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Case Report

A rare case of orbital melanoma: chronicle of the eye

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ABSTRACT

Melanoma is a malignancy of melanocytes and can present as a primary malignancy, secondary malignancy due to local extension, or metastatic lesion from a distant primary lesion. A 75 years old male visited the eye OPD and reported experiencing painful loss of vision in his right eye and proptosis for the past 2 months. Visual acuity of right eye was no perception of light. There was abaxial proptosis (inferomedial) of right eye with gross lid edema along with conjunctival chemosis. MRI scan orbit reveals large heterogeneously enhancing soft tissue density lobulated mass of approximate size 4.4×3.4× 2.9 cm noted in right orbit, involving posterior wall of right eyeball, displacing it medially and causing proptosis, with hyperintensese areas within the vitreous chamber of right eyeball. Patient underwent right orbital subtotal exenteration and the specimen sent for histopathological examination revealed malignant melanoma of right orbit involving choroid and infiltrates all structure of globe. Patients who present with imaging evidence of a well-defined tumour and unilateral proptosis should be evaluated for the rare cancer known as primary orbital melanoma. An aggressive and multidisciplinary approach is needed in management of this tumour.

Keywords: Orbital melanoma, Malignant melanoma, Tumour of eye

INTRODUCTION

Melanoma is a malignancy of melanocytes and can present as a primary malignancy, secondary malignancy due to local extension, or metastatic lesion from a distant primary lesion. Melanoma makes up 5-10% of secondary orbital malignancies, but represents only a small fraction of primary orbital neoplasms.[1] The number of reported cases of choroidal melanomas has gone up in recent years, potentially because of increased UV light exposure, or potentially because of better detection.[2] Proptosis is typically painless in patients with primary orbital melanoma; however, there may also be related alterations in visual acuity, diplopia, or discomfort.[3] Due to the disease's rarity, diagnosis takes longer than expected, and treatment calls for a multidisciplinary team that includes ophthalmologists, pathologists, and radiation oncologists. Here we discussed a case of malignant melanoma of right orbit involving sinuses and part of optic nerve.

CASE REPORT

A 75 years old male visited the eye OPD and reported experiencing painful loss of vision in his right eye and proptosis for the past 2 months. The patient does not have diabetes or hypertension and is cachexic. There is no significant family history. The patient underwent uneventful cataract surgery on both eyes 10 years ago.

On examination

Visual acuity of right eye was no perception of light. There was abaxial proptosis (inferomedial) of right eye measuring about 24 mm on Luedde exophthalmometer. There was gross lid edema along with blackish discoloration of periocular skin along with conjunctival chemosis of right eye with dark reddish hue. Cornea was opaque with superficial and deep vascularization and rest

other intraocular structures could not be examined (Figure 1).

MRI scan orbit reveals large heterogeneously enhancing soft tissue density lobulated mass of approximate size 4.4×3.4×2.9 cm noted in right orbit, involving posterior wall of right eyeball, displacing it medially and causing proptosis, with hyperintensese areas within the vitreous chamber of right eyeball. The mass was involving intraconal and extraconal compartments, abutting lacrimal gland laterally (Figure 2). CT orbit shows bony attenuation of floor of right orbit and inner table of right frontal sinus. However no obvious intrasinus extension seen. No gross extension in brain/superior orbital fissure seen in present scan.

Ultrasonography of orbit shows, well defined iso-hypoechoic lesion of approximate size 4.4×3.7 cm, noted in right orbit, replacing whole right eye ball with peripheral vascularity on colour doppler imaging.

The patient was advised to get a PET scan because he couldn't afford one due to his poor income. However, USG abdomen and chest X-ray revealed no obvious extension of metastatic cells to liver and lungs.

Patient underwent right orbital subtotal exenteration and the specimen sent for histopathological examination revealed malignant melanoma of right orbit. The tumour involves choroid and infiltrates all structure of globe. The tumour was solid, dark brownish multinodular mass infiltrating and involving fat and extraocular muscles. The tumour cells show predominant B cell (75%), along with small population of epithelioid cells (20%) and intermediate cells (5%) (Figure 3). Tumour involves optic nerve and showed extraorbital extension.

Patient sent to radiotherapy department for further management. Currently patient is receiving plaque brachytherapy with iodine-125. Till now, the patient is monitored for medication toxicity and local recovery.

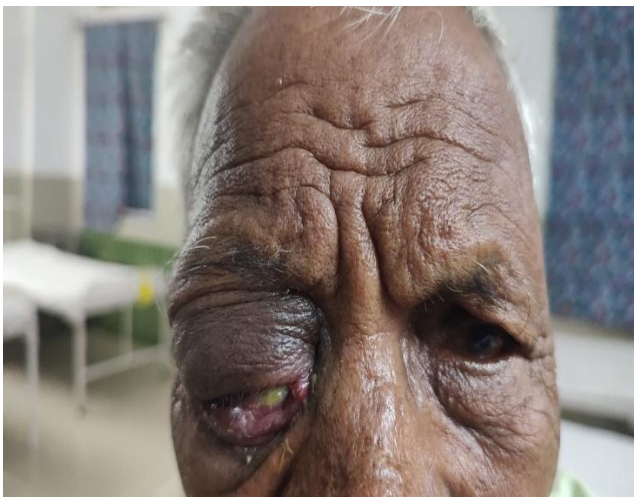

Figure 1: Presentation of patient with proptosis.

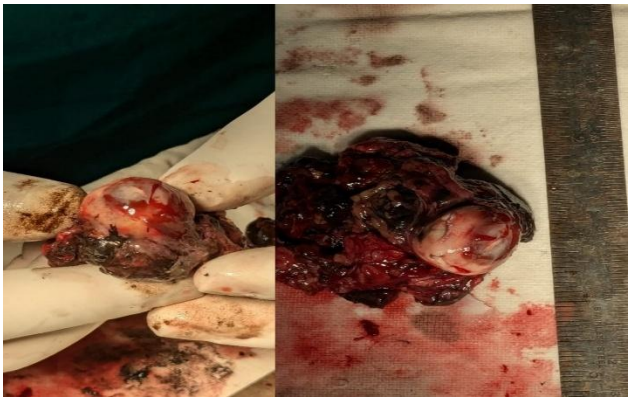

Figure 2: Specimen of right eyeball exenterated with tumour mass.

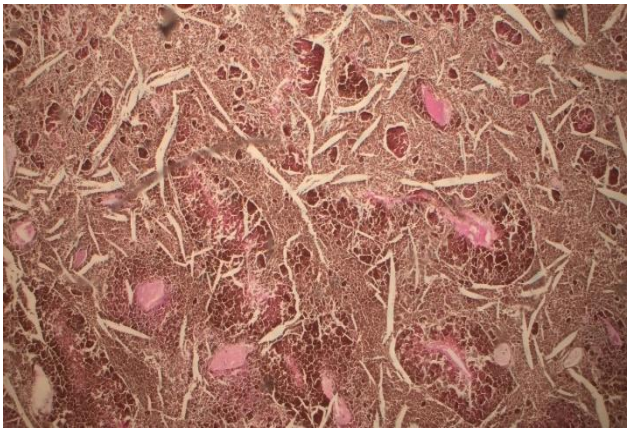

Figure 3: Large sheets of epitheloid cells and some spindle melanoma cells with pale chromatin and large nucleoli seen.

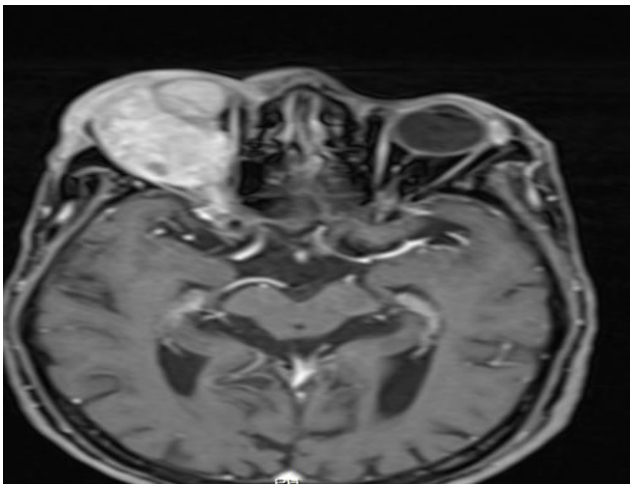

Figure 4: MRI of hetergenous lobulated mass in posterior wall of right orbit displacing globe medially involving optic nerve and surrounding structure.

DISCUSSION

The origin of melanocytes is from the neural crest cells, which also differentiate to form the cranial skeleton.[4] The

majority of the cranial neural crest cells undergo a transformation into ectomesenchyme, which is responsible for forming the head's skeleton, muscles, and dermis. As the remaining neural crest cells migrate through the dermis, they undergo a transformation into melanoblasts and eventually become melanocytes. This suggests that there is a possibility that certain melanoblasts do not detach from the ectomesenchyme during differentiation and subsequently undergo malignant transformation.[5]

The uncommon occurrence of primary orbital melanoma can cause delays in diagnosis and treatment due to the unspecified radiographic features and the lack of ocular melanosis and sometimes lack of dermatological features.[6] Tellada et al published the biggest series of orbital melanomas, reviewing 264 cases during a 40-year span. Only 21 (8%) of these individuals had primary orbital melanoma identified. At the time of diagnosis, the patient's mean age was 42 years, with a 3:1 male to female ratio. Five patients (24%) and sixteen patients (76%) respectively reported with diplopia and blurred vision. Seven patients (33%) had orbital tumor removal without orbital exenteration, while fourteen patients (67%) received orbital exenteration as part of their treatment.

They reported 13 live patients (63%), 2 of whom had received adjuvant radiation therapy or chemotherapy, after a mean follow-up of 4.5 years. The author also described about positive association of younger presentation with better survival and negative association with congenital melanosis.[3] Interestingly, our patient had orbital exenteration with postoperative radiation therapy and was 33 years older with no presentation of congenital melanosis.

A larger series from Shields et al. reported 10 instances (0.8%) had primary orbital melanoma. The cases were categorized as either intraneural or extraneural tumours, and histologic subtypes resulting from congenital melanosis, blue nevus, or de novo formation were also noted. They found that painless proptosis was the most typical appearance. Their main suggestion was to remove the tumor entirely, reserving total exenteration for emergencies.[7]

The research lacks sufficient information to draw conclusions about the advantages of chemotherapy or further radiation therapy in cases where chemoradiation was primary treatment.[8]

CONCLUSION

Though a rare case, it is possible to uncover a risky diagnosis by using meticulous orbital dissection and histological examination techniques. Patients who present with imaging evidence of a well-defined tumour and unilateral proptosis should be evaluated for the rare cancer known as primary orbital melanoma. An aggressive and multidisciplinary approach is needed in management of this tumour.

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