Orbital melanoma with calcification: A diagnostic dilemma

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Primary orbital melanoma is rare and has varied initial presentation. A 28-year-old female presented with proptosis and decreased vision in the left eye. Computed tomography scan showed an orbital mass with contrast enhancement and calcification around the optic nerve leading to a diagnosis of meningioma. The patient chose to be on observation. Loss of vision with an increase in proptosis was seen at 6 months follow-up. On surgical exploration, a well-defined pigmented mass was seen encasing the optic nerve. Histopathological analysis revealed a malignant melanoma. Metastatic workup was negative. Left eye lid sparing exenteration was done. A high index of suspicion is necessary in a rapidly growing suspected optic nerve sheath meningioma and a differential diagnosis including orbital melanoma be considered.

Key words: Calcification, meningioma, orbital melanoma

Primary orbital melanoma (POM) remains an enigma, with variable clinical and radiological features.

We report a case of POM, which presented with radiological evidence of calcification, leading to an erroneous diagnosis of an optic nerve sheath meningioma.

Case Report

A 28-year-old female presented with a 6 months history of painless, progressive diminution of vision in the left eye. On examination, her best-corrected visual activity was 6/6 in the right eye and hand movement perception in the left eye. There were no signs of ocular melanocytosis or oculodermal melanocytosis. The left eye examination showed axial proptosis of 2 mm, with relative afferent pupillary defect. Anterior segment examination was unremarkable, with no conjunctival or scleral pigmentation. Fundus examination showed a pallid disc edema with choroidal folds. Opto-ciliary shunt vessels were absent. No pigmented lesion was noted in the fundus. There was no restriction of extraocular movements. The right eye examination was normal. Computed tomography (CT) scan showed evidence of a hyperdense lesion with calcification and brilliant enhancement with contrast. The lesion was involving the retrobulbar, intraconal compartment and encircling the optic nerve, but not extending to the orbital apex [Fig. 1]. A preliminary diagnosis of optic nerve sheath meningioma was made based on the radiological appearance. The patient was counseled as regards the management. The patient refused surgery for histopathological confirmation and was willing for regular follow-up. She presented 6 months later, with an increase in the left eye proptosis (an increase of 6 mm of proptosis), no light perception in the left eye and a left afferent pupillary defect [Fig. 2]. Anterior segment examination was normal. On fundus examination, there was disc pallor. No pigmented lesion was noted on fundus examination. Restriction of extraocular movements was present in all gazes. Given the rapid progression of the lesion, that is uncharacteristic of a typical meningioma and the absence of other signs of meningioma (absent optociliary shunt vessels, no tram track sign on imaging) the preliminary diagnosis was doubted and surgical exploration was planned. A well-defined, pigmented mass encasing the optic nerve was seen intraoperatively. Histopathologically, biopsy of the mass was interpreted as malignant melanoma, showing epithelioid cells arranged in sheets with marked pigmentation. A metastatic workup including a thorough dermatological assessment, CT head, chest X-ray, abdominal ultrasound and liver function test was negative. Ultrasound examination of the right eye did not show any uveal or choroidal mass. Lid sparing exenteration was done, histopathology of which was confirmatory of an orbital melanoma with no involvement of the optic nerve head or juxtapapillary choroid, infiltrating the sclera, and the extraocular muscles. Oval to spindle shaped cells with prominent nucleoli and marked cytoplasmic pigmentation were seen arranged in fascicles and nests with interspersed mitosis [Figs. 3 and 4]. Serial sectioning did not show any involvement of the uvea or choroid. Postoperatively, the patient was doing well with a healthy socket.

She presented 20 months later with a pigmented growth on the left lateral orbital rim. CT scan showed heterogeneously enhancing mass extending into the left infratemporal fossa measuring 2.3 cm × 1.9 cm. Incision biopsy of the mass was positive for recurrence of malignant melanoma. Metastatic

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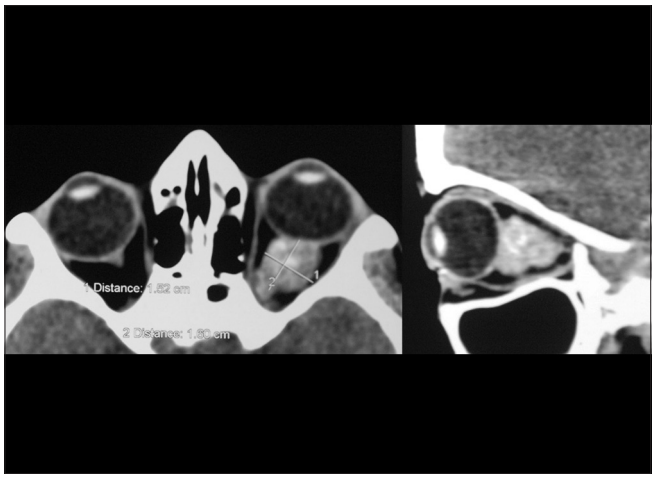

Figure 1: Hyperdense lesion with calcification involving retrobulbar, intraconal compartment and encircling the optic nerve in left orbit on axial and sagittal computed tomography scan

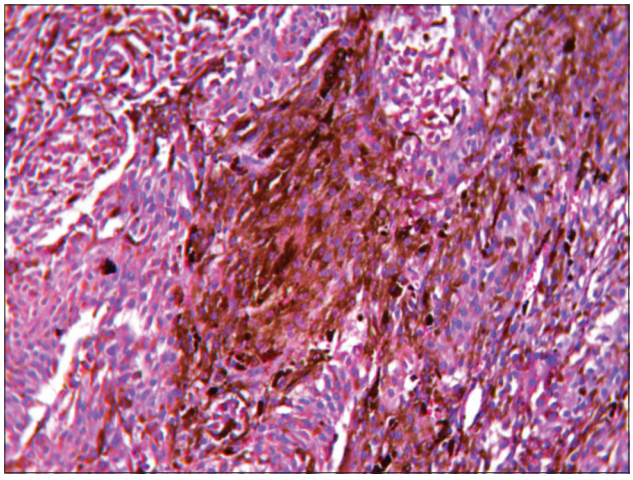

Figure 3: Sheet of oval to spindle pigmented cells with pleomorphism, interspersed necrosis and angiogenesis (H and E, ×200)

workup was negative for any distant focus of melanoma. The patient was treated with local radiotherapy followed by chemotherapy (dacarbazine 300 mg with cisplatin 70 mg for six cycles). One year postchemotherapy, patient presented with a headache. CT scan showed intracranial metastasis. Patient declined any further treatment and expired after 1 month.

Discussion

Primary melanomas may rarely arise in the orbit, where melanocytes are usually absent. They are thought to represent metastasis from an unknown primary or to arise from ectopic melanocytes (nevus of ota or blue nevus). If there is no evidence of ocular melanocytosis, and no demonstrable primary melanoma elsewhere, the lesion can be classified as a presumed de novo POM.[1]

The usual presentation of POM is proptosis. However, it may present as orbital varix,[2] orbital inflammatory pseudotumor,[3] eyelash poliosis,[4] meningioma,[5] or an orbital vascular anomaly.[6]

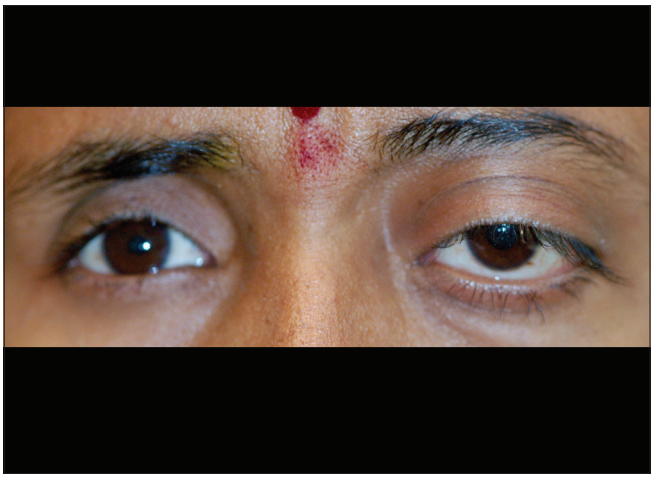

Figure 2: Left eye proptosis with left afferent pupillary defect

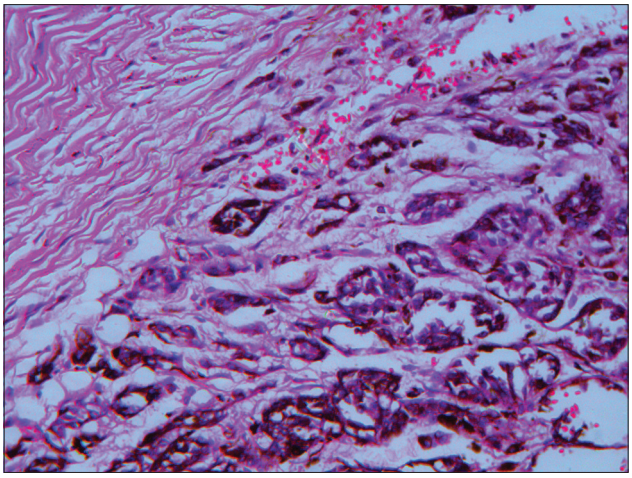

Figure 4: Irregular islands of infiltrating melanoma cells. Vague pseudo glandular pattern also seen (H and E, ×200)

Although calcification is encountered in cases of intraocular melanoma postdestructive therapeutic interventions,[7] spontaneous calcification is very rare. Only three cases of spontaneous calcification in intraocular melanoma have been reported in the literature.[8]

Spontaneous calcification in an orbital melanoma is rarer still with only one case being reported in the literature by Froula *et al*.[9]

In our case, there was no clinical or histopathological evidence of any melanocytoma, choroidal melanoma, nevus of ota or blue nevus. The CT appearance showed calcification in a mass surrounding the optic nerve, which led to the radiological diagnosis of a meningioma. With this presentation, treatment was delayed as the patient refused surgery on learning about the slow growing nature of the suspected diagnosis (optic nerve sheath meningioma).

The patient presented with a local recurrence 20 months following surgery. No distant focus of melanoma evolved during this duration, giving credence to the de novo origin of the orbital melanoma. To conclude, orbital melanoma although rare should be considered as a differential diagnosis in an orbital mass presenting with calcification. A suspected optic

nerve sheath meningioma with a rapid growth phase should be viewed with suspicion and an alternative diagnosis may be considered.

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Conflicts of interest

There are no conflicts of interest.

References

1. Shields JA, Shields CL. Orbital malignant melanoma: The 2002 Sean B. Murphy lecture. Ophthal Plast Reconstr Surg 2003;19:262-9.
2. Löffler KU, Witschel H. Primary malignant melanoma of the orbit arising in a cellular blue naevus. Br J Ophthalmol 1989;73:388-93.
3. Shields JA, Shields CL, Eagle RC Jr., De Potter P, Oliver GL. Necrotic orbital melanoma arising *de novo*. Br J Ophthalmol 1993;77:187-9.
4. Alsuhaibani AH, Alhumayed M. Primary orbital melanoma with poliosis and a palpable mass. Arch Ophthalmol 2011;129:1382-3.
5. Mohindra S, Mohindra S, Dhawan A, Saikia U, Mankoo RS. Primary malignant melanoma of the optic nerve sheath: A case report. Surg Neurol 2009;71:372-5.
6. Lee V, Sandy C, Rose GE, Moseley IM, Cree I, Hungerford JL. Primary orbital melanoma masquerading as vascular anomalies. Eye (Lond) 2002;16:16-20.
7. Kiratli H, Bilgiç S. Calcification in choroidal melanoma after transpupillary thermotherapy. Am J Ophthalmol 2001;132:939-41.
8. Csákány B, Tóth J. Spontaneous calcification of a choroidal melanoma. Acta Ophthalmol 2009;87:575-6.
9. Froula PD, Bartley GB, Garrity JA, Forbes G. The differential diagnosis of orbital calcification as detected on computed tomographic scans. Mayo Clin Proc 1993;68:256-61.