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A Rare Case of Primary Orbital Melanoma and its Neurosurgical Approach

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I. INTRODUCTION:

Orbital tumours include a wide variety of lesions. They can be classified based on origin as primary, secondary and metastatic tumours.(1) Primary tumours originate from orbital structures itself. The most common primary orbital tumours are lymphoproliferative lesions of the orbit and adnexa.(2)Whereas secondary tumours arise from the structures outside the orbit extending into orbital cavity. To name a few common secondary orbital tumours are basal cell eyelid carcinoma, squamous cell conjunctival carcinoma, and retinoblastoma. (3)

Metastatic orbital tumours as the name defines are tumours that spread from distant primary tumours. Among children, the most common include neuroblastoma, Ewing tumour, chloroma, and Langerhans cell histiocytosis. In adults, primary tumours that commonly metastasize to bone (eg: breast, lung and prostate cancers) may form orbital metastatic tumours.(1)

Orbital melanoma is a malignancy of melanocytes. They can present as primary, secondary (due to local extension)or metastatic lesions. Orbital melanomas are relatively uncommon as a whole with secondary orbital melanomas being more common than primary orbital melanomas. Secondary orbital melanomas constitute 5-15% of all metastatic orbital tumour.(4) Secondary orbital melanomas are due extension from a uveal, conjunctival or eyelid primary melanoma, or as a metastatic disease. (5) Primary orbital melanoma constitutes only less than 1% of primary orbital tumours (6) and around 1% of all orbital tumours (7), thus highlighting its extremely rare occurrence. As per Anna et.al,2017, only approximately 50 cases were reported to that date. (8)

Since melanocytes are neural crest cell derived, they may present in places of neural crest cell migration.(9) Primary ocular melanomais believed to originate from melanocytic cells of the leptomeninges or ciliary nerve or from ectopic intra-orbital nests of melanocytes. (8) Primary orbital melanoma is usually associated with predisposing pigmented lesion such as hypercellular blue naevi, oculomelanocytosis, oculodermalmelanocytosis (also known as naevus of Octa) or orbital melanocytosis.(10) Hypercellular blue naevus presents as congenital, thickened, periocular, pigmented naevus of slate grey-blue colour whereas ocular melanocytosis presents as flat grey-blue area of cutaneous pigmentation associated with episcleral pigmentation. (11)These pre-existing lesions may also be an origin for primary orbital melanomas thus should be monitored regularly. (6,11)

The typical presentation of primary orbital melanoma is gradual painless proptosis. (11) There can be associated diplopia, blurring of vision, and ptosis. (6)In terms of treatment, surgery is the mainstay and some surgical options available are local resection, debulking or exenteration. Local adjuvant radiotherapy and adjuvant immunotherapy are other available modalities. (5)

Hereby we present a rare case of a patient with primary orbital melanoma involving a 33 year old gentleman whom was treated with local excision.

II. REPORT:

A) History

A 33 year old gentleman presented with diplopia particularly on looking to the right with gradual worsening for the past three years. It was disturbing his daily activities of life especially on using motor vehicles, hence he presented to the ophthalmologist. Otherwise, there is no blurring of vision, decreased visual acuity and no pain experienced. Patient was treated for craniopharyngioma with radiosurgery before with no recurrence on follow-up. There is no history of other malignancies. There is no history of trauma or infection involving the eyes.

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B) Examination

On examination, there is restricted right eye abduction (Grade -3 on a 9 point scale). There is no proptosis. There are no pigmented ocular lesions seen. Other ocular and neurological examination are unremarkable.

C) Investigation

Blood investigations were unremarkable. Patient was subjected for MRI (Magnetic Resonance Imaging) of the brain and orbits and MRI revealed right intraconal lesion with slight extension to superior right lateral intraconal space. The lesion showed enhancement post contrast. Patient was then referred to neurosurgery team for co-management.

D) Management

Patient was counselled for right orbital exenteration, however patient refused and only opted for tumour excision. Hence, tumour resection was planned via the right orbito-zygomatic craniotomy approach. Intra-operatively, bluish-black tumour was seen in the intraconal region with poor tumour margins. Tumour was seen to infiltrate the surrounding extra-ocular muscles and optic nerve. In order to preserve these structures, tumour was excised and removed in piece-meal fashion. Dura was repaired and the bone flap was placed back.

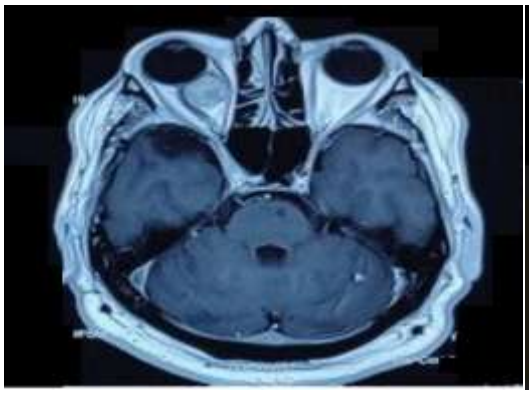
Histopathological examination of intra-operative specimen revealed neoplastic cells of epithelioid and oval to spindle-shape with abundant brownish pigments with peri-neural invasion. These neoplastic cell were stained positive for S100 and Melan-A thus, confirming the diagnosis of primary orbital melanoma. A negative finding for total body skin examination further confirmed its primary origin.

E) Outcome

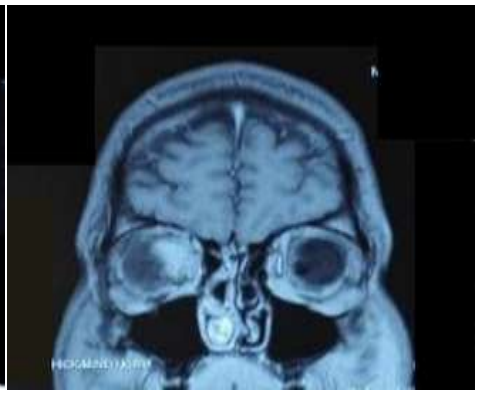
Post-operative, patient developed ecchymosis and ptosis however subsided spontaneously with two weeks. His right eye abduction improved thus reducing diplopia. Post-operative CT showed minimal hematoma. Patient is currently under ophthalmology follow-up. Six months follow-up showed no worsening signs and symptoms and no radiological evidence of recurrence.

Images:

Pre-operative MRI:

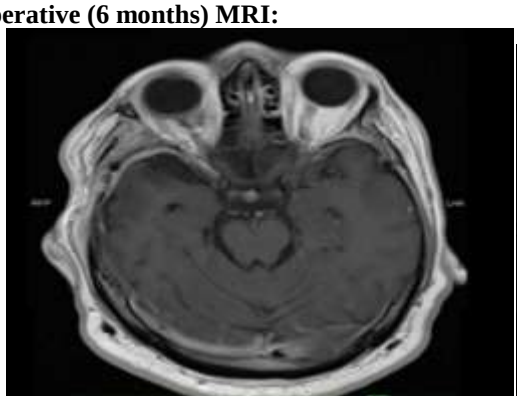


T1-Gado Axial



T1-Gado Coronal

Post-operative (6 months) MRI:



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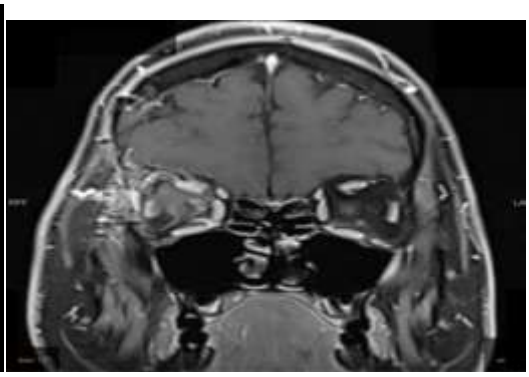

T1-Gado Coronal

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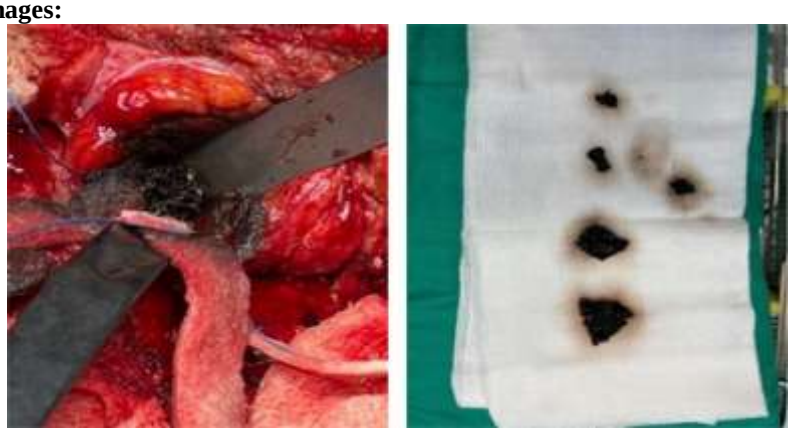
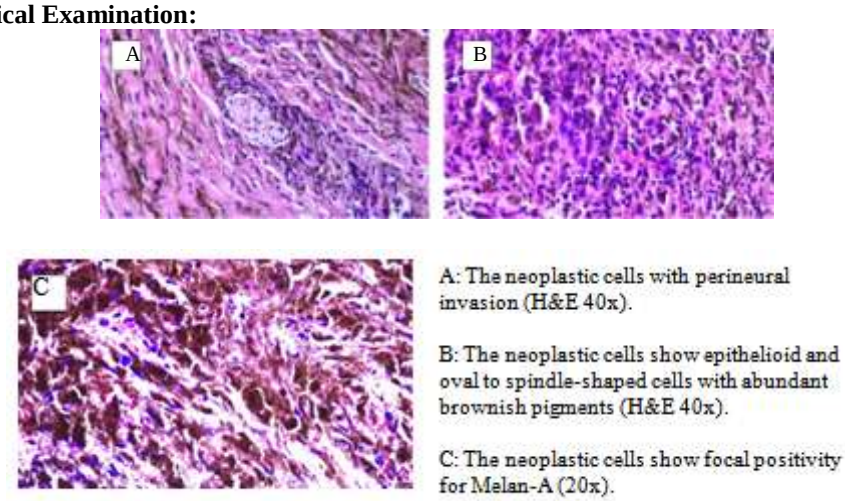
Intra-operative images:



Histopathological Examination:



A: The neoplastic cells with perineural invasion (H&E 40x).

B: The neoplastic cells show epithelioid and oval to spindle-shaped cells with abundant brownish pigments (H&E 40x).

C: The neoplastic cells show focal positivity for Melan-A (20x).

III. DISCUSSION:

Primary ocular melanoma usually presents with gradual, painless proptosis (11). Modupe et al. 2020 study showed 73% presented with unilateral proptosis.(5)However this typical presentation was absent in our patient. This is due to the lesion's small size which has yet to exert mass effect. Also, patient did not have any pre-disposing pigmented lesions suggestive primary ocular melanoma thus making it more difficult for us to diagnose. His only presenting complaintwas diplopia due to reduced range of right eye abduction. This can be explained due to the infiltration of the tumour into the extra-ocular muscles particularly right lateral rectus thus impairing their motility function.(1)

Second Malignant Neoplasm (SMN) is a known entity referring to late complications arising after exposure to genotoxic therapies, which include radiotherapy and some chemotherapeutic agents.(12)Majority of SMNs are radiation-induced tumours. Examples are meningiomas and gliomas arising following radiation for leukaemia and cranial nervous system tumours. Keeping this mind, we believe patient's previous history of exposure to radiosurgery for craniopharyngioma could be a pre-disposing risk factor for development of primary orbital melanoma.In view of close proximity of intraconal region and diaphragmatic sellae, we postulate effects of the radiation could have triggered genetic mutation of the hidden dormant melanocytic cell in the in the intraconal region. However further studies are needed to validate our hypothesis.

Nevertheless, we advise for a high suspicion index of orbital tumours among patients with ocular signs and symptoms and history of radiosurgery involving the cranium. An imaging of MRI of the brain and orbits would be suitable to look for any growth and this is proven effective in our study as the MRI was able to detect the primary orbital melanoma. We opted MRI instead of Computerised Tomography (CT) because it is more sensitive to the different soft-tissue characteristics of orbital lesions. Also, CT has the disadvantage in the field of ophthalmology because its radiation may cause cataract. (1)

Orbital exenteration with adjuvant radiotherapy is the widely accepted treatment in primary orbital melanoma. (5,11,13)Orbital exenteration involves removal of orbital with or without periorbital contents; including the globe, adnexa, and part of the bony orbit as well as the periorbita and surrounding face. (14) This is a disfiguring procedure and requires a second surgery for cosmetic reconstruction. In our case, this was proposed to the patient however he refused for the same. Hence, to respect patient autonomy we only performed tumour de-bulking.

However, this proved to be effective too.Patient improved symptomatically and till date no recurrence is noted. In our literature review, we realised Rose AM et all 2017 also did not use orbital exenteration routinely because primary tumour debulking still effectively addresses the disease. (8)More studies are definitely needed to decide on the type surgical intervention needed in primary orbital melanoma. Till then, we suggest to decide surgical treatment on case-to-case basis considering both patient factor and the extent of lesion.

Understanding of the anatomical division of orbital tumours is vital before discussing on the neurosurgical approach. The orbital muscle cone has a conical shape with the globe acting as the base and the optic canal as its apex.The muscle cone itself constitutes extraocular rectus muscles; namely superior rectus, inferior rectus, medial rectus and lateral rectus. The intermuscular septae form the division of intraconal and extraconal space.(1)The intracanalicular orbital tumours are the ones that are at least partially within the optic canal. (15)

Two major types of surgical approaches in removal of orbital tumours are transorbital approach and transcranial approach.(15) Mostly, anterior lesions are treated via transorbital approach whereas posterior lesions (involving intraconal and extraconal regions) are removed via trans-cranial approach. Transcranial approach helps to visualise lesions in orbital apex well through lateral wall of orbit. Since the MRI of our patient showed tumour residing in the intraconal region with superolateral extension, we chose the transcranial surgical routevia single piece of right orbitozygomatic craniotomy for more fine access. This approach ensured maximum exposure and helped us to remove maximum lesion possible with minimal bone loss. Not only that, this approach also provides a short straight distance between surgeon and the tumour and minimalize retraction thus reducing iatrogenic injury. (16) The other advantage is reflex bradycardia does not occur as this approach avoids compression on the eyeball. From our study, this surgical approach helped us to achieve good surgical resection with good cosmetic results. However in this surgical route, it is pertinent to careful during the freeing of all remaining periorbita and other soft tissue attachments to avoid damage to levator palpebrae muscle, superior rectus muscle and trochlear nerve.

We followed-up our patient at regular intervals with clinical examination and repeated CT and MRI of the brain and orbit. We also did a CT Thoracic-Abdomen-Pelvis which did not show any distant metastasis. Positron Emission Tomography (PET) scan is a better modality to look for distant metastasisin the follow-up period, however due to lack of resources in our setting, it was not done. PET scan is important because orbital melanomas metastasize hematogenouslyas the orbit lacks in lymphatics. This is not applicable in eyelid or conjunctival melanomasas they metastasize via lymphatic channels. (17)

IV. CONCLUSION:

Primary ocular melanoma is an extremely rare entity, nevertheless should be considered as differential in diagnosing space-occupying orbital lesions. Higher index of suspicion should be applied in patient with exposure of radiation especially to cranial and orbital regions. Proptosis may not be present in early lesions hence anabsence of it should not rule out this entity.

From our experience, tumour de-bulking gave a good prognosis hence should be considered rather than exenteration in managing primary orbital melanoma, however further studies are needed to verify this. A transcranial approach via orbito-zygomatic craniotomy is ideal for good operative field, better cosmesis and lesser risks and complication.

Since most cases are diagnosed by ophthalmologists and neurosurgical approaches offer a wide and safe surgical window(16), a multi-disciplinary team approach (particularly oculoplastic and neurosurgery teams) is highly recommended for the better planning and management of such cases.

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