Moyamoya Disease Associated with Tuberculum Sellae Meningioma and Cavernous Sinus Hemangioma

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Key words
- Cavernous sinus
- Meningioma
- Moyamoya disease
- Tuberculum sellae

Abbreviations and Acronyms
ACA: Anterior cerebral artery
ECA: External carotid artery
EDMS: Encephaloduromyosynangiosis
EMS: Encephalomyosynangiosis
GH: Growth hormone
ICA: Internal carotid artery
IGF-1: Insulin-like growth factor 1
MCA: Middle cerebral artery
MRA: Magnetic resonance angiography
MRI: Magnetic resonance imaging
PCA: Posterior cerebral artery
STA: Superficial temporal artery

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BACKGROUND: **Primary moyamoya disease associated with skull base tumors has been reported only rarely in the literature. Surgical treatment can be complicated due to the compensatory collateral circulation through meningeal and leptomeningeal anastomosis. A standard frontotemporal craniotomy may interrupt critical transdural anastomoses.**

CASE DESCRIPTION: **We report a case of primary moyamoya disease coexisting with tuberculum sellae meningioma and left cavernous sinus hemangioma. Simultaneous management of tuberculum sellae meningioma and moyamoya disease was performed using a left modified pterional incision. Two separate bone windows were opened to protect the transdural anastomosis via the middle meningeal artery. The tuberculum sellae meningioma was successfully removed through a small frontal craniotomy, and encephaloduromyosynangiosis was used to treat moyamoya disease through a temporoparietal craniotomy. Finally, CyberKnife radiotherapy was used to treat the left cavernous sinus hemangioma at 6 weeks after the initial operation. The patient recovered well without complications. This is the first report of moyamoya disease associated with tuberculum sellae meningioma and cavernous sinus hemangioma.**

CONCLUSIONS: **With careful bone flap design, moyamoya disease and skull base tumors can be treated simultaneously. Care should be taken to avoid interruption of critical dural-pial collaterals and injury to fragile moyamoya vessels.**

INTRODUCTION

Moyamoya disease is a chronic cerebral vasculopathy characterized by progressive stenosis and occlusion of the terminal portion of bilateral internal carotid arteries (ICAs) accompanied by moyamoya vessels. Although moyamoya disease has been associated with various treatments and diseases, such as cranial therapeutic irradiation,[1-3] Down syndrome,[4] neurofibromatosis,[5] and sickle cell anemia,[6] its etiology remains unclear. Primary moyamoya disease associated with various brain tumors, including craniopharyngioma,[2,7-9] pituitary adenoma,[10,11] and glioma,[12-14] has been reported in the literature; however, primary moyamoya disease coexisting with skull base meningioma has been reported only rarely. Ogama et al.[15] reported a case of moyamoya syndrome associated with basal meningioma that was successfully treated via a modified transsphenoidal approach; however, preoperative angiography was not performed, and moyamoya syndrome was not treated.

Here we report the first case of moyamoya disease with tuberculum sellae meningioma and left cavernous sinus hemangioma. Simultaneous surgical management of tuberculum sellae meningioma and moyamoya disease was performed using a left modified pterional incision. Tuberculum sellae meningioma resection plus the combination of direct and indirect revascularization was planned. The tuberculum sellae meningioma was successfully removed through a unilateral subfrontal approach, and encephaloduromyosynangiosis (EDMS) was used to treat moyamoya disease through a temporoparietal craniotomy. Finally, the left cavernous sinus hemangioma was treated with CyberKnife radiotherapy at 6 weeks after the craniotomy.

CASE PRESENTATION

A 46-year-old woman was admitted to our hospital with progressive visual loss over the previous 6 months. Ophthalmologic examination showed right temporal hemianopsia with severe impairment of visual acuity (left 20/20, right finger counting/50 cm). No other neurologic deficit besides the severe visual disturbance was detected. Endocrinologic examination revealed normal basal levels of pituitary hormones. Sagittal T1-weighted magnetic resonance imaging (MRI) demonstrated an isointense mass extending from the planum sphenoidale to the diaphragm sellae (Figure 1A). On contrast-enhanced T1-weighted MRI, the

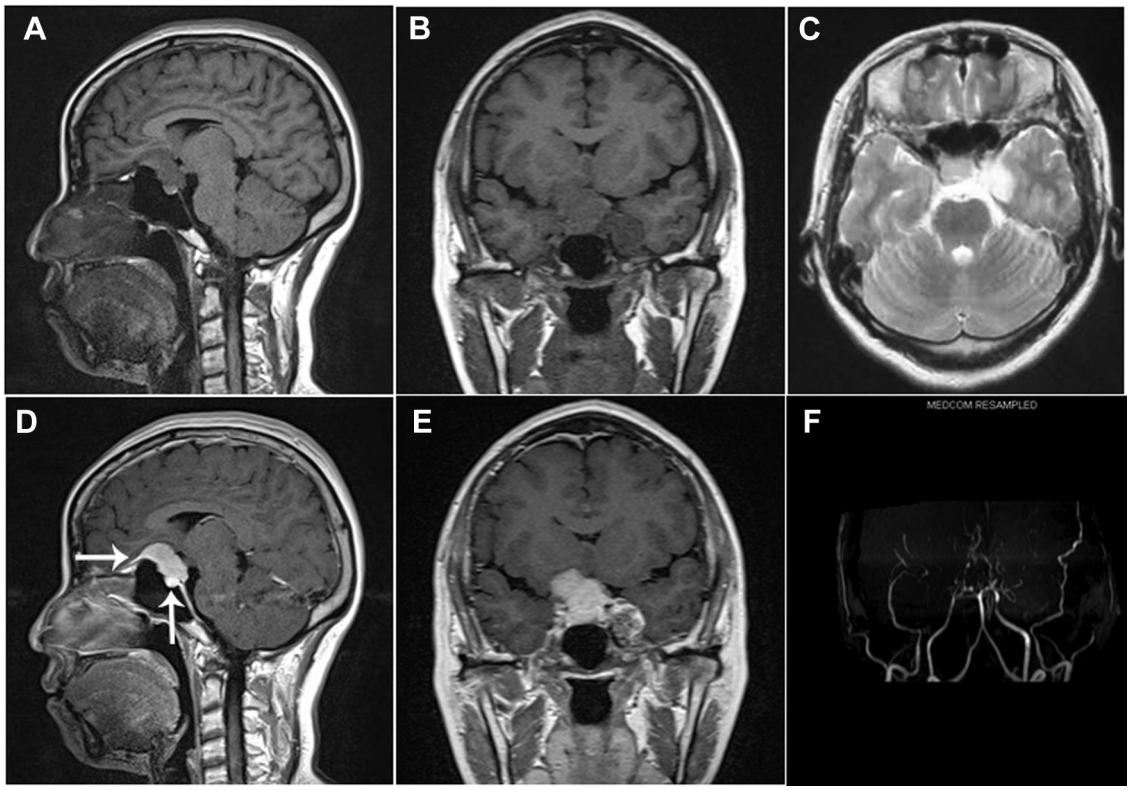

Figure 1. (**A**) Sagittal T1-weighted magnetic resonance imaging (MRI) demonstrating an isointense mass extending from the planum sphenoidale to the diaphragm sellae. (**B**) Coronal T1-weighted MRI showing another hypointense mass in the left cavernous sinus with homogeneous marked high signal intensity on axial T2-weighted MRI. (**C**) The mass involving the tuberculum sellae was homogenously enhanced after contrast administration, whereas the mass in the left cavernous sinus was heterogeneously enhanced. (**D**) Interesting view of "beak of Kiwi bird" enhancement for tuberculum sellae meningioma (*left arrow*), with the pituitary enhanced below (*right arrow*). (**E**) Coronal postcontrast MRI. (**F**) Magnetic resonance angiography showing occlusion of the bilateral internal carotid artery terminals.

tumor was homogenously enhanced with an obvious dural tail sign (Figure 1E), characterized as looking like the beak of a Kiwi bird. A T1-weighted coronal MRI showed another lesion involving the left cavernous sinus with hypointensity and heterogeneous contrast enhancement (Figure 1B and D); however, a homogeneous marked hyperintensity on T2-weighted MRI (Figure 1C) suggested the possibility of cavernous hemangioma.

We then performed a magnetic resonance angiography (MRA) scan to further explore the relationship of tumors with the bilateral ICAs and their branches. The MRA showed occlusion of the bilateral ICA terminals, suggestive of moyamoya disease (Figure 1F). A left carotid angiogram (Figure 2A–D) showed complete occlusion of both the proximal ACA and MCA with a small amount of moyamoya vessels and extracranial collaterals via the middle meningeal artery (ICA stage V).[16] A right carotid angiogram (Figure 2E–H) showed complete occlusion of the ICA just distal to the origin of the ophthalmic artery with supply of brain from the external carotid artery (ECA) (ICA stage VI). Single-photon emission computed tomography examination demonstrated decreased cerebral blood flow in the left side. Therefore, tuberculum sellae meningioma resection plus left superficial temporal artery (STA) to MCA bypass was planned. To avoid the risk of collateral vessel damage, careful preoperative planning, including bone flap design, was performed.

Operation

After obtaining written informed consent from the patient and his family, we performed the surgery via a left modified pterional approach. The skin incision was curved posteriorly, with the posterior branch of the STA included in the flap (Figure 3A). The incision was extended by 1.0–1.5 cm above the superior temporal line. The temporal muscle was incised along the posterior margin of the skin flap. The temporal muscle was separated using a periosteal elevator to expose the entire deep temporal artery network on the deep surface of the temporal muscle (Figure 3B). Two separate bone windows were opened to protect spontaneous anastomosis between the middle meningeal artery and cortical artery (Figure 3C). We preferred a unilateral subfrontal approach to remove the tuberculum sellae meningioma, using a small frontal craniotomy. The dura was opened parallel to the orbit. The left carotid cistern was opened, and a retractorless technique was used. The tumor originated from the tuberculum sellae, extending posteriorly into the diaphragm sellae. After the basal blood supply was interrupted, the