partially occipitalized atlas.

Although C2 segmental VA by itself is usually asymptomatic and found incidentally (as in our case), occasionally it may cause rotational verteobasilar insufficiency (Bow Hunter’s syndrome), recurrent embolic strokes, or rarely compressive cervical myelopathy [6–8].

For diagnosis or preprocedural evaluation, axial CT and MRI may show findings related to C2 segmental VA. Flow-void of VA with anomalous course may be observed on T2–MRI-weighted images. CT scan may show hypoplasia of the unilateral transverse foramen [9]. MRA has been studied for VA anomalies, and although it is easier and less invasive than other modalities, it lacks the ability to show clear osseous images or allow visualization of the reciprocal anatomy from the surgeon’s point of view [9]. However, 3D CTA has more advantages; in addition to being able to accurately define the course of VA, it can also delineate the surrounding osseous anatomy with multiplanar image reconstruction, allows evaluation of reciprocal anatomy, and can be done in less time than conventional angiography or MRA [9]. Conventional angiography can show the anomalous course of the VA; however, even when combined with reconstructed CT images it cannot show the osseous tissue simultaneously or clearly with the artery, or allow evaluation of reciprocal anatomy; thus, the use of conventional angiography may be limited to cases that need interventional procedures [9]. Thus, CTA can be one-stop evaluation in these cases, for better delineation of the osseous anomalies along with detection of anomalous VA on the contrast scan, and in our case, it allowed the detection of anomalous VA on the other side. This being true in many cases, in some cases with verteobasilar insufficiency upon head motion or rotation, evaluation may require dynamic imaging like digital subtraction angiography to reproduce the symptoms while visualizing the reduction in blood flow upon head rotation [7].

If C2 segmental artery is symptomatic by itself, decompression surgery may be attempted. Our patient had partially occipitalized atlas and stabilizing surgery was being contemplated; thus, detecting any anomalous VA was critical. To our surprise and unexpectedly, the C2 segmental artery was seen on the unfused side, and whether this represents an incidental finding related to the incidence in general population or is a consequence of occipitalized atlas is yet to be known. Interestingly, opposite VA formed a bony canal through the fused and/or occipitalized atlas elements.

In conclusion, C2 segmental type of VA by itself, or more commonly due to associated osseous CVJ anomalies, can be associated with compressive symptoms, verteobasilar insufficiency, embolic strokes, and other possible neurologic problems. Although it may remain undiagnosed for years and is sometimes asymptomatic, it could potentially put the patient at risk during procedures in the CVJ area. A radiologist should be vigilant about the course of VA when reviewing imaging for the craniovertebral region, especially in patients who also have CVJ osseous anomalies. Both VAs should be carefully evaluated regardless of the bony fusion status in cases of occipitalized atlas. CTA is exceptionally valuable and an accurate modality, with many advantages over other modalities for detection and evaluation of such anomalous VAs with simultaneous reciprocal evaluation of bony anatomy at the same time.

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