

PERSISTANT PRIMITIVE TRIGEMINAL ARTERY WITH VERTEBRO-BASILAR HYPOPLASIA WITH VERTEBRO-BASILAR INSUFFICIENCY- A CASE REPORT

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ABSTRACT

Persistent primitive trigeminal artery (PPTA) is one of the persistent carotid-vertebro-basilar anastomosis. It is usually unilateral and seen in 0.1 - 0.6 % of cerebral angiograms. It is associated with other anomalies like ectasia and fenestration of the ACA, absence of CCA, the absence of both ICAs, hypoplastic basilar artery and vertebral arteries, bilateral occlusion of the vertebral arteries, primitive otic artery, intracranial aneurysms at the circle of Willis, carotid cavernous fistula, miscellaneous conditions like NF1, Klippel Feil syndrome, Moya-Moya disease, supratentorial and infratentorial AVMs. It can present as vertebro-basilar insufficiency due to steal phenomenon and due to micro emboli originating from atherosclerotic carotid artery which travel via PPTA to posterior circulation. We present cases of PPTA causing vertebro-basilar insufficiency with cerebellar signs due to steal phenomenon and flattening of ventral surface of pons on the left side due to long standing pulsations of the PPTA, and pontine infarct. Striking finding in one of our cases is agenetic appearance of bilateral vertebral arteries in intracranial portion along with proximal two third of basilar artery.

Keywords: Persistent primitive trigeminal artery, Tau sign, vertebro-basilar insufficiency, persistent carotid - vertebrobasilar anastomosis.

Introduction

In dates back to the year 1844 the earliest description of persistent trigeminal artery was given. Persistent trigeminal artery (PTA) is the most common of the primitive carotid-basilar anastomoses with an estimated incidence of 0.1 to 1.0% on cerebral angiograms that persist into an adult. A number of cases of the PTA and its variants were found at autopsy on cerebral angiograms and on MRI and MRA scans demonstrating the anatomy of this primitive anastomosis and its relationship to various vascular diseases.¹

Case Report

A 55 year old male patient presented with gait ataxia and lower limb weakness since 1 day. He was a known hypertensive. On examination, there was no nystagmus. Finger nose ataxia and Romberg's signs were positive. There was swaying towards the left. Deep tendon reflexes were exaggerated. Power in both lower limbs was 4/5 and upper limbs was 5/5. Sensations were normal. Higher functions and speech were normal. Clinical suspicion of posterior circulation stroke was made. MRI Brain with MR Angiography of the brain and neck was done. There were few small, scattered hyperintense areas in bilateral frontal and

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parietal white matter on T2WI and FLAIR not showing restricted diffusion on DWI suggestive of chronic white matter ischemic changes. No acute infarct or intracerebral hemorrhage was noted. There was a flattening of pons on left side (Fig. 1). Both cerebellar

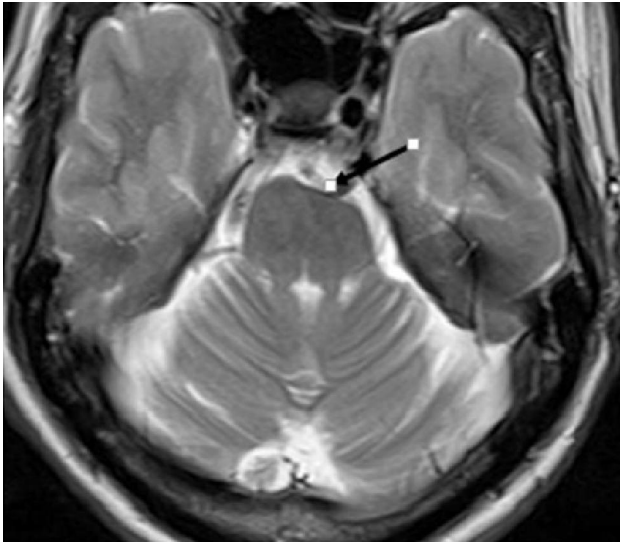


Figure 1: Axial T2 image showing PTA on left side with flattening of ventral surface of pons on left side (marked by the black arrow).

hemispheres appeared normal (Fig. 2). MR Angiography of the neck revealed normal CCAs, ICAs, ECAs and cervical vertebral arteries. MR angiography of brain showed markedly hypoplastic V3 and V4 segments of bilateral vertebral arteries and proximal and mid portion of basilar arteries. The persistent primitive trigeminal artery was seen on left side indenting and flattening ventral surface of left half of

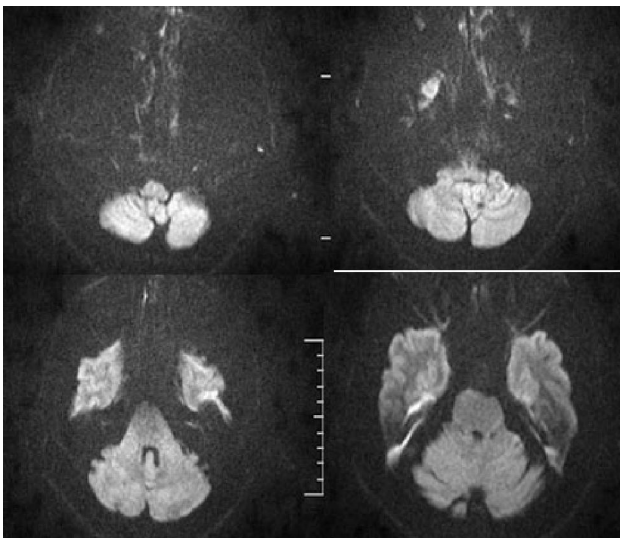


Figure 2: Diffusion Image showing no acute infarct in cerebellum and brain stem.

the pons. PCOM on the left side was not seen. A1 segment of right ACA was hypoplastic with both ACAs filling from the left side. MCAs, PCAs were normal (Fig. 3 and 4).

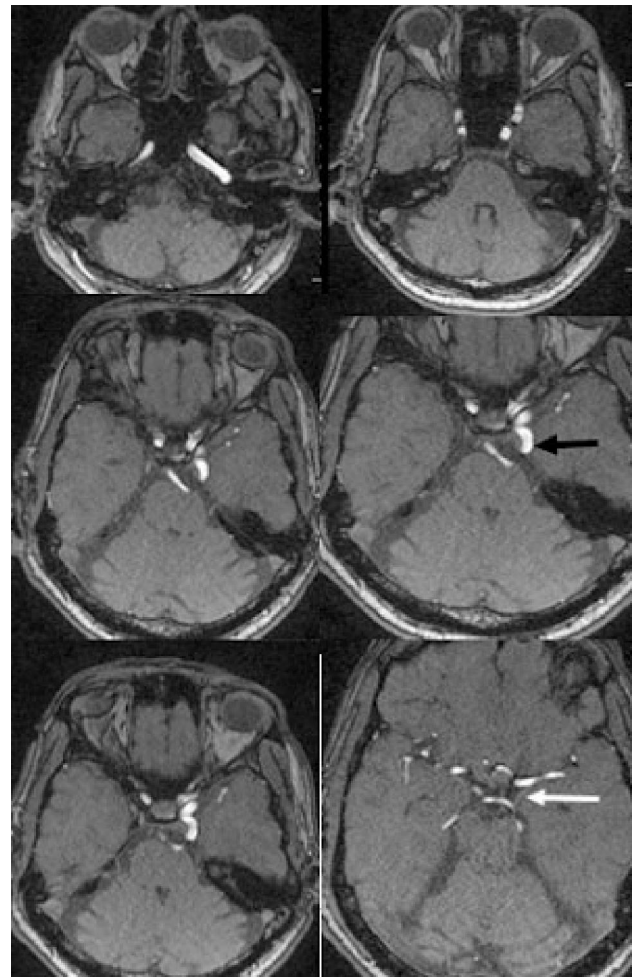


Figure 3: MR Angiography brain showing bilateral hypoplastic vertebral arteries, hypoplastic basilar artery in proximal half, PTA on left side (marked by black arrow) with hypoplastic left PCOM (marked by the white arrow).

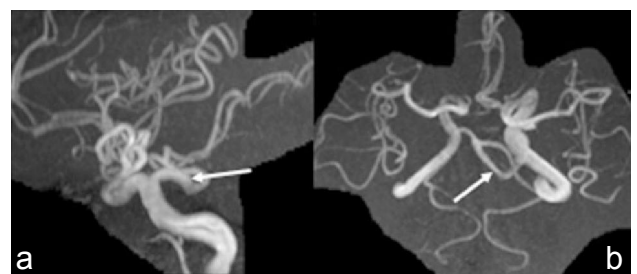


Figure 4a: Lateral MRA shows Tau sign – Trident shape (marked by white arrow).

4b: MRA showing bilateral hypoplastic vertebral arteries and basilar artery, the hypoplastic A1 segment of right ACA, a persistent primitive trigeminal artery on the left side (marked by white arrow), absent PCOM on left side.

Another 61 year old female patient presented with acute onset nausea and vomiting, slurring of speech, inability to move left upper and lower limbs and fall while trying to walk for 6 hours. The power was 1/5 in the left upper and lower limbs and reflexes were absent. MRI brain showed acute infarct in pons on the right side showing restricted diffusion on DWI (Fig. 5) with persistent primitive trigeminal artery on the right side on MR angiography of the brain (Fig. 6).



Figure 5: Another patient with diffusion weighted image showing acute infarct in pons on right side with restricted diffusion (marked by white arrow).

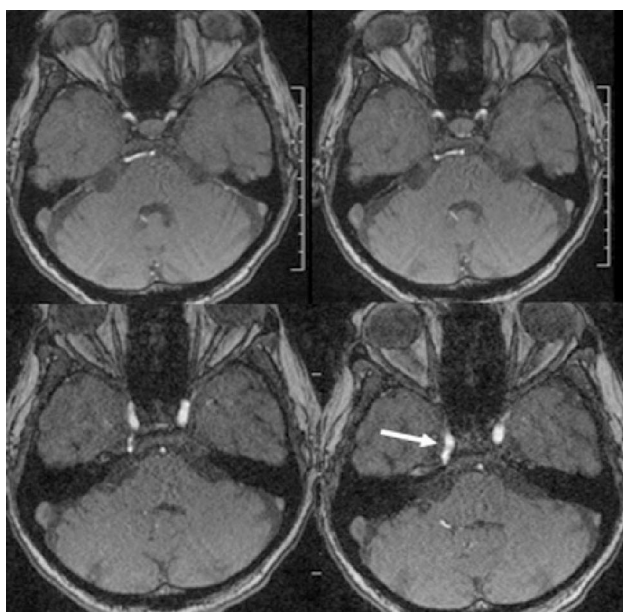


Figure 6: MRA brain showing lateral type PPTA on right side (marked by white arrow).

Discussion

During embryogenesis (about 35 days of gestational age), anastomosis rise between carotid and vertebro-basilar system at 4 sites - trigeminal, otic, hypoglossal and proatlanto intersegmental. These anastomosis persists for 1 week and regress with subsequent development of the vertebral arteries and posterior communicating arteries.² Persistent primitive trigeminal artery (PPTA) is one of the persistent carotid-vertebro-basilar anastomosis. It is usually unilateral and seen in 0.1 - 0.6 % of cerebral angiograms. It is the most cephalad and common of persistent fetal anastomosis. It arises from presellar ICA as it exits the carotid canal and enters the cavernous sinus. It extends posteriorly usually between the origins of superior and anterior inferior cerebellar arteries and join the mid basilar artery.³ Flow in the anastomosis is from the ICA to the basilar artery.

Sagittal MR image shows anomalous vessel as an abnormal flow void originating from the posterior aspect of cavernous ICA with horizontal course terminating in basilar artery seen as a Tau sign (combination of vertical and horizontal segments of the ICA and the proximal portion of the trigeminal artery creates an outline of Greek letter Tau). The part of the basilar artery caudal to the anastomosis with the trigeminal artery is usually hypoplastic.⁴

PTA is of two types (Salas classification) - lateral and medial (both are equally common).⁵ In the lateral type (Petrosal type), it courses posterolaterally with the trigeminal nerve while medial type (intrasellar, sphenoidal or transhypophyseal persistent trigeminal artery) has transhypophyseal or intrasellar course, coursing postero-medially from its origin, with compression of pituitary gland and penetrating the dorsum sellae. Medial or intrasellar location of PPTA is must to be diagnosed in patient undergoing transphenoidal surgery for pituitary adenoma as accidental transection of the artery can cause life threatening hemorrhage.⁶ PPTA is classified according to the configuration of the ipsilateral posterior cerebral artery. In Saltzman Type 1, posterior communicating artery is absent or poorly opacified and PCA is normal. PPTA is the main supply to the distal basilar artery, posterior cerebral artery and superior cerebellar artery territories. In Saltzman Type 2, P1 segment of PCA is absent with ipsilateral PCA arising directly from the ICA (fetal



origin of PCA). Basilar artery is usually hypoplastic in both types caudad to the anastomosis.⁷ In Saltzman Type 3, (called Saltzman variants), PTA variant directly joins to a cerebellar artery. It arises from the ICA and anastomoses directly with superior cerebellar artery (Saltzman type 3a), anterior inferior cerebellar artery (Saltzman type 3b) or with posterior inferior cerebellar artery (Saltzman type 3c) without anastomosing with basilar artery (Fig. 7 and 8). Type 3b variant is most

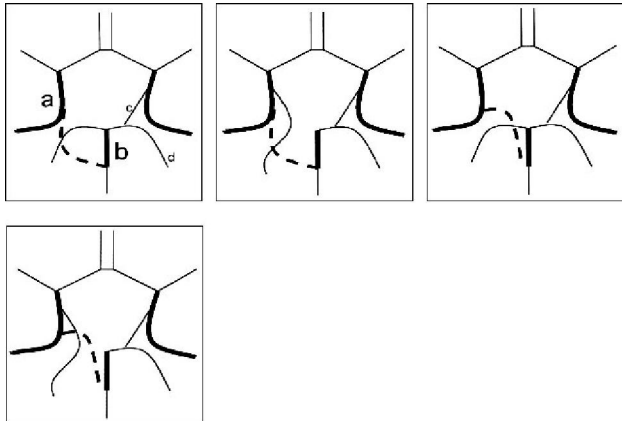


Figure 7: Drawings shows four types of persistent trigeminal artery in inferosuperior projection. A, Drawing shows lateral type, Saltzman type 1. a = internal carotid artery, b = basilar artery, c = posterior communicating artery, d = posterior cerebral artery. B, Drawing shows lateral type, Saltzman type 2. C, Drawing shows medial type, Saltzman type 1. D, Drawing shows medial type, Saltzman type 2.⁸

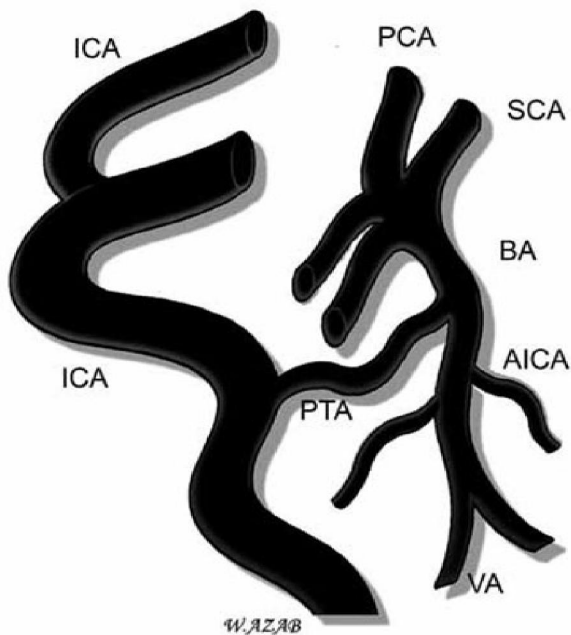


Figure 8: Schematic representation of origin of PTA from ICA, coursing between SCA and AICA and entering basilar artery.⁵

common. PTA variants are uncommon with an incidence of approximately 0.18 %. Detection of Saltzman is difficult as these are small in caliber.^{8,9}

The presence of embryonic vessel in adult life is indicative of disturbed cerebro-vascular development. Hence PPTA is associated with other anomalies like ectasia and fenestration of the ACA, absence of CCA, the absence of both ICAs, hypoplastic basilar artery and vertebral arteries, bilateral occlusion of the vertebral arteries, primitive otic artery, intracranial aneurysms at the circle of Willis, carotid cavernous fistula, miscellaneous conditions like NF1, Klippel Feil syndrome, Moya-Moya disease, supratentorial and infratentorial AVMs. It can present as vertebrobasilar insufficiency with resultant brain stem and cerebellar infarct due to micro emboli originating from atherosclerotic carotid artery, which travel via PPTA to posterior circulation or due to occlusion of the PPTA with resultant decreased flow in the basilar artery.² Lateral variant of PTAS is associated with brainstem ischemia, trigeminal neuralgia and ophthalmoplegia. Medial variant is associated with posterior fossa symptoms due to steal phenomenon. PTA rarely causes trigeminal neuralgia due to their intracranial course. They run medially and join basilar artery and are less likely to compress root entry zone of the trigeminal nerve. They can cause trigeminal neuralgia only when their course is long and tortuous. PTAVs (type 3) enter posterior fossa through the Meckel's cave or isolated dural foramen, do not join basilar artery and course dorsally and run near the root entry zone of the trigeminal nerve and hence can cause trigeminal neuralgia.¹⁰

Striking finding in our case is agenetic appearance of bilateral vertebral arteries in intracranial portion along with proximal two third of the basilar artery.

Conclusion

Identification of PPTA is essential in cases of vertebrobasilar insufficiency and brain stem infarct. Detection of agenetic / hypoplastic PCOM and hypoplastic P1 segment of PCA helps in classifying PPTA in Saltzman type 1 and type 2 respectively.

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