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Special Article

High-Resolution CT of Lesions of the Optic Nerve

Robert G. Peyster¹
Eric D. Hoover
Beverly L. Hershey
Marvin E. Haskin

The optic nerves are well demonstrated by high-resolution computed tomography. Involvement of the optic nerve by optic gliomas and optic nerve sheath meningiomas is well known. However, nonneoplastic processes such as increased intracranial pressure, optic neuritis, Grave ophthalmopathy, and orbital pseudotumor may also alter the appearance of the optic nerve/sheath on computed tomography. Certain clinical and computed tomographic features permit distinction of these nonneoplastic tumefactions from tumors.

The advent of high-resolution computed tomographic (CT) scanning made it possible for optic nerve lesions to be well evaluated in both the axial and coronal planes. Optic gliomas and sheath meningiomas are well documented causes of enlargement of the optic nerve/sheath. Several nonneoplastic conditions may also affect the optic nerve/sheath and simulate these tumors on CT. The more common of these are increased intracranial pressure associated with pseudotumor cerebri, optic neuritis, Graves ophthalmopathy, and orbital pseudotumor. Certain distinguishing features allow their differentiation from the neoplasms of the optic nerve.

Materials and Methods

Cases with verified causes for enlargement of the optic nerve/sheath were selected from more than 10,000 patients undergoing cranial CT scanning during a 2 year period. All studies were performed on a GE/8800 scanner. Unless contraindicated, iodinated contrast material (Conray 60, 150 ml drip infusion; Mallinckrodt, St. Louis) was used in all cases. Routine orbital scanning consisted of 5 mm axial and coronal sections. When necessary, 1.5 mm sections were used to optimize visualization of the optic nerves/sheaths.

Normal Anatomy

The anatomic course of the optic nerve is oblique posteriorly, superiorly, and medially from its retinal insertion to its exit from the orbit through the optic canal. Occasionally, the nerve describes a gentle curve, but it is usually quite straight in the axial plane (fig. 1A). Often, due to its course through the orbit, the nerve will appear segmentally on several axial sections, especially with very thin slices [1]. Gaze position may alter the appearance of the optic nerve. An axial scan plane angled 20° below the orbitomeatal line with the eyes in up-gaze position may permit visualizing the optic nerve in a single 5 mm section [2]. The optic nerve is normally homogeneous in size and density throughout its course and is about 4.5 mm wide [1]. Our own measurements of 100 normal optic nerves confirm this figure for axial scans, but indicate a slightly larger (5 mm) diameter in the coronal view (Peyster RG, unpublished data). In coronal sections obtained perpendicular to the orbitomeatal line, the nerve can be seen inserting on the globe medially and slightly above the posterior pole as a round area of high density. Posteriorly, the optic nerve has a more oval appearance due to its oblique course in the posterior orbit (fig. 1B).

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¹ All authors: Department of Radiology, Hahnemann Medical College & Hospital, 230 N. Broad St., Philadelphia, PA 19102. Address reprint requests to R. G. Peyster.

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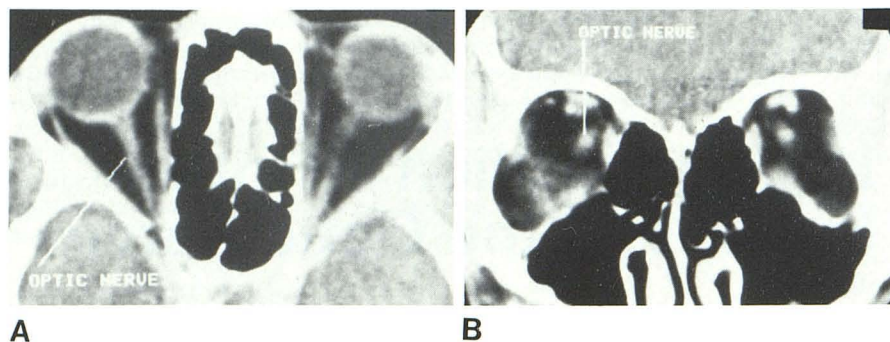


Fig. 1.—Normal optic nerves. A, 5-mm-thick axial section. Both optic nerves/sheaths are visualized. B, Coronal section. Oval appearance of optic nerves/sheaths due to their oblique course through orbit. Note: In both axial and coronal scans throughout this article, with the exception of fig. 1B, the right orbit appears on the reader's left.

TABLE 1: Conditions Associated with Optic Nerve Enlargement

Neoplasms:

Optic nerve glioma
Meningioma
Neuroma
Hemangioblastoma
Metastasis
Leukemia

Nonneoplastic:

Increased intracranial pressure
Optic neuritis
Graves disease
Orbital pseudotumor
Toxoplasmosis
Tuberculosis
Sarcoidosis
Central retinal vein occlusion
Traumatic hematoma of the optic nerve sheath

Lesions of the Optic Nerve/Sheath

Both neoplastic and nonneoplastic conditions may cause abnormalities of the optic nerve/sheath detectable by CT. Most of these conditions are detected because of enlargement of the optic nerve/sheath and are listed in table 1.

Tumors of the Optic Nerve/Sheath

Optic Nerve Glioma

Optic nerve glioma is a rare tumor predominantly occurring in children. Eighty percent occur within the first decade [2]. There is a strong association between optic nerve gliomas and neurofibromatosis, in which bilateral tumors may be found [3, 4]. Clinically, gliomas are difficult to distinguish from other orbital tumors, with the patient's age being one of the most important factors in diagnosis. Visual loss, often insidious, is the first symptom to develop, followed by proptosis, which usually develops rapidly. On examination, optic atrophy and a Marcus-Gunn pupillary reaction with varying visual field cuts are observed [2]. Optic gliomas grow very slowly and do not metastasize [4].

The CT appearance of optic nerve gliomas is variable. Fusiform enlargement of the optic nerve silhouette is most often seen with smaller lesions, but larger masses may be eccentric or multilobulated [2]. The baseline density of smaller lesions is about the same as the normal optic nerve, but larger masses may present higher density values, probably due to elimination of partial volume averaging of orbital

fat [2]. Enhancement after intravenous contrast administration varies from imperceptible to moderate, but is generally less intense than in meningiomas [2, 4, 5]. Calcifications are seen occasionally [4]. Most importantly, the optic nerve cannot be seen separately from the mass (figs. 2A and 2B) [4]. CT is especially valuable in demonstrating intracranial extension of orbital gliomas [4–6]. Enlargement of the optic canal is often demonstrable on CT (fig. 2C), obviating conventional tomography. Metrizamide cisternography may aid in detecting subtle involvement of the intracranial optic nerve, chiasm, and optic tracts (figs. 2D–2F). Our personal experience with intraorbital optic nerve gliomas is limited to the one case illustrated in figure 2, which was typical in all respects.

Optic Nerve Sheath Meningioma

Meningiomas primarily in the optic nerve sheath are even rarer than gliomas [3]. They are seen most often in middle-aged women [7]; in children, they are more common than intracranial meningiomas [3]. Bilateral optic nerve sheath meningiomas are rarely encountered, most often in association with neurofibromatosis [8, 9]. Clinically, the predominant early feature is visual loss. Proptosis occurs later and is usually mild. Physical examination reveals an abnormal optic disk, which may be swollen or atrophic, visual field cuts, often a central scotoma, and restriction of eye movement [2]. These tumors tend to recur after surgery and to extend intracranially.

Our experience with nine cases of optic nerve sheath meningioma is the basis for much of the following description of characteristic CT findings. The figures in parentheses refer to the number of our cases in which the various findings were present.

Segmental (1/9) or diffuse (8/9) thickening of the optic nerve sheath is the usual appearance of these tumors on CT. The enlargement may be fusiform (2/9) (fig. 3), but uniform (6/9) thickening of the sheath is more common (fig. 4). High-resolution scanning often allows visualization of the normal optic nerve running through the tumor, giving rise to the "tram-track" appearance (8/9) of the low-density nerve surrounded by the higher density tumor on axial views (figs. 3 and 5). The corresponding finding in coronal views is a "donut" (7/9) configuration, with a ring of high density around the nerve (fig. 3B). Very thin sections (e.g., 1.5 mm) may be necessary to elicit tram-tracking (fig. 5); this finding distinguishes meningioma from glioma [10]. Calcification

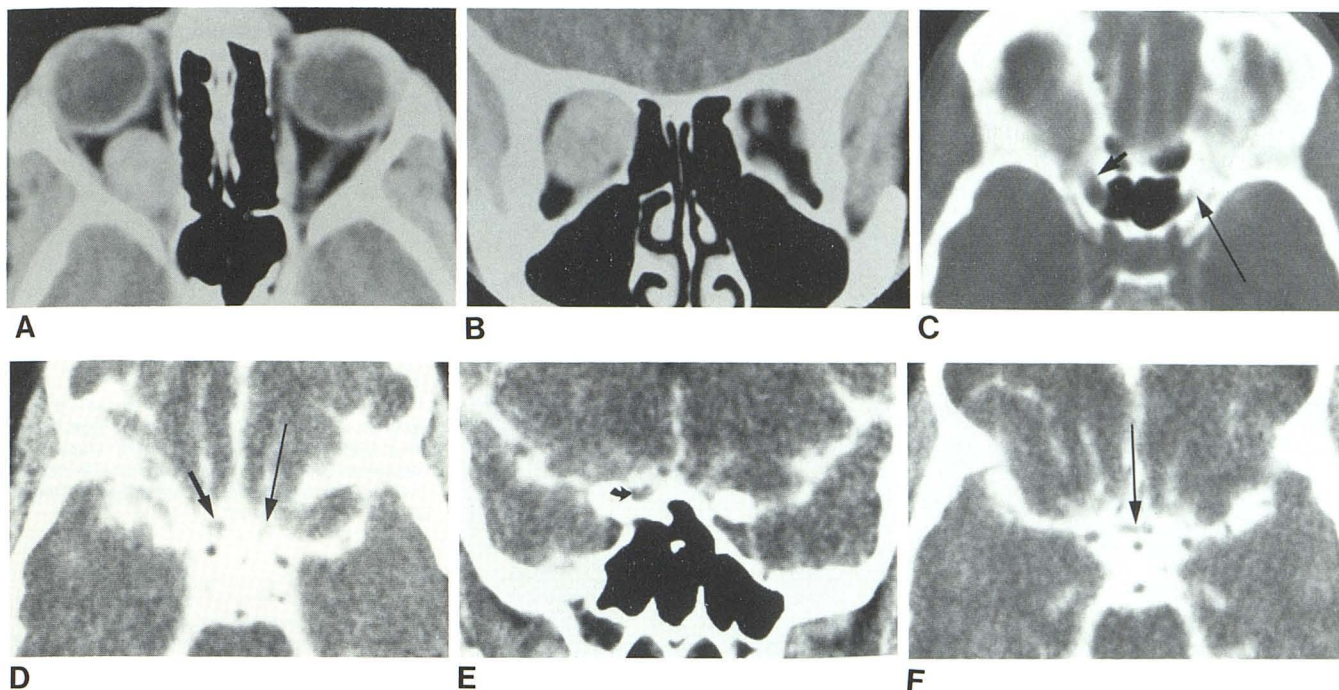


Fig. 2.—Optic glioma. **A**, Axial scan with contrast. Midline fusiform intracanal lesion in expected position of optic nerve. **B**, Coronal scan with contrast. Mass fills most of posterior orbit and is homogeneously dense. **C**, Axial scan. Right optic canal (*short arrow*) is enlarged compared with normal left canal (*long arrow*). **D**, Metrizamide cisternogram, axial view. Subtle

enlargement of right intracranial optic nerve (*short arrow*) compared with normal right nerve (*long arrow*) was confirmed at surgery. **E**, Metrizamide cisternogram, coronal view. Again, involvement of the right intracranial optic nerve (*arrow*). **F**, Metrizamide cisternogram, axial view. Optic chiasm (*arrow*) appears normal. However, infiltration of chiasm was noted at surgery.

Fig. 3.—Optic nerve sheath meningioma, fusiform type. **A**, Axial scan with contrast. While entire orbital part of optic nerve/sheath appears involved, posterior part is enlarged in fusiform fashion. Lower density optic nerve (*arrow*) within thickened nerve sheath causes "tram-track" appearance. **B**, Coronal scan. Optic nerve (*arrow*) within high-density tumor.

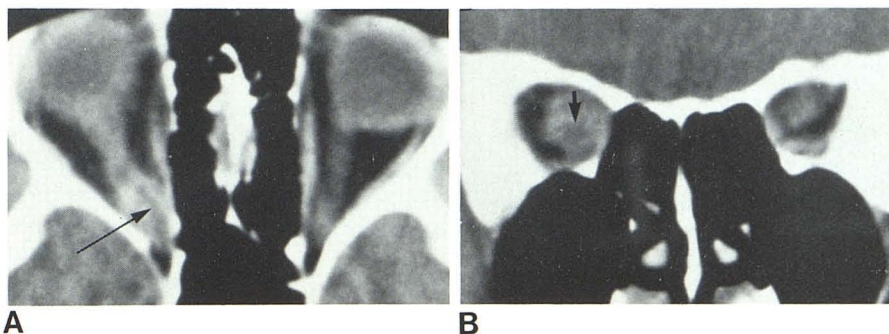


Fig. 4.—Optic nerve sheath meningioma. Diffuse thickening of optic nerve sheath while lateral and medial margins are, in some areas, higher in absorption than center.

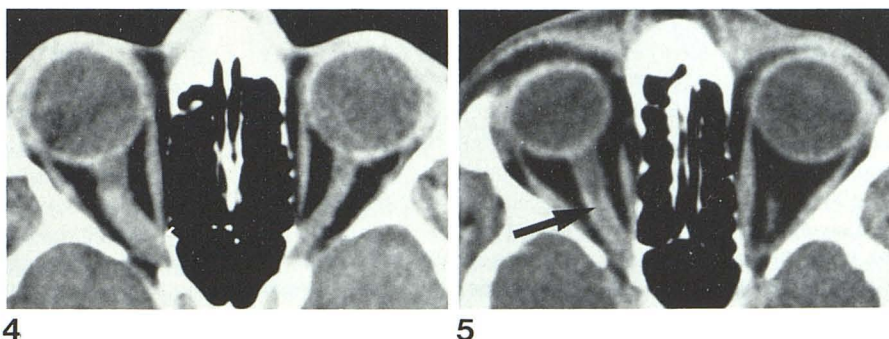


Fig. 5.—Optic nerve sheath meningioma with tram-track sign. Axial CT reveals enlargement of optic nerve/sheath with lower absorption nerve (*arrow*) encased by thickened sheath.

(0/9) is uncommon but is more frequent than in gliomas, and may produce a CT appearance of a sleevelike case around the nerve [2]. Optic nerve sheath meningiomas enhance with contrast (9/9), although less intensely than

intracranial meningiomas [2]. As with optic gliomas, optic canal widening may be demonstrated, as was seen in two of the five cases in our series in which the canals were well visualized. Typical hyperostosis of adjacent bones may be

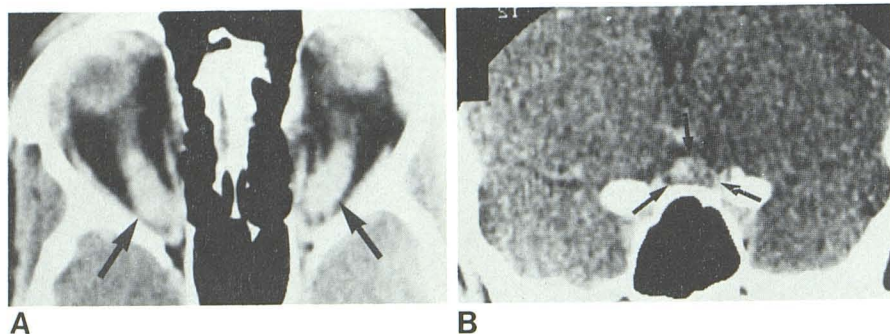


Fig. 6.—Bilateral optic nerve sheath meningiomas. A, Axial CT. Enlargement of posterior halves of both optic nerves (arrows). B, Coronal view at level of tuberculum. Enhancing mass (arrow) that, at surgery, involved optic chiasm and was continuous with optic nerve lesions.

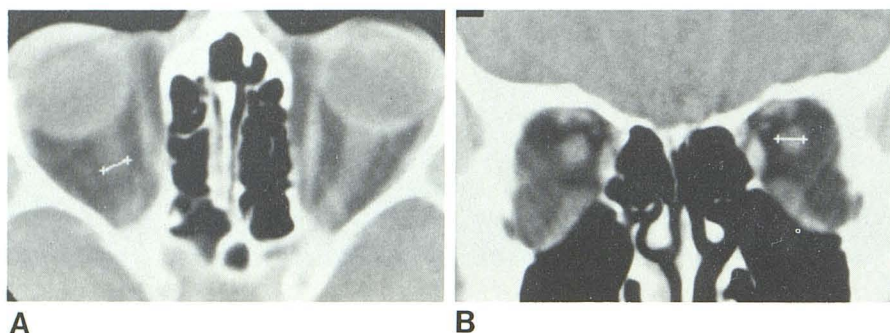


Fig. 7.—Bilateral papilledema in patient with pseudotumor cerebri. A, Axial CT. Both optic nerves/sheaths markedly enlarged. Transverse measurement of right optic nerve was 6 mm. B, Coronal scan again shows bilateral enlargement of optic nerves/sheaths.

seen (1/9). Intracranial extension of tumor (2/9) is best evaluated with contrast material (fig. 6). Fine-needle biopsy of optic nerve meningiomas, as well as gliomas, has been performed under CT guidance [11].

Other Tumors

Neuromas of the optic nerve sheath are rare lesions that are often associated with neurofibromatosis [2, 5]. Isolated case reports of hemangioblastoma [12] and metastasis and leukemia [13] producing optic nerve enlargement on CT have also been reported.

Nonneoplastic Involvement of the Optic Nerve Sheath

Increased Intracranial Pressure

Bilateral enlargement of the optic nerve/sheath has been noted in patients with increased intracranial pressure and papilledema [14–16]. The optic nerve sheath contains all meningeal layers, and the intracranial subarachnoid space is continuous with that of the nerve sheath. Dilatation of this space would render the same appearance on CT as enlargement of the optic nerve itself. It is postulated that increased intracranial pressure is transmitted to the subarachnoid space within the optic nerve sheath causing this space to enlarge [14–16]. Metrizamide has been noted to enter the optic nerve sheath during cisternography [15, 16]. This method could be used to confirm dilatation of the sheath's subarachnoid space in these cases.

We encountered enlarged optic nerve silhouettes in three patients with chronic pseudotumor cerebri (fig. 7), but have not seen this with papilledema secondary to hydrocephalus and brain tumors. Since we make CT sections through the

orbits only incidentally in patients with evidence of increased intracranial pressure, we have no idea of the relative frequency of this finding.

Optic Neuritis

Optic neuritis is an acute inflammation of the optic nerve. While it may be associated with other inflammatory diseases, it is usually idiopathic, and is often a manifestation of multiple sclerosis [17]. Enlargement and contrast enhancement of the optic nerve can occasionally be seen secondary to edema and increased vascular permeability [18] (fig. 8). These changes are reversible as the neuritis subsides with steroid therapy. Although we have examined about 10 patients with the clinical diagnosis of optic neuritis, we have seen definite abnormalities on CT in only two.

Graves Disease

Enlargement of the intraorbital structures in Graves disease occurs primarily in the chronic phases and is mainly due to an increase in mucopolysaccharides, collagen, and glycoproteins, and to water binding by these substances [19]. Optic nerve/sheath enlargement may be seen but occurs only during the advanced stages when marked muscle enlargement is readily identified by CT [5, 20] (fig. 9). This finding was seen in five of 25 cases of orbital Graves disease reviewed for this article, always in association with severe muscle enlargement.

Orbital Pseudotumor

Orbital pseudotumor is an idiopathic disease that may involve any of the intraorbital structures. Pathologically, a

Fig. 8.—Optic neuritis, left eye, in 25-year-old man with sudden loss of vision in left eye, which improved rapidly on steroids. A, Axial CT with contrast. Enlargement of left optic nerve (7 mm) compared with right (4 mm). B, Coronal scan. Enlargement of left optic nerve.

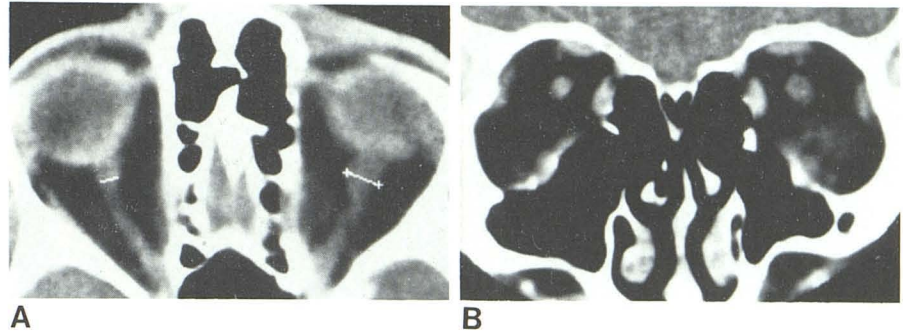


Fig. 9.—Graves disease with bilateral optic nerve/sheath enlargement. Axial scan shows bilateral enlargement of optic nerves/sheaths. Bilateral extraocular muscle involvement with proptosis is characteristic of Graves disease.

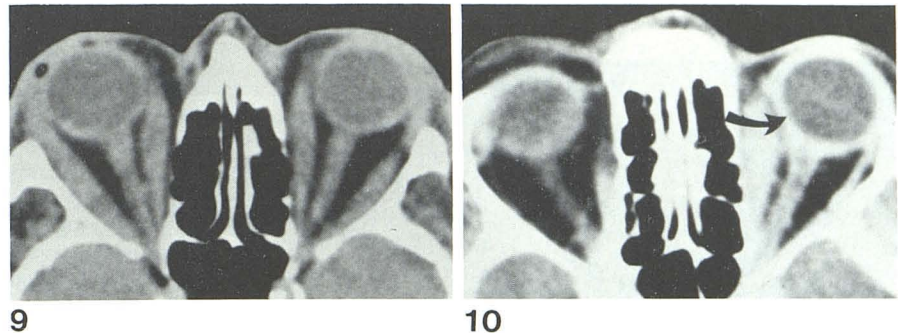


Fig. 10.—Orbital pseudotumor with enlargement of the optic nerve/sheath. Axial scan reveals thickened enhancing left optic nerve/sheath with tram-track appearance. Involvement of medial rectus and sclera (arrow) distinguishes this condition from meningioma.

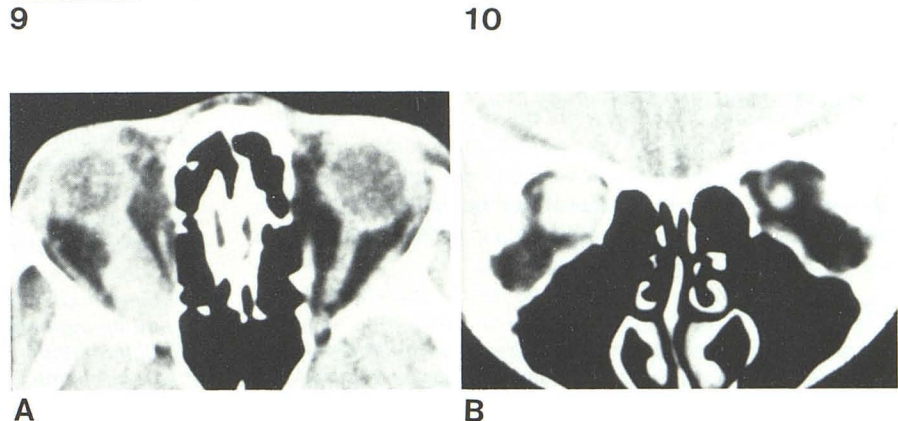


Fig. 11.—Orbital pseudotumor with isolated optic nerve/sheath involvement. A, Axial scan with contrast. Diffuse thickening of right optic nerve/sheath. Tram-tracking not noted. It would be difficult to distinguish this process from optic glioma or sheath meningioma based on scan alone. Biopsy revealed orbital pseudotumor. B, Coronal scan. Massive enlargement of posterior part of optic nerve/sheath.



nonspecific inflammatory infiltrate is seen [19]. Enlargement and contrast enhancement of the optic nerve/sheath may occur in orbital pseudotumor, usually in association with other abnormalities, which suggest the proper diagnosis [20, 21] (fig. 10). When the optic nerve/sheath alone is involved, it is not possible to distinguish pseudotumor from optic glioma or meningioma by CT scan (fig. 11).

Review of the CT scans of 17 patients with orbital pseudotumor (three with bilateral disease) showed definite optic nerve involvement in four of 20 orbits and equivocal involvement in five others. In only the case illustrated (fig. 11) was an abnormal optic nerve seen as an isolated finding. Tram-tracking was also seen in one instance (fig. 10).

Other Causes of Optic Nerve/Sheath Enlargement

Toxoplasmosis, tuberculosis, sarcoidosis [13], central retinal vein occlusion, and traumatic hematoma [16] (fig. 12) have been reported to cause optic nerve/sheath enlargement on CT scan.

Optic Nerve Drusen

Drusen, or hyaline bodies, are congenital and of unknown origin. Their presence within the optic nerve head may produce "pseudopapilledema" especially in children [22]. They often calcify, and can be easily recognized on CT as punctate high densities at the junction of the optic nerve and globe [21, 23] (fig. 13).

Discussion

High-resolution multiplane CT scanning can provide exquisite detailed images of the optic nerve permitting earlier detection of optic nerve lesions and greater discrimination between various abnormalities. While enlargement of the optic nerve/sheath by optic glioma and meningioma has been emphasized in the literature, it is important to recognize that nonneoplastic processes may produce a similar or identical CT appearance. Tram-tracking, in particular, while highly suggestive of meningioma is not pathognomonic. Due

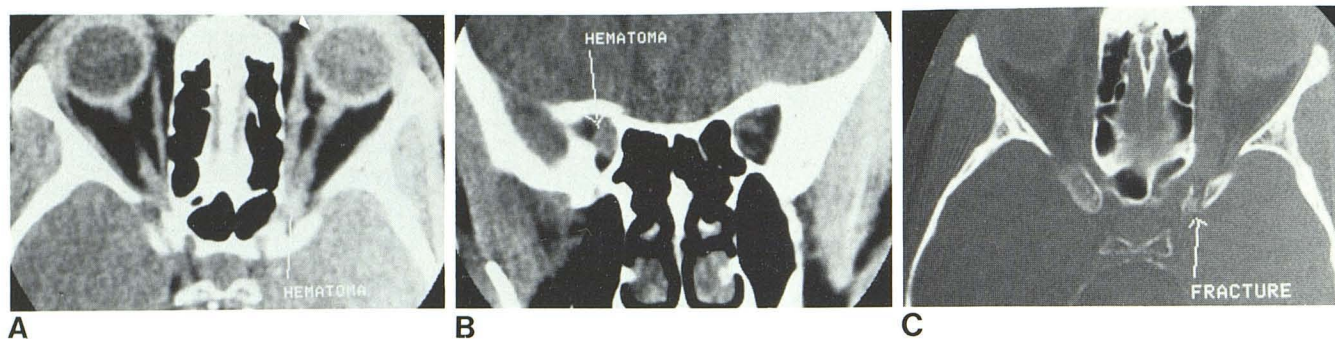


Fig. 12.—Optic nerve sheath hematoma. **A**, Axial scan. Posterior aspect of left optic nerve/sheath is enlarged. **B**, Coronal views at orbital apex. Marked enlargement of left nerve/sheath. **C**, "Bone window" view clearly identifies fracture through left anterior clinoid process.

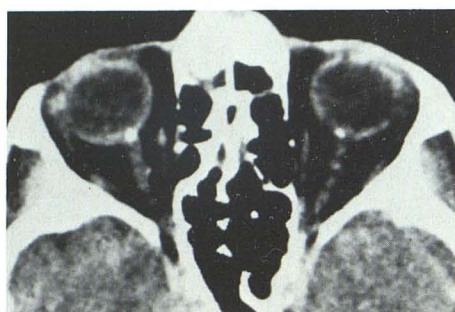


Fig. 13.—Optic nerve drusen. Axial scan shows bilateral calcifications at insertion of optic nerves on globes.

consideration of the associated abnormalities and clinical data will lead to the correct diagnosis in most cases.

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Addendum

Since preparation of this paper, we have encountered "tram-tracking" in a part of an optic nerve that was otherwise extensively involved with a malignant lacrimal gland tumor.