

Neuroimaging Highlight

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Intrinsic Arteriovenous Malformation of the Trigeminal Nerve

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A 45-year-old man presented with a one-year history of left facial pain compatible with trigeminal neuralgia. There was an electric shock-like pain that moved from just in front of his ear to the side of his cheek at the inferior aspect of his jaw. These episodes of pain lasted only seconds, and occurred in clusters of a week or two, every couple of months. He took ibuprofen during the episodes. The patient also had occasional left-sided headaches with light sensitivity and flashing lights. He was otherwise well. His only regular medication was a cholesterol lowering drug. On exam, motor and sensory function of the trigeminal nerves was normal. Hearing and facial nerve function were normal.

Magnetic resonance imaging (MRI) utilizing a three-dimensional fast imaging employing steady state acquisition (FIESTA) sequence allowed direct visualization of the trigeminal nerve root within the prepontine cistern (Figure 1). Vascular flow voids were seen around the left nerve root and the nerve root fibers were splayed with flow voids interposed between the fibers. A large left lateral mesencephalic vein was identified behind the abnormal cluster of flow voids and extended superiorly in the ambient cistern. The appearance was compatible with an intrinsic arteriovenous malformation (AVM) of the left trigeminal nerve.

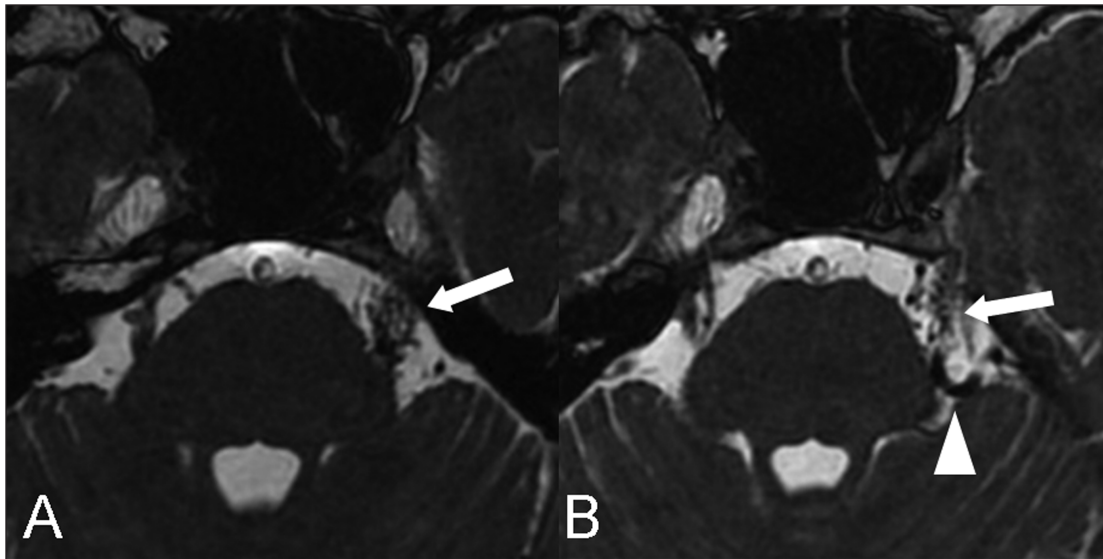


Figure 1: Axial MRI FIESTA images demonstrate a cluster of vascular flow voids within and around the left trigeminal nerve root (arrows, A and B). The root fibers are splayed by the flow voids. There is an enlarged left lateral mesencephalic vein arising posterior to the cluster of flow voids and extending superiorly in the ambient cistern (arrowhead, B).

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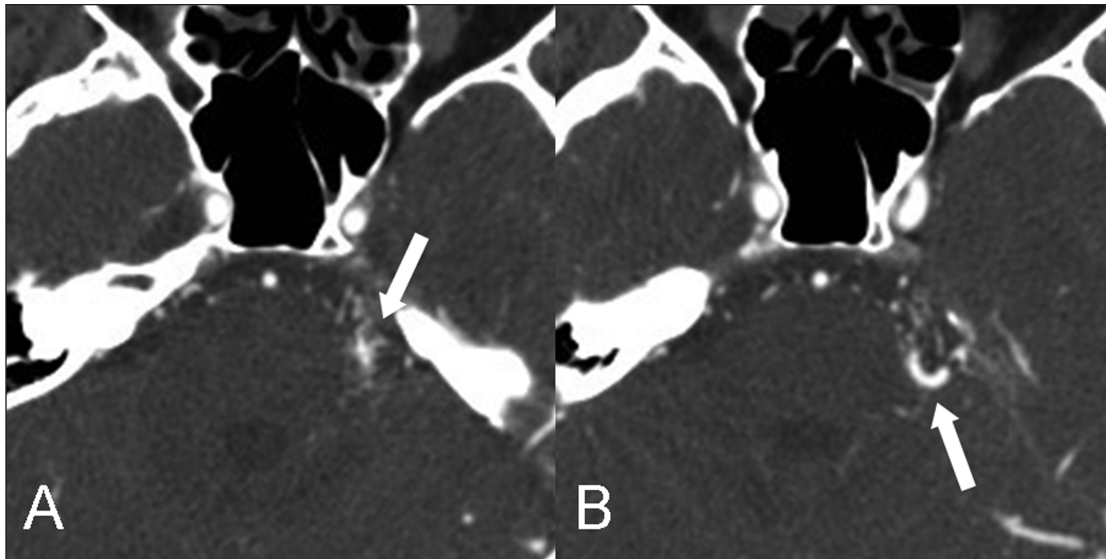


Figure 2: Axial CTA images demonstrate a cluster of vessels within and around the left trigeminal nerve root (arrow, A). There is an enlarged left lateral mesencephalic vein arising posterior to the cluster of vessels and extending superiorly in the ambient cistern (arrow, B).

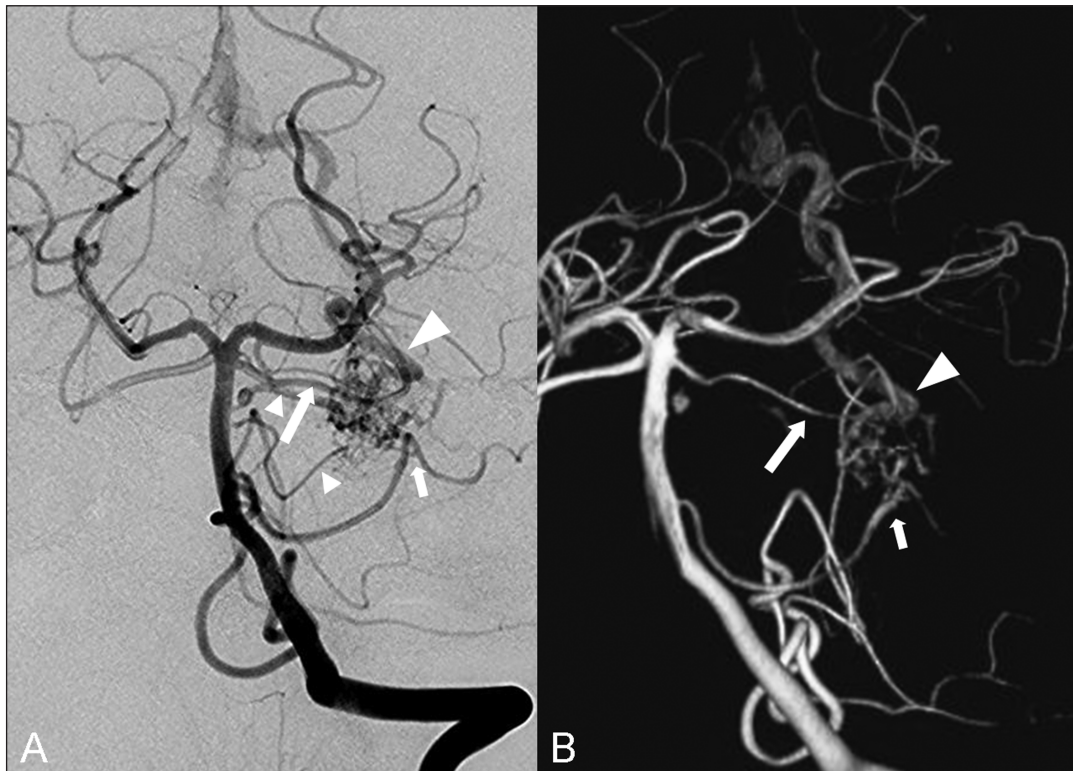


Figure 3: Conventional angiography (A) and 3D rotational angiography (B) demonstrates an AVM of the trigeminal nerve which is directly supplied by the left superior cerebellar artery (long arrow, A and B), and supplied en passage by the left anterior inferior cerebellar artery (short arrow, A and B). There is also supply by two enlarged circumflex arteries from the basilar, seen best on the conventional angiogram (small arrowheads, A). An enlarged left lateral mesencephalic vein is seen draining the AVM and extending superiorly to drain into the basal vein of Rosenthal (large arrowhead, A and B).

Computed tomography angiography (CTA) demonstrated a small cluster of vessels in the left prepontine cistern surrounding and within cisternal portion of the trigeminal nerve (Figure 2). A large left lateral mesencephalic vein extends posteriorly from the cluster of the vessels. The left superior cerebellar artery and anterior inferior cerebellar artery are both enlarged and course into the abnormal cluster of vessels.

Conventional angiography performed with a left vertebral artery injection confirmed an AVM of the trigeminal nerve measuring 15 by 17 by 22 mm (Figure 3). The AVM is supplied directly by an enlarged left superior cerebellar artery. It is also supplied en passage by an enlarged left anterior inferior cerebellar artery. There is also contribution by enlarged circumflex arteries directly from the basilar. There was minimal dural supply from a small left middle meningeal artery branch (not shown). The large left lateral mesencephalic vein seen on the CTA and MRI is identified on angiography, and it extends superiorly into the basal vein of Rosenthal.

DISCUSSION

We present an intrinsic AVM of the cisternal portion of the left trigeminal nerve, characterized using MRI, CTA, and conventional angiography. Arteriovenous malformations of cranial nerves are rare, the optic nerve being the most common location. Most AVMs that cause trigeminal neuralgia are posterior fossa AVMs that extrinsically compress the nerve rather than involve the nerve itself. There are many reported AVMs that extrinsically compress the trigeminal nerve¹. Arteriovenous malformations intrinsically involving the trigeminal nerve have been rarely reported²⁻⁴. While AVMs of the optic pathway and nerve have been associated with the cerebrofacial arteriovenous metamerism syndrome (Wyburn-Mason syndrome), to our knowledge no syndromic predilection for trigeminal lesions has been reported. Trigeminal AVMs are only seldom identified on conventional CT and MRI imaging, and are usually detected during surgical exploration (2,4). Newer high resolution imaging techniques such as CTA, FIESTA, and three-dimensional spoiled gradient recalled acquisition at steady state³ may improve detection. Based on surgical exploration, the nidus has been described as being present within the root entry zone² and cisternal portion³ of the nerve, however may also be predominantly subpial, and can involve the adjacent pons⁴.

The natural history of this rare lesion is not well defined. It is unclear whether the long term risk of hemorrhage is similar to other brain AVMs where the reported annual risk is on the order of 2-4%⁴. The treatment is controversial. The usual treatment options for AVMs include surgical resection, stereotactic radiosurgery and, sometimes, embolization when feasible. Preservation of trigeminal nerve function is an important consideration. Surgical resection poses a high risk of loss of nerve function³. Stereotactic radiotherapy presents an attractive option in that it provides satisfactory outcomes both for AVMs, and for the treatment of trigeminal neuralgia, with a relatively low risk of complications⁵. However, the disadvantage is that the patient is exposed to risk of hemorrhage during the time to obliteration. One group has reported microvascular decompression for treatment of neuralgia followed by radiosurgery⁴. In our case, the patient was treated by gamma knife stereotactic radiosurgery.

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