Coincident intrasellar persistent trigeminal artery and craniopharyngioma: case report and implications for transsphenoidal surgery☆

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Abstract

The persistent trigeminal artery (PTA) is the largest and most commonly occurring type of remnant fetal arteries, typically originating from the posterior bend or lateral wall of the intracavernous carotid artery. There are no published reports of coexisting PTA and midline epithelial tumors. We describe a coincident case of craniopharyngioma associated with an adjacent PTA traversing through the sella turcica, which are both developmental midline skull base anomalies. The caliber and location of the PTA precluded an endoscopic endonasal operation; the tumor was safely resected via a supraorbital keyhole approach. Anatomic implications pertaining to surgical approach and treatment are also discussed.

Introduction

The persistent trigeminal artery (PTA) is the largest and most common type of remnant fetal carotid–basilar anastomotic arteries, seen in 0.1%—1.0% of cerebral angiograms [1,2]. PTAs are known to be associated with a wide range of pathology [3]. In this article, we describe the first known case of craniopharyngioma associated with a PTA. We also review the literature and discuss the impact of anatomic variation on surgical planning.

Case report

History

A 56-year-old man presented with severe visual loss in the right eye and bitemporal hemianopsia. MRI demonstrated a large midline suprasellar mass adherent to vessels of the circle of Willis and compressing the optic chiasm and hypothalamus. Fig. 1 shows a large flow void that was noted directly inferior to the tumor on MRI. Figs. 2 and 3 show a large fetal-type PTA coursing along the left and midline aspects of the sella turcica, exiting via a foramen in the dorsum sellae and connecting the left internal carotid artery to the basilar artery on CT angiogram. The PTA was Saltzman type I, with the left vertebral artery terminating as the PICA and hypoplastic right vertebral and basilar arteries.

Operation

Due to the caliber and course of the PTA, a right supraorbital keyhole approach was selected for tumor resection. The tumor was internally debulked from the left optic nerve, carotid artery, and optic chiasm. The pituitary stalk, infiltrated with tumor, was sacrificed at the level of the diaphragma sellae. The PTA was seen coursing immediately below the diaphragma sellae and confirmed using micro-Doppler ultrasonography. Tumor dissection between the left P1 artery and the basilar artery was successful; only a small area of residual tumor remained firmly attached to the basilar artery and was deliberately left in place.

Pathological findings

Pathology confirmed that the suprasellar mass was an adamantinomatous craniopharyngioma.

Postoperative course

As expected based on deliberate resection of the tumor and pituitary stalk, panhypopituitarism with diabetes insipidus requiring full hormone replacement developed postoperatively. Stereotactic
radiosurgery was administered for the residual tumor. At one year, vision improved and no new focal neurological deficits developed.

Discussion

Craniopharyngiomas and PTA

Craniopharyngiomas are rare midline epithelial tumors with an incidence rate in the United States of 0.18 per 100,000 person-years [4]. These tumors typically originate in the suprasellar region and often affect the hypothalamus and the optic pathways. It is generally accepted that they are developmental neoplasms arising from remnant squamous epithelial cells of Rathke’s pouch [5].

PTAs normally originate from the intracavernous ICA. The trigeminal artery first appears in the embryo at the 3 mm stage [3]. During the 4–5 mm stage of the embryo, a portion of the ICA begins to supply this region, eventually developing into the posterior communicating artery. This results in regression of the trigeminal artery [2,3]. Persistent trigeminal arteries are evaluated by the Saltzman classification as type I, II, or III. Type I involves the PTA supplying the SCA and PCA, a hypoplastic proximal basilar, and an absent ipsilateral posterior communicating artery. A PTA is classified as a Type II if it only supplies the anterior superior cerebellar arteries and the PCAs arise from the posterior communicating arteries. Finally, Type III describes a PTA connected with another fetal remnant, the primitive paired longitudinal neural artery, and supplying only one ipsilateral cerebellar artery without connection to the basilar artery. PTAs are also associated with other regional vascular abnormalities, such as hypoplastic or absent vertebral arteries. Type III is the most common, at 60% of cases, with Type I following at 24% and Type II at 16% [6]. As briefly noted above, our case was a Saltzman Type I.

The prevalence of PTA is roughly 0.1%–0.6% of the general population; while rare, PTAs have been linked to a variety of clinical conditions in the literature, most commonly aneurysms [1,2]. However, there have been no reports describing coincident PTA and craniopharyngioma. Although confirmation of any definitive connection is beyond the scope of this article, this case report is interesting because it describes two concurrent midline developmental anomalies located adjacent to each other.

The PTA and surgical planning

Although the transsphenoidal approach has been successfully performed in patients with PTA, we chose to use an endoscopic supraorbital approach for this patient, as it offers nearly equivalent tumor exposure with decreased risk of injury to the PTA and no requirement for manipulating the pituitary gland or diaphragm sellae. Our surgical planning reflects the need for all surgeons performing transsphenoidal procedures to be vigilant for vascular anomalies on MRI, as well as fully explore them using noninvasive vascular imaging. The consequences of not fully appreciating these anomalies prior to surgery could be catastrophic.

Conclusions

This case, the first published report of a coincident PTA and craniopharyngioma, is a meaningful demonstration of coexisting...
anatomical variations present in the midline skull base. However, it also stands as an example of how vitally important it is to identify and appreciate vascular abnormalities during surgical planning. We hope that our successful management of this complicated case can provide a blueprint for how to approach similar situations.

References


