



Case Report

Primary malignant melanoma of orbit

J J Kuli¹, Bikram Dam^{1,*}, Sunanda Nandi¹

¹Dept. of Ophthalmology, Assam Medical College & Hospital, Dibrugarh, Assam, India



ARTICLE INFO

Article history:

Received 21-04-2020

Accepted 24-04-2020

Available online 30-06-2020

Keywords:

Malignant melanoma

Eye neoplasia

Lid exenteration

Pigmentation.

ABSTRACT

Primary malignant melanoma may rarely arise in the orbit, where melanocytes are usually absent. A 43- year old man presented with painful, profuse, unilateral, eccentric proptosis for 1 year. Computed tomography scan showed intra-ocular mass in posterior chamber of right globe with coarse calcification and contiguous retrobulbar mass. The patient gave consent for surgical exploration where a well-defined pigmented mass was seen encasing the optic nerve. Histopathological workup revealed it to be malignant melanoma. Metastatic workup came out to be negative. Right eye lid sparing exenteration was done. Hence in patients presenting with massive unilateral proptosis, primary malignant melanoma though extremely rare should be one of the suspects.

© 2020 Published by Innovative Publication. This is an open access article under the CC BY-NC license (<https://creativecommons.org/licenses/by-nc/4.0/>)

1. Introduction

Ocular melanoma is a rare type of tumor and the exact cause is unknown. Most malignant melanomas in the orbit are secondary tumors spreading from an intraocular choroidal primary site or from the conjunctiva. Primary melanomas are extremely rare but can arise from a cellular blue nevus or in the setting of oculodermal melanocytosis. Occasionally malignant melanoma may metastasize to orbital structures from distant sites. In advanced cases proptosis, ptosis, chemosis, ophthalmoplegia and orbital pain may be seen.¹ The primary orbital melanoma represents less than 1% of the primary orbital neoplasia.²

2. Case Report

A 43-year-old man presented to out-patient -department with a history of gradual progressive bulging of the right eye for 1 year associated with a 7-day history of periorbital pain, swelling associated with proptosis of the same eye. The proptosis was significant and restricting the right eye movement in all directions. There was no history of any thyroid disorder or trauma to the right eye.



Fig. 1: Patient presenting with unilateral proptosis

On physical examination, the right eyeball was deviated downward and laterally with significant proptosis and restricted right eye movements. The proptosis was non pulsatile without ocular bruit. It measured around 38mm in luedde exophthalmometer as shown in Figure 1. The difference was around 15mm when compared with the left eye. Visual acuity of right eye showed perception of light to be negative. In left eye visual acuity was 6/12. Right eye revealed chemosis with conjunctival injection along with tense upper eyelid. Cornea was hazy with exposure keratopathy which caused non-visualization of both anterior

* Corresponding author.

E-mail address: doctorvikramdam@gmail.com (B. Dam).

and posterior segments.

2.1. Investigations

Routine Blood investigations: TLC, DLC, Platelet Count, Random Blood Sugar, Hb, ESR, LFT all WNL. HbsAg, HIV, HCV- ab Test were all WNL.

USG-B-SCAN of R/E revealed tumour mass protruding into the vitreous and pushing lens and iris forward as shown in Figure 2



Fig. 2: USGb-scan picture

CECT SCAN shows a 3.6 x 3.5 x 2.1 cm mildly enhancing intra-ocular mass in posterior chamber of right globe with coarse calcification and contiguous retrobulbar mass, inseparable superior and lateral rectus muscle, displacing optic nerve towards the left side as shown in Figure 3.

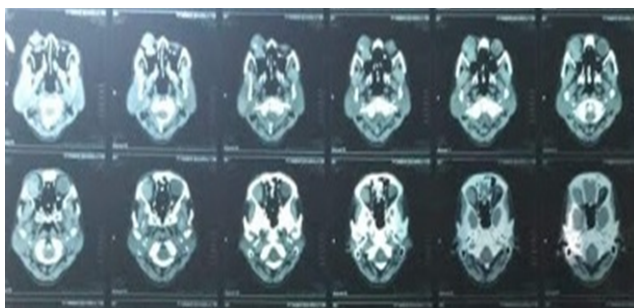


Fig. 3: CTscan picture

Calcification of lens of right globe was also seen.

A surgical exploration was planned and a well-defined pigmented mass encasing the optic nerve was seen intra-operatively. On histo-pathological analysis the biopsy was interpreted as Malignant melanoma, showing epithelioid cells arranged in sheets with marked pigmentation as shown in Figure 4.

A complete metastatic workup was done which included a thorough dermatological assessment, CT scan of Brain,

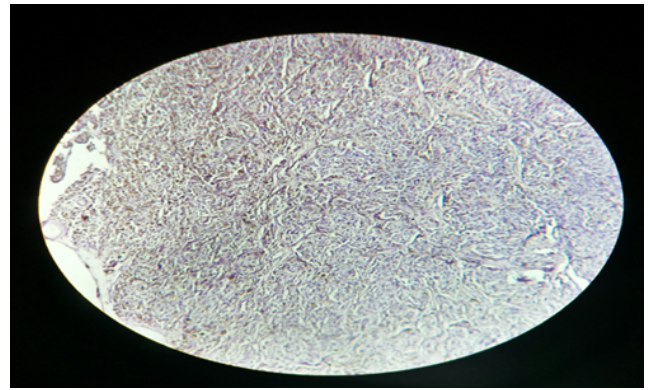


Fig. 4: Histopathological picture

chest X-ray, USG abdomen and found to be negative.

The ultrasound examination of the left eye did not show any uveal or choroidal mass.

We did lid sparing exenteration with adjuvant radiotherapy and the histopathological analysis of whose was confirmatory for orbital melanoma.

Oval to spindle shaped cells with prominent nucleoli and marked cytoplasmic pigmentation were seen arranged in fascicles and nests with interspersed mitosis. Serial sectioning did not show any involvement of uvea or choroid.

The patient was doing well with a healthy socket at the follow-up at 4 months.

3. Discussion

Primary melanoma may rarely arise in the orbit, where melanocytes are usually absent. They are thought to be representing metastasis from an unknown primary or to arise from ectopic melanocytes. If there is no evidence of ocular melanocytosis, and no demonstrable primary melanoma elsewhere, the lesion can be classified as a presumed as de novo primary orbital melanoma.³

The usual presentation of primary orbital melanoma is proptosis. However, it may present as orbital varix,⁴ orbital inflammatory pseudotumor,⁵ eyelash poliosis⁶ meningioma⁷ or an orbital vascular anomaly.⁸

Treatment of choice is based on exenteration, that is complete removal of the orbital content. Radiotherapy and chemotherapy have been used as additional treatment with uncertain results⁹

In our case there was no clinical or histopathological evidence of any melanocyte, choroidal melanoma, nevus of ota or blue nevus. CECT Scan showed coarse calcification and contiguous retrobulbar mass.

The patient presented to us 1-year post development of symptoms and no distant focus of melanoma evolved during this duration giving credence to the denovo origin of the orbital melanoma.

4. Source of Funding

None.

5. Conflicts of Interest

None.

References

1. Rumelt S, Yanoff MO, Dukerjs. Elsevier. 2013.
2. Surakiatchanukul T, Sioufi K, Pointdujour-Lim R, Jr RCE, Shields JA, Shields CL, et al.. Caruncular Oncocytoma Mimicking Malignant Melanoma; 2017.
3. Bains S, Kim U, Shanti R. Orbital melanoma with calcification: A diagnostic dilemma. *Indian J Ophthalmol.* 2016;64:932.
4. Figueira E, Rajak S, McKelvie P, Kalantzis G, Ismail A, Gonzales M, et al. Primary orbital melanoma: a case series and literature review. *Orbit.* 2018;37(5):352–7.
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(3):372–5.
6. Lee V, Sandy C, Rose GE, Moseley IM, Cree I, Hungerford JL. Primary orbital melanoma masquerading as vascular anomalies. *Eye.*

2002;16(1):16–20.

7. Kiratli H, Bilgiç S. Calcification in choroidal melanoma after transpupillary thermotherapy. *Am J Ophthalmol.* 2001;132:939–41.
8. Csakany B, Tóth J. Spontaneous calcification of a choroidal melanoma. *Acta Ophthalmol.* 2009;87(5):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 Proce.* 1993;68(3):256–61.

Author biography

J J Kuli Professor and HOD

Bikram Dam Resident

Sunanda Nandi Resident

Cite this article: Kuli JJ, Dam B, Nandi S. **Primary malignant melanoma of orbit.** *IP Int J Ocul Oncol Oculoplasty* 2020;6(2):136-138.