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Radiologic-Pathologic Correlation
Meningioma of the Optic Nerve Sheath

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Clinical History

A 50-year-old woman presented with a 1-year history of severe diplopia associated with proptosis and slightly decreased vision involving the right eye (OD 20/40; OS 20/20). Physical examination revealed proptosis of the right eye associated with limited upward gaze.

Magnetic resonance (MR) imaging of the orbit showed a right intraorbital mass involving the optic nerve sheath and resulting in mild proptosis. The mass had a fusiform shape and measured 2.5 × 1.5 × 2 cm. It was located eccentrically relative to the optic nerve, with the bulk of the lesion along the nasal aspect of the optic nerve, displacing the proximal half of the medial rectus muscle medially. The mass involved the entire length of the intraorbital segment of the optic nerve, extending from the posterior aspect of the globe to the orbital apex. Abnormal signal was seen within the nerve sheath (Fig 1), suggesting a small endodural component; however, there was no evidence of direct extension into the optic nerve. The mass did not extend into the globe or optic canal. It showed intermediate signal intensity, equivalent to that of the temporal lobe cortex, on T1-weighted (600/20/2 [repetition time/echo time/excitations]), balanced (2000/30/2), and T2-weighted (2000/80/2) axial MR images (Fig 1A–C). The mass could be distinguished from the optic nerve on balanced and T2-weighted images (Fig 1B and C). A cleavage plane between the mass and the medial rectus muscle could be seen on the T1-weighted image (Fig 1A).

The mass showed prominent and homogeneous enhancement after the intravenous administration of gadopentetate dimeglumine, as seen on T1-weighted, fat-suppressed axial and coronal images (Fig 1D–F). The degree of enhancement was similar to that of the extraocular muscles. The normal nonenhancing optic nerve was readily identified within the substance of the enhancing mass. The left optic nerve and the remainder of the optic pathway appeared within normal limits.

The patient underwent a right orbitotomy and subfrontal craniotomy with excision of the mass and intraorbital optic nerve. The gross surgical specimen consisted of an ovoid mass of firm tissue measuring approximately 3.0 × 2.5 × 1.5 cm that encircled the optic nerve (Fig 1G). A macrosection showed the neoplasm to extend through the dura of the optic nerve, compress the optic nerve parenchyma focally, and infiltrate the surrounding orbital fat widely (Fig 1H). The cells composing the neoplasm were arranged in nests and whorls. The individual cells had indistinct margins and relatively uniform, vesicular nuclei (Fig 1I). Some of the neoplastic

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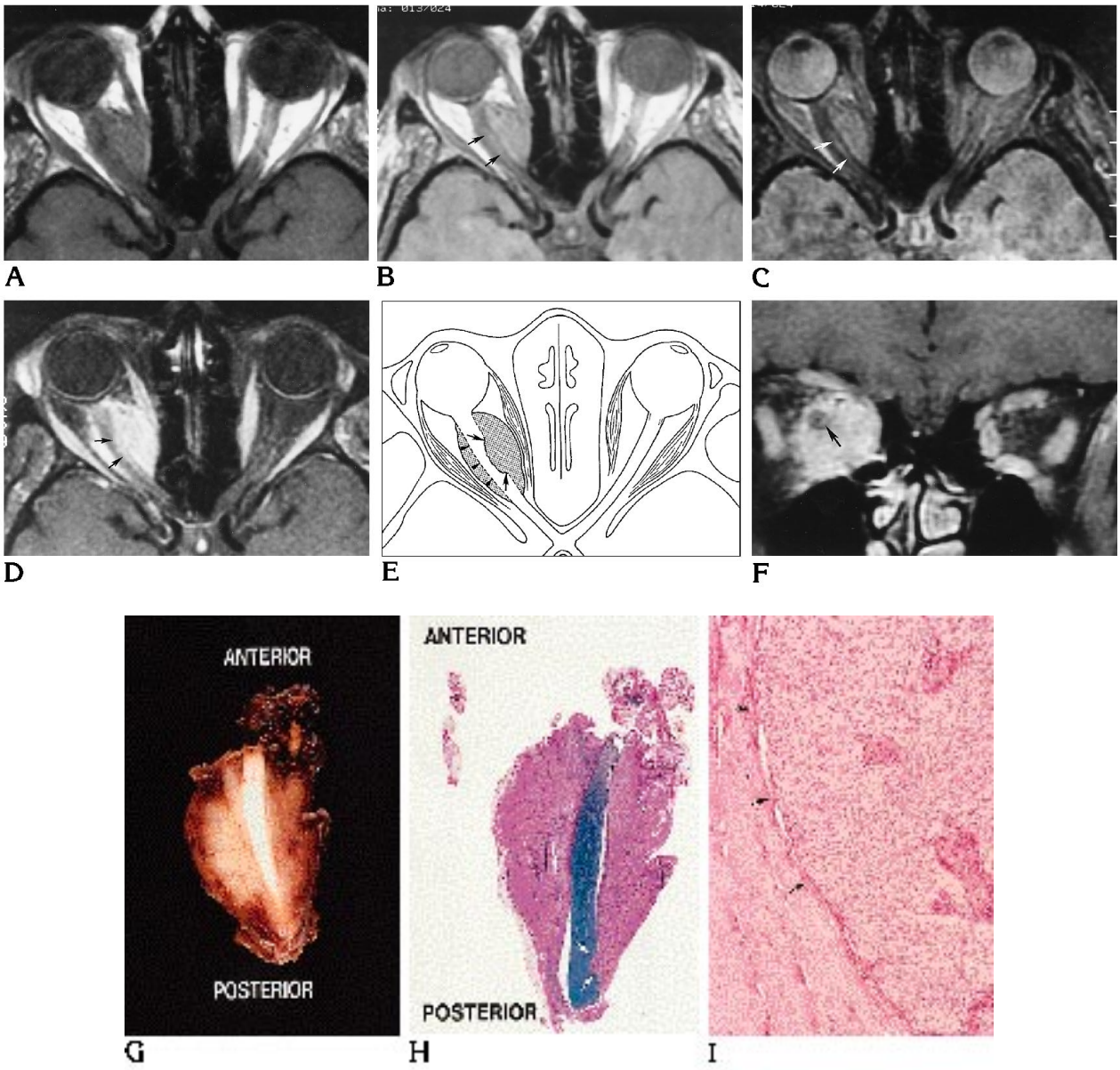

Fig 1. *A*, T1-weighted axial MR image of the orbits shows a fusiform mass of intermediate signal intensity involving the right intraorbital optic nerve. The mass encroaches on the medial rectus muscle. Mild right proptosis is present.

B, Balanced axial MR image of the orbits shows that the mass is isointense to the temporal lobe cortex and can be distinguished from the optic nerve. Note the intradural extension of the tumor (*arrows*).

C, The mass remains isointense to the temporal lobe cortex on the T2-weighted axial MR image of the orbits. Intradural extension of the relatively hyperintense tumor is again noted (*arrows*).

D, Fat-suppressed contrast-enhanced T1-weighted axial MR image of the orbits shows uniform enhancement of the mass, which extends from the posterior aspect of Tenon's capsule to the orbital apex. The nonenhancing optic nerve is seen within the substance of the enhancing mass. The endodural component of the tumor is also seen (*arrows*).

E, Axial diagram of orbits shows tumor surrounding the right intraorbital optic nerve (*arrowheads*) and extending through the optic nerve sheath to encroach on the optic nerve (*arrows*).

F, Fat-suppressed contrast-enhanced T1-weighted coronal MR image of the orbits shows the enhancing tumor surrounding and encroaching on (*arrow*) a nonenhancing optic nerve.

G, Gross surgical specimen cut longitudinally through the optic nerve shows neoplastic infiltration of orbital fat.

H, Macrosection of the surgical specimen shows infiltration of the orbital fat, extension through the optic nerve dura (*arrows*), and compression of nerve parenchyma, which is still well myelinated (ie, stained dark blue) (Luxol fast blue myelin stain).

I, Microsection shows a typical meningothelial meningioma composed of relatively small, polygonal neoplastic cells arranged in small nests and indistinct whorls. The neoplasm is compressing the optic nerve focally (*arrows*).

cell nuclei contained pseudoinclusions. The optic nerve showed minimal demyelination or axonal degeneration.

The diagnosis was optic nerve sheath meningioma

Discussion

Primary orbital meningiomas are benign neoplasms that arise directly from arachnoid cap cells within the optic nerve sheath (ONS). These tumors rarely arise separately from the ONS complex. When they do, the neoplasm is thought to arise from ectopic rests of arachnoid cells (1). Four percent of primary orbital meningiomas do not involve the ONS (2). Secondary orbital meningiomas represent the intraorbital extension of neoplasm from sites immediately adjacent to the orbit and account for 90% of orbital meningiomas (2). This group of lesions includes sphenoid, frontal, olfactory, and parasellar region meningiomas. These neoplasms arise from the meninges that are adjacent to the orbit and enter the orbit through either the optic canal, superior orbital fissure, or direct bony extension (1).

Epidemiology

Meningiomas involve the orbit in 0.4% to 1.3% of cases, and 3% to 9% of orbital neoplasms are meningiomas (1–3). Primary orbital meningiomas account for 10% to 33% of orbital meningiomas (1, 2). Of the primary ONS neoplasms, meningiomas are second to gliomas in frequency, representing one third of the neoplasms in this location (2). The peak incidence of ONS meningiomas is in the fourth and fifth decades of life; the mean age at clinical presentation is 40.8 years (2). ONS meningiomas have a female predilection of 61% to 84% of cases (1–3). In children, ONS meningiomas are more common than intracranial meningiomas and have an equal sex distribution (1, 4).

ONS meningiomas tend to involve a single orbit. This is the case in 95% of reported cases, whereas bilateral involvement is observed in 5% of reported cases (2). Bilateral involvement may be caused by tumor extension along the optic canal into the optic chiasm to involve the contralateral ONS. Less commonly, bilateral ONS involvement is attributable to the presence of multiple meningiomas (1). An association exists between bilateral ONS meningiomas and neurofibromatosis (2–4). Rarely, ONS meningiomas have been reported in association with prior radiation therapy (1).

Clinical Presentation

ONS meningiomas typically present with a gradual loss of vision and proptosis. The extent of visual impairment and proptosis depends on the location of the neoplasm along the course of the ONS (3). Orbital apex tumors are associated with more vision loss and relatively less proptosis, whereas lesions located more distally along the ONS result in less vision loss and more proptosis (3). Decreased visual acuity is seen in 96% of patients with ONS meningiomas and is characteristically painless and slowly progressive (2). When bilateral ONS meningiomas are present, unilateral vision loss is initially noted (1). Vision loss may become more manifest during pregnancy (3). Gaze-evoked amaurosis can occur (1). Color vision and contrast sensitivity may be affected early in the clinical course (1, 2). Proptosis is observed in 59% to 90% of patients and usually occurs later in the patient's clinical course (2, 3). The degree of proptosis is proportional to the duration of symptoms, averaging 2 to 5 mm on exophthalmometry (2, 3). Proptosis is caused directly by the space-occupying effects of the neoplasm or indirectly by hyperostosis.

Optic disk changes, either swelling or atrophy, are frequently seen during fundoscopic examination and have been reported in 98% of patients (2, 4). Optociliary shunt vessels may be seen on the disk surface; these vessels represent the collateral circulation between the compressed central retinal vein and the ophthalmic veins via the choroidal and ciliary veins (1, 2, 5). The optociliary shunt vessels tend to involute as optic atrophy progresses (1). Visual field defects, peripheral or central, occur in 83% of patients and are associated with visual impairment and optic atrophy (2). Ocular motility may be limited, particularly with upward gaze (2). This is attributed to stiffening of the ONS.

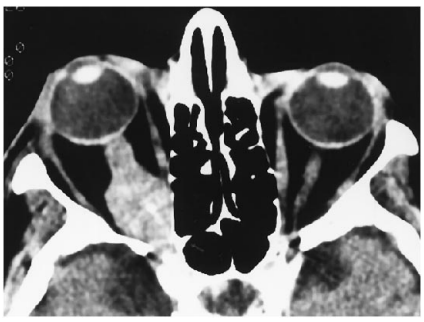

Fig 2. Unenhanced axial CT image of the orbits in a case of ONS meningioma shows a fusiform calcified mass involving the right intraorbital optic nerve. The mass causes proptosis.

Less-specific symptoms include orbital pain and headache.

Radiology

Cross-sectional imaging with computed tomography (CT) and MR is extremely useful in characterizing and fully delineating ONS meningiomas. The involved ONS complex is enlarged. Diffuse thickening of the ONS is observed more frequently than segmental thickening (4). Furthermore, three patterns of ONS enlargement have been described: tubular, fusiform, and excrescent or globular (2, 6). The tubular pattern is present in 64% of cases; the fusiform and excrescent patterns are present in 10% and 25% of cases, respectively (2). This morphologic appearance reflects the pattern of tumor growth. Because meningiomas originate within the arachnoid, the initial growth is within the arachnoid space, beneath the dura. The tumor gradually expands the dura and will eventually invade and extend through the dural sleeve of the ONS complex, as in the current case. All ONS meningiomas have a homogeneous texture and a smooth or slightly lobulated peripheral margin. The meningioma is usually isodense to the optic nerve on the unenhanced CT examination. However, increased density may be observed secondary to calcification (Fig 2). This calcification may form a sleevelike case around the optic nerve (4). ONS meningiomas are isointense to the optic nerve on T1- and T2-weighted MR images (5, 7, 8). These neoplasms may demonstrate decreased signal intensity on T1-weighted images and increased signal intensity on T2-weighted images (7).

ONS meningiomas demonstrate homogeneous enhancement, comparable to the enhancement seen within the extraocular muscles, after intravenous contrast administration on both CT and MR. The increased density of the calcified or enhancing tumor is readily distinguished from the less dense and uninvolved optic nerve. The “tram track” sign consists of a relatively lucent optic nerve in the center of an enlarged ONS complex with peripheral enhancement (7). This sign is not pathognomonic for meningioma; it has been reported in association with orbital pseudotumor and optic neuritis (7). On MR, the increased intensity of the enhancing tumor is readily separated from the nonenhancing optic nerve. Enhanced studies are also useful for the evaluation of intracanalicular, intracranial, and contralateral ONS extension and/or involvement. When combined with fat suppression techniques, enhanced MR images yield optimal lesion detection and delineation (Fig 3) (9). Perioptic cysts are bulbous dilatations of the nerve sheath containing trapped cerebrospinal fluid, typically seen between the globe and the anterior aspect of the meningioma (Fig 4). These perioptic cysts can be detected with MR (10). Osseous changes can sometimes be observed, especially with CT. These bony alterations include widening of the optic canal, when the canalicular segment of the ONS is involved, and hyperostosis if the lesion arises in the appropriate location (4). CT does not reliably dif-

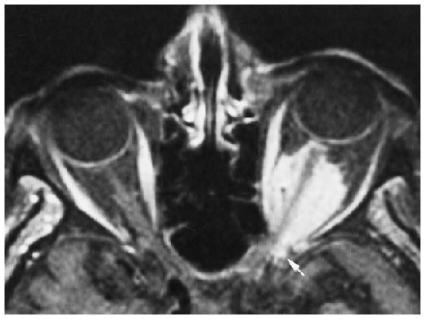

Fig 3. Fat-suppressed contrast-enhanced T1-weighted axial MR image of the orbits in a case of ONS meningioma shows an enhancing mass that extends from the posterior aspect of the globe to the intracanalicular segment of the left optic nerve (*arrow*).

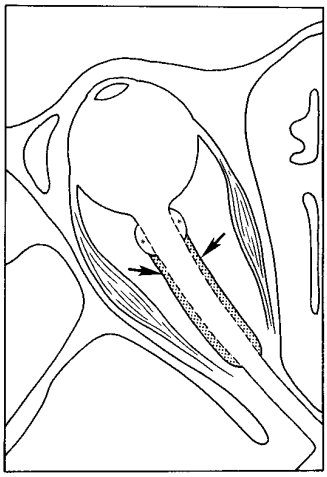

Fig 4. Axial diagram of orbits shows a perioptic cyst (*asterisks*) between the globe and a meningioma (*arrows*).

ferentiate between true hyperostosis and tumor infiltration in the bone (5).

Pathology

Grossly, optic nerve meningiomas usually encircle the optic nerve, extend through the optic nerve dura, and infiltrate the orbital tissues. The nerve often undergoes atrophy from compression, but the parenchyma is rarely invaded by the neoplasm. Although the orbital fat and extraocular muscles are often infiltrated by the tumor, invasion through the sclera into the globe is unusual (11).

Microscopically, optic nerve meningiomas show a spectrum of histologic features similar to intracranial meningiomas; however, meningothelial meningiomas are the type most commonly encountered in the orbit (11). These neoplasms are composed of polygonal cells with indistinct cytoplasmic margins and relatively small ovoid nuclei that often contain pseudoinclusions. These are small vacuoles that are thought to result from intranuclear herniation of cytoplasm. Mitotic figures are rarely seen. Transitional and fibroblastic meningiomas are encountered much less frequently in the orbit.

Differential Diagnosis

Several neoplasms are encountered along the ONS complex. These include optic nerve glioma, schwannoma, neurofibroma, hemangioblastoma, hemangiopericytoma, reticuloendothelial sarcoma, metastasis, and leukemia. The most common tumor that involves the ONS and requires differentiation from the ONS meningioma is the optic nerve glioma. These neoplasms can also involve both orbits and may be associated with neurofibromatosis type 1 in 10% to 50% of cases (12). On imaging studies, optic nerve gliomas show fusiform enlargement of the optic nerve, with normal surrounding dura. A kinked, tubular shape may be observed. These tumors show decreased or intermediate signal intensity on T1-weighted MR images and increased signal intensity on T2-weighted MR images and exhibit variable contrast enhancement (7, 8). They are not associated with calcification or perioptic cysts. The ONS may be thickened because of arachnoid hyperplasia, but does not enhance (8). Optic nerve gliomas can enlarge the optic canal, but typically do not cause hyperostosis.

A number of nonneoplastic lesions can cause enlargement of the ONS, including optic neuritis, orbital pseudotumor, toxoplasmosis, tuberculosis, sarcoidosis, syphilis, juvenile xanthogranuloma, arachnoid cyst, and hematoma (1, 4). These lesions tend to be unilateral and can be differentiated from ONS meningiomas by their clinical and imaging presentation.

Treatment

There is considerable controversy regarding the treatment of ONS meningiomas. The natural course of these tumors consists of slow progression. Visual loss may progress without accompanying changes on imaging examinations (1). The major goals of therapeutic intervention are to restore or preserve vision and to prevent intracranial spread and involvement of the opposite eye. Surgery is indicated for large extensive neoplasms, for cosmetically disfiguring proptosis, and for the prevention of intracranial or transchiasmal spread (2, 3). Transorbital and/or transcranial approaches have been used with surgical complication rates of 30% and 9%, respectively (1, 2). Enucleation is indicated for severe proptosis; exenteration may be performed for extensive lesions.

Complete removal of intraorbital tumor and the optic nerve is possible without removal of the eye (3). Surgical resection has met with dismal results with respect to the preservation of vision. It has proved difficult to preserve optic nerve function with surgery because meningiomas involve the pial blood supply of the optic nerve. Therefore, attempts at removing a tumor surrounding the optic nerve carry a high risk of causing blindness (1). Opening the dural sheath to decompress the optic nerve offers no benefit (2). The recurrence rate after surgery is high: 22 (25%) of 88 patients experienced a recurrence in one series (2). Surgical intervention is even more debatable when the patient's vision is relatively good (3).

Radiation therapy has been used in the treatment of ONS meningiomas with some response reported at a dose of 4500 to 5000 cGy (1). Radiation therapy may help to preserve visual function. Improvements in vision after this treatment may not be accompanied by changes on imaging evaluations (13).

Prognosis

Important prognostic factors of ONS meningiomas include the location of the neoplasm, the size and manner of growth, and the proximity to important structures (3). The morbidity and mortality of orbital meningiomas is related to the progressive involvement of more and more intracranial structures. Primary orbital meningiomas carry a better prognosis overall than secondary orbital meningiomas because the former are more accessible, making complete surgical removal feasible (3).

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