Intracranial extension of inflammatory pseudotumor of the orbit

Case report

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Orbital pseudotumors are inflammatory lesions of the orbit of unknown etiology, which initially mimic an orbital neoplasm. Involvement of the other orbit is uncommon, and extension into the paranasal sinus rare. Intracranial extension of pseudotumor of the orbit is previously unreported. This is a case report of a patient with pseudotumor of the orbit with intracranial extension. The literature of the subject is reviewed.

Key Words • orbital pseudotumor • exophthalmos • inflammation • orbit

Orbital pseudotumors are inflammatory lesions of unknown origin within the orbit, which initially simulate a true neoplasm. Although Birch-Hirschfeld,2 who introduced the term, included all non-neoplastic disorders, ophthalmologists generally limit the term to those lesions that are of an idiopathic inflammatory nature.3,4,5,6 The lesion usually presents as an orbital tumor. Involvement of the lacrimal gland and muscle cone is common, but extension into the periosteum is rare.7 Although involvement of the paranasal sinus has been infrequently noted,1,11,12 intracranial extension has not been reported to our knowledge. We present a case of pseudotumor of the orbit with intracranial extension.

Case Report

This 71-year-old man first presented in 1979 with swelling around his right eye associated with increased lacrimation. Besides mild hypertension, he had no other medical problems. His left eye had been amblyopic from a young age, with vision restricted to counting fingers at a distance of 2 feet.

First Admission. Visual acuity in the right eye, corrected, was 20/30 with full visual fields and normal funduscopy. There was a nontender mass on the superomedial aspect of the right orbit with slight proptosis of the right eye. Pupillary reactions and eye movements were normal.

Plain x-ray films of the orbit and paranasal sinuses showed slight blurring of the bone margins on the medial aspect of the orbit, and computerized tomography (CT) showed a right orbital mass. A chest x-ray film was normal. The orbit was explored through an ethmoidectomy and the mass biopsied. The histological report was inflammatory pseudotumor, and this was confirmed at three institutions including the Cleveland Clinic Foundation and the Armed Forces Institute.

Second Admission. One year later, following an increase in the size of the mass, the patient was referred to the Cleveland Clinic Foundation. At that time there was restriction of movements of the right eye in all directions, especially laterally. The visual acuity had not changed. A CT scan showed increased size of the tumor and extension into the paranasal sinuses and the left orbit (Fig. 1).

Fig. 1. Enhanced computerized tomography scan through the orbits showing the lesion extending from the right orbit into the paranasal sinuses and left orbit.
FIG. 2. **Upper:** Enhanced computerized tomography (CT) scans demonstrating enlargement of the lesion in the orbits and paranasal sinuses, and extending into the anterior cranial fossa with moderate hydrocephalus. **Lower:** Semi-croronal CT scan showing that the floor of the anterior fossa is intact.

The mass was again biopsied, confirming the diagnosis of orbital pseudotumor. However, due to the extension of the mass into the paranasal sinuses, it was thought that the tumor might be a variant of Wegener's granulomatosis or a lethal midline granuloma, despite the histology and absence of any clinical findings suggestive of these entities. Treatment with steroids (prednisolone, 100 mg/day in divided doses) combined with cytotoxic agents was commenced, starting initially with nitrogen mustard (to a total dose of 24 mg) and followed by a course of methotrexate (12.5 mg/week for 10 weeks) with only slight temporary diminution of the size of the mass clinically and on CT scan.

**Third Admission.** The patient presented again in 1982. The mass had continued to enlarge, but the visual acuity in the right eye remained at 20/30. A course of deep x-ray therapy was initiated: 4000 rads in 25 incremental doses was given over 4 weeks. Following this there was a temporary slight decrease in the size of the tumor.

In February, 1983, the patient was referred to the Neurosurgery Department at the Cleveland Clinic Foundation with a 6-month history of progressive dementia, difficulty in walking, failure of vision, and urinary incontinence. General examination was normal. There was proptosis of both eyes, the right more than the left, and restricted gaze in all directions on the right side; both pupils reacted sluggishly to light. In the right eye the visual acuity was 20/400, and the visual field was concentrically diminished. The right corneal reflex was depressed and sensation decreased in the first and second divisions of the trigeminal nerve on the right side. The fundus was normal. The patient was moderately demented and had an ataxic gait, but no focal neurological signs.

The CT scan now showed considerable further growth of the lesion into the paranasal sinuses and the left orbit. There was also a large intracranial mass and the patient had moderate hydrocephalus (Fig. 2). The chest x-ray film was normal, as was a full laboratory investigation including blood examination and urinalysis.

**Operation.** In March, 1983, bifrontal craniotomy was performed. A tumor was found arising from the floor of the anterior cranial fossa on both sides and extending up under both frontal lobes. The tumor extended from the anterior margin of the cribiform plate posteriorly over both optic nerves and optic chiasms. A total intracranial excision was performed. The tumor was firm, fibrous, and only slightly vascular. Although there were small pits in the bone of the floor of the anterior cranial fossa, the bone was grossly intact with no frank defect extending into either the orbit or the paranasal sinuses.

**Postoperative Course.** The postoperative course was uneventful save for transient slight increase in confusion. The patient was discharged home, and 1 month postoperatively the visual acuity on the right side had improved to 20/60.

**Pathological Examination.** At operation the tumor was hard, fibrous, and relatively avascular. On histological examination it consisted of small lymphocytes, plasma cells, and some macrophages embedded in a fibrous stroma (Fig. 3). The lymphocytes had normal morphology, and Russell bodies were also present. Immunoperoxidase stains, using both frozen and paraffin
Intracranial extension of orbital pseudotumor

sections, showed that the plasma cells had physiological proportions of kappa and lambda chains. There was no evidence of necrotizing granuloma or vasculitis.

The diagnosis was of an inflammatory pseudotumor. The specimen was similar to that seen at the previous biopsies, with no histological evidence of either Wegener's granulomatosis or a lethal midline granuloma. Extensive bacteriological and fungal cultures were negative.

Discussion

The original description of orbital pseudotumor by Birch-Hirschfeld in 1905 included any non-neoplastic inflammatory lesions. Ophthalmologists now generally limit the term to those lesions mimicking an orbital tumor which are of an idiopathic nature. Orbital pseudotumors have been classified and grouped by various authors, either according to their supposed etiology, position within the orbit, or type of infiltrate. A distinction is made between the orbital non-neoplastic tumors, in which there is a definite diagnosis, and orbital pseudotumor. Localized non-neoplastic orbital swellings can result from either infections (fungal, parasitic, or bacterial), foreign-body granulomas, amyloid, or monostotic fibrous dysplasia. Alternatively, the orbital swelling may be the result of a known systemic disorder such as Wegener's granulomatosis, polyarteritis nodosa, sarcoid, histiocytosis, or an endocrine exophthalmos.

As the list of defined diseases giving rise to orbital swelling increases, the number of idiopathic inflammatory lesions decreases. Difficulty may arise in differentiating idiopathic orbital pseudotumor from ocular manifestations of generalized diseases, particularly lymphoproliferative disease and Wegener's granulomatosis. In the latter condition, although there is a classical triad of clinical findings, the concept of a limited form of Wegener's granulomatosis has been described and should be especially considered in patients with bilateral lesions. However, those patients have characteristic pulmonary findings and the pathology shows necrotizing granuloma and vasculitis, all of which were absent in this patient.

A lethal midline granuloma is a slowly progressive destructive ulceration of the tissues of the nose, sinus, or pharynx. There is considerable debate as to whether lethal midline granuloma should be classified as a separate clinical and pathological entity, or whether it is part of a spectrum of diseases consisting of Wegener's granulomatosis and limited Wegener's granulomatosis. This case, however, had neither the clinical nor pathological findings of a lethal midline granuloma.

Orbital pseudotumor can be classified according to the portion of the orbit involved. Rootman and Nugent described five variations of orbital involvement, each type having a particular clinical presentation and CT findings. Histological classifications differ, but most are based on the cellular components of the tumor. However, the interpretation and meaning of the cellular patterns vary so much that histopathological classification has been confusing and has failed to show any correlation between clinical and pathological features. Coop classified the pseudotumor of unknown etiology as an orbital lipogranuloma because the granulomatous process starts as fat necrosis. Reese described five histological categories. Blodi and Gass grouped their cases into nine histological varieties and, as they noted no difference in the histological groups as regards symptoms, signs, bilateral incidence, or prognosis, assumed that the condition was a single entity and that the histological picture varied with the stage of the disease at which the biopsy was performed.

Henderson proposed a classification that included two types of inflammatory pseudotumors, either of which could be acute, subacute, or chronic, depending on the amount of fibrosis present. Type I was less common and the basic pathological processes were
characterized by vasculitis and a cellular response of a diffuse polymorphic infiltration, with eosinophils being the most prominent cellular component in the acute phase and lymphocytes and macrophages playing a lesser role. In Type II, the necrotizing vasculitis is lacking and the cellular response in the acute phase is generalized lymphocytic infiltration, which may initially have a perivascular distribution. Orbital tumor is replaced by fibrosis in the chronic stage. The lesion in our case was histologically a Type II inflammatory pseudotumor according to this classification.

Steroids, surgery, and radiotherapy have been used to treat orbital pseudotumor. Although most patients have a good response to steroid therapy,19,24,28 some do relapse7,20,27-29 as in this case, and steroid unresponsiveness has been noted in the sclerosing pseudotumors.1 Excellent results have been reported with the use of radiotherapy,10,18,23,24 although improvement has not been universal20,29 and this patient had only transient response to radiotherapy. Coop8 noted that the best results occurred when there was a cellular predominance rather than fibrotic tissue. Although only three of his nine patients treated with radiotherapy improved, the series consisted of a heterogeneous group of cases and the details of those treated are not mentioned. Severe visual deterioration may occur1 and has been reported in approximately 5% of cases.3,4 Occasionally, orbital decompression is necessary,25 although surgical manipulation in the posterior orbit may have a poor prognosis.20

Orbital pseudotumors are nearly always restricted to the orbit. Although the findings relating to the eye are usually unilateral, bilateral involvement is now revealed more often on CT scanning12 and is associated with no particular histological characteristic.3,4,7 Donaldson, et al.10 reported a series of five cases, three of which had bilateral involvement. Paranasal sinus involvement with pseudotumor has been reported infrequently.11,13 Jellinek21 found involvement of the first and second division of the trigeminal nerve in half of his patients. Pseudotumor may also invade the cavernous sinus and, when this occurs, the process might be considered part of the Tolosa-Hunt syndrome.11

In a case report by Eshagian and Anderson13 involving paranasal sinus extension by pseudotumor, the bone walls of the sinus were intact but pitted, a finding which closely resembles the appearance of the bone of the floor of the anterior fossa in our case. Presumably, the tumor penetrates the bone through microscopic channels.

The present case is most unusual. The original presentation was of a pseudotumor of the orbit (that is, it was a swelling in the orbit mimicking a tumor that was found on histological examination to be an inflammatory pseudotumor). There was only a transient response to steroid and deep x-ray therapy, and the pseudotumor continued to grow as evidenced both clinically and on the CT scans. The original treatment with cytotoxic agents was based on the knowledge that the invasive nature of the mass noted on CT scanning made a diagnosis of orbital pseudotumor unlikely, so that the tumor might represent an unusual form of Wegener's granulomatosis or lethal midline granuloma. However, the histology confirmed that it was a pseudotumor, and there were none of the stigmata of Wegener's granulomatosis or lethal midline granuloma. As the original presentation was of a mass with the histology of an inflammatory pseudotumor involving the orbit, it is warranted to refer to this lesion as an "orbital pseudotumor." However, the subsequent course with growth into the paranasal sinuses and the other orbit is unusual, and the intracranial extension is unique.

References

Intracranial extension of orbital pseudotumor


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