CASE REPORT

Primary Intraorbital Ectopic Meningioma

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ABSTRACT—We report a case of intraorbital meningioma. Operative findings and histopathological examination revealed the tumor to be meningothelial meningioma and to be located entirely outside the optic dura. This case demonstrates the occurrence of primary intraorbital ectopic meningioma, and the tumor was removed through a modified Dolenc approach. The primary intraorbital ectopic meningioma is discussed and the surgical approach to the orbital apex region is reviewed.

Primary intraorbital meningioma arising from the optic nerve sheath is rare and is estimated to account for 5 to 10% of expanding lesions of the orbit.[1–6] Primary intraorbital meningiomas originating from structures other than the optic nerve sheath have been termed primary intraorbital ectopic meningioma.[7–11] The incidence of this tumor is extremely low and its existence has remained a matter of controversy.[2,5] We have experienced a case of intraorbital ectopic meningioma that was confirmed by the histopathological examination of the tumor removed in en bloc, including the optic nerve overlying its dorsal surface.

We report a case of meningioma located within the muscle cone and in the orbital apex inferomedial to the optic nerve; the surgical approach to the orbital apex region is discussed.

CASE STUDY

A 71-year-old man noticed a progressive visual disturbance of the right eye about 6 months before his visit to the outpatient clinic of the Department of Ophthalmology of our hospital on June 23, 1997. Ophthalmological examinations revealed a pale optic disc and severely impaired visual acuity in the right eye with ability to count fingers 20 cm in front of the right eye. Computed tomography (CT) and magnetic resonance imaging (MRI) revealed a tumor in the right orbital apex inferomedial to the optic nerve. The patient refused surgery because of his age. He lost vision in his right eye in December, and repeated MRI revealed that the tumor had grown larger. The patient was referred to

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our department for surgical removal of the tumor and was admitted to the hospital on February 2, 1998.

Neurological and Neuro-ophthalmological Findings

Neurological and neuro-ophthalmological examination revealed mild exophthalamos of the right eye (right eye 20 mm and left eye 15 mm on a Hertel exophthalmometer), a pale optic disc, and atrophy of the optic nerve on funduscopy, no direct light reflex in the right eye, and moderately disturbed lateral gaze of the right eyeball. MRI revealed a 1.5 × 2 cm spherical tumor in the apex inferomedial to the optic nerve, located within the muscle cone and not invading into the optic canal (Figs. 1 and 2). Bone density axial and coronal CT imaging revealed no hyperostosis or erosion of the optic canal or orbital wall. Angiography revealed no tumor staining via the ophthalmic and ethmoidal arteries. The results of these neuro-ophthalmological and imaging studies suggested a diagnosis of intraorbital meningioma.

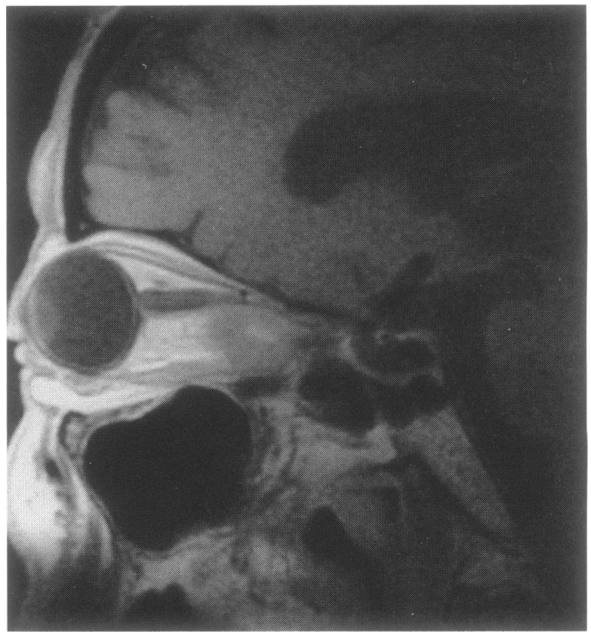

Figure 2. Gadolinium-enhanced, T1-weighted sagittal image. The tumor was located inferomedial to the optic nerve, and the nerve was compressed upward.

Operation

The tumor was removed through a modified Dolenc approach on February 23, 1998. Osteoplastic craniotomy was performed with removal of the frontotemporal bone as well as the supraorbital limb with the bone flap en bloc. The roof and lateral orbital wall were removed with a rongeur to expose the orbital apex and the superior orbital fissure. The anterior clinoid process was drilled off, and the dura propia of the optic nerve and the distal ring of the internal carotid artery were exposed. The periorbital sheath and the annulus of Zinn were incised, and the tumor was approached by separating the muscles between the superior and the medial rectus muscles. The tumor did not involve the optic canal and was located in the apex inferomedial to the optic nerve, where the optic nerve was compressed and forced upward (Fig. 3). The tumor was removed in en

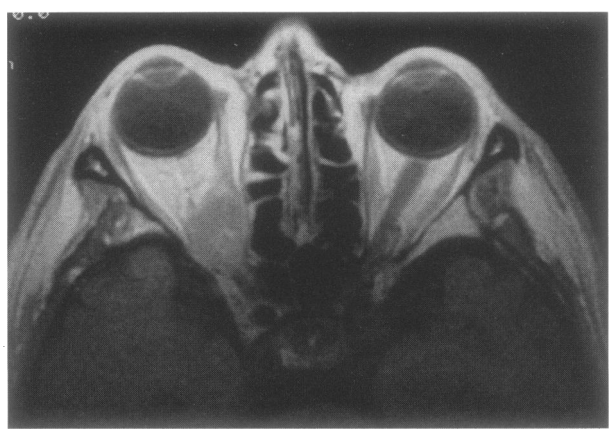

Figure 1. Gadolinium-enhanced, T1-weighted axial image. A tumor located in the right orbital apex and within the muscle cone that has not extended into the optic canal.

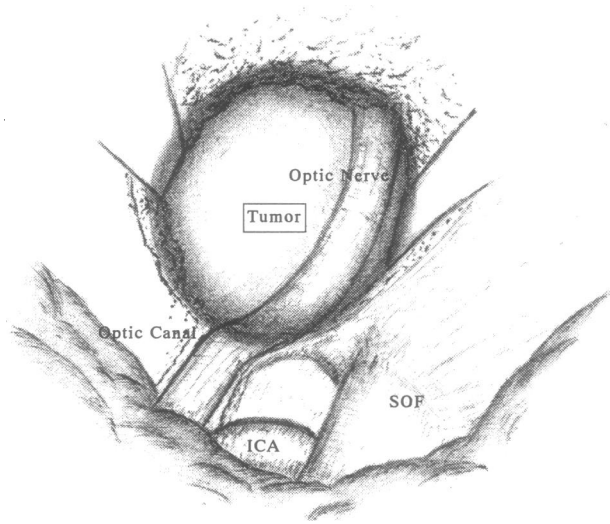

Figure 3. Schematic drawing of the intraoperative findings. A modified Dolenc approach provided a good operative field and identification of the anatomical relations between the tumor and important orbital structures of the optic nerve and superior orbital fissure.

bloc with the optic nerve overlying its dorsal surface. The operation revealed that the tumor was located infero-medial to the optic nerve with no apparent involvement of the optic nerve dura.

Histopathological examination with high magnification (×66) revealed tumor cells with whorl formation, findings compatible with meningothelial meningioma (Fig. 4). Low magnification demonstrated that the subarachnoidal space and dura of the optic nerve contained no tumor cells, and that the tumor was entirely located outside of the optic dura (Fig. 5).

DISCUSSION

Most intraorbital meningiomas are secondary, that is, they extend into the orbit from an intracranial site of origin, while primary intraorbital meningiomas are rare, with an incidence of 5 to 10%.[1-6] The majority of primary intraorbital meningiomas arise from the optic nerve sheath because arachnoidal cells are present in the optic nerve sheath.[1-3,56] Although no arachnoidal cells have been found outside the dural sheath of the optic nerve in the orbit, 13 cases of meningiomas with no connections to the optic nerve sheath and the intracranial meninges have been reported and have been referred to as "ectopic" or "extradural" meningioma of the orbit.[7-11] The three ectopic meningiomas of the fibroblastic type were considered extradural, and to have possibly originated from the periorbita because the periorbita is composed of fibrous tissue.[7-11] The remaining 10 cases were of the meningothelial type, and the origin of these tumors remains undeterminated. Although these tumors are speculated to have originated from arachnoidal cell "rests" in the periorbita or within the muscle and from ectopic meningeal tissue that was pinched off within the orbit during embryonal life,[7,8,10] not all authors are convinced of their existence.[2,5]

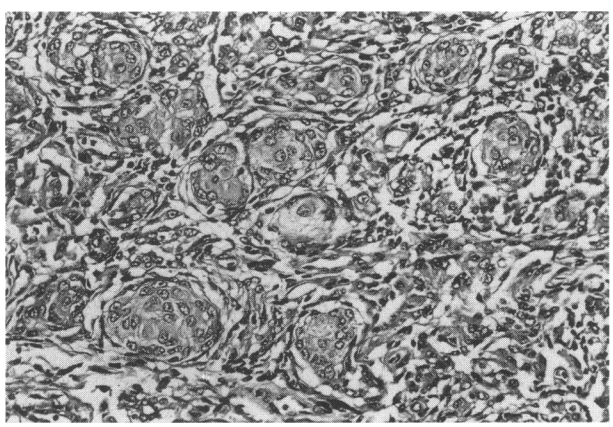

Figure 4. Higher magnification showing typical tumor cells and whorl formation compatible with meningothelial meningioma. (Hematoxylin and eosin ×66.)

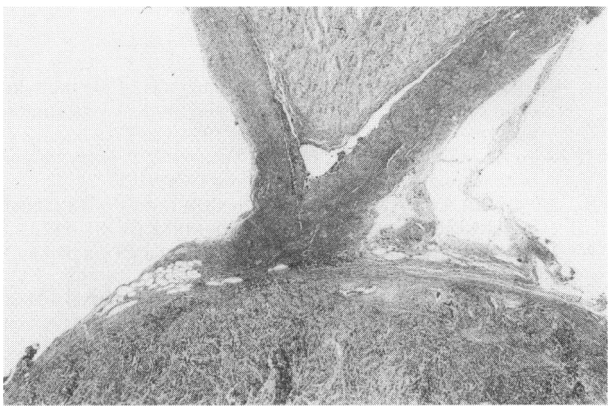

Figure 5. Low magnification showing the optic nerve, its dura, and the tumor. The tumor has no apparent connections to the dura or the optic nerve sheath. (Hematoxylin and eosin ×66.)

All cases were pre-MRI era, and their anatomical localization in the orbit was mainly based on the intraoperative findings. Our patient is the first in which ectopic meningioma was seen on MRI that clearly revealed the location of the tumor in the orbit in relation to the optic nerve and the optic canal. Because the patient was blind preoperatively, the tumor was removed in en bloc, including the optic nerve overlying its dorsal surface. The histopathological sections disclosed the anatomical relations between the tumor and the optic nerve sheath more clearly, that is, that the tumor has no direct connection with the subarachnoidal membrane or the dural sheath of the optic nerve. This strongly indicates that the tumor originated from intraorbital structures outside the dural sheath of the optic nerve, that is, that the meningothelial meningioma originated from the ectopic site in the orbit. However, these histopathological examinations do not provide information with regard to the origin of the tumor.

A single-piece craniotomy including the frontal convexity, the roof, and lateral wall of the orbit as well as the frontozygomatic process was reported by Maroon et al,[1,12] and this approach has been most commonly applied to tumors in the orbital apex. Although it allows good access to the orbital apex through the frontal base, it does not provide a sufficient operative field to expose the optic canal and the superior orbital fissure. We applied a modified Dolenc approach to remove this tumor because it allows postero-lateral access to the orbital apex and good identification of the optic canal and the superiororbital fissure.[13,14] By removing the anterior clinoid process and the lateral wall of the orbit as well as the roof, the optic canal dura and the superior orbital fissure were easily exposed. As shown in our patient, this approach allows easy tumor removal with better identification of the anatomical relations between the tumor and the optic nerve and easy suture of the transected annulus of Zinn. We believe that all tumors in the orbital apex can be best manipulated by this approach.

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