Retroorbital Hemangiopericytoma and Cavernous Sinus Schwannoma

—Case Report—

Atul GOEL, Dattatraya MUZUMDAR, Ketan DESAI, and Aadil CHAGLA

Department of Neurosurgery, King Edward VII Memorial Hospital and Seth G.S. Medical College, Parel, Mumbai, India

Abstract

An 18-year-old male presented with severe proptosis and blindness in the right eye. Neuroimaging revealed a large and hypervascular tumor in the right retrobulbar region and a large tumor in the left cavernous sinus. Angiography showed the right tumor was extensively vascular, fed by a hypertrophic ophthalmic artery, and the left tumor was moderately vascular, fed by a large middle meningeal artery. Following embolization of the feeder vessels, the right retrobulbar hemangiopericytoma and the left cavernous sinus schwannoma were uneventfully and successfully resected. Such combinations of different pathological lesions present unusual therapeutic challenges.

Key words: hemangiopericytoma, cavernous sinus, orbit, schwannoma

Introduction

Cases of multiple but different pathological lesions in the brain of the same patient are rare. We report a patient with a rare combination of a retrobulbar hemangiopericytoma in the right eye and a schwannoma in the dural confines of the lateral wall of the left cavernous sinus.

Case Report

An 18-year-old male was admitted with complaints of gradual progressive proptosis and worsening of vision in the right eye for 8 months. On admission, he had severe proptosis and complete loss of vision in the right eye. The eyeball was hanging almost out of the socket and there was marked conjunctival chemosis. All extraocular movements were severely restricted. The left eye function was normal. Neurological examination found no other abnormalities. There was no family history or cutaneous stigmata suggestive of neurofibromatosis 1. Magnetic resonance (MR) imaging showed a large retrobulbar lesion on the right and a large cavernous sinus lesion on the left (Fig. 1). The MR imaging characteristics of both lesions were similar. The lesions appeared isointense on both T1- and T2-weighted images and had multiple linear hypointense streaks suggestive of flow voids. Angiography revealed that the right retrobulbar tumor was extensively vascular and was fed by a hypertrophic ophthalmic artery (Fig. 2). The left cavernous sinus lesion was moderately vascular and was supplied by a large middle meningeal artery.

Embolization of the ophthalmic artery feeders of the right retroorbital lesion and the middle meningeal artery feeders of the left cavernous sinus lesion was carried out using glue. A lateral orbitotomy was performed and the large, vascular, and relatively well-defined extraconal tumor was completely resected, and all the nerves in the vicinity were preserved (Fig. 3). Histological examination showed a moderately cellular tumor comprising of ovoid cells with vesicular nuclei oriented around blood vessels in an ‘antler-horn’ configuration (Fig. 4). A few compressed slit-like blood vessels were also seen. Reticulin stain showed diffuse pericellular reticulin investing individual cells. The findings were consistent with hemangiopericytoma.

A left basal temporal craniotomy was then performed. Using an extradural interdural approach, the large tumor located within the confines of the lateral dural wall of the cavernous sinus was completely excised. Postoperative MR imaging showed no trace of either of the tumors (Fig. 5). Histological
examination showed a moderately cellular tumor comprising of spindle cells arranged in compact whorls and bundles with nuclear palisading (Antoni A) consistent with schwannoma (Fig. 6). Immunohistochemical examination of the tissue was not done.

The patient tolerated both the operations well. After 10 months, the right eye had normal movements, but without vision. The left eye was entirely normal, with preserved vision and extraocular movements. There was no sensory or motor loss in the distribution of the fifth cranial nerve.

**Discussion**

In the present case, both the retroorbital hemangiopericytoma and the cavernous sinus schwannoma were complex therapeutic and surgical challenges. The right retroorbital hemangiopericytoma was highly vascular but had a well-defined plane of dissection. The tumor was only removed with difficulty despite the successful embolization. The similar neuroimaging appearance of the right and left lesions suggested that the left lesion was also a hemangiopericytoma. As there was normal vision and intact extraocular movements in the only functioning eye, surgery on a vascular lesion located...
Fig. 4 Photomicrograph of the hemangiopericytoma showing a moderately cellular tumor comprising of ovoid cells with vesicular nuclei oriented around blood vessels in an ‘antler-horn’ configuration. A few compressed slit-like blood vessels are also seen. HE stain, ×160.

Fig. 5 Axial T1-weighted magnetic resonance image after the second surgery showing complete excision of both the right retroorbital hemangiopericytoma and the left cavernous sinus schwannoma.

Fig. 6 Photomicrograph of the schwannoma showing a moderately cellular tumor comprising of spindle cells arranged in compact whorls and bundles with nuclear palisading (Antoni A). HE stain, ×160.

tive of fifth cranial nerve or any other cranial nerve dysfunction either before or after surgery. Considering the location, one of the fibrils of the fifth cranial nerve appears to be the most likely origin. After successful resection of both the tumors, one of which was malignant and the other was benign, cranial radiotherapy was withheld as the retroorbital hemangiopericytoma was remarkably localized and was completely resected. Although the long-term outcome of the patient remains uncertain, the postoperative outcome was encouraging.

References


Address reprint requests to: Prof. A. Goel, Head, Department of Neurosurgery, King Edward VII Memorial Hospital, Parel, Mumbai-400012, India. e-mail: atulgoel62@hotmail.com.