Intracranial Extension of an Orbital Pseudotumor Accompanied by Internal Carotid Artery Occlusion

—Case Report—

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Abstract

A rare case of histologically verified orbital pseudotumor with intracranial extension in a 33-year-old male is reported. Preoperative radiographs showed the mass extending beyond the orbit and involving the bilateral cavernous sinuses. The intracranial lesion at biopsy surrounded the right internal carotid artery above the right cavernous sinus. Cerebral angiography showed complete occlusion of the cervical portion of the right internal carotid artery. Possibly, multiple lesions had extended contiguously through the venous system around the cavernous sinus, with subsequent involvement of the internal carotid artery. This case suggests that orbital pseudotumor may not be a separate clinical entity, but a part of the syndrome caused by inflammation of the venous system around the cavernous sinus.

Key words: pseudotumor, exophthalmos, inflammation, orbit

Introduction

Orbital pseudotumor is a term generally accepted as describing a nonspecific idiopathic inflammatory lesion of the orbital tissues, which initially mimics a true neoplasm.^{3,5)} Orbital pseudotumors are usually confined to the muscular cone, and rarely extend to the paranasal sinuses or cavernous sinus.^{4,7,9)} We present a rare case of histologically verified orbital pseudotumor with intracranial extension, accompanied by complete occlusion of the internal carotid artery.

Case Report

A 33-year-old male first noted double vision in June, 1982. Recurrent, severe pain in the right orbital, frontal, and occipital regions began 1 week later. He underwent surgery for chronic paranasal sinusitis at another institution, but his condition did not improve significantly. In July, 1988, he developed

gradually progressive visual loss on the right over 3 months. The pain was refractory to conservative therapy. He was eventually admitted to our hospital in July, 1990.

On admission, he was blind on the right. Physical examination found hyperesthesia in the distribution of the first and second divisions of the trigeminal nerve and the second and the third cervical nerves on the right. No decrease in facial sensation could be detected. No pupil abnormality, proptosis, ptosis, restriction of ocular movements, conjunctival injection, or chemosis was present. Ophthalmoscopy showed right optic atrophy. Blood, urine, and cerebrospinal fluid analyses gave normal results except for an increased antinuclear antibody level. A chest x-ray film was normal.

Computed tomographic (CT) scans demonstrated a dense, soft tissue mass in the right orbit extending posteriorly to the bilaterally enlarged cavernous sinuses (Fig. 1). Magnetic resonance (MR) imaging showed the lesion to be hypointense to fat but

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Fig. 1 CT scan, showing the lesion extending from the right orbit into the bilateral cavernous sinuses (arrows).

Fig. 2 Both T₁- (left) and T₂-weighted (right) MR images, showing the lesion hypointense to fat but isointense with muscle (arrows).

isointense with muscle on both T₁- and T₂-weighted images (Fig. 2). The signal intensity of the lesion in the orbit did not change on entering the cavernous sinus. The intracavernous internal carotid artery flow void was absent on the right. Bilateral carotid angiograms revealed complete occlusion of the cervical portion of the right internal carotid artery (Fig. 3), with stenosis of the cavernous portion of the left internal carotid artery. A transfrontal orbital venogram disclosed complete occlusion of the bilateral superior ophthalmic veins in the orbital apices and no filling of the cavernous sinus on either side.

Steroid treatment was begun, but achieved neither relief of the pain nor diminution of the mass on serial CT scans. Because of his persistent severe symptoms and the lack of a definitive diagnosis, a right frontotemporal craniotomy with orbital unroofing was performed in August, 1990.

A firm, fibrous, and relatively avascular tumor was found in the orbital apex with posterior extension through the right optic canal and the right superior orbital fissure. Intracranial tumor was present under the right internal carotid artery and optic nerve, and wrapped around the internal carotid artery above the right cavernous sinus. The right optic nerve was dark red and degenerating. Multiple orbital and intracranial biopsy specimens all showed proliferation

of dense, hyalinized, fibrous tissue with minimal cellularity, compatible with a diagnosis of healed inflammatory pseudotumor (Fig. 4). No biopsy specimens were obtained from within the cavernous sinus. According to the rapid histological diagnosis of pseudotumor by frozen sections, no further tumor removal was performed. A right superficial temporal artery biopsy showed no evidence of temporal arteritis.

Postoperatively, his signs and symptoms were essentially unchanged. He received with nonsteroidal anti-inflammatory drugs such as mefenamic acid. His pain improved slowly and he was discharged 1 month after surgery.

Fig. 3 Right carotid angiogram, demonstrating complete occlusion of the cervical portion of the internal carotid artery (arrow).

Fig. 4 Photomicrograph of the tumor biopsy specimen, showing dense, hyalinized, fibrous tissue with minimal cellularity. HE stain, ×100.

Discussion

Birch-Hirschfeld²⁾ introduced the term orbital pseudotumor in 1905 to cover all orbital masses other than neoplasms. The term is now generally accepted as indicating a nonspecific inflammatory lesion of the orbital tissues of unknown etiology.^{3,7)} Such lesions have been classified according to the part of the orbit involved¹³⁾ or the histology.^{3,7)} However, the histological classifications are confusing and show no correlation between the clinical and pathological features. Blodi and Gass³⁾ assumed that orbital pseudotumor is a single entity with the histological appearance varying at different stages. Non-neoplastic orbital swellings are also associated with systemic diseases, *e.g.* Wegener's granulomatosis, sarcoidosis, and endocrine exophthalmos.⁷⁾ In our case, however, there was no clinical or pathological evidence of any systemic disease, except for the increased antinuclear antibody level.

CT and MR imaging can demonstrate lesions located in the orbit and the cavernous sinus directly.^{1,6,9,13,14)} CT demonstrates orbital pseudotumor as a soft tissue mass in the orbit, with enlargement of the extraocular muscles or uveal and scleral thickening.¹³⁾ Atlas *et al.*¹⁾ reported that the MR appearance of an orbital pseudotumor is hypointense to fat and isointense with muscle on T₁-weighted images, and isointense or slightly hyperintense compared to fat on T₂-weighted images. This contrasts with other disease entities, including malignancy and hematoma, which are markedly hyperintense compared to fat on T₂-weighted images. The lesion in our patient was hypointense compared to fat on T₂-weighted images. Irisawa *et al.*⁹⁾ found similar MR features and histological appearance of prominent fibrosis. The hypointensity on the T₂-weighted images presumably reflected the relatively low-resonating proton density of fibrous tissue.

Orbital pseudotumors are usually located within the muscular cone. However, Kaye *et al.*¹⁰⁾ described a pseudotumor with extension into the paranasal sinuses and the anterior cranial fossa. These lesions may also extend to the cavernous sinus. Orbital pseudotumor closely resembles the Tolosa-Hunt syndrome.^{4,12,14)} Patients with the Tolosa-Hunt syndrome demonstrate extension from the cavernous sinus into the orbital apex on high-resolution CT scans and MR images,^{8,11,14)} or at autopsy.⁴⁾ This suggests an overlap between orbital pseudotumor and the Tolosa-Hunt syndrome.

Orbital pseudotumor and inflammation around the cavernous sinus are apparently part of a similar disease process, although histological data is limited.

In our case, preoperative radiographs and the operative findings revealed the mass located in the orbit and the optic canal, superior orbital fissure, cavernous sinus, and around the right carotid siphon. Multiple orbital and intracranial biopsy specimens all showed the features of healed inflammation. Possibly, these inflammatory lesions spread contiguously through the venous system around the cavernous sinus, with subsequent involvement of the internal carotid artery. This case suggests that orbital pseudotumor may not be a separate clinical entity, but a part of the clinical spectrum of disease caused by inflammation of the venous system around the cavernous sinus, presenting with different signs and symptoms according to the site.

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