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Clinical pathologic reviews

Ectopic orbital meningioma: report of two cases and literature review

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ABSTRACT

Ectopic orbital meningioma is a rare tumor usually affecting the medial orbit. We present two cases that occurred in a 56-year-old woman and a 28-year-old man. The tumors in both patients were subtotally excised via orbitotomy surgery and were located in the superior quadrant in one of our patients and in the temporal quadrant in the other. Following histopathologic diagnosis, external beam radiotherapy (EBRT) was administered to one patient and intensity modulated radiotherapy to the other. We identified 12 other well-documented cases of ectopic orbital meningioma previously reported. Ectopic meningioma should be considered in the differential diagnosis of medial as well as lateral and superior orbital tumors. The tumor is usually well circumscribed but can be ill defined in imaging studies. There are intralesional calcifications and sclerosis of adjacent bone in some cases. Ectopic orbital meningioma can recur after incomplete excision. Based on the efficacy of EBRT in optic nerve sheath meningioma, we used this treatment to decrease the risk of recurrence in our two patients and found no tumor recurrence at follow-ups of 24 and 74 months, but one patient had severe vision loss from radiation retinopathy.

Orbital meningiomas originating from the arachnoid around the optic nerve are known as primary optic nerve sheath meningiomas (ONSMs). Intracranial meningiomas originating from the sphenoid wing can also invade the orbit and are considered to be secondary orbital meningiomas. A third group of orbital meningiomas occur apart from the optic nerve without any demonstrable connection with the intracranial meninges and are classified as ectopic meningiomas.[1,2,4,6,7,9,10,12,13] We report two rare cases of ectopic orbital meningioma and review the literature.

1. Case reports

1.1. *Case 1*

A 56-year-old woman presented with 3 months of slowly progressive proptosis and eyelid swelling. She had no signs of neurofibromatosis. Best corrected visual acuity was 20/20 in both eyes. There was 5 mm of proptosis OD by Hertel exophthalmometry (Fig. 1A). There was no relative afferent pupillary defect. Intraocular pressure was 16 mm Hg in both eyes. Anterior segment and fundus examinations were

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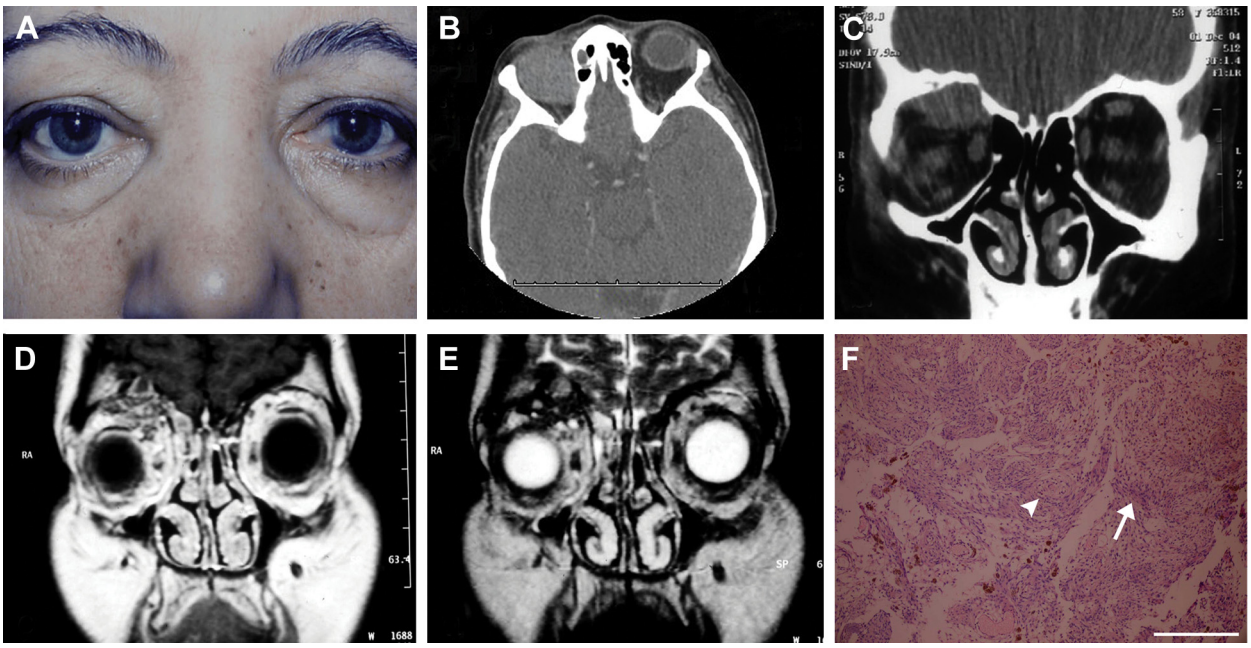

Fig. 1 – *A:* Facial photograph demonstrates right proptosis in a 56-year-old woman. *B:* Axial CT shows a superiorly located orbital mass on the right side. *C:* Coronal CT shows a superiorly located orbital mass producing thinning of the overlying bone on the right side. *D:* T1W contrast enhanced orbital MRI reveals an ill-defined in the right superior orbit with marked contrast enhancement. *E:* The tumor is hypointense with respect to orbital fat and cerebral gray matter on T2 weighted images. This hypointensity on T1W images is due to the accumulation of blood breakdown products in the tumor. *F:* Histopathologic examination shows that the tumor consists of round to oval cells with indistinct cytoplasmic borders and vesicular nuclei. Mitosis or prominent nucleoli are not observed. The tumor cells seemed to form a predominantly meningothelial (snycytical) pattern (*arrow*) and cellular arrangement reminiscent of whorling pattern (*arrowhead*). The histopathologic findings were consistent with the diagnosis of orbital meningothelial meningioma. (Hematoxylin and eosin ×200; scale bar: 250 μm.)

unremarkable. Orbital computed tomography revealed a superiorly located mass producing thinning of the overlying bone (Fig. 1B,C). Orbital magnetic resonance imaging (MRI) showed an ill-defined mass in the right superior orbit. The tumor was isointense with respect to the orbital fat and cerebral gray matter on T1-weighted images, hypointense on T2-weighted images, and demonstrated moderate contrast enhancement (Fig. 1D,E).

The patient underwent a superonasal skin incision orbitotomy. Only a subtotal resection could be performed because of the friable nature of the tumor. No connections to optic nerve was detected during surgery but the tumor was in close proximity to the periorbita. On gross examination, it had a gray-yellow color. On histopathology, the tumor consisted of round to oval cells with indistinct cytoplasmic borders and vesicular nuclei. The cells seemed to form a predominantly meningothelial (snycytical) pattern (long arrow) and cellular arrangement reminiscent of whorling pattern (short arrow) (Fig. 1F). Mitosis or prominent nucleoli were not observed. Hemosiderin aggregates were seen within the tumor. Hypointensity on T2-weighted MR images was attributed to the presence of hemosiderin and blood breakdown products. On immunohistochemistry, the tumor stained negative for epithelial membrane antigen (EMA), HMB-45, Melanin A, S-100, glial fibrillary acidic protein, and carcinoembryonic antigen. The diagnosis was ectopic intraorbital meningioma, despite EMA negativity. The patient underwent conventional external beam radiotherapy (EBRT [50 Gy]) to the right orbit. At that time, she did not have access to stereotactic fractionated radiotherapy or a more precise approach such as cyberknife radiosurgery. She was seen postoperatively at irregular intervals. Four years after diagnosis, the right eye had radiation maculopathy and proliferative radiation retinopathy and underwent one intravitreal triamcinolone acetonide injection and three sessions of laser photocoagulation over a 10-month period. At 74 months follow-up, there was no tumor recurrence, and visual acuity OD was counting fingers at 2 meters.

1.2. *Case 2*

A 27-year-old man presented with slowly progressive proptosis of the left eye over 6 months (Fig. 2A). He did not have neurofibromatosis. Visual acuity was 20/20 in both eyes. There was 12 mm of proptosis on the left side. The motility of the left eye was limited on elevation and abduction. Conjunctival edema and injection were present over the lateral rectus muscle insertion in the left eye. Both fundi were normal. Orbital MRI showed a well-defined tumor laterally in the orbit. The tumor was isointense on T1-weighted images, hyperintense on T2-weighted images, and demonstrated moderate