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CASE REPORT - 1

Title : Choroidal melanoma :

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Abstract : A 61-year-old male was referred to the ocular oncology clinic with an one month history of diminision of vision in the right eye. Patient was apparently alright 1 month back, when he developed diminision of vision; it was sudden in onset, gradually progressive and painless. On examination vision in right eye was 6/60 and left eye 6/36. On SLBE Anterior segment was within normal limits. Patient was investigated for the above complaints. On B-scan it showed convex polypoidal mass in right eye. Colour Doppler revealed vascularity in mass. MRI was done to confirm the findings.

Key word : Slit-lamp bio-microscopy (SLBE), Normal size reactive to light (NSRTL), choroidal melanoma.

Introduction : The incidence of choroidal melanoma in the world is 0.02–0.06%[1]. Choroidal melanoma and other uveal melanomas most often affect Caucasians of Northern European descent. Incidence of choroidal melanoma among blacks is extremely rare. Those of Hispanic and Asian origin are thought to have a small risk compared with whites. In most series, the median age at diagnosis is about 55 years. Choroidal melanoma is found slightly more frequently in men for all age groups, except in the group from 20 to 39 years, where a small predilection exists for women [2]

Case-report : A 61-year-old male was referred to the ophthalmology clinic with one month history of diminision of vision in the right eye. Patient was apparently alright one month back, when he developed sudden onset, painless, progressive diminision of vision in right eye. Diminision of vision was more in central visual field than periphery. Patient had history of curtain falling appearance in front of eye. Patient also had a history of wavy appearance of straight line. No history of flashes of light, pain, redness, watering ordiplopia in both eyes. No history of pain in abdomen, nausea, vomiting, hemoptysis, fever and weight loss suggestive of metastasis.

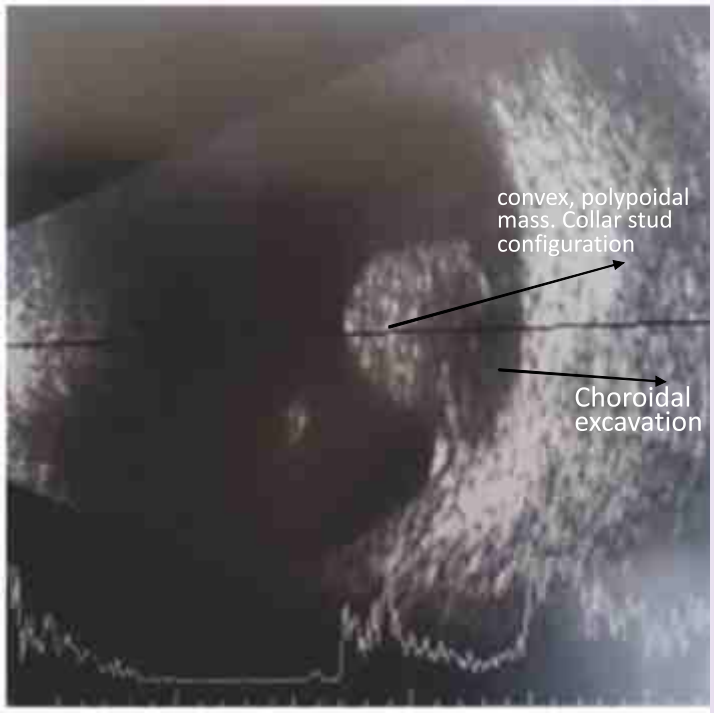
Table 1 : Eye Examination

Vriables	R/E	L/E
Vision	6/60	6/36
Ocular Movements	Normal	Normal
Eyebrow	Normal	Normal
Eyelid	Normal	Normal
Eyelash	Normal	Normal
Conjunctiva	Normal	Normal
Cornea	Clear	Clear
Anterior ChamberIris	N Content & Depth	N Content & Depth
PupilLens	N Colour Pattern	N Colour Pattern
	NSRTL	NSRTL & Pseudoexfoliation
	Grade 1 Nuclear Sclerosis	Grade 1 Nuclear Sclerosis

On Fundus examination : Right eye revealed a elevated, sub retinal black mass with large retinal detachment. with juxtapapillary choroidal melanoma surrounded by creamy yellow choroidal spots.

Left eye was within normal limits.

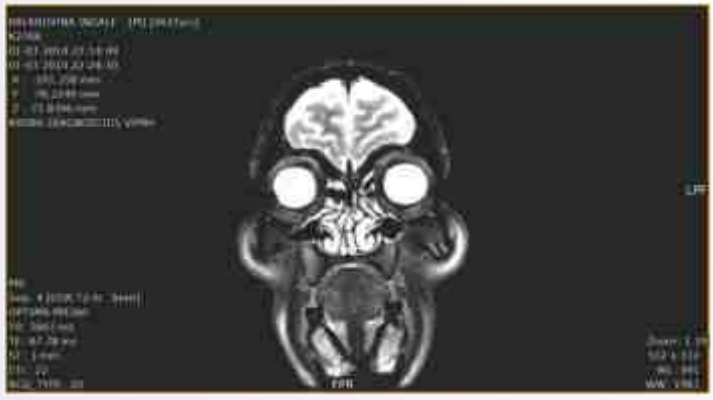
Investigations : 1. USG B-SCAN : Right eye showed convex, polypoidal mass. Collar stud configuration was present. Choroidal excavation was present. There were no signs of orbital shadowing.



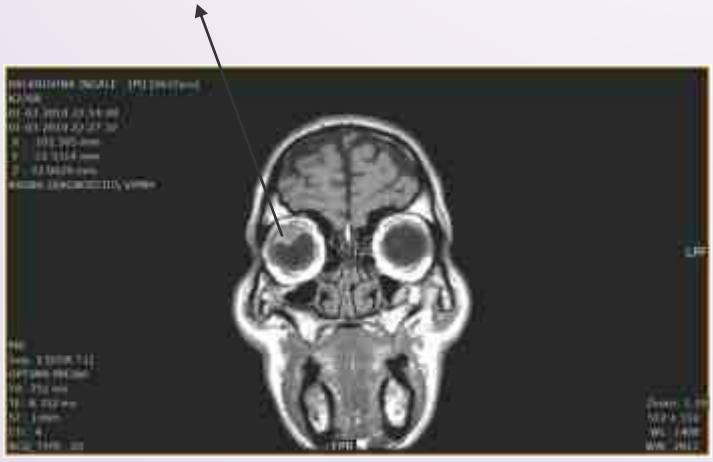
2. **Color Doppler** reveals tumour vessels as pulsating channels or lakes of color entering the base of the lesion.

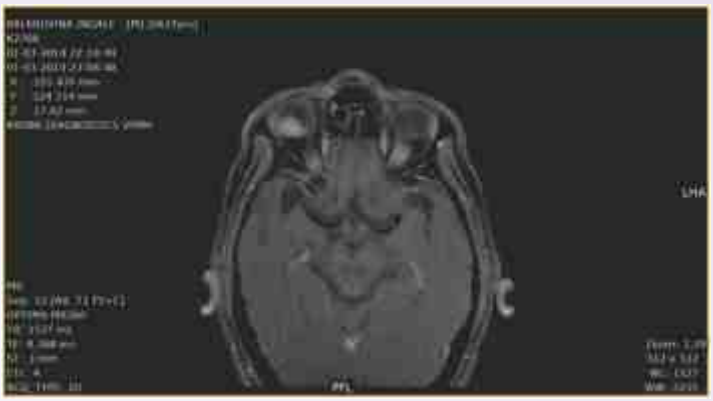
3. **MRI SCAN :**

- Accompanying MRI scans revealed well defined, enhancing, altered signal intensity right intra-ocular lesion in postero-supero-lateral aspect. It measures approx 13 x 11 x 11mm (ML X AP X SI).
- The left eyeball is normal with normal uveo - scleral attachment.
- Optic nerves are normal bilaterally, in course & caliber, with normal signal intensities within them. Optic chiasm and supra – sellar regions appear normal.
- Extra-conal compartments are normal. Extra-ocular muscles are normal bilaterally.
- Soft tissues around the eyeball or orbit are normal.



Well define enhanceing, altered signal intensity





Discussion : The uveal tract is the highly vascular and densely pigmented layer of the eyeball, lying between the sclera (superficial to it) and the retina (deep to it). The anterior, visible portion is the iris. This extends back into the ciliary body (at the level of the lens) and then extends round to the posterior pole. This fundus portion is known as the choroid. Choroidal melanoma is the most common primary malignant intra-ocular tumour and accounts for 80% of all melanomas of the uveal tract.[3]

Primary choroidal melanoma arises from melanocytes within the choroid. It is thought to develop from pre-existing melanocytic naevi , although de novo growth may occur. The colour varies from darkly pigmented to amelanotic. It is usually dome-shaped. If it breaks through Bruch's membrane (which effectively forms a blood/neural tissue barrier between the vascular choroid and the retinal layer) as it grows, it looks like a mushroom. It can also be bilobular, multilobular and diffuse in shape. Occasionally, there may be a number of small lesions in one or both eyes (although bilateral involvement is generally rare).

Choroidal melanomas remain asymptomatic for prolonged periods of time so they are diagnosed lately. Sometimes they may be found accidentally during ophthalmoscopy. In general, the more anterior is their origin,the longer the delay of any symptoms. Choroidal melanoma may present with the following symptoms:

1. Blurred visual acuity
2. Paracentral scotoma
3. Painless and progressive visual field loss
4. Floaters
5. Severe ocular pain

6. History of weight loss, marked fatigue, cough, or change in bowel or bladder habits

Histologic evaluation of the tumor after enucleation can confirm the diagnosis and determine the prognosis[4].The modified Callender classification is the most commonly used histologic classification for classifying uveal melanomas, which divides uveal melanocytic tumors into the following groups.[5]

1. Spindle cell nevi
2. Spindle cell melanomas
3. Necrotic melanomas
4. Epithelioid cell melanomas
5. Mixed cell melanomas

Fine-needle biopsy and incisional biopsy are not usually performed but can be done in difficult cases, particularly for distinguishing amelanotic melanomas from metastatic tumors, and in situations where the results of other ancillary tests are equivocal. Fine-needle biopsy is increasingly being performed for prognostic purposes. Metastasis is also common in choroidal melanoma. Several studies have found several tumor features to correlate with increased mortality, including larger size, anterior location, transscleral extension, growth through the Bruch membrane, optic nerve extension, lack of pigmentation, and histologic characteristics (eg, mitotic activity and cell type)[6]. Metastases are most commonly to the liver (this is the sole site of metastasis in 80% of cases)[7].Less frequently, choroidal melanoma can metastasise locally into the orbit, the conjunctiva or the maxillofacial bones.[8]

Uveal melanomas are genetically homogenous, with few tumor-specific cytogenetic aberrations. Some of these aberrations correlate with the metastatic potential of the tumor, resulting in metastatic disease followed by death. Recurrent aberrations in uveal melanomas concern loss of 1p, monosomy of chromosome 3, loss of 6q and 8p, and gain of 6p and 8q.Loss of chromosome 1p was observed in metastases,[9]and concurrent loss of 1p and chromosome 3 is associated with decreased survival[10,11]. Furthermore, monosomy 3 is considered to be an early event in UM, and several studies have shown that it is a strong predictor of survival[12,13,14]. Loss of chromosome 3 is frequently associated with amplification of 8q, often seen as isochromosome 8, q-arm.

The available literature suggests that risk of choroidal and ciliary body melanomas associated with nevi of the uveal tract is low[15]. Hormonal influences are suspected to be a factor in cutaneous melanoma based on reports of an increased risk for women in their child-bearing years[16,17]. Acute or intense exposure to ultraviolet light might increase the risk of uveal melanoma, but the role of acute or chronic sunlight exposure remains inconclusive[18,19].

Biopsy :

The biopsy showed long, fusiform cells with prominent nucleolus i.e. spindle-B cells. Large round epitheloid cells with eosinophlilic cytoplasm were also present confirming the diagnosis of primary Choroidal melanoma.

Treatment :

Right eye enucleation was done under local anesthesia and the eyeball was sent for histopathology. One of the two clinical trials of the collaborative ocular melanoma study (COMS) compared preoperative external beam radiation therapy plus enucleation to enucleation alone in patients with large choroidal tumors to address the concern that enucleation might precipitate tumor metastasis and shorten survival[20].

Prognosis and survival :

Despite the availability of alternative treatment modalities, the survival rates of patients with uveal tract melanoma have not changed in 30 years. Common sites of metastases include liver (90%), lung (24%), and bone (16%).Patients with metastases confined to extra hepatic locations have longer survival (19–28 months).

Conclusion :

Treatment for a small choroidal melanoma in the posterior fundus ranges from observation to several treatment options, including laser photocoagulation, plaque radiation therapy, external beam charged particle radiation therapy, transpupillary thermotherapy, location tumor resection, and enucleation. Plaque brachytherapy is the most frequently used eye-sparing treatment for these lesions.

But, radiation therapy is about twice as expensive as enucleation and there appear to be no significant quality of life differences between patients treated with radiation or enucleation.

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