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Radiologic Evaluation of Tumors of the Optic Nerve

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The case material of 42 patients with primary tumors of the optic nerve and chiasm examined at our institution within a 10 year period was reviewed. In each case, the radiologic data and certain clinical information were compiled to determine the role of radiology in the workup of patients with these tumors and to compare the results of various imaging methods. Although different types of information are obtained from each method, late-generation computed tomography provides the most useful and complete information about optic nerve gliomas and intraorbital optic nerve meningiomas. Angiography is also helpful in the evaluation of intracranial optic nerve meningiomas.

In the past, radiologic evaluation of the optic nerve depended on indirect signs on plain films, optic foramen views or tomograms, and, in some cases, angiograms. The optic nerve itself was not imaged unless pneumoencephalography was performed, and then only the intracranial part or the optic chiasm was demonstrated. Clinicians depended primarily on information from the patient's history and clinical examinations, such as visual field testing. By imaging the optic nerve directly, computed tomography (CT) has provided an important new method for tumor detection. Several recent studies have reported the usefulness of CT in evaluating orbital tumors, including optic nerve tumors [1–3], and in demonstrating the normal anatomy of the optic nerve and chiasm [4–6]. Herein we evaluate the role of radiology in the workup and diagnosis of optic nerve tumors in the last decade and compare the results of various imaging methods.

Materials and Methods

Clinical and radiologic information was reviewed in 42 patients with optic nerve tumor initially examined and treated at our institution between 1970 and 1980. (One of the patients elected to have a surgical procedure elsewhere; her pathologic slides were sent to us for review.) Clinical information of interest included age, gender, and neuroophthalmologic symptoms, such as decreased visual acuity, diplopia, proptosis, and pain. Proptosis was considered to be present when there was more than two units of difference in the Krahn measurements. Pain was of recent onset and was located in the orbital or frontal area. Results of visual field testing were also reviewed. All scotomas, including complete blindness, were considered to be visual field defects. Clinical data are summarized in table 1.

The radiologic tests included skull films, optic foramen views, limited optic canal tomography and pneumoencephalography, orbital angiography, and early- and late-generation CT. All of the radiographs were available for review.

Skull films consisted of our standard series of posteroanterior, Caldwell, and stereoscopic lateral views. Optic foramen views were done in the posteroanterior position with the foramen projected into the lower outer quadrant of the orbit. Fluoroscopic spot films were necessary in some young patients.

Tomograms of the optic canals were taken in the axial plane on a high-resolution unit with use of trispiral motion. Pneumoencephalograms were obtained with use of anteropos-

TABLE 1: **Optic Nerve Tumors: Summary of Clinical Data**

	Intraorbital tumors		Intracranial tumors	
	Gliomas	Meningiomas	Gliomas	Meningiomas
No. patients	12	10	14	6
Age range (years)	1–24	13–70	2–62*	18–53
Gender	7F, 5M	6F, 4M	7F, 7M	4F, 2M
Symptoms:				
Decreasing visual acuity	9	8	10	6
Pain	6	6	6	4
Proptosis	7	8	1	1
Diplopia	3	3	5	...
Visual field defects:				
Scotoma on affected side	3	2	7	3
Blind on affected side	8	7	4	3
Operation	9	10	11	6
Surgical biopsy	6	9	9	6
Radiation therapy	8	...	13	...
Course:				
Complications from recurrence	2	1	3	...
Well (after 2 years)	8	6	7	4
No follow-up	2	3	4	2
Neurofibromatosis	4	2	6	...

* This group included only one patient older than age 28 years.

terior and lateral linear motion tomography after the fractional injection of air intrathecally and patient positioning for anatomic evaluation of the suprasellar cistern and anterior third ventricle. Selective internal carotid angiography was performed with use of the direct carotid or transfemoral approach, magnification views, and subtraction developing techniques.

Early-generation CT was performed initially on a Mark I and an EMI 1005 scanner and later on an EMI 5005 scanner with use of 5 mm to 1 cm slices parallel to the orbitomeatal line. Late-generation CT scans were obtained on a GE CT/T 8800, and one of two methods was used for examining the optic nerve. One method was to scan directly in the axial and coronal planes through the orbits and optic chiasm with 5 mm slices and technique factors of 400 mA, 576 pulses/3.3 msec, and 9.6 sec of scanning time. The alternate method was to scan through the orbits and optic chiasm parallel to the orbitomeatal line at 1.5 mm intervals with use of 600 mA, 576 pulses/3.3 msec, and 9.6 sec of scanning time. Multiplanar reformatting was then used to image in the coronal and sagittal planes.

Contrast studies in both early- and late-generation CT examinations were done with slow intravenous infusion of a contrast agent containing 42 g of iodine.

Results

We divided the surgically proven cases into tumors of either primary intraorbital or intracranial origin by using information from the surgical exploration. Tumors that clearly arose within the orbit or tumors that arose within the optic canal and extended into the orbit were defined as primary intraorbital. Gliomas that arose within the intracranial part of the optic nerve or chiasm were defined as

primary intracranial. After careful exclusion of meningiomas of the sphenoid ridge or planum sphenoidale, six cases of meningioma were encountered that arose within the proximal optic canal near the inner meatus and extended only intracranially. For our purposes, we also defined these as intracranial tumors.

In 20 of the 26 patients with gliomas in this series, surgical inspection or biopsy was performed. The patients who did not undergo a surgical procedure had strong clinical and radiologic evidence to support the diagnosis of optic nerve glioma, including continuous or progressive proptosis and visual field defects, abnormal findings on optic foramen views, or thickened nerves on CT. The location of the tumor was judged from clinical and radiologic evidence. These patients also had had at least 2 years of follow-up, as it is known that optic neuritis may temporarily mimic optic nerve tumor [7]. Five of the six unoperated patients with gliomas received radiation therapy. All of the meningiomas in our series were surgically proven lesions.

Intracranial extension to the chiasm or optic tract was common in the primary intraorbital tumors, occurring in five of 10 intraorbital meningiomas and five of 12 intraorbital gliomas. Among the 14 cases of primary intracranial gliomas, intraorbital extension had occurred in five.

Pneumoencephalograms, obtained in two patients with intraorbital optic nerve gliomas (table 2), demonstrated a mass in the suprasellar region, an indication of intracranial extension.

Of the eight patients with intracranial optic nerve gliomas examined by CT, extension posterior to the chiasm was demonstrated in four, and ventricular obstruction was present in two of these four. These findings were confirmed at operation.

CT showed no evidence of intracranial extension in six patients with intraorbital meningioma, and, surgically, only one of the six was found to have extension to the chiasm. CT was used to evaluate six of the nine cases of surgically proven intraorbital glioma. This examination accurately predicted two cases of intracranial extension of the three found at operation and correctly identified three cases confined to the orbit or optic canal.

Of the 31 patients who returned to our institution for follow-up, six had recurrent tumor; this group included five patients with gliomas, two of whom required ventricular shunting for management of obstructive hydrocephalus. The gliomas recurred 2, 4, 4, 5, and 10 years after operation or radiation therapy. One patient with an intraorbital meningioma was found to have recurrent tumor 2 years postoperatively.

Discussion

Morphologically, the optic nerve is a white fiber tract of the central nervous system. It contains neuroglial cells and, within the orbit, is sheathed by the leptomeninges (fig. 1). Gliomas arise from glial cells, most commonly astrocytes and less commonly oligodendrocytes. Meningiomas arise from arachnoid cells. Basically, these two tumors—the glioma and the meningioma—are the neoplasms most commonly associated with the optic nerve and its sheath.

TABLE 2: **Optic Nerve Tumors: Summary of Radiologic Findings**

	Intraorbital tumors		Intracranial tumors	
	Gliomas	Meningiomas	Gliomas	Meningiomas
Skull films:				
Hyperostosis	...	1	1	...
Bony erosion or destruction	2	...	7	...
Tumor calcification	...	1	1	...
Totals*	2/12	2/8	9/13	0/6
Optic foramen views; canal enlarged with:				
Sclerotic margins	5	...	5	...
Indistinct margins	4	...	...	...
Bony hyperostosis	...	1	...	...
Totals	9/10	1/6	5/9	0/2
Optic canal tomograms	...	1/2	1/1	0/2
Pneumoencephalograms	2/2	...	...	...
Angiograms:				
Arterial displacement	2	3	4	1
Tumor stain	...	...	...	1
Arterial encasement	1	1	...	...
Totals	2/3	4/4	4/5	2/3
Early-generation CT:				
Thickened nerve	3	4	...	...
Orbital or suprasellar mass	4	2	7	...
Tumor enhancement	2	1	4	...
Totals	7/9	5/8	7/7	0/4
Late-generation CT:				
Thickened nerve	4	5	...	...
Orbital or suprasellar mass	...	2	5	...
Tumor enhancement	1	4	3	...
Totals	4/4	5/5	5/5	...

* Totals represent no. patients with abnormal findings/total no. patients who underwent this radiologic examination. Some patients had more than one specific finding in each category.

Gliomas may affect any part of the visual pathway from the nerve fiber layer of the retina through the optic nerve, chiasm, optic tract, optic nerve radiation, and occipital cortex. "Optic nerve gliomas" are considered to arise from the anterior visual pathways—that is, the optic nerve, chiasm, and tract. Most often, they become manifest in the first decade of life and have an increased incidence in patients with neurofibromatosis [8, 9]. Histologically, most of these tumors are of low-grade malignancy (grade I or II) [10]. The histologic picture may vary from a predominantly fibrillar pattern to one that is microcystic (fig. 2). Rosenthal fibers, eosinophilic bodies formed from the cytoplasm of degenerating cells, are a frequent finding (fig. 3). As the tumor grows, the meninges undergo reactive proliferation and thereby contribute to the increased size of the nerve. Gliomas occurring in adults have been reported to be more aggressive and of higher grade malignancy than those occurring in young patients [11].

Meningiomas usually arise from arachnoid cells situated along the sheath; rarely, they arise from ectopic arachnoid cells within the muscle cone or the walls of the periorbita [12, 13]. Histologically, the cells of the orbital meningioma are usually of the meningotheliomatous type (fig. 4) but may occasionally be transitional [15]. This tumor most often occurs in adult patients but can be seen in young patients
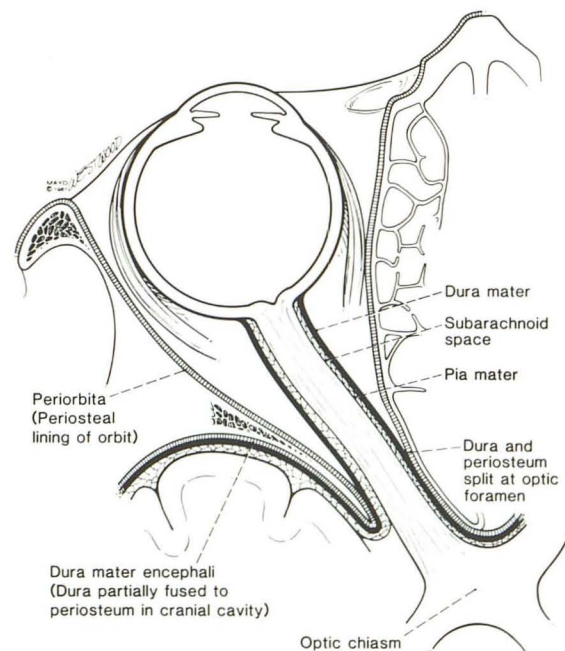

Fig. 1.—Optic nerve and its coverings. Dura mater is fused with periorbita only at apex of orbit, and intraorbital part of optic nerve is covered by dura, arachnoid, and pial layers.
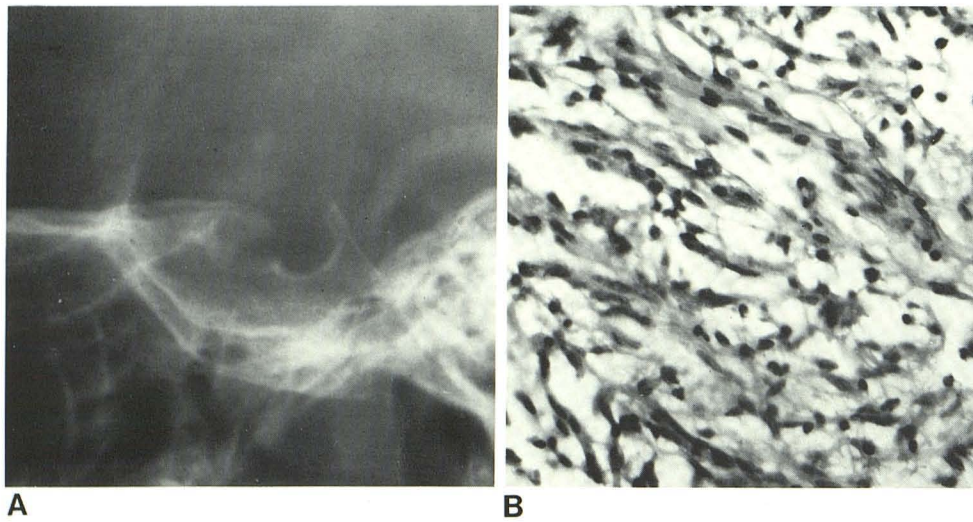
Fig. 2.—Glioma of optic chiasm in 3-year-old boy. **A,** Although chiasmatic sulcus can be prominent radiographically as a normal variant, this patient has a particularly eroded and demineralized sulcus. **B,** Astrocytoma, grade 1; microcystic pattern. (H and E ×400.)
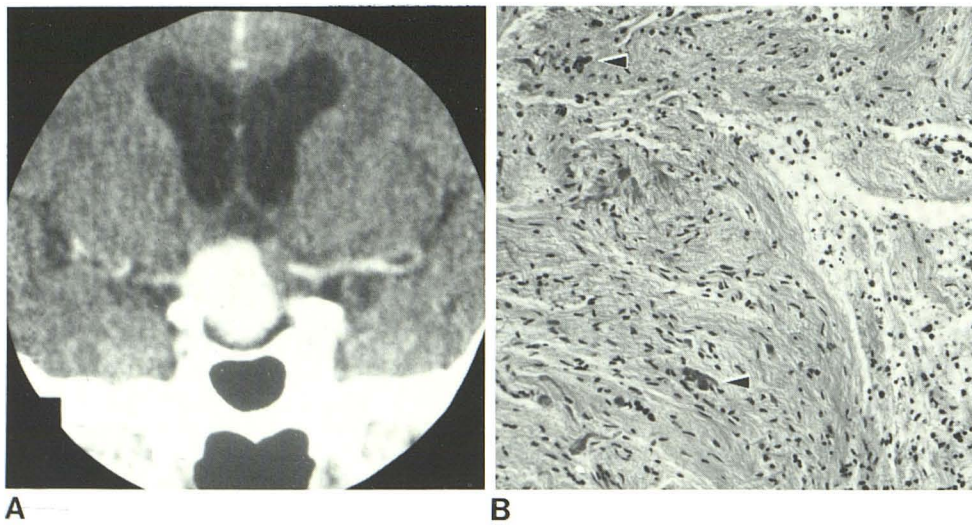

Fig. 3.—Glioma of optic chiasm in 23-year-old woman. **A,** After injection of contrast agent, coronal late-generation CT scan shows enhanced, enlarged globular mass in chiasm. **B,** Astrocytoma, grade 1. Tumor cells are spindle-shaped. Numerous Rosenthal fibers (*arrowheads*) scattered throughout. (H and E ×160.)
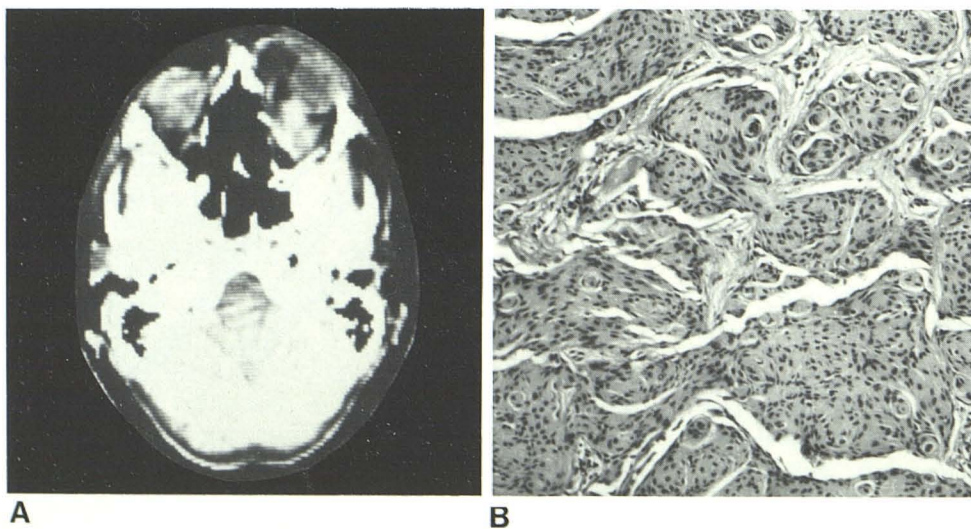

Fig. 4.—Intraorbital meningioma in 40-year-old woman. **A,** Early-generation CT scan. Large mass in apex of orbit. (Reprinted from [14]). **B,** Cells are rounded and lie in nests, characteristic of meningotheliomatous pattern. (H and E ×160.)

with neurofibromatosis. There is an increased incidence in women.

Radiologic examination is important in the diagnosis and evaluation of optic nerve tumors. We have used the commonly used imaging methods, which range from the easily obtained and relatively inexpensive skull films to the complex and invasive angiographic examination. The information gained from the different examinations also varies, with a relatively low yield from the plain skull films to a high yield from late-generation CT and angiography.

In our series, information was gained from skull films somewhat less often than has been reported for all orbital lesions [2]; we noted abnormal findings in 13 (33%) of 39 patients and abnormal findings indicative of optic nerve tumor in nine (23%). Gliomas most often produced erosion of the anterior clinoid processes or chiasmatic sulcus (fig. 2). This finding was present in two intraorbital gliomas that had extended intracranially and in five intracranial gliomas. In addition, two cases of intracranial glioma produced erosion of the dorsum; at surgery, both were found to have extended posteriorly and caused obstructive hydrocephalus. One intracranial glioma produced hyperostosis of the planum sphenoidale. One intraorbital meningioma produced hyperostosis of the anterior clinoid process by extending intracranially (fig. 5). Intracranial meningiomas produced no bony changes, presumably because growth was upward and away from the bony structures of the middle fossa. Tumor calcification was rare, seen in only one patient with an intracranial glioma and one patient with an intraorbital meningioma who had neglected medical attention and in whom the tumor had become quite large. In most cases, the information gained from plain films was obtained from the lateral view.

Optic foramen views were most helpful in evaluating both intracranial and intraorbital gliomas; widened foramina were detected in 14 (74%) of 19 cases (fig. 6). This included six patients with neurofibromatosis and widened foramina. It has been noted that caution should be exercised in interpreting widened foramina in patients with neurofibromatosis

because of dysplastic bony changes [16]. In these six patients, optic nerve gliomas were confirmed at operation in four and by strong CT evidence in the other two.

With use of optic foramen views, changes were disclosed in only one (12.5%) of eight meningiomas, perhaps because meningiomas involved a smaller part of the optic nerve than did gliomas and were less likely to cause visible enlargement of the foramen. In this single case, the meningioma produced both enlargement of the canal and surrounding bony hyperostosis.

We have used tomograms of the optic canal infrequently, usually when optic foramen views yielded no information. In those cases, we used axial plane tomograms. In our series, they added information in one case in which an intracanalicular meningioma had widened only the proximal part of the optic canal. Abnormal results in another case only confirmed findings seen on optic canal views. Recently, Strother et al. [17] reported the usefulness of tomography in the axial projection of the optic canal in primary optic nerve meningiomas.

Pneumoencephalography was used in only two patients in our series who were examined in the early 1970s. At that time, it was a valuable alternative imaging method; currently, it is used rarely in our practice to supplement or confirm CT findings.

In many patients able to undergo an invasive procedure, angiography demonstrated findings characteristic of the presence of a lesion, the most common of which was displacement of the ophthalmic or supraclinoidal internal carotid arteries (fig. 7). Two cases of arterial encasement were demonstrated. The uniform tumor stain reportedly produced by meningioma in some studies [1, 18] was seen in only one case.

Many of the angiograms were obtained preoperatively in patients with clinical information suggestive of large or extensive tumors. Of the 12 patients with abnormal findings on angiograms in our series, nine had had abnormalities detected on plain films or CT before the angiograms were obtained. Angiography, however, is a useful procedure for

Fig. 5.—Intraorbital meningioma with intracranial extension in 15-year-old girl. A, Lateral skull film. Thickened bone and sclerosis in region of right anterior clinoid process. B, Pathologic classification was psammomatous meningioma. Numerous psammoma bodies (H and E ×160.)

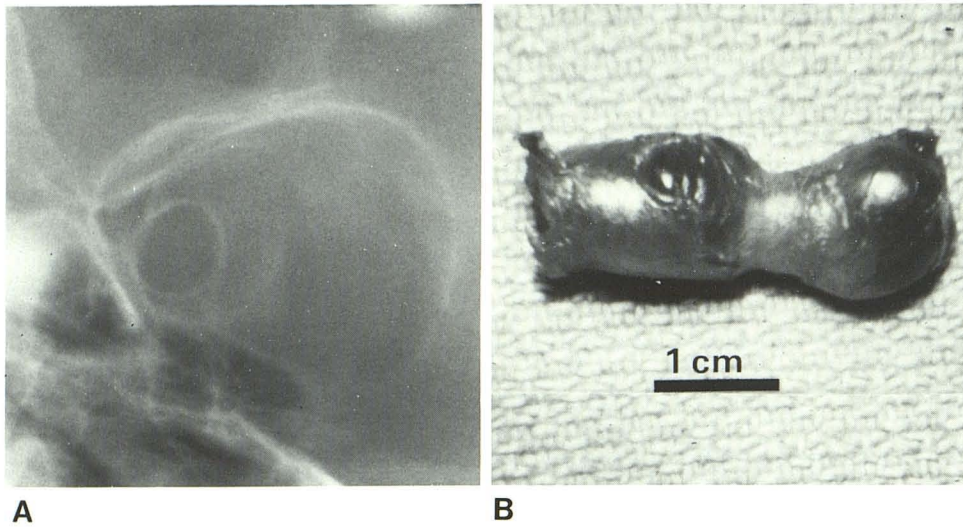A   B
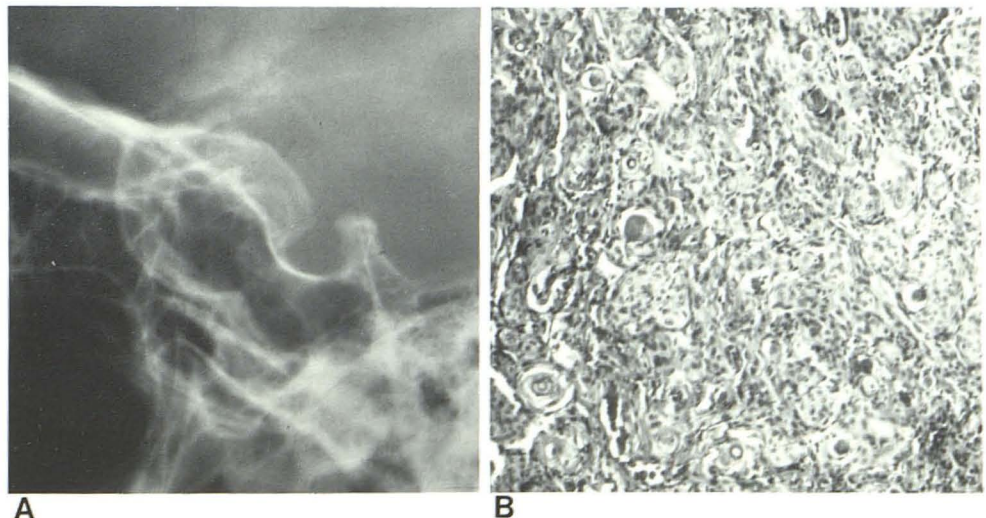
Fig. 6.—Glioma of optic nerve in 1-year-old girl with neurofibromatosis. **A,** Right optic foramen markedly enlarged. **B,** Surgical specimen. Diffusely enlarged right optic nerve. Circumferential enlargement limited only in intracanalicular portion.
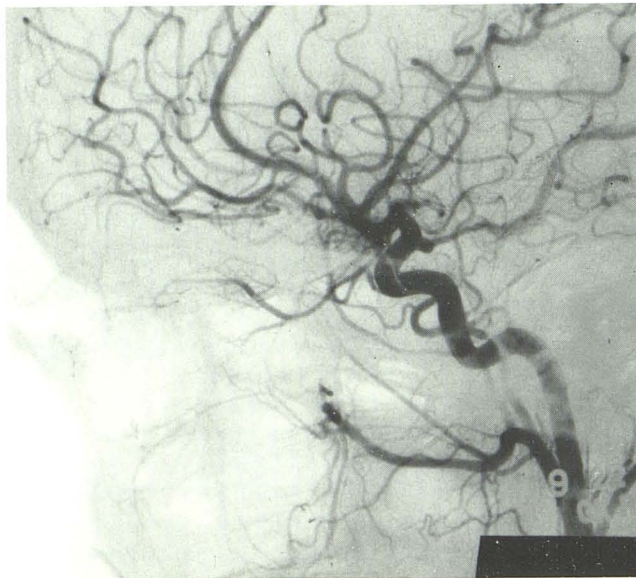

Fig. 7.—Intraorbital optic nerve meningioma in 50-year-old woman. Angiogram. Ophthalmic artery displaced downward and narrowed in proximal part. At surgery, meningioma was 1 × 0.5 cm.

Fig. 8.—Intraorbital optic nerve meningioma in 13-year-old boy with neurofibromatosis. Early-generation CT scans. **A,** Thickening of optic nerve. **B,** Enhancement after injection of contrast agent.

Fig. 9.—Glioma of optic chiasm. Early-generation CT scans without (**A**) and with (**B**) contrast agent. Enhancing mass in suprasellar region.

the diagnosis of intracranial meningiomas. In our six patients with intracranial meningioma arising from the intracanalicular optic nerve, results of all radiologic examinations were normal except for the angiographic findings in two patients. This series of examinations included skull films in all six patients, optic foramen views and optic canal tomograms in two patients each, and early-generation CT in four patients (None of the six patients underwent evaluation by late-generation CT.)

Early-generation CT proved valuable in evaluating all optic nerve tumors except intracranial meningiomas arising from the intracanalicular optic nerve. It provided information not only about tumor location but also about size and extension of the lesion, relati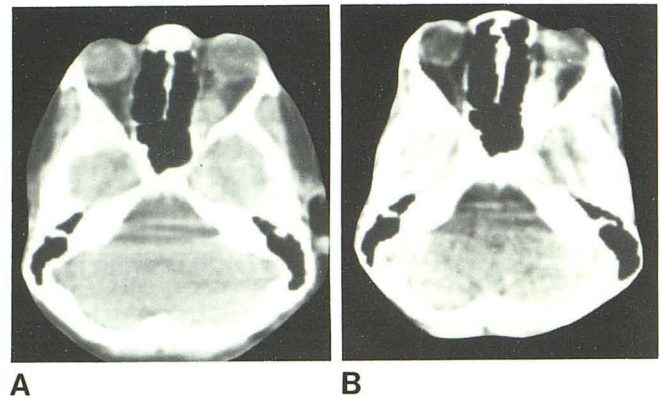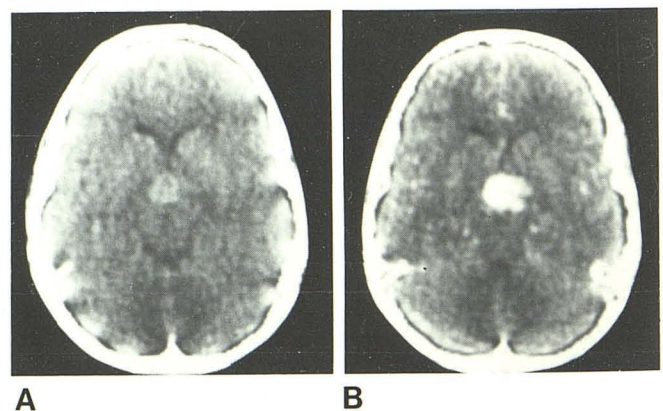on of the tumor to other orbital or cranial structures, and secondary pathologic signs such as ventricular dilatation. We found the most specific sign of optic nerve tumor to be a thickened nerve with enhancement when a contrast agent was used (fig. 8). A mass in the

orbital apex was also seen in cases of intraorbital tumor (fig. 4), and an enhancing mass in the suprasellar region was evident in the cases of glioma of the chiasm (fig. 9).

Late-generation CT revealed abnormalities in all 14 patients in whom this examination was done. We believe this is the current procedure of choice for suspected intraorbital meningiomas and both types of optic nerve gliomas. This examination can often be definitive for diagnosing an optic nerve tumor and excluding other types of intraorbital neoplasms not arising from the optic nerve. The anatomic position of the tumor and the extent of involvement among the globe, optic canal, and intracranial compartment can also be assessed. Because 1.5 mm slices can be obtained and multiplanar reformatting can be used, mistakes caused by

a partial volume effect in determining the caliber and the course of the nerve [4] can be minimized. In four cases of optic sheath meningioma, peripheral enhancement of a thickened nerve was present (figs. 10–12); this is a suggestive sign of meningioma. In four patients with glioma of the optic chiasm, the chiasm was enlarged and globular (figs. 3 and 13), findings that correspond to those reported in a recent study [5].

The exact role of late-generation CT in distinguishing optic nerve neoplasms from optic neuritis or nonneoplastic swelling of the nerve must be further defined. The tumors in this series that were studied with late-generation CT have been assessed by operation or biopsy. In general, CT revealed a prominent, diffusely enlarged, and sometimes en-

Fig. 10.—Intraorbital optic nerve meningioma in 15-year-old girl. Late-generation CT scan after injection of contrast agent. Thickened right optic nerve with peripheral enhancement. (Same case as in fig. 5.)
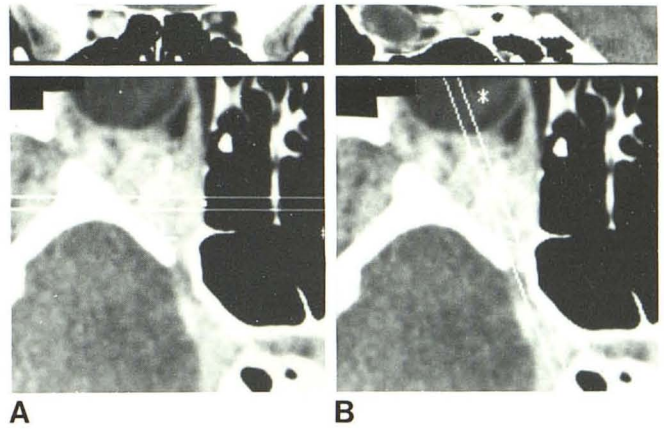

A   **B**

Fig. 12.—Recurrent intraorbital optic nerve meningioma in 31-year-old woman. Late-generation CT scans. Reformatted images in coronal plane (**A**) and along axis of optic nerve (**B**).

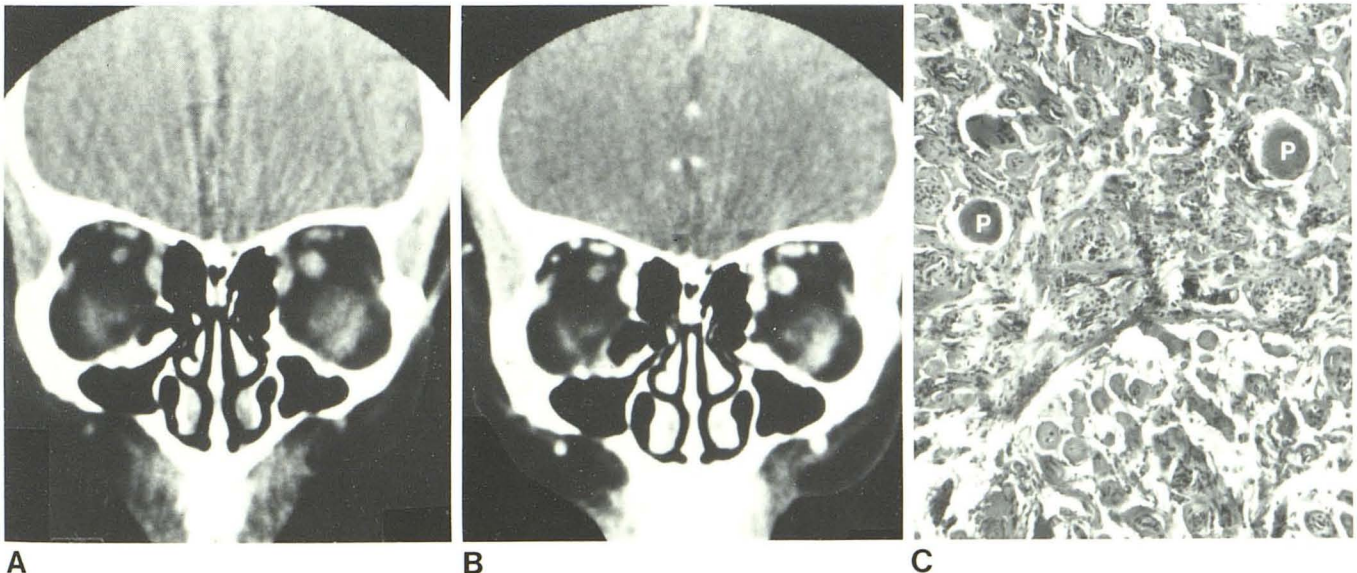**A**   **B**   **C**

Fig. 11.—Intraorbital meningioma in 67-year-old woman. Coronal late-generation CT scans through orbits. **A**, Before administration of contrast agent. Thickened right optic nerve. **B**, After administration. Peripheral en-

hancement. **C**, Pathologic classification was psammomatous meningioma with numerous psammoma bodies (P) scattered throughout. (H and E ×400.)
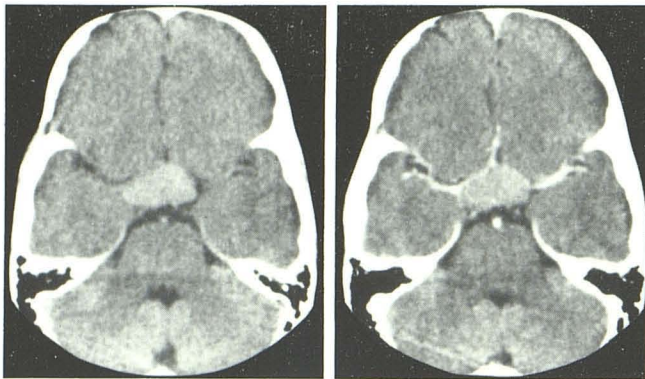
Fig. 13.—Late-generation CT scans of surgically proven glioma of optic chiasm in 22-year-old man. **A,** Chiasm enlarged by a globular mass. **B,** Tumor enhances slightly after administration of contrast material.

hanced nerve in axial, coronal, and sagittal reformatted planes, exclusive of abnormal changes in the rectus muscles—features strongly suggestive of a neoplasm of the optic nerve. Less extensive swelling and focal enhancement may be seen in cases of optic neuritis, particularly with use of early-generation CT techniques. Nevertheless, when the pathologic changes are subtle, some overlap exists in the appearance of inflammatory swelling and neoplasms of the optic nerve. The extent to which late-generation CT can confidently distinguish these conditions remains to be investigated.

Our experience, then, is that angiography and CT play important and specific roles in the evaluation of optic nerve tumors. Late-generation CT is becoming increasingly important because of the associated high information yield and relatively low risk to the patient. Late-generation CT used with metrizamide cisternography may be the most definitive means in future studies for resolving subtle questions about intracanalicular and chiasm extension of known optic nerve tumors. Optic foramen views deserve mention for their simplicity and sensitivity for certain tumors.

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