Persistent trigeminal artery supply to an intrinsic trigeminal nerve arteriovenous malformation: A rare cause of trigeminal neuralgia

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ABSTRACT

Infratentorial arteriovenous malformations (AVM) associated with the trigeminal nerve root entry zone are a known cause of secondary trigeminal neuralgia (TN). The treatment of both TN and AVM can be challenging, especially if the AVM is embedded within the trigeminal nerve. A persistent trigeminal artery (PTA) can rarely supply these intrinsic trigeminal nerve AVM. We present a 64-year-old man with TN from a right trigeminal nerve AVM supplied by a PTA variant. The patient underwent microvascular decompression and a partial resection of the AVM with relief of facial pain symptoms. His residual AVM was subsequently treated with CyberKnife radiosurgery (Accuray, Sunnyvale, CA, USA). A multimodality approach may be required for the treatment of trigeminal nerve associated PTA AVM and important anatomic patterns need to be recognized before any treatment. Herein, we report to our knowledge the third documented patient with a posterior fossa AVM supplied by a PTA and the first PTA AVM presenting as facial pain.

1. Introduction

Type I classic trigeminal neuralgia (TN) is described as an intermittent, sharp lancinating pain in the trigeminal distribution secondary to vascular compression of the nerve root entry zone by a superior cerebellar artery loop in 80% of cases and, less commonly, by an anterior inferior cerebellar artery (AICA) loop, vein, or intraneural vessel [1]. Tumors, vascular malformations, and other skull base anomalies may also trigger TN, demanding a more multimodal approach than traditional microvascular decompression [2]. Treatment of such lesions is important for symptomatic relief and hemorrhage prevention.

We describe a posterior fossa arteriovenous malformation (AVM) supplied by a persistent trigeminal artery (PTA) presenting as facial pain. We discuss the anatomical relationships of PTA with the trigeminal nerve and management paradigms important for treatment of these lesions.

2. Case report

A 64-year-old man presented with a 2 month history of sharp, severe right-sided facial pain within a V1/V2 distribution. His clinical examination was within normal limits.

MRI demonstrated several vascular flow voids near the trigeminal nerve complex, which were initially interpreted by a general radiologist as AICA loops impinging upon the cisternal segment of the trigeminal nerve. Additionally, abnormal high signal within the basal vein of Rosenthal on magnetic resonance angiography (MRA) sequence was not initially appreciated (Fig. 1). Given his poor response to medical therapy and MRI findings suggestive of vascular compression at the trigeminal nerve root entry zone, the patient was recommended to undergo craniotomy and microvascular decompression.

Intraoperatively, an AVM was found embedded within the sensory root of trigeminal nerve and superficial brainstem. The arterial loops of the AVM compressing the trigeminal nerve were coagulated to decompress the nerve. A partial obliteration of the AVM was performed without attempted complete resection.

A postoperative cerebral angiogram demonstrated a 12 × 8 mm residual AVM in the right cerebellopontine angle primarily supplied by a PTA variant and a right AICA branch (Fig. 2). The patient experienced complete symptomatic relief following microvascular decompression. He underwent further CyberKnife stereotactic radiosurgery (Accuray, Sunnyvale, CA, USA) for the residual AVM (Fig. 3) and continues to remain asymptomatic.

3. Discussion

In rare instances, an AVM may result in TN, with a cited prevalence of up to 1.5% [3]. These AVM are typically located in the cerebellum, cerebellopontine angle, or brainstem, but occasionally may be embedded within the trigeminal nerve [4].

Careful review of preoperative imaging studies is important in order to identify uncommon causes of TN. Herein, the increased flow voids near the cisternal segment of the right trigeminal nerve were misinterpreted as loops arising from the AICA, which was positioned more posteriorly in the right cerebellopontine angle. Furthermore, increased signal on the MRA within the right basal vein of Rosenthal, representing arteriovenous shunting, was not appreciated.

The PTA occasionally serves as an anomalous source of arterial supply to posterior fossa AVM. The artery serves as an embryological structure connecting the precavernous carotid artery and basilar artery until the seventh week of gestation. It typically regresses but remains postnatally in 0.1–0.6% of the population [5]. The PTA

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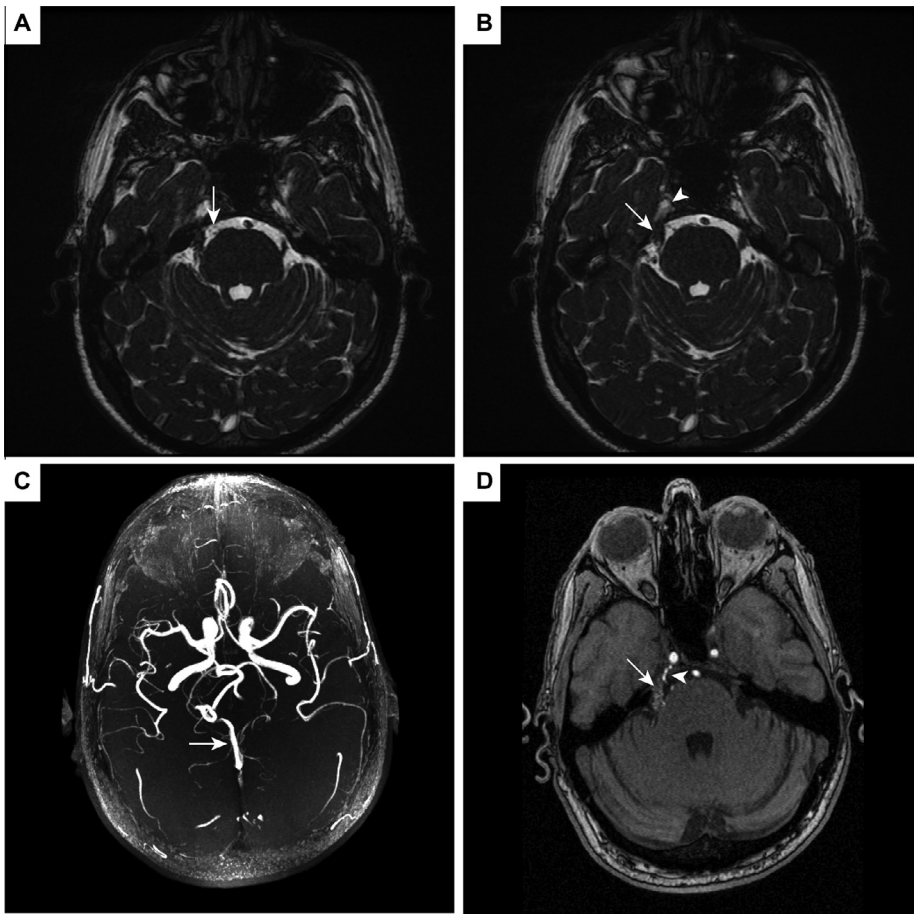

Fig. 1. Trigeminal nerve arteriovenous malformation (AVM) identified by MRI. (A) Axial T2-weighted image at the level of the pons demonstrates multiple small flow voids (arrow) in the right lateral prepontine cistern adjacent to the right cranial nerve V. (B) Axial T2-weighted image at the level of Meckel's cave (arrowhead) demonstrates enlargement of the cisternal segment of the right cranial nerve V (arrow) proximal to the right Gasserian ganglion. (C) Axial maximal intensity projection image from brain magnetic resonance angiography (MRA) demonstrates abnormal high signal in the right basal vein of Rosenthal that is consistent with arteriovenous shunting related to the right trigeminal nerve AVM. (D) Axial image from brain MRA demonstrates a persistent trigeminal artery arising from the cavernous segment of the right internal carotid artery (arrowhead). Abnormal increased signal is present within an enlarged cisternal segment of the right cranial nerve V (arrow), which corresponds to an AVM.

usually originates separate from and proximal to the meningohypophyseal trunk.

Given its anatomic location, the PTA can uniquely supply feeders to both anterior and posterior circulation AVM. A PTA variant connecting to the AICA can feed posterior fossa AVM. There are four prior reports of a PTA supplying infratentorial AVM to our knowledge, all presenting with intracerebral or subarachnoid hemorrhage [6]. To our knowledge, this report is the first to describe a PTA variant that supplies an intrinsic trigeminal nerve AVM presenting with TN.

Treatment of intrinsic trigeminal nerve AVM can be challenging, as completely resecting the AVM may compromise normal function (Table 1). In our case, surgical decompression and partial obliteration of the AVM was followed by adjunctive radiosurgery to target the intrinsic nerve component of the AVM.

Endovascular transarterial embolization of cerebellopontine angle and trigeminal AVM using liquid embolics can help reduce the nidus size and vessel pulsations by decreasing flow, which may resolve TN [7]. Transarterial embolization of an AVM supplied by a PTA or a variant is dangerous, placing two circulations at risk. In our patient, the PTA was not large enough to allow safe catheterization and, therefore, no endovascular therapy was pursued. Additionally, perforating pontine branches from the cisternal segment of a PTA may not be angiographically visible [8]. Non-targeted embolization of perforators can result in brainstem infarction with devastating consequences [9]. Therefore, we do not recommend transarterial embolization of AVM with a PTA supply.

4. Conclusion

Intrinsic AVM of the trigeminal nerve can be supplied by a PTA. The AVM nidus and the PTA can both cause trigeminal nerve root compression related to their anatomic relationships. A multimodal approach utilizing surgery and radiosurgery may be helpful for the treatment of these challenging lesions.

Conflicts of Interest/Disclosures

Dr. Steinberg serves on the Neuroscience Advisory Board at Medtronics, Inc.

The other authors declare that they have no financial or other conflicts of interest in relation to this research and its publication.