Acute Orbital Pseudotumors: Classification and CT Features

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Acute Orbital Pseudotumors:
Classification and CT Features

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Acute orbital pseudotumors are inflammatory lesions of unknown etiology that may affect part or, less often, all of the tissue within the orbit. A retrospective computed tomographic (CT) study of 16 patients demonstrated that these lesions occur in one of five specific anatomic patterns: anterior, posterior, diffuse, lacrimal, or myositic. The most common location was lacrimal followed by anterior pseudotumors. Posterior, diffuse, and myositic pseudotumors were equally frequent. Localization on the basis of clinical features correlated with the CT localization. Illustrative cases of each of the five types are included. The role of CT in evaluating the therapeutic response is discussed.

Orbital pseudotumors are reactive lesions of unknown etiology that clinically mimic neoplasms or inflammatory diseases of the orbit, particularly thyroid ophthalmopathy. Microscopically these lesions demonstrate a wide spectrum of inflammatory and pseudoneoplastic features. Lymphocytic infiltration is a frequent histologic feature; consequently pseudotumors may mimic lymphomas [1]. However, pseudotumors show histologic features of a reactive rather than a neoplastic process; thus, the diagnosis can be confirmed from the histology. A correlation between histologic features and response to treatment as well as prognosis has been described [2].

Clinically, acute orbital pseudotumors typically have a rapid onset and frequently produce proptosis, lid swelling, chemosis, pain, and limitation of ocular movement [3, 4]. Papilledema and optic neuropathy may also occur. The specific clinical features reflect the location of the pseudotumor [5]. We tried to determine the location and character of acute orbital pseudotumors on computed tomographic (CT) scans and to assess the correlation of specific clinical features with the CT scan location.

Materials and Methods

Patients were classified as having acute pseudotumors by virtue of rapid clinical onset and development (days to weeks) of any combination of chemosis, injection, lid edema, pain, tenderness, papilledema, and ureitis associated with inflammatory features in the orbit or in the globe. In addition, the patients did not have either clinical or laboratory evidence of infective cellulitis, thyroid eye disease, or systemic inflammatory disorders. All the lacrimal pseudotumors were biopsied; the biopsies showed diffuse lymphocytic infiltrates with destruction of the gland.

Sixteen patients were studied. They were 6–79 years old; 10 were male and six were female. Six were under age 20 years. CT was performed either on an EMI 1010 or a GE 8800 scanner. Axial scans were obtained on all patients and eight had coronal views. The scans were examined retrospectively, with particular regard for location of the pseudotumor. Specific involvement of the globe, optic nerve, or extraocular muscles was recorded. Most of the patients had at least one follow-up scan. Contrast enhancement was used in six patients.

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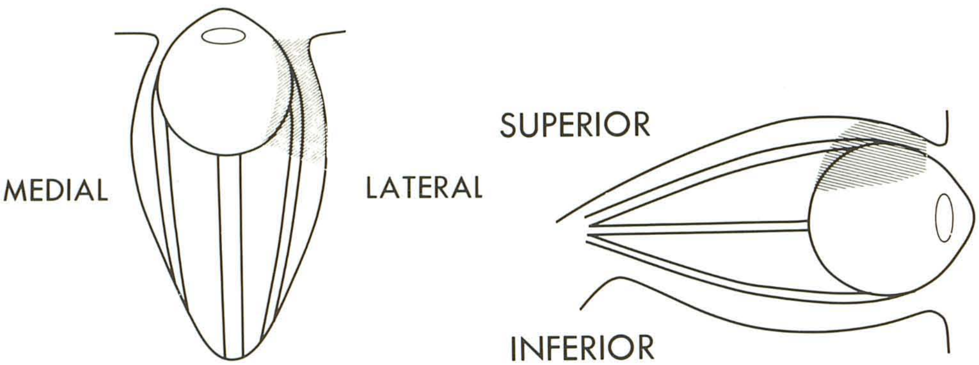

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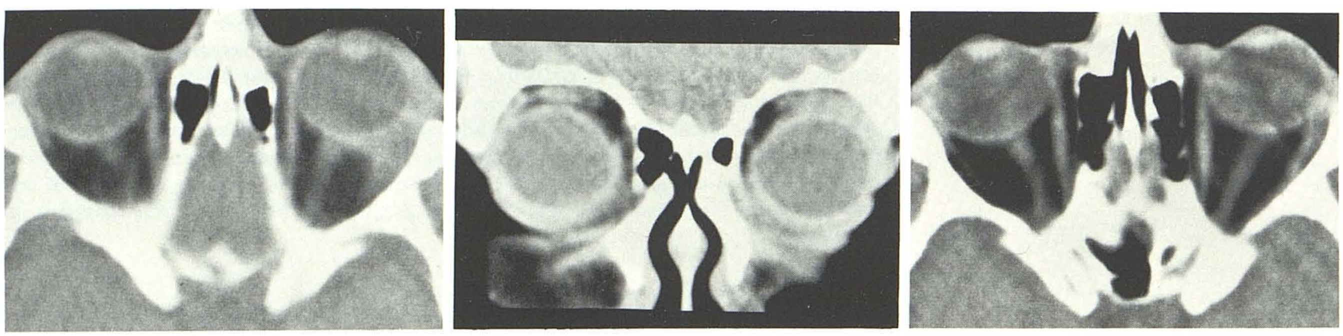
Fig. 1.—Lacrimal pseudotumor, axial (**A**) and sagittal (**B**) views.



A B C

Fig. 2.—Lacrimal pseudotumor. **A**, Axial, unenhanced scan. Soft tissue mass in anterolateral aspect of orbit obscures lateral aspect of globe and produces medial deviation of globe. **B**, Coronal, unenhanced scan. Mass in superolateral aspect of orbit produces medial and inferior displacement of globe. **C**, Steroids produced clinical resolution within 1 month. Only minimal prominence of lacrimal gland after 3 months.

Results

Acute orbital pseudotumors were seen to occur in one of five anatomic patterns on CT scan (table 1). The characteristic clinincal and CT features and the response to therapy are described below. There were no false-negative scans. The pseudotumor was bilateral in only one patient. Enhancement was seen in all of the six patients given intravenous contrast material, but it was not useful in further defining the lesion.

Lacrimal Pseudotumors (fig. 1)

All six patients with acute lacrimal pseudotumors typically developed sudden onset of pain, tenderness, and injection of the lid and conjunctiva in the area of the lacrimal gland in association with a palpable lacrimal gland and an S-shaped deformity of the upper lid with pouting of the lacrimal ducts. In addition, there was minimal proptosis with a downward and inward displacement of the globe. None of these patients had optic neuropathy and three demonstrated restriction of ocular movements in the direction of the swollen gland.

On CT, pseudotumors of the lacrimal gland were confined

TABLE 1: **Clinical Features of Orbital Pseudotumors**

	CT Location				
	Lacrimal	Anterior	Posterior	Diffuse	Myositic
No. Patients	6	4	2	2	2
General features:					
Pain	+	++	+	+++	++
Proptosis	+[*]	++	+	+++	+
Lid swelling	++[†]	++	+	++	–
Conjunctival injection	++[‡]	++	–	++	++[§]
Decreased range of muscle motion	+[‖]	++	–	+++	++[¶]
Palpable mass	+++	–	–	–	–
Optic neuropathy	–	++	++	++	–
Ocular:					
Choroiditis	–	++	–	++	–
Papillitis	–	++	–	+++	–
Retinal detachment	–	+	–	–	–

Note.—– = none; + = infrequent/minimal; ++ = common/moderate; +++ = marked/characteristic.
* Inferomedial.
† Upper outer.
‡ Supertemporal.
§ Over muscle insertion.
‖ Superolateral gaze.
¶ Indirection of involved muscle.

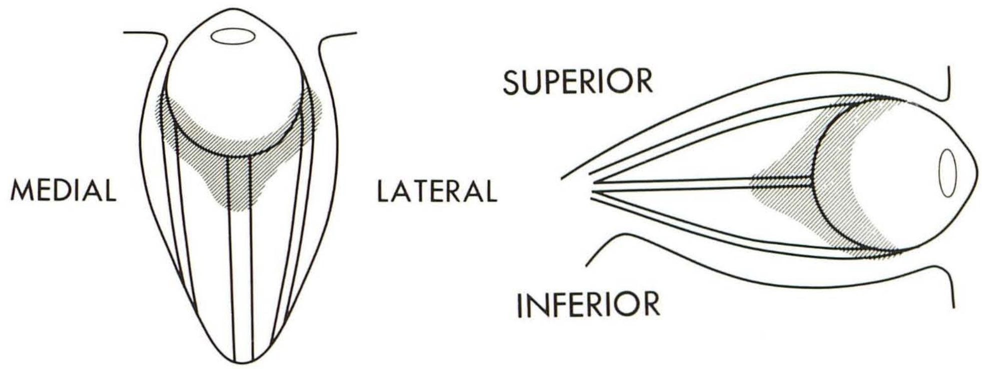


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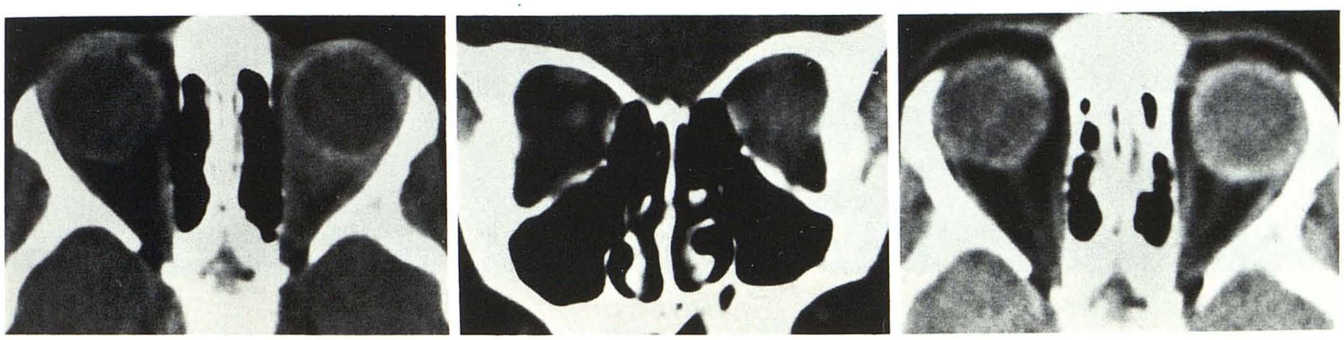
Fig. 3.—Anterior pseudotumor, axial (**A**) and sagittal (**B**) views.



A B C

Fig. 4.—Anterior pseudotumor. **A**, Axial, unenhanced scan. Wedge-shaped area of soft tissue density immediately behind globe, extending along optic nerve. Sclera is thickened and poorly defined. **B**, Coronal, unenhanced scan. Soft tissue mass obscures normal definition of optic nerve and extraocular muscles at plane just posterior to globe. The patient had progressive clinical improvement with steroid therapy. **C**, Unenhanced scan 9 months after initial examination. No residual soft tissue mass. Junction of optic nerve and globe is well defined.

to the superolateral aspect of the orbit, adjacent to and obscuring the lateral aspect of the globe. Characteristically, inferomedial displacement of the globe occurred (figs. 2A and 2B). With steroid therapy the mass size diminshed and the lateral aspect of the globe and anterior part of the lateral rectus muscle were then better visualized (fig. 2C).

Anterior Pseudotumors (fig. 3)

There were four anterior pseudotumors, one of which was bilateral. In addition to pain, diplopia, and lid swelling, these acute pseudotumors affected the globe producing uveitis in three, papillitis in two, optic neuropathy in three, and an exudative inflammatory retinal detachment in one.

On CT, these lesions were intimately related to the posterior aspect of the globe and produced thickening of the posterior sclera and/or choroid, obscuring the junction of the optic nerve with the globe. In addition, they extended a variable distance posteriorly along the optic nerve (figs. 4A and 4B). As the mass resolved with steroid therapy, the junction of the optic nerve and globe could again be visualized on CT (fig. 4C).

Posterior Pseudotumors (fig. 5)

The two acute posterior pseudotumors both produced less proptosis, pain, and inflammation than either the anterior or diffuse types but were associated with early development of clinical optic neuropathy. On CT, they were confined to the apical part of the orbit, characteristically obscuring the anatomy of the extraocular muscles and optic nerve at this site. Anterior extension along the course of a muscle or optic nerve was seen (figs. 6A and 6B). With therapy, the extension of the lesion along the muscles and optic nerve initially resolved with later clearing of the soft tissue at the apex (fig. 6C).

Diffuse Pseudotumors (fig. 7)

Acute diffuse pseudotumors had clinical features very similar to those of anterior pseudotumors but were more severe. Each of two patients had evidence of papillitis, choroiditis, and optic neuropathy. On CT, these pseudotumors involved the entire orbit with a soft tissue mass extending from the apex to the posterior margin of the globe. The optic nerve and extraocular muscles were obscured to a variable extent (figs. 8A and 8B). With resolution, the normal fat density was restored and the extraocular muscles and optic nerve were well defined on CT (fig. 8C).

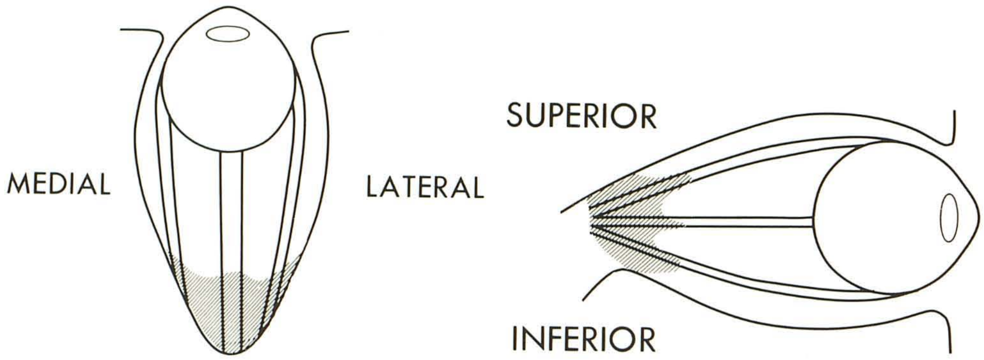


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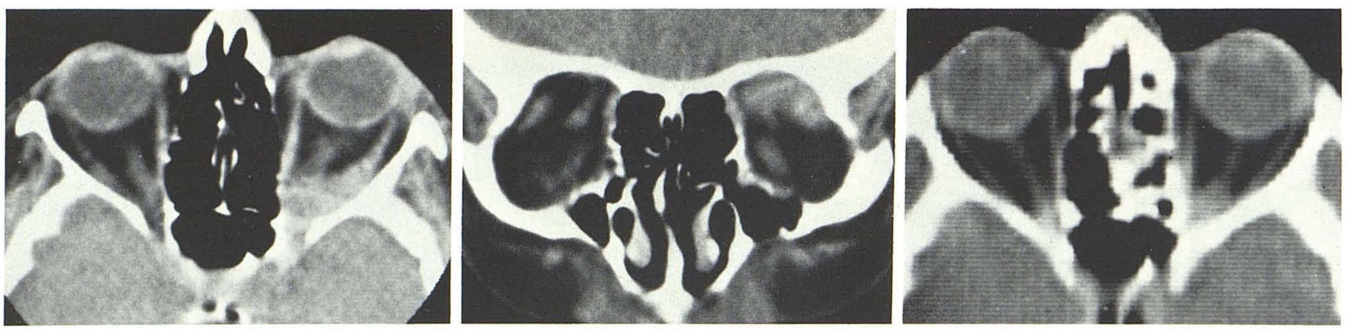
Fig. 5.—Posterior pseudotumor, axial (**A**) and sagittal (**B**) views.



A B C

Fig. 6.—Posterior pseudotumor. **A**, Axial, unenhanced scan. Soft tissue mass fills apex of orbit extending along posterior aspect of lateral rectus muscle. Posterior part of optic nerve is obscured. **B**, Coronal, unenhanced scan. Mass in apex of orbit obscures normal definition of optic nerve with partial obscuring of extraocular muscles. Lateral rectus muscle appears thickened. The patient improved with prednisone but because of several recurrences as therapy was tapered, orbital irradiation was given. **C**, Unenhanced scan 6 weeks later. Almost total resolution of pseudotumor.

Myositic Pseudotumors (fig. 9)

Both of these lesions were characterized by their sudden onset of pain with eye movement, associated with localized injection of the globe and reduced ocular motility. On CT, these pseudotumors involved an extraocular muscle, with relatively diffuse enlargement of the muscle (fig. 10). The acute myositic pseudotumors responded readily to steroid therapy with evidence on CT of a reduction in size of the involved muscle.

Discussion

Recent advances in CT scanning permit a more precise anatomic assessment of orbital lesions [6, 7], allowing greater diagnostic accuracy. In correlating the clinical features of acute orbital pseudotumors with the associated CT abnormalities, we have found that five anatomic groups exist and that the clinical features correspond with the site of involvement. The only correlation between the CT location and the response to therapy was a tendency for lacrimal and myositic pseudotumors to resolve more rapidly with steroids than did the anterior, posterior, or diffuse pseudotumors.

Inflammation of the globe, including uveitis, papillitis, choroidal effusions, and retinal detachments, are features of anterior or diffuse pseudotumors. This is consistent with the intimate relation of the pseudotumors with the posterior aspect of the globe.

Optic neuropathy may occur with anterior or diffuse pseudotumors but is a particularly important early feature of posterior pseudotumors. The optic nerve is not involved in lacrimal or myositic pseudotumors and thus optic neuropathy is not a feature of these lesions.

The localized extent of lacrimal pseudotumors seen on CT accounts for the specific nature of many of the clinical features. The conjunctival injection involves the superotemporal part of the globe and the lid swelling involves particularly the upper outer aspect of the eyelid. In addition, the proptosis tends to be inferomedial as expected with the superolateral location of the lesion. Restriction of movement is characteristically most marked with superolateral gaze. A tender palpable gland confirms lacrimal involvement.

Myositic pseudotumors produce several localized features that depend on the muscle involved. Conjunctival injection occurs at the site of muscle insertion and the major restriction of eye movement occurs in the direction of the involved muscle. Proptosis is variable and is often in a direction away from the involved muscle. Swelling of the eyelid is not a particular feature of these pseudotumors.

The CT features of acute orbital pseudotumors are not pathognomonic but may be highly specific, particularly when the clinical features are considered. Scleral thickening has been reported as CT evidence of an orbital pseudotumor

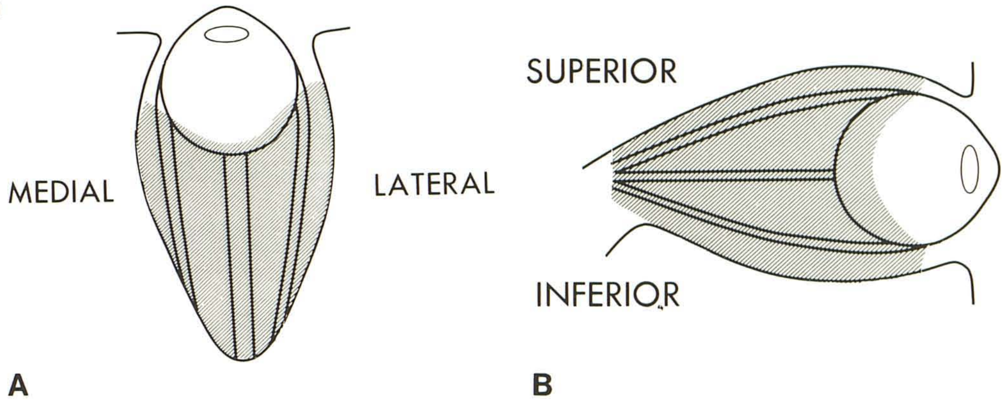


Fig. 7.—Diffuse pseudotumor, axial (**A**) and sagittal (**B**) views.

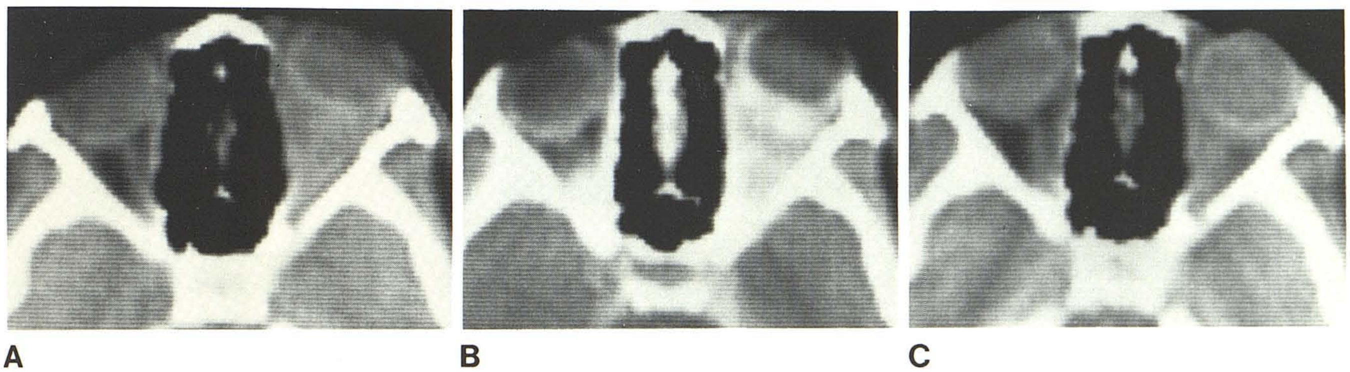


Fig. 8.—Diffuse pseudotumor. **A**, Axial, unenhanced scan. Soft tissue mass fills entire orbit from apex to globe with proptosis of globe. Normal fat density is lost and optic nerve and extraocular muscles are poorly visualized. **B**, Axial, enhanced scan. Obvious enhancement throughout pseudotumor is not entirely uniform. **C**, Unenhanced scan 3 weeks after initiation of prednisone. Partial resolution with some restoration of fat density.

[8]. This is a nonspecific feature of inflammation involving the sclerouveal rim and, as we have shown, does not occur with posterior or myositic pseudotumors. In addition, thickening of the sclera is not an isolated finding but occurs in association with soft tissue involvement of the adjacent optic nerve, muscles, or lacrimal gland. The sclera may be obscured with diffuse pseudotumors and in this situation the sign is not useful. Orbital cellulitis or vasculitis may produce scleral thickening as well.

Orbital pseudotumors require differentiation from other orbital processes. Thyroid ophthalmopathy is usually bilateral [9] but may mimic a myositic pseudotumor on CT if only one extraocular muscle is involved. The muscle enlargement with pseudotumor has been reported as an irregular contour of the muscle, unlike the smoother enlargement demonstrated with thyroid ophthalmopathy [10].

Lacrimal gland tumors not associated with bone changes on CT may be impossible to distinguish from lacrimal pseudotumors [11]. Optic gliomas, meningiomas, or neuromas in the apex of the orbit can mimic a posterior pseudotumor. Unlike these other entities, a posterior pseudotumor is not well delineated, totally obscures the soft tissue definition in the apex of the orbit, and often produces soft tissue extension along an extraocular muscle as well as the optic nerve.

CT is an excellent method for assessment of therapy in these patients [12] since diminution of the soft tissue mass with increased definition of contiguous orbital structures can be readily appreciated. Most acute orbital pseudotumors respond readily to steroids whereas the chronic orbital pseudotumors show a mild or poor response and often require radiation therapy. In the absence of clinical information, acute and chronic orbital pseudotumors in our experience cannot be differentiated by CT unless follow-up scans are obtained.

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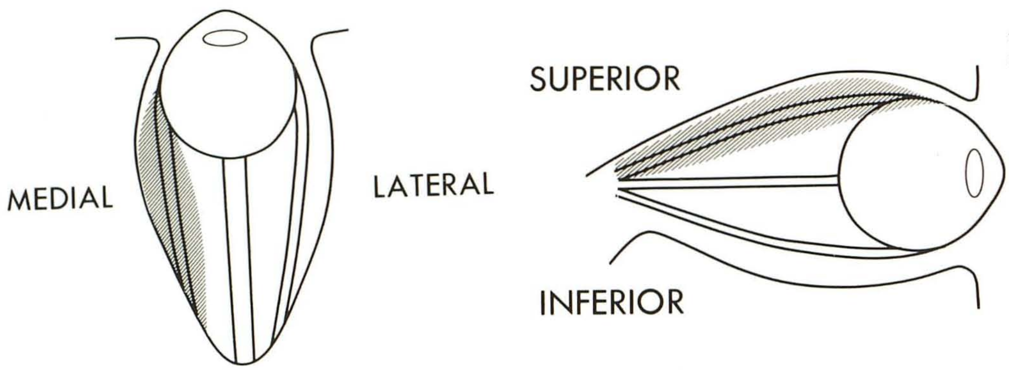

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Fig. 9.—Myositic pseudotumor involving medial (**A**, axial view) and superior (**B**, sagittal view) rectus muscle.

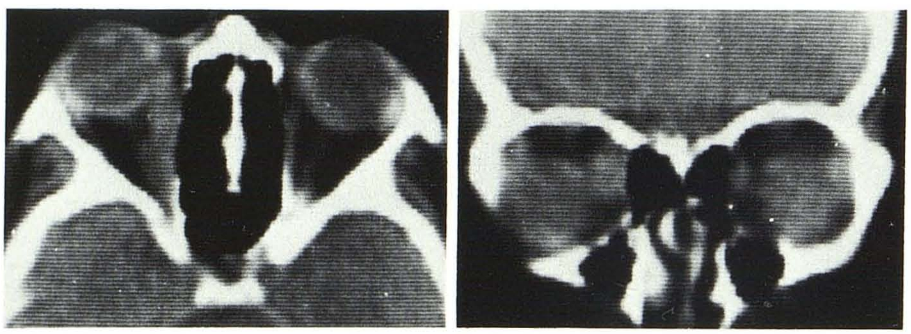

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Fig. 10.—Myositic pseudotumor. **A**, Axial scan. Soft tissue thickening along entire course of medial rectus muscle. **B**, Coronal scan. Marked thickening of medial rectus muscle.

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