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Case Report

Trigeminal neuralgia associated with a variant of persistent trigeminal artery [☆]

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ABSTRACT

A persistent trigeminal artery variant (PTAV) is an anomalous vessel that originates from the internal carotid artery and directly supplies the cerebellum without interposition of the basilar artery. This anatomic variant is usually found incidentally on imaging but can rarely have clinical implications. We describe a case of a 74-year-old woman presenting with multiple years of lower jaw pain attributed to trigeminal neuralgia, unresolved with medication. A persistent trigeminal artery variant compressing the trigeminal nerve was identified on magnetic resonance imaging and magnetic resonance angiography. The characteristic imaging findings of PTAV are essential for identifying an etiology of medically refractory trigeminal neuralgia and may assist with preoperative planning.

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Introduction

The primitive trigeminal artery (PTA) is the most common persistent carotid-basilar anastomosis [1]. While the PTA communicates with the basilar artery, the persistent trigeminal artery variant (PTAV) arises from the internal carotid artery (ICA) and supplies the cerebellum without interposition of the basilar artery. PTA or PTAV compression of the trigeminal nerve is reported to be the cause of 0.2%–0.6% of trigeminal neuralgia cases [2]. We report a case of trigeminal neuralgia associated with a persistent trigeminal artery variant.

Case report

A 74-year-old Asian woman presented with 12 years of worsening paroxysmal pain in the left jaw. The patient underwent extraction of multiple teeth from the left mandible without resolution of the pain. Her symptoms were attributed to trigeminal neuralgia, but no significant improvement was achieved with various doses of carbamazepine. Otherwise, the physical exam was unremarkable.

MR and MR angiography were performed to evaluate for secondary causes of trigeminal neuralgia. MR angiography

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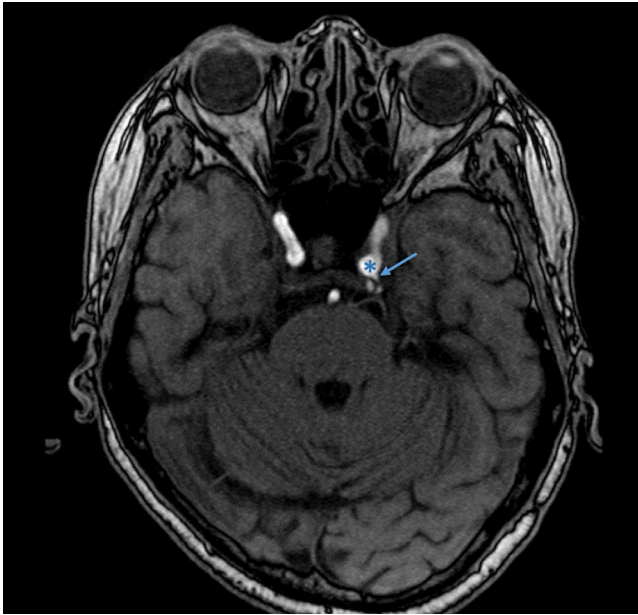


Fig. 1 – 3D Time-of-flight magnetic resonance angiography image in the axial plane demonstrates an anomalous vessel, the PTAV (arrow), originating from the left cavernous ICA (asterisk). (Color version available online.)

demonstrated an anomalous vessel originating from the cavernous left internal carotid artery (ICA) and terminating into the left anterior inferior cerebellar artery (AICA) (Fig. 1). High-resolution 3D T2 sequences showed the vascular loop causing mass effect upon the inferior aspect of the left trigeminal nerve at the root entry zone (Fig. 2). Additionally, CT angiography redemonstrated a tortuous PTAV arising from the left ICA (Fig. 3).

Given her persistent symptoms and minimal success achieved with medical therapy, the patient was offered sur-

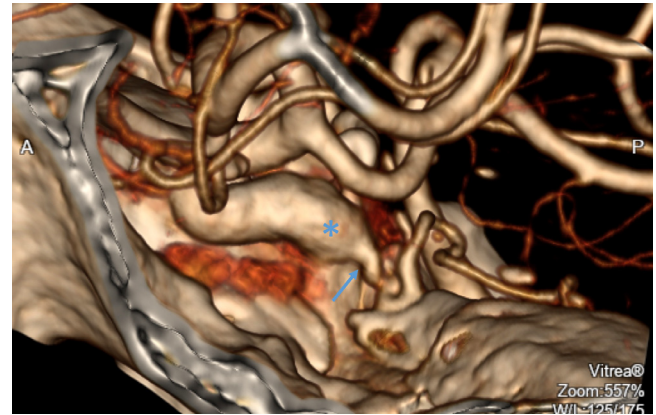


Fig. 3 – Lateral 3D reformatted CTA image demonstrating origin of PTAV (arrow) from the cavernous segment of the ICA (asterisk), proximal to the posterior genu. The tortuous PTAV is seen coursing posteriorly and eventually supplies the AICA territory. (Color version available online.)

gical intervention for trigeminal neuralgia. Treatment options including continued medical therapy, gamma knife radiosurgery, and microvascular decompression were discussed with the patient. Ultimately, the patient decided not to pursue neurological surgery and instead continue with medical management.

Discussion

Etiology and demographics

During embryonic development, 4 major anastomoses arise between the carotid and vertebrobasilar systems: The primitive trigeminal, hypoglossal, otic, and proatlantal

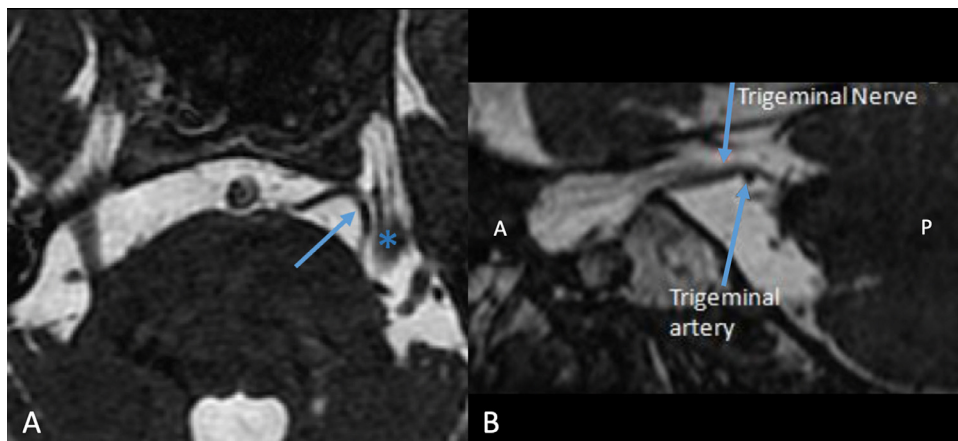


Fig. 2 – MR demonstration of PTAV. Axial 3D T2 sequence (A) reveals a vascular loop of the PTAV (arrow) abutting the root of the left trigeminal nerve (asterisk) at its origination from the lateral pons. As the vessel courses posteriorly, it travels inferior to the trigeminal nerve. Sagittal 3D T2 sequence (B) shows a PTAV (lower arrow) deflecting the left trigeminal nerve (upper arrow) from the undersurface of the nerve, where the V3 fibers are located. This finding is consistent with the patient's pain in the left V3 distribution. (Color version available online.)

intersegmental arteries. These anastomoses provide the main blood supply via the internal carotid artery to the hindbrain during the early embryonic stage. Of the four carotid-basilar anastomoses, the primitive trigeminal artery (PTA) is the last to involute at the 7–14 mm embryonic stage, after the development of the posterior communicating artery (PCOM) [1]. Rarely, these anastomoses persist into adulthood. The most common persistent primitive carotid-basilar anastomosis is the PTA, which originates from the cavernous segment of the ICA, based on the Bouthillier classification of the ICA, and communicates with the basilar artery [3,4].

The persistent trigeminal artery has been classified into 2 types first described by Saltzman [5]. In Saltzman Type I, the PTA joins the basilar artery at the level between the superior cerebellar artery (SCA) and the anterior inferior cerebellar artery, and the PTA supplies the upper basilar artery, including posterior cerebellar and superior cerebellar artery territories. The proximal basilar artery and PCOM are hypoplastic. In Saltzman Type II, the PTA joins the basilar artery at the origin of the SCA and supplies the SCA territory, while the patent PCOM supplies the posterior cerebral arteries [1]. In both types of PTA, basilar artery formation from midline fusion of the paired longitudinal neural arteries is complete [6].

A variant of the persistent trigeminal artery, as in this case, is classified as Saltzman type III [1]. The variant is characterized by the presence of a primitive trigeminal artery arising from the cavernous ICA and terminating as a cerebellar artery, without the interposition of the basilar artery. It has been postulated that this is a result of incomplete fusion of the paired longitudinal neural arteries that form the basilar artery [6]. However, the exact sequence of embryonic development of the PTAV is not well understood.

The reported incidence of PTA and PTAV is 0.1%–0.6% and 0.18%–0.76%, respectively [1]. There is no sex predilection in PTAV [3]. The PTAV may terminate as the SCA, AICA, or posterior inferior cerebellar artery, but most commonly terminates as the AICA [7]. In this case, the artery terminates in an AICA, which results in mass effect on the cisternal component of the left trigeminal nerve.

Clinical and imaging findings

Radiological diagnosis of persistent trigeminal artery variant was made on MRA and CTA in this case. On both modalities, the left PTAV was shown to arise from the proximal aspect of the cavernous segment of the left ICA, coursing posteriorly in a tortuous path along the left trigeminal nerve and terminating as the left AICA. Visualization and identification of PTAVs may be difficult because they are usually small in caliber [8].

Although PTA and PTAV are typically incidental findings, they may have clinical implications, such as trigeminal neuralgia. In our case, the vascular loop of the PTAV compresses the left trigeminal nerve from the inferior surface, specifically where the V3 fibers course. This corresponds with the patient's presentation of pain in the V3 distribution.

Trigeminal neuralgia is thought to be caused by neurovascular compression in the trigeminal root entry zone, most frequently by the superior cerebellar artery, which causes nerve root atrophy or displacement [9]. Although infrequent, a PTAV may cause trigeminal neuralgia due to the proximity of its

course to the root entry zone, as in this case. After the PTAV enters the posterior fossa through the Meckel cave or an isolated dural foramen, it courses dorsally and runs near the root entry zone of the trigeminal nerve. Classic-type PTAs are less likely to cause trigeminal neuralgia because they tend to course medially to join the basilar artery after entering the posterior fossa, rather than the more lateral course of PTAVs [1]. Those PTAs that do result in trigeminal neuralgia may have a long and tortuous course.

Other clinical associations with PTAVs include cerebral vascular disease, such as vertebrobasilar embolic ischemia and trigeminal cavernous fistulas [1]. These fistulas present clinically with ocular symptoms. Intracranial aneurysms may not be associated with PTA or its variants, according to a recent analysis of a large number of MRA studies (>16,000 patients) [3]. Their reported incidence of intracranial aneurysms coexisting with PTA and PTAV was 4.2% and 4.0%, respectively, which was similar to the prevalence of aneurysms in the general population ($3.7 \pm 0.7\%$) [3].

Treatment and prognosis

Treatment options for vascular loop compression trigeminal neuralgia range from conservative management with medications to surgical intervention. Microvascular decompression surgery provides the most complete and durable relief from trigeminal neuralgia [10]. During microvascular decompression, most PTA and PTAV loops are found to compress the medial aspect of the root entry zone and branch into several perforating arteries that continue to the brainstem around the entire nerve root [2]. Therefore, caution should be taken to avoid avulsion or kinking of the perforating branches when repositioning these vessels during surgery. In several cases of trigeminal neuralgia caused by PTA or PTAV, facial pain resolved after the surgery [2,10,11].

Another treatment option for vascular loop compression trigeminal neuralgia is gamma knife radiosurgery, a noninvasive stereotactic radiosurgical technique that utilizes a focused beam of radiation to target the root of the trigeminal nerve. This option may be preferred in elderly patients because of the lower complication rate [12]. Other available minimally invasive interventions for trigeminal neuralgia include radiofrequency rhizotomy, percutaneous glycerol rhizotomy, and percutaneous balloon compression.

Conclusion

Knowledge of PTAV is useful in the general interpretation of cranial MR angiography. Although rare and usually asymptomatic, it is important to recognize PTA and PTAV as potential causes of trigeminal neuralgia, especially in medically refractory cases where surgical treatment is considered.

Patient consent statement

Patient consent was not obtained; however, any and all identifiable patient information has been removed.

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