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Orbital Pseudotumor:
Association of Orbital Vein Deformities and Myositis

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Twenty adult patients with orbital pseudotumor were studied with orbital venography and magnification arteriography. Computed tomography (CT) scans were obtained in 14 patients. Occlusive and constrictive changes of the proximal segments of the superior ophthalmic veins were the most frequent vascular abnormalities. The affected ophthalmic arteries were often attenuated but not occluded. Myositis and surrounding connective tissue inflammatory changes are thought to result in sufficient mass effect to cause the venous stasis with narrowing and subsequent occlusion. Such venographic changes are not distinctive for orbital pseudotumor but may also be seen in cases of thyrotoxicosis, Tolosa-Hunt syndrome, and orbital tumors.

Orbital pseudotumor is an inflammatory process of unknown etiology, found in adults as a common cause of unilateral proptosis. Wilner et al. [1] recently produced an experimental model in rabbits comparable to inflammatory pseudotumor of the orbit in humans. They indicated that myositis is an important consideration in the pathogenesis of the experimental pseudotumor. The anatomic proximity of various ocular muscles to important venous structures is of significance when myositis is a prominent feature and may cause narrowing or occlusion of the veins. We report the association of the myositis of orbital pseudotumor with abnormalities of the ophthalmic veins.

Materials and Methods

Twenty patients believed to have orbital pseudotumor were investigated at our institution during a 5 year period. Most of the patients had the classical clinical triad of proptosis, tissue swelling, and impaired ocular motility. All the patients had unilateral symptoms and in no case was bilateral disease detected.

Fourteen patients were scanned with either the EMI Mark I scanner or the CT 1010 unit. Criteria for computed tomographic (CT) diagnosis included proptosis, uveoscleral thickening, and a nonspecific orbital density, sometimes enhancing with iodine and involving mostly the orbital apex, but also involving the globe and peripheral intraorbital muscles.

All 20 patients underwent magnification orbital angiography and venography as an initial part of their workup. Venography was performed in the classical manner by percutaneous puncture of a midline forehead frontal vein with manual compression of both angular veins [2]. Biplane filming was used, although the frontal projection was more useful than the corresponding lateral view. Potential venographic changes included diffuse deformity of the involved superior ophthalmic veins, as well as narrowing of the veins and venous occlusions [3].

Magnification internal and external carotid angiography in the lateral plane was obtained with the Siemens 0.2 mm focal spot tube without the use of a grid. Over 3 times

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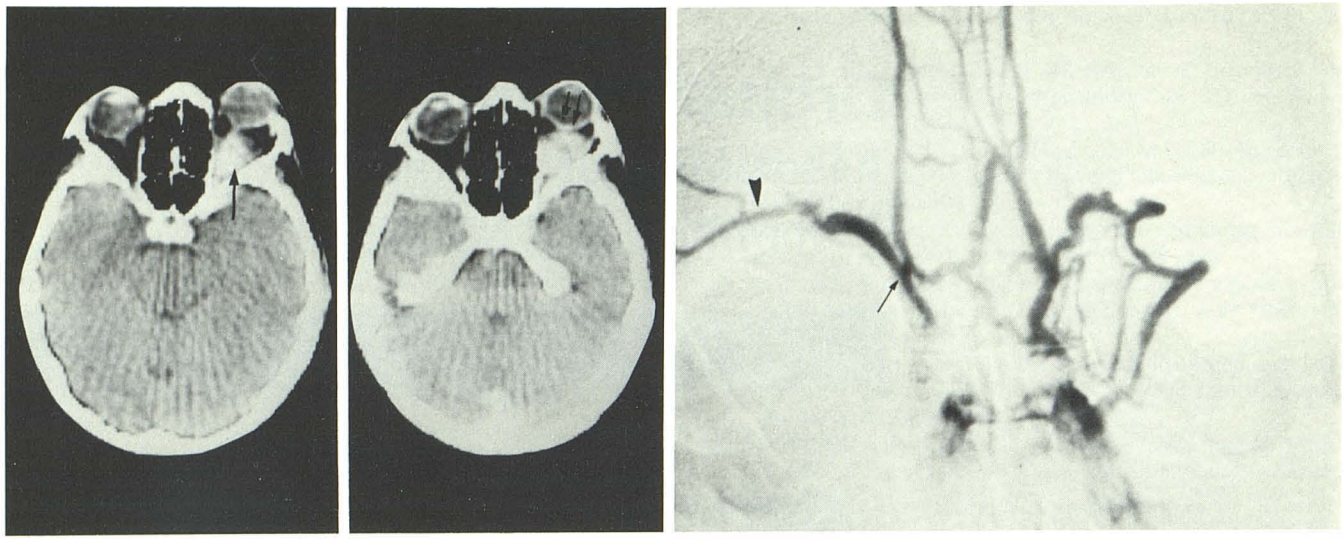


Fig. 1.—Without (**A**) and with (**B**) iodine enhancement. Radiodense right intraorbital mass (*black arrow*), primarily involving apex, is slightly enhanced. Proptosis is present; uveoscleral thickening mostly appreciated with intravenous contrast infusion (*white arrows*). **C**, Frontal venogram. Right superior ophthalmic vein (*arrow*) does not fill despite normal appearance of comparable left vein. Collateral circulation (*arrowhead*) through right supraorbital vein. Magnification arteriography was normal. Marked right ocular improvement was noted with conservative management.

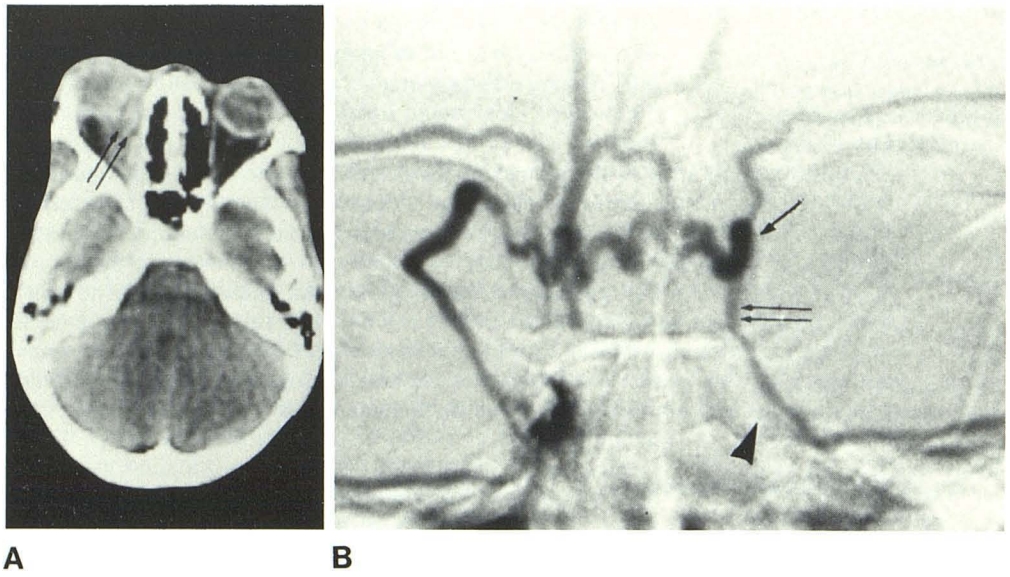


Fig. 2.—**A**, After iodine enhancement. Entire left orbital apex is dense (*arrows*). Abnormal left medial rectus muscle; left lateral rectus muscle apparently normal in peripheral part. Marked left proptosis and uveoscleral thickening. Normal right orbit. **B**, Frontal venogram. Complete occlusion of left superior ophthalmic vein (*single arrow*). Collateral flow through left angular vein (*double arrows*). Diminished visualization of left cavernous sinus (*arrowhead*). Magnification orbital arteriography showed stretching of left ophthalmic arterial branches with no neovascularity. No surgery was performed; patient improved with steroid therapy.

magnification was achieved in all of our patients. The caliber of the involved ophthalmic arteries, and the presence of proptosis, were assessed. Only nine of the patients had a tissue diagnosis consistent with pseudotumor, whereas the diagnoses in the other patients were established by clinical improvement after antiinflammatory therapy.

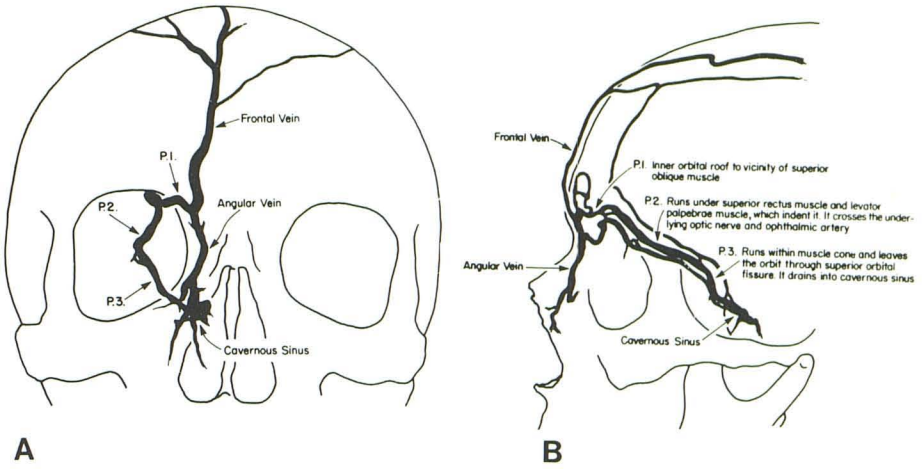
Results

The patients were 27–76 years old (mean, 51.9). All 20 patients had unilateral clinical and radiographic changes. Proptosis was present in 17 patients; diplopia, pain, and soft-tissue swelling were less frequent. No bilateral disease occurred either initially or after follow-up evaluation.

CT was not performed initially since no scanner was available. In the 14 patients who had CT, orbital densities with minimal enhancements, involving primarily the affected orbital apices, were the most common abnormalities (figs. 1A, 1B, and 2A). The muscle origins at the orbital apices were probably involved in all patients. Peripherally, individual muscles (medial and lateral rectus muscles) were involved in 10 patients. Proptosis and uveoscleral thickening were seen in 11 patients (figs. 1A, 1B, and 2A).

Venographic changes were often diffuse and were found

Fig. 3.—Anatomy of orbital veins and their relation to anatomy of adjacent muscles.



to varying degrees in all 20 patients. Narrowing or occlusion of the distal segment of the superior ophthalmic vein (P1 segment) was present in 14 patients (figs. 1C, 2B, 3, 4, and 5C). Similar changes in the P2 and P3 segments were also seen, but were less frequent. The superior ophthalmic veins were completely occluded in five patients. No case of clinical or venographic occlusion of the cavernous sinus was seen.

No arteriographic occlusions were noted. Stretching of the ophthalmic artery was seen in 11 patients (figs. 5A and 5B) and narrowing of the artery in four patients. Proptosis of the choroidal blush was seen in nine patients.

All nine biopsied cases had inflammatory changes in the connective tissue and muscle. Plasma cells, increased lymphocytes, and myositis were microscopically present. Therapy with steroids reduced inflammation, but often some visual impairment persisted. Recurrences of the proptosis and diplopia were not infrequent especially if the patient discontinued the steroids prematurely.

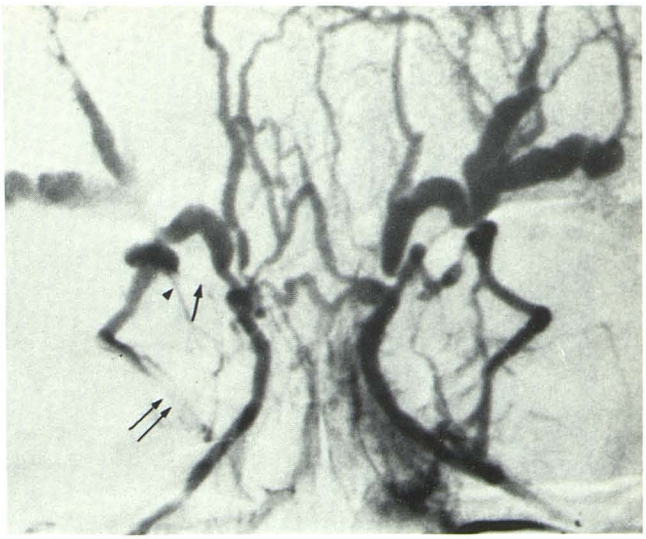


Fig. 4.—Frontal venogram. Excellent filling of normal left superior ophthalmic vein. Narrow right P1 segment (*single arrow*). Slightly dilated right P2 segment compared with corresponding left P2 segment. Collateral venous return to P2 from right median connecting vein (*arrowhead*). Right P3 segment also narrowed (*double arrows*); poor filling of right cavernous sinus. CT was abnormal with nonspecific right orbital density and associated proptosis. Surgical biopsy showed nonspecific inflammatory tissue within right orbit and pronounced diffuse myositis.

Discussion

In a 1909 review, Birch-Hirshfield et al. [4] were the first to use the term pseudotumor of the orbit. By 1923, Benedict and Knight [5] had found 25 reported cases. Ingalls [6] indicated that pseudotumor is a most important cause of young adult intraorbital mass. Blodi and Gass [7] showed an unusually high incidence in the seventh and eighth decades of life.

Pathologically, severe inflammation similar to the Arthus type of vasculitis with vessel wall necrosis and resultant fibrinoid changes can be found. Fibrous connective tissue is noted as the disease progresses. Chronic changes include diffuse lymphocytic and plasma cell infiltration with occasional macrophages and eosinophils [1, 6, 7]. Myositis and connective tissue involvement are found [8].

Experimental production of the Arthus type reaction in the rabbit was a definite aid to understanding the relation of chronic inflammation to the resultant clinical findings [1]. Proptosis, soft-tissue swelling, impaired ocular motility, diplopia, decreased vision, and papillitis were noted.

Enzmann et al. [9] described the CT changes of orbital pseudotumor. They found diffuse soft-tissue densities that varied considerably in size, shape, and location throughout the orbit. Optic nerve involvement and uveoscleral thickening were present. Though bilateral pseudotumor was found in six of the nine cases of Enzmann et al. [9], this is distinctly unusual [6, 7]. In our 14 cases that had CT scans of the orbit, the appearances were considered compatible with the diagnosis of orbital pseudotumor.

Orbital venography was performed in all 20 patients. Occlusive and constrictive changes, particularly of the distal segments of the involved superior ophthalmic veins, were the most common vascular abnormalities. Similar changes were described by Castan et al. [10], who studied 18 cases of orbital pseudotumor by phlebography and found scat-

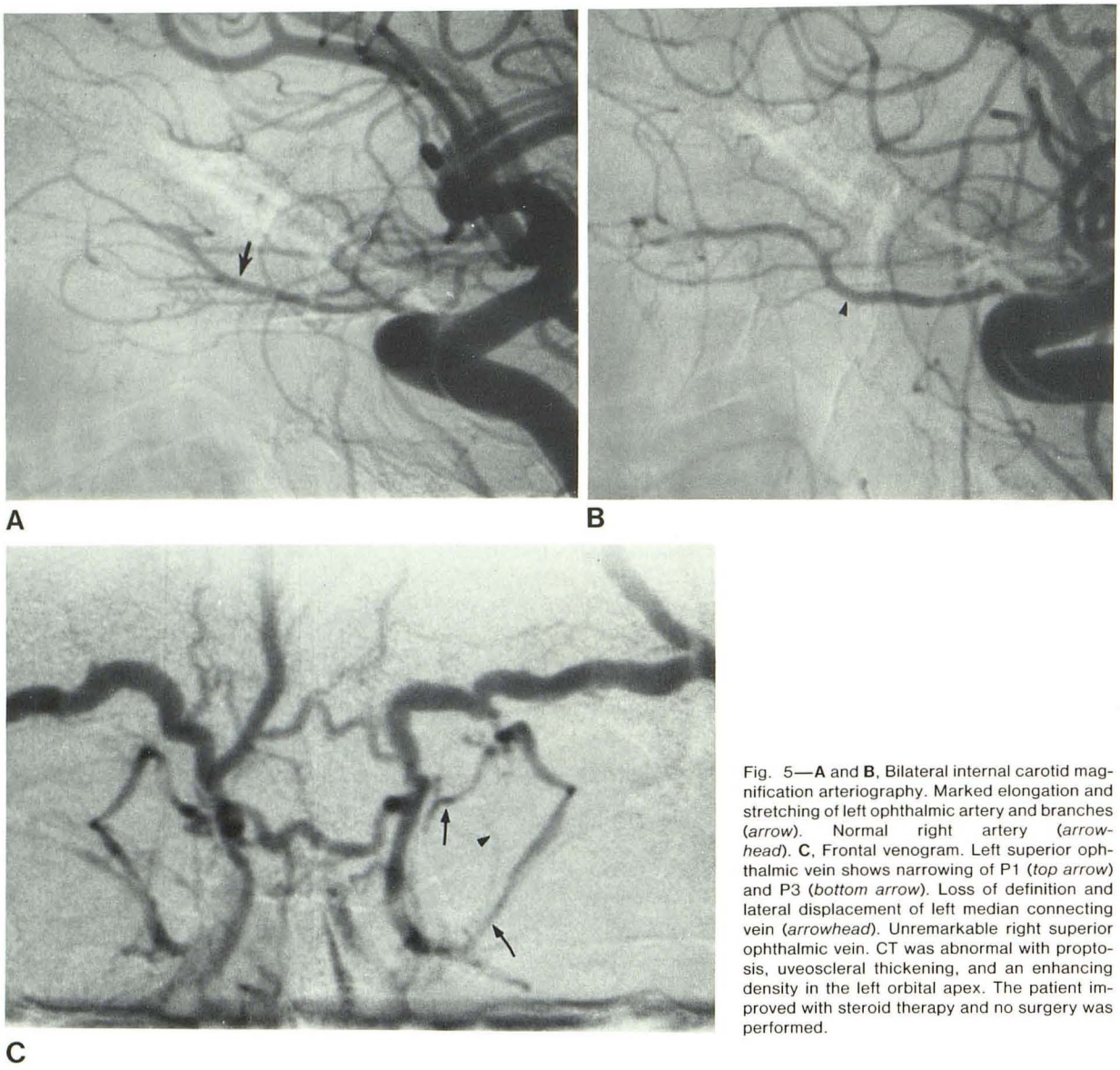


Fig. 5—**A** and **B**, Bilateral internal carotid magnification arteriography. Marked elongation and stretching of left ophthalmic artery and branches (*arrow*). Normal right artery (*arrowhead*). **C**, Frontal venogram. Left superior ophthalmic vein shows narrowing of P1 (*top arrow*) and P3 (*bottom arrow*). Loss of definition and lateral displacement of left median connecting vein (*arrowhead*). Unremarkable right superior ophthalmic vein. CT was abnormal with proptosis, uveoscleral thickening, and an enhancing density in the left orbital apex. The patient improved with steroid therapy and no surgery was performed.

tered venous occlusive changes and intraorbital edema to be the most common abnormal findings.

The superior ophthalmic vein lies close to muscle tissue [11] (fig. 3). Although we were unable to directly correlate the site and type of venographic changes (occlusion or attenuation of the superior ophthalmic veins) with the CT appearances, it is conceivable that the myositis and surrounding connective tissue inflammation can encroach on the superior ophthalmic veins, resulting in various degrees of stasis with narrowing and subsequent occlusion. The discrepancy between the marked venous narrowing and occlusion and the minimal arteriographic narrowing is notable. A possible cause could be the intraluminal pressure differences between the ophthalmic artery and the ophthalmic vein. The vein could be more susceptible to an increase in intraocular pressure with subsequent stasis and occlusion.

Orbital venographic abnormalities have also been found in other diseases. Russell and Miller [12] described orbital venographic changes in eight cases of thyromyopathy and in one case of pseudotumor. They found two main venous abnormalities: lack of visualization of the affected superior ophthalmic veins and attenuation of venous caliber. They stated that the bilaterality of these abnormalities made the diagnosis of endocrine exophthalmos more likely. There is a close histologic resemblance between endocrine exophthalmos and myositic orbital pseudotumor [5–7]. Hyperthyroidism, hypothyroidism, thyroiditis, and thyroid myopathy all may have a clinical and pathologic picture similar to that of orbital pseudotumor.

Ophthalmic vein abnormalities may also be seen in the Tolosa-Hunt syndrome. This was shown by Muhletaler and Gerlock [13], who described occlusion of the third segment of the superior ophthalmic vein.

With the development of high resolution CT scanners and routine coronal orbital scanning, correlation of the involvement of individual intraorbital muscles with specific venous changes may be possible. Conclusive evidence that the

inflammatory changes of orbital pseudotumor result in orbital venous occlusions will come only after careful experimental and/or postmortem studies. In the meantime, a growing body of circumstantial evidence links the myositis with adjacent orbital vein abnormalities.

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