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Case report

Orbital melanocytoma: When a tumor becomes a relieving surprise

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ABSTRACT

Purpose: Melanocytomas are rare pigmented tumors that arise form melanocytes and have been reported in the central nervous system. Orbital melanocytomas "also known as blue nevus" are rarely reported. The occurrence of choroidal melanoma and orbital melanocytomas has never been described.

Observations: This is a case of orbital melanocytoma in a 34 year old female who presented with left proptosis and ecchymosis. She has the right eye enucleated to treat a large choroidal melanoma, 6 years earlier. Orbital metastasis was suspected. After orbital imaging and systemic evaluation, incisional biopsy was planned yet the mass could be totally excised and it turned out to be melanocytoma. The condition was not associated with nevus of Ota and the patient is not known to have any predisposing condition for melanocytic lesions.

Conclusion and importance: Melanocytoma and malignant melanoma share the same cell of origin. The benign course, the well differentiated cells, absence of anaplasia and the positive reaction to Human Melanoma Black-45 (HMB-45) and S-100 proteins established the diagnosis of the former. Such diagnosis was a relief for this one eyed patient.

(HMB-45:human melanoma black-45).

1. Introduction

Diffuse melanocytosis and neurocutaneous melanosis, melanocytoma and malignant melanoma represent a spectrum of lesions that originate from melanocytes.[1] (see Table 1)

Melanocytomas are rare pigmented primary tumors. They are usually discrete, solitary well differentiated and show slow growth yet there is a high probability of recurrence.[2,3] Local and systemic dissemination as well as malignant transformation are rarely reported.[4]

Orbital malignant melanomas as well as melanocytomas have been reported. Primary malignant melanomas constituted less than 1% of orbital tumors[5,6] while only 7 cases of orbital melanocytomas were described to date.[1,3,4,6–9]

We report the eighth case of orbital melanocytoma and the first case associated with a contralateral previously treated choroidal malignant melanoma.

2. Case report

A thirty four year old female presented in 2016 with left proptosis of 6 months duration and recent ecchymosis and extensive subconjunctival hemorrhage following sudden rise of blood pressure (Fig. 1a). Further clinical evaluation revealed intact ocular motility, best corrected visual acuity (BCVA) was 0.9, normal anterior and posterior segments examination with no signs of optic nerve involvement.

The right orbit was anophthalmic following enucleation (by Nasr. HE) in 2010 for a large intraocular choroidal mass measuring 22 × 22 × 14 mm. The mass showed acoustic criteria of malignant melanoma with secondary exudative retinal detachment and no extraocular extension. This was further confirmed by histopathological studies and metastatic work up. By that time, examination of the Left eye and orbit was unremarkable. The patient received no adjuvant treatment and remained free from local recurrence and systemic spread for 2 years before she dropped her follow ups.

On her recent presentation, orbital computed tomography (CT) showed a well-defined isodense intraconal lesion measuring 2.6 × 2.5 × 2.4 mm with intrinsic areas of high density. The lesion was located infero-medially between the optic nerve and the inferior rectus pushing the former supero-laterally and the latter inferiorly as well as scalloping the lamina papyracia. The right orbit showed no signs of local recurrence.

Magnetic resonance imaging (MRI) confirmed the mass location and showed it to be hyper-intense in T1-weighted images (Fig. 1b&c) that became hypointense in T2-weighted images (Fig. 1d). General examination and metastatic work-up showed no evidence of metastases.

Incisional biopsy was planned anterior inferomedial

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Table 1
Summary of the clinical findings, associations, treatment modalities and follow up periods of all the reported isolated orbital melanocytomas in comparison to the currently reported case.

Case	De Tella[1] 2003	Mathai[7] 2008	Tsugu[6] 2009	Sato[8] 2009	Ortiz[9] 2013	Tregango[3] 2014	Placilli[4] 2016	Current case
Age	35years	40years	51years	49years	68years	28years	26 years	34 years
Gender	M	M	M	M	M	M	M	F
Side	Rt	Rt	Rt	Rt	Lt	Rt	Rt	Lt
Primary presentation	Proptosis	Proptosis	Proptosis Diplopia	Diplopia	Proptosis	Proptosis	Proptosis Diplopia	Proptosis Ecchymosis
Location	Intraconal, around ON	Intraconal Superior	Intraconal	Intraconal Apex	Intraconal Apex	Rxtraconal	Intraconal around ON	Intaconal Infero-medial
Intracranial extension	Yes	Yes						
Associations		Recurrence after 17 y of previous removal				Ipsilateral Nevus of Ota	Lt cavernous hemangioma removed 13 y earlier	Rt Choroidal melanoma/ enucleation 6 years earlier
Treatment	Subtotal resection Radiotherapy Chemotherapy	Resection	Resection	Subtotal resection Chemotherapy	Resection	Resection	Resection	Resection
Follow up	5 y	6 m	15 m	12 y	3 y	7 m	3 years	1 y

M, male; F, female; y, year; m, month; Rt, right; Lt, left, 1st; first.

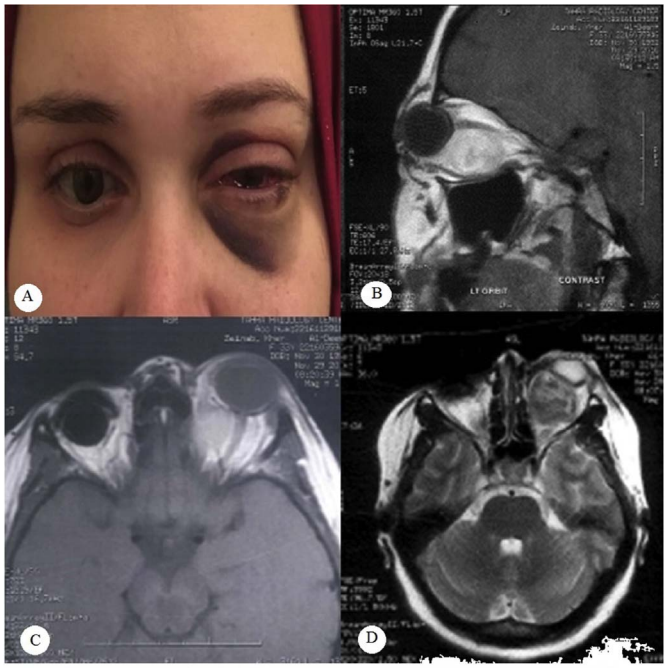

Fig. 1. Clinical appearance and MRI images of the patient.
a-Clinical appearance at presentation showing Lt proptosis, ecchymosis and subconjunctival hemorrhage. The Rt eye is fitted with ocular prosthesis.
b- T1 weighted sagittal MRI image showing a hyperintense intraconal well circumscribed mass that lies inferiorly and surrounds the optic nerve with no clear line of separation.
c- T1 weighted axial image showing the masd pushing it temporally with no evidence of orbital apex involvement. The Rt orbit is anopthalmic with implant.
d- T2 weighted axial image showing the hypointense appearance of the previously described mass.
(Rt:right,Lt:left,MRI:magnetic resonance imaging).

transconjunctival orbitotomy with medial rectus muscle disinsertion by (Nasr HE). Intraoperatively, a dark brown round lesion was identified. It was surrounded by a thin capsule of fibrous tissue and had no firm adhesions to the optic nerve or the surrounding structures. Hence, the surgeon was able to deliver it as one intact mass measuring 2.5 × 2.5 × 2.7mm. The patient had an uneventful postoperative course with preserved preoperative vision and ocular motility.

Histopathological examination revealed a heavily pigmented lesion that was composed of polygonal cells sheets with few scattered lymphocytes, blood vessels and extravasated red blood cells (Fig. 2a–f). The cells had obscured cytological details due to heavily packed cytoplasm with melanin pigments yet they did not show anaplastic features (Fig. 2a–f). Immunohistochemistry revealed positive reaction for protein S-100 (Fig. 2a–f) and HMB-45monoclonal antibodies (Fig. 2a–f). Based on the previous findings, the diagnosis of melanocytoma was confirmed. The patient did not receive any adjuvant treatment and she is still tumor free after one year of follow up.

3. Discussion

Melanocytomas are borderline tumors between cellular blue nevus and spindle melanomas.[1,7] They originate from melanocytes that are derived from the neural crest.[3] They are usually well-differentiated with a benign nature, therefore, they show no tissue invasion and they present as space occupying lesion with corresponding signs and symptoms according to their site.[5,6,10]

In literature, around 110 cases of meningeal melanocytomas have been reported with female predilection especially in the 5th decade of life.[1,6,7] They were reported in masses related to leptomeninges where melanocytes show high concentration.[1,7]

Intraocular melanocytomas have been described in all parts of the uveal tract, sclera and conjunctiva yet few reports are available in literature about orbital presentation.[4] Our report is the first to describe the occurrence of orbital melanocytoma and malignant melanoma in the same patient.

During the patient's preliminary evaluation metastases was the primary presumption due to the history of enucleation done 6 years earlier to treat large choroidal melanoma. Systemic evaluation revealed no evidence of distant metastasis. We came to the conclusion that the presenting isolated intraconal orbital mass still could be a solitary metastasis or a new lesion.

Malignant melanoma has variable clinical and histological appearance, hence it is difficult to clinically differentiate it from melanocytoma as well as other neuroectodermal tumors with melanin content such as melanotic meningioma, schwannoma and neurofibroma. All of them should be considered in the differential diagnosis.[2,7]

Metastases from choroidal melanoma to the contralateral orbit are rarely reported.[11] Although, orbital metastases have been described following systemic spread from choroidal melanoma,[12] there are few reports about orbital secondary lesions to be the first sign of distant spread.[11,13,14]

Radiological studies can be of use in diagnosing melanin-containing lesions but they are not conclusive. Computed Tomography (CT) scans show melanocytic tumors as well defined isodense to slightly