

Proptosis and complete oculomotor ophthalmoparesis due to a persistent trigeminal artery

Proptosis y oftalmoparesia oculomotora completa secundaria a una arteria trigeminal persistente

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Abstract

The persistent trigeminal artery (PTA) is a rare vascular anomaly which represents a persistent communication between the internal carotid artery and vertebrobasilar system. This anastomosis is important because it provides the main source of blood supply during the development of the embryonic brain in its most critical stage. The diagnosis, in the majority of cases, concerns an incidental finding. The 25% of the persistent trigeminal arteries are linked to some vascular brain diseases, especially with intracranial aneurysms, and neuroophthalmologic involvement is not frequent. This case reports a male patient who developed proptosis and a complete ophthalmoparesis of the third cranial nerve with an anatomical variant of the normality consisting of a PTA detected by the magnetic resonance imaging.

Key words: Oculomotor nerve. Proptosis. Angiography. Diplopia. Ophthalmoplegia.

Resumen

La arteria trigeminal persistente es una anomalía vascular poco común que se presenta como una comunicación entre la arteria carótida interna y el sistema vertebrobasilar. Esta anastomosis es importante debido a que asegura el correcto aporte sanguíneo del cerebro durante el desarrollo embrionario, que es la etapa más crítica. El diagnóstico, en la mayor parte de los casos, es un hallazgo accidental. En el 25% de los casos, la arteria trigeminal persistente se relaciona con algunas enfermedades cerebrales de tipo vascular, especialmente con aneurismas intracraneales, mientras que la repercusión neurooftalmológica no es frecuente. Se reporta el caso clínico de un paciente varón que desarrolla una proptosis, así como una oftalmoparesia completa del tercer par craneal con una variante anatómica de la normalidad, que consiste en una arteria trigeminal persistente detectada mediante resonancia magnética.

Palabras clave: Nervio oculomotor. Proptosis. Angiografía. Diplopía. Oftalmoplejía.

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Introduction

In the embryonic stage, the posterior circulation, represented by the primitive vertebrobasilar system, depends largely on the established anastomosis with the primitive internal carotid. One of this anastomosis is represented by the persistent trigeminal artery (PTA) (it represents 85% of the permanent vasculature cases in adult age). It is so designated due to its link with the closest cranial nerve, and connects the cavernous portion of the internal carotid artery (ICA) with the basilar system. This anastomosis is important because they provide the main source of blood supply during the development of the embryonic brain in its most critical stage¹⁻³.

It is originated during the 4th week intra-uterine life, and suffer a process of regression in the 8th week, period in which the correct blood supply toward the posterior fossa is assured^{1,3}.

There are various anatomical classifications from PTA according to its route, which were previously named after Saltzman. The most important one from the clinical perspective originates from the cavernous sinus, and it flows proximal to the sensorial trigeminal nerve, lateral to the sella turcica (Saltzman type 2). This variant has most often been linked to ischemic cerebrovascular diseases, neuralgia and paralysis of the trigeminal, and oculomotor common paresis or paralysis, while the medial variant (less frequent than the previous one and called Saltzman type 1) has been related with symptoms caused by posterior fossa steal syndromes due to the fact that it is connected to basilar and hypoplastic posterior communicating arteries^{2,3}.

The diagnosis, in the majority of cases, concerns an incidental finding in cerebral vasculature through neuroimaging, found in 0.1% to 0.6% of the cerebral angiographies performed. However, there are reported cases in which its direct etiology has been proposed with several pathological processes such as neurovascular compressions and vertebrobasilar symptoms³⁻⁸. The 25% of the persistent trigeminal arteries are linked to some vascular brain diseases, especially with intracranial aneurysms, which are frequently found in the bifurcation of the internal carotid^{2,6,8}.

We present a case report about a male patient with this pathology that manifested itself with ophthalmological signs that although they are described in the literature, recent cases have not been described.

Case report

A previously healthy 63-year-old patient male was referred with a 4-year history of proptosis development in

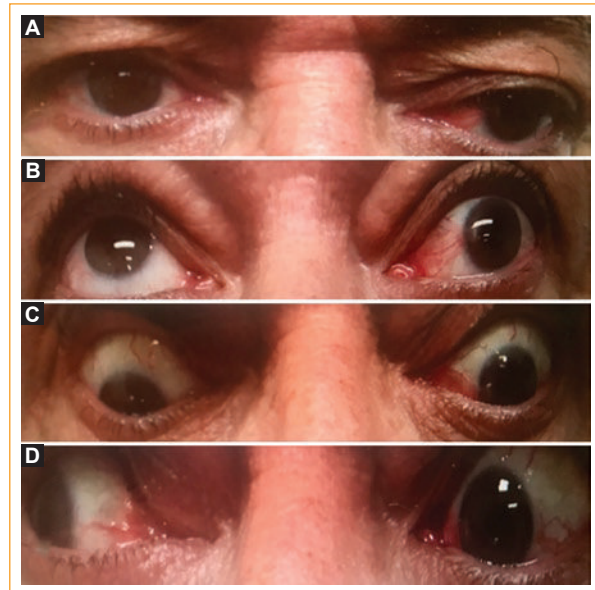


Figure 1. External photograph. **A:** Conjunctival hyperemia, proptosis, ptosis, and exotropia of the left eye; **B:** Limitation to supraversion; **C:** Limitation to infraversion; **D:** Limitation to adduction.

the left eye (OS), accompanied by diplopia, conjunctival chemosis, and palpebral ptosis (Fig. 1). In the ophthalmological explorations, the visual acuity was 20/40 in the right eye (OD) and 20/63 in the OS, apart from a limitation in the oculomotor motility to the adduction, supraversion, and infraversion (Fig. 1). In the anterior segment, hyper-reactive mydriasis was seen. The ocular fundus was normal in both eyes, but the abnormal visual field of the OS showed a large generalized scotoma.

A soft tissue density injury is detected in computed axial tomography of the orbits (CTA). It occupies the orbital apex, the optic foramen, and the left cavernous sinus, so therefore, a magnetic resonance imaging showed an anatomical variant consisting of a PTA which is conducted from the left carotid loop to the middle third of the basilar artery, and generates an left carotid artery size increase of the same secondary side to an outflow, without the identification of neither possible aneurysms nor inflammatory disturbances at a cavernous sinus level (Fig. 2).

Visual evoked potentials were done with the result of a delay in the conduction of the left retrobulbar visual pathway with diminished amplitude.

Once the cause of a complete ophthalmoparesis of the third cranial nerve by compression of PTA at mesencephalic level is confirmed, it is decided to perform a selective carotid angiography, where elongation and

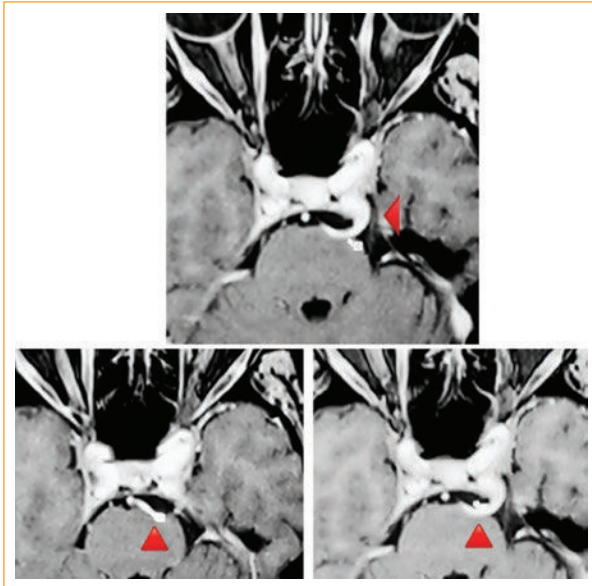


Figure 2. Cranial magnetic resonance imaging: T1-weighted axial section showing left tubular image corresponding to the persistent trigeminal artery which is conducted from the left carotid loop to the middle third of the basilar artery.

severe tortuosity of supra-orthotics trunks are found, severe stenosis in the outgoing of the left vertebral artery; which makes its catheterization impossible (Fig. 3).

Discussion

The PTA is an anastomosis of the embryonic vascular cerebral system, which flows between the ICA and the basilar artery, and therefore, connects the anterior circulation with the posterior one. Its presence disrupts the usual organization of the cranial nerves within the cavernous sinus; consequently, the oculomotor nerves could be displaced in any direction¹⁻³.

It is generally presented in an asymptomatic manner and usually identified as incidental in certain angiographic studies due to other reasons. Within the clinical picture, the affection of the ophthalmic nerves is found in third place, together with trigeminal neuralgia for compressive reasons. Within the oculomotor involvement, nerve VI is the most affected (up to 4 out of 6 cases), followed by nerve III (as it is in our case)⁹.

The oculomotor nerve (nerve III) is originated in the mesencephalic nuclei. Previous to its entry into the orbit, it penetrates through the lateral wall of the cavernous sinus together with the pathetic nerve, abducens

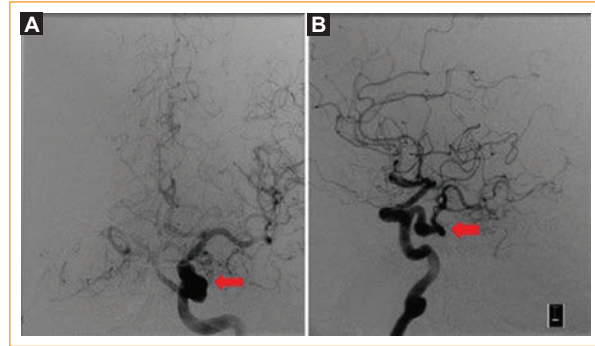


Figure 3. (A and B) Left selective angiography: persistent trigeminal artery origin and its journey to the basilar artery.

nerve, and the first branch of the trigeminal nerve. The nerve compression could be produced at a midbrain level, in its pars sellaris and/or at a cavernous sinus level^{9,10}.

Its injuries are manifested with binocular diplopia, droopy eyelid, paralysis of the elevation or view descent, and abduction alterations; if it was due to compressive reasons, the parasympathetic innervation of the ciliary muscle would be affected, therefore resulting in non-reactive mydriasis^{9,10}. Our patient is compatible with a complete left ophthalmoparesis of the third cranial nerve.

The paralysis of the third nerve are classified as infranuclear and supranuclear. The first is acquired and caused for the most part by vascular diseases, vascular malformations, tumors, and among others. As the trigeminal artery is frequently associated with cerebral vasculature alterations, it can produce ophthalmoparesis for own compressions, hemorrhage, aneurysms and even fistulas, in which case eye pain, chemosis, proptosis, and ophthalmoplegia may be caused. In the case presented, the alterations are produced in themselves by the own artery as it presents a significant fusiform dilatation.

All patients must be evaluated neuroradiologically. The first test must be a CTA examination, which will allow to visualize the trigeminal artery and subsequently an angiographic magnetic resonance or angiography to confirm the diagnosis^{1,3}. If the patient presented at anytime severe headaches, besides the other accompanying symptoms, it would be indicative of an ictal case or stroke, so the measures to be carried out would become invasive.

In conclusion, PTA is a rare anatomical variant, its diagnosis in most cases is typically accidental because

it does not present clinical manifestations, and if it did present any, the most common picture would be cerebrovascular. Oculomotor paralysis would occupy, this way, the third place in frequency. Due to this, it is a pathology that is taken into account whenever carrying out a differential diagnosis in response to these alterations. Patients should be evaluated neuroradiologically.

In case of concluding the diagnosis, a thorough anatomical-vascular study is required due to its high relation with vascular-cerebral morphology alterations and aneurysms which could require endovascular/neurosurgical treatment in case it becomes symptomatic.

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Ethical disclosures

The next manuscript has been evaluated favorably as appropriate by the ethics and research committee of the Doctor Negrin University Hospital, with the following approval code: 170072.

Protection of human subjects and animals in research. The authors declare that the procedures followed were in accordance with the regulations of the responsible Clinical Research Ethics Committee and accordance with those of the World Medical Association and the Helsinki Declaration.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors must have obtained the informed consent of the patients and/or subjects mentioned in the article. The author for correspondence must be in possession of this document.

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Conflicts of interest

The authors declare no conflict of interest.

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