Compression of the C-2 root by a rare anomalous ectatic vertebral artery

Case report

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The authors report a symptomatic congenitally anomalous ectatic vertebral artery not passing through the transverse foramen of the atlas (C-1), but instead piercing the dura mater below the posterior arch of the C-1 in the atlantoaxial (C1-2) interlaminar space. This occurrence is exceptionally rare, but in this case it was uniquely associated with occipital neuralgia due to vascular compression of the C-2 root. Microvascular decompression was curative. Neuroradiological and surgical findings are presented and their implications discussed.

Key Words • anomalous artery • ectasia • neurovascular compression • occipital neuralgia • microvascular decompression

Vertebrobasilar ectasia causing neurovascular compression within the posterior fossa and cervicomedullary region is an uncommon but well-recognized clinicopathological entity. Symptomatic cases present with a wide range of clinical syndromes such as hydrocephalus, trigeminal neuralgia, hemifacial spasm, cerebellopontine angle syndrome, glossopharyngeal neuralgia, spasmodic torticollis, and brain-stem or cervicomedullary compression. A case of an ectatic (but otherwise normal) vertebral artery of the cervical region presenting as a worsening occipital neuralgia has also been reported.

We report a patient with a rare congenitally anomalous ectatic vertebral artery associated with a Klippel-Feil syndrome (C2-3), presenting as occipital neuralgia. Neuroradiological and surgical findings are presented and the clinical implications are discussed.

Case Report

This 75-year-old woman presented with a 2-year history of sharp, stabbing pains aggravated by neck movements and followed by discomfort and tightness in the left suboccipital region. She had an 18-month history of progressive tingling and clumsiness of both hands, and difficulty in walking with a tendency to fall. Her medical history was insignificant and there was no family history of neurofibromatosis. On admission to another hospital in April 1990, neurological examination showed spastic quadriplegia (Medical Research Council power Grade 4/5), generalized hyperreflexia, upgoing plantar reflexes, bilaterally impaired position sense, impaired touch and pain sensations below the C-4 level, and tenderness over the left suboccipital region just medial and below the mastoid process.

Examination. Plain x-ray films of the cervical spine showed congenital fusion of the C2–3 vertebral bodies and their laminae (Klippel-Feil syndrome), gross enlargement of the left C-2 transverse foramen as well as the left C2-3 intervertebral foramen, and cervical spondylotic changes at multiple levels. Plain computerized tomography (CT) of the spine was interpreted as showing a soft-tissue lesion at the C1–2 level on the left side within the spinal canal and extending laterally. A space-occupying lesion such as schwannoma of the left C-2 root was suspected. The patient was transferred to the Neurosurgery Department at the Royal Preston Hospital with her neurological status unchanged. Magnetic resonance (MR) imaging revealed significant spinal canal stenosis from C-4 to C-7, explaining her myelopathic symptoms.

To eliminate the likelihood of schwannoma of the left C-2 root, the patient underwent CT myelography, revealing a rounded filling defect at the C1–2 level on the left side (Fig. 1 left). Interestingly, following intra-
venous contrast infusion, postmyelographic CT scans suggested that the filling defect was a vascular structure rather than a neoplasm (Fig. 1 right). There was no evidence of canal stenosis or cord compression at C1-2 (Fig. 1).

Vertebral angiography confirmed a congenitally anomalous ectatic vertebral artery on the left that did not pass through the transverse foramen of the atlas but entered the C1–2 interlaminar space below the posterior arch of the atlas and, making a loop around the cervicomedullary region, became continuous with the basilar artery without joining the right vertebral artery (Fig. 2). The latter was small in caliber, normal in its course, and ended in the right posterior inferior cerebellar artery itself.

Operation. An extensive C-2 to C-7 decompressive laminectomy was performed. At surgery, severe stenosis was noted at C4–7. The fused C2–3 laminae complex was excised to expose a large ectatic left vertebral artery coursing posteromedially between the atlas and axis over the dorsal aspect of the left C-2 root, then appearing in the C1–2 interlaminar space. It pierced the dura mater posteriorly below the posterior arch of the atlas between C-1 and C-2. Microvascular techniques were used to separate the artery from the left C-2 root, and a Surgicel plug was kept between the two structures. The C-2 root was flattened and thinned out due to chronic pulsatile compression.

Postoperative Course. The patient's postoperative course was uneventful. She was relieved of her occipital neuralgia and made a gradual neurological recovery. She was virtually asymptomatic 1 $\frac{1}{2}$ years later except for some residual spasticity in the lower limbs.

Discussion

Normal Anatomy

Usually the vertebral artery arises from the first part of the subclavian artery and enters the transverse foramen of the C-6 vertebra (first segment). It then ascends through the transverse foramina from C-6 to the atlas (second segment). Up to the transverse foramen of the C-3 vertebra the artery's course remains vertical, then it turns laterally in the transverse foramen of the axis under the superior articular facet and, bending upward, enters the transverse foramen of the atlas which is more lateral than the others. In its third segment, the artery emerges on the superior surface of the atlas and curves horizontally over the lateral and posterior surfaces of the superior articular process of C-1. It creates a groove in this process and the root of the posterior arch and passes medially in front of the posterior atlanto-occipital membrane. In its fourth segment, the artery pierces the dura mater and arachnoid below the occipital bone and atlas and enters the cranial cavity through the foramen magnum. At the pontomedullary junction, it forms the basilar artery by joining with the opposite vertebral artery.

Anomalous Anatomy

In a series of 300 vertebral angiograms, Tokuda, et al., found a 2% to 3% incidence of vertebral artery anomalies, including two cases of vertebral artery running under the posterior arch of C-1. They also emphasized associated malformations such as Klippel-Feil syndrome, as in our case. In this patient, the left vertebral artery described a rare anomalous course except in its first segment; it turned posteromedially between the atlas and axis over the dorsal aspect of the left C-2 root and presented in the C1–2 interlaminar space in its third segment (Fig. 2). It pierced the dura mater and arachnoid below the posterior arch of the atlas between C-1 and C-2 (Fig. 3) and, making a loop around the cervicomedullary region, became continuous with the basilar artery in its fourth segment.

Six similar cases have been reported previously in the literature where the vertebral artery pierced the dura mater below the posterior arch of the atlas in the C1–2 interlaminar space. Of these cases, one patient...
Anomalous vertebral artery and occipital neuralgia

was symptomatic and presented with spasmotic torticollis as a result of compression of the accessory nerve by the anomalous vertebral artery.14 In this case, microvascular decompression was effective. Our patient is unique, presenting with occipital neuralgia due to compression of the C-2 root by an anomalous ectatic vertebral artery; surgical decompression of the C-2 root resulted in a successful outcome. The features of cervicothoracic myelopathy in our case were due to the canal stenosis rather than to an anomalous vertebral artery (Fig. 1).

Arterial Ectasia

Ectasia is a diffuse dilation of the artery over its entire length. Its pathogenesis remains controversial.5 Some authors believe it is a result of a degenerative process of the arterial wall either in itself or associated with hypertension, while others believe atherosclerosis may not play a role in its pathogenesis.7 In the study on ectatic basilar arteries, Hegedus found severe reticular fiber defects in the elastic lamina. According to the literature, the left vertebral artery is larger than the right in 42% to 51% of persons, and the right is larger than the left in 32% to 41%.6,10,13 It is difficult to be sure in our case as to whether the ectasia of the left vertebral artery was a congenital anomaly or due to a degenerative etiology. However, we presume it to be due to the former in the absence of signs of atherosclerosis and hypertension and also because the involved vessel was a dominant artery without being joined by the opposite vertebral artery. Associated vertebral abnormalities in our case also favor congenital etiology.

Occipital Neuralgia

Occipital neuralgia is commonly seen in cases where there is injury or irritation of the nerve root of either C-2 or C-3 or the greater or lesser occipital nerve caused by cervical spine trauma, cervical spondylosis, schwannoma of the C-2 or C-3 root, or congenital anomaly such as Arnold-Chiari malformation.9 A rare case of deteriorating occipital neuralgia due to an ectatic vertebral artery has been described by Bossi and Caffaratti.2 However, the unique feature of our case was an anomalous ectatic vertebral artery in the C1–2 interlaminar space causing a pulsatile compression of the C-2 root and presenting as occipital neuralgia. The neuralgia was relieved following microvascular decompression of the left C-2 nerve root.

Clinical Implications

The importance of this anomalous course of the vertebral artery is the danger to the vessel during a lateral C1–2 puncture for myelography or during a cervical decompressive laminectomy. Such a vertebral artery can mimic a neoplasm within the spinal canal. If surgical exploration is undertaken without vertebral angiography (which is not routinely performed in the investigation of a cervical cord neoplasm), there is potential danger to the vertebral artery if the diagnosis was unsuspected and the spinal cord explored on the assumption that the lesion is a tumor. Therefore, the possibility of an anomalous vertebral artery should be borne in mind when a lesion lateral to the spinal cord at the C1–2 region is demonstrated radiologically. Enlargement of the C2–3 intervertebral foramen was also deceptive in our case as this further indicated the possibility of a cervical root schwannoma. The role of conventional vertebral angiography or MR angiography cannot be overemphasized in suspected cases. Our report adds one more cause to the differential diagnosis of the occipital neuralgia and one more example in support of the theory of neurovascular compression.

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References

1. Abe K: [A rare abnormal case of the vertebral artery showing no passing through the foramen transversarium of the atlas.] Acta Anat Nippon 43:393–394, 1968 (Jpn)
6. Jamjoom AB, Rawlinson JN, Coakham HB: Multiple

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