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Primary choroidal melanotic malignant melanoma; Ultrasound and MR imaging features

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Abstract
Choroidal malignant melanoma is the most common primary adult ocular malignancy. Primary choroidal melanoma arises from melanocytes within the choroid. Patients with choroidal melanoma may present with or without visual symptoms. Asymptomatic tumors are more frequent and often are detected only at routine eye examinations. Melanotic melanoma has intrinsic T1and T2 weighted shortening effects, aiding histological diagnosis, whereas Amelanotic melanoma or mildly pigmented lesions of melanoma donot demonstrate these characteristic Magnetic Resonance imaging features.
We report a 58 year old male patient with progressive visual impairment and loss of vision of the right eye that had been evolving for 1 year, evaluated clinically and radiologically by real time grey scale B-Mode ultrasonography (USG) & Magnetic Resonance imaging (MRI).

Keywords: B-Mode ultrasound, Choroidal melanoma, Magnetic resonance imaging, Malignant melanoma, Uveal melanoma

1. Introduction
The uveal tract is the highly vascular and densely pigmented layer of the eyeball, lying between the sclera and the retina and consists of anteriorly iris & posteriorly ciliary body & choroid.
Uveal melanoma is a serious life-threatening intraocular malignancy, primarily involving the choroid (90%), ciliary body (7%) or iris (2%) [1].
98% of cases occur in Caucasians [1].
It is rarely found among black people. Those of Hispanic and Asian origin are thought to have a small risk compared with white people. The rate of occurrence in the general population varies between five and eight cases per 1 million people [2].
There is a slight predisposition for males and uveal melanoma is most often diagnosed in the 6th decade [3]. They are the second most common intraocular neoplasm overall, after metastasis. Ocular melanomas are generally unilateral, although bilateral melanomas occur sporadically [4]
The 5-year survival rate for patients is 84% with small choroidal melanomas, 68% with medium choroidal melanomas, and 47% with large choroidal melanomas [1].
We report a 58 years old male patient with pathologically proven primary Choroidal melanotic malignant melanoma of right orbit.
The Clinical, pathological, specific MRI and B-Mode ultrasound findings in the diagnosis of primary Choroidal melanotic malignant melanoma are discussed.

2. Case Report
A 58 year old male presenting with progressive visual impairment and loss of vision of the right eye that had been evolving for 1 year was referred for B-Mode ultrasound (USG) & magnetic resonance imaging (MRI) evaluation of the orbits & Brain.
At ophthalmologic examination, absent vision was noted in the right eye and visual acuity of 20/20 in the left eye. At indirect ophthalmoscopic examination, a pigmented mass was seen in the posterior portion of the fundus of the right eye. The blood tests including complete blood count (CBC), hemoglobin, liver function tests (LFT), and renal function tests (RFT) were all within normal limits.
On B-Mode trans-orbital ultrasound (Voluson GE) examination using a high frequency transducer revealed a large lobulated domed mass arising from the choroid. The lesion measured 7 mm thick and 25 mm in diameter and was classified as a large tumor (Fig.1A-D). Low level echoes seen in the anterior vitreous cavity suggestive of vitreous hemorrhage.

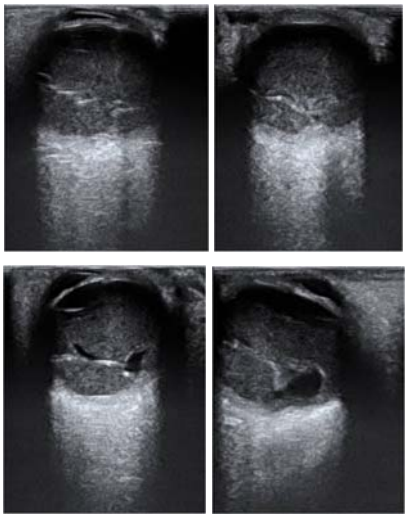

Fig 1: (A-D) Choroidal Melanoma. B-scan ultrasound: Shows a dome-shaped large lobulated solid mass posteriorly, with posterior acoustic shadowing and choroidal excavation.

The MRIs were performed using 1.5 T magnets (Signa GE). Standard T1- and T2-weighted Spin-echo pulse sequences were used.

MR imaging was performed with 2-mm sections by using a 5-cm surface coil. The patient was examined with T1 Weighted fat saturation sequences, after administration of intravenous Gadolinium (0.1mmol /kg) in axial, coronal and sagittal planes.

On MRI scan of the right orbit, demonstrated a single dome shaped intraocular mass measuring 10× 25 × 7 mm in the posterior quadrant and arising from the choroid of the eye. The mass was depicted as hyperintense on T1-weighted MR images (Fig.2A-D) and hypointense on T2-weighted MR images (Fig.3A-D) and enhanced moderately after injection of gadolinium contrast (Fig.4A-D). No extraocular extension was apparent. These findings are compatible with a typical melanotic malignant melanoma of the choroid. Mild hyperintense anterior vitreous seen on T1W sequences suggestive of subacute vitreous hemorrhage. No associated retinal detachment was evident. Left globe appear normal.

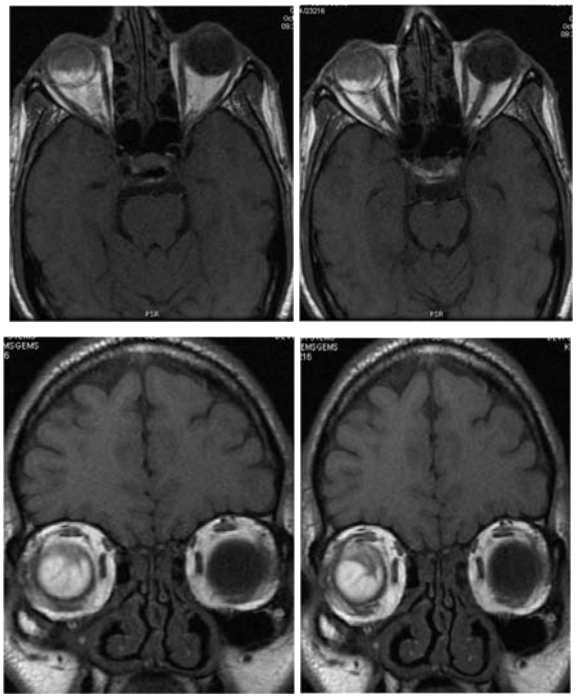

Fig 2: Axial (A,B) & Coronal (C,D) T1W MR without contrast demonstrates a large mass along the posterior choroid, that is moderately hyperintense relative to vitreous.

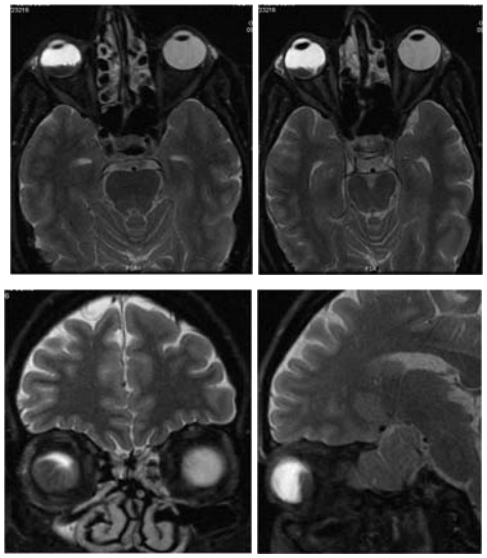

Fig 3: Axial (A, B), Coronal (C) & Sagittal (D) T2W MR demonstrates a large lobulated dome shaped mass along the posterior choroid, that is hypointense relative to vitreous.

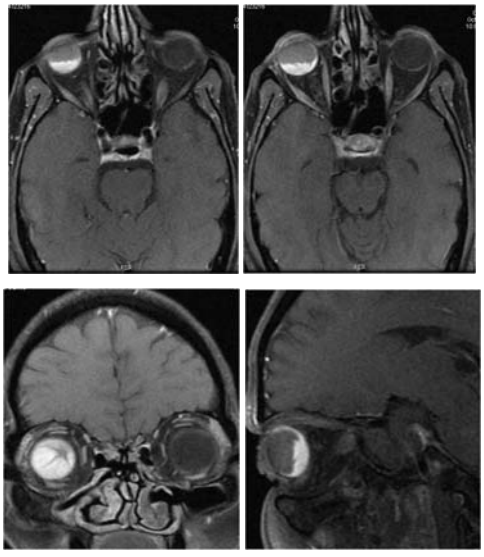

Fig 4: Axial (A, B), coronal (C) & sagittal (D), T1W gadolinium Contrast enhanced MR demonstrates a well-defined lobulated moderate contrast enhancing mass (choroidal melanoma) within the posterior quadrant of the right globe.

The investigation to look for distant metastasis and systemic disease included a brain MR imaging examination, a chest radiograph, a CT scan of the abdomen and pelvis, and liver function tests. The findings from all of these examinations were normal.

The patient underwent Enucleation of the right eye, followed by a bioceramic orbital implant insertion to receive the eye prosthesis.

Histopathology of the mass confirmed pigmented spindle a tumor cells of melanotic melanoma.

3. Discussion

Melanomas are malignant neoplasms of melanocytes. The commonest orbital malignant melanoma (MM) is uveal melanoma. This is because the uvea is the most vascularised portion of the eye, hence a substrate for primary and metastatic neoplasm [5].

Uveal melanoma is sub-classified into anterior uveal melanomas when the tumour arises from the iris and posterior melanomas when it arises from the choroid or ciliary body. The choroidal sub-type is the commonest MM among adults. WHO and International agency for Research on Cancer (IARC) reported 0.1-2.3 per 100,000 world-wide [6].

Melanoma rarely involves the conjunctiva, eyelid, or nasolacrimal duct [7].

Ocular melanoma is not considered an inherited disease, although recent studies suggest a genetic component. Possible predisposing factors include preexisting nevi, other melanocytic conditions such as ocular melanosis and nevus of Ota, impaired immunity, light-colored irides, occupational exposure (eg,welding), and trauma [3, 7].

Choroidal melanomas are usually located posteriorly and are classified by size as, Small (less than 10 mm), Medium-sized (11 to 15 mm) and larger tumors (greater than 15 mm). Small tumors are typically discoid and confined to the choroid. Medium sized tumors usually have a classic collar-button or mushroom appearance. This is secondary to tumor herniation through a ruptured Bruch membrane, the transparent layer between the retinal pigment epithelium and the choroid. Large tumors can fill the globe and extend through the sclera.

Ciliary melanomas tend to be small and may invade the anterior angle and iris. Iris melanomas are also small and can extend into the ciliary body or seed the anterior chamber [8].

Microscopically MM are classified into 3 cell types; Spindle A, Spindle B and epithelioid.

Patients with uveal melanoma may present with or without visual symptoms. The typical symptoms, when present, are visual field defects, visual loss, and photopsia (perceived flashes of light) [9]. These are usually caused by tumor encroachment on the central retinal fovea, contact with the lens, or exudative retinal detachment. Severe pain is rare with melanoma unless associated with extraocular extension, inflammation, or neovascular glaucoma, and such pain is due to ciliary nerve involvement [10].

Diagnosis of choroidal melanoma is based mainly on the findings from indirect ophthalmoscopy, the clinical history, transillumination, or US and its sequential evaluation.

At USG examination, A-mode US demonstrates a tumor with low-to-medium internal reflectivity spikes on the tumor surface and vascular oscillations. B-mode US demonstrates a domed, lobulated, or mushroom-shaped mass [11]. Large tumors show echolucency in the tumor base because of sound attenuation and can be heterogeneous secondary to hemorrhage or necrosis [12]. Coexisting retinal detachment is commonly seen extending from the tumor margins and over the tumor apex.

The typical imaging feature of melanoma is choroidal excavation. An area of excavation under a small posterior wall mass is indicative of melanoma, although this feature is not always present. At color Doppler US, they are usually seen to be vascular. Metastases demonstrate higher flow than melanoma at Doppler US.

On CT, Ocular melanoma appears as a focal sharply marginated, elevated mass hyperdense to vitreous with mild to moderate contrast enhancement after intravenous administration of contrast material. The appearance on CT is nonspecific [13, 14].

The MRI characteristics of orbital melanoma have been mainly attributable to paramagnetic properties of melanin. This melanin shortens T1 and T2 relaxation times leading to T1W hyperintense orbital melanoma which is a hypointense on T2W with respect to the hyperintense vitreous. However the presence of tumour necrosis containing water, presence of blood degradation products and iron content may explain the varying combinations of signal intensities such as decreased signal on T1W and/or increased signal on T2W [15].

Fat suppression techniques help to improve the conspicuousness of the tumour and in differentiations from pseudo-melanomas and assess orbital extension [16]. Fat suppression technique combined with Gadolinium enhanced MRI images help to detect small intra-ocular mass with thickness of >1.8mm [15, 16]. Amelanotic melanoma exists, behaving just like any other tumour with hypointense or isointense on T1W and hyperintense/isointense T2W sequences.

The main differential diagnosis is choroidal metastases (from lung, breast, hypervascular, and hematologic malignancies), which are frequently bilateral (33%) and multiple. At MR imaging, choroidal metastases tend to be iso- to hyperintense on T1-weighted images and hyperintense on T2-weighted images, with heterogeneous enhancement. Obtaining the clinical history of the patient is therefore mandatory.

The combination of T1W hyperintensity and T2W hypointensity signals have been seen in other intra-ocular lesions like serous retinal detachment, secondary sub-retinal fluid, uveal melanocytoma, choroidal osteoma, sub-acute choroidal / retinal haemorrhagic detachment, Retinoblastoma, Retinal capillary haemangioma, Focal retinal gliosis, Medulloepithelioma, and Inflammatory granuloma. This is due to proteinacious content or blood degradation product [15]. Post-contrast T1W with fat suppression may help to differentiate these masses from MM as retinal detachment do not enhance and Choroidal haemangioma with high vascular flow and enhancement exhibit isointensity to slight hyperintensity on T1W and hyperintense T2 weighting which is isointense to vitreous.

Extrascleral local extension occurs via scleral emissary channels for neurovascular structures and is associated with increased risk of recurrence [13] and hematogenous dissemination [8]. Metastasis usually occurs hematogenously and is more common with choroidal melanomas [3]. The liver is the most common remote site of metastasis [8].

The 5-year survival rate for patients is 84% with small choroidal melanomas, 68% with medium choroidal

melanomas, and 47% with large choroidal melanomas [1].
Accurate and early diagnosis of choroidal melanoma by any means is important because of the direct correlation between tumor size and mortality. Tumor size not only influences prognosis but may also determine subsequent management. Newer treatment methods, such as radioactive plaque therapy, laser photocoagulation, proton beam irradiation, and en bloc resection, now offer an alternative to enucleation.
Our patient underwent Enucleation of the right eye, followed by a bioceramic orbital implant insertion to receive the eye prosthesis.

4. Conclusion

Choroidal melanoma is the second most frequent ocular malignancy after metastasis and the most common primary ocular malignant neoplasm in adults. The diagnosis is usually made from clinical examination and ocular ultrasound. CT and MR may be helpful for further evaluation. Postcontrast T1-weighted images were most sensitive in detecting uveal melanoma by MR examination and helpful in differentiating tumor from subretinal fluid collection. Findings from contrast-enhanced MR imaging, added to clinical data and results from other imaging techniques, provide further diagnostic information and allow confirmative diagnosis, thereby obviating unnecessary interventional procedures.

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