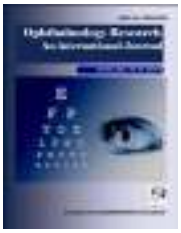

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Panophthalmitis Masquerading as Ocular Melanoma-an Eye Opener

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Authors' contributions

This work was carried out in collaboration between all authors. Author SA writing the manuscript, literature search. Author RNG Pathology work up of the enucleated globe. Author KPK Clinical workup. Author PS Laboratory Investigation including imaging studies. All authors read and approved the final manuscript.

Case Study

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ABSTRACT

Introduction: We report a clinicopathological correlation of a 45 year-old female who manifested as melanoma clinically and radiologically but turned out to have acute panopthalmitis on histopathology examination. We feel that there is significant clinical implication in sharing the knowledge of this clinico-pathological correlation.
Case Presentation: A 45 year- old female presented with complaints of sudden painful diminution of vision in left eye for five days. Examination revealed proptosis of left eye with neovascularization of iris, seclusiopupillae, and fundus details could not be visualized. Computerized Tomogram (CT) examination, 99m TC mibi whole body scan and Single-photon emission computed tomography (SPECT CT) were done. SPECT CT showed focal area of increased tracer uptake in anterior and superolateral aspect of choroid in left eye. Avid mibi tracer uptake in choroid of left eye suggested a high possibility of ocular tumor with involvement of retrobulbar portion of left optic nerve. Histopathological examination revealed panophthalmitis with dense neovascularization of the choroid and ciliary body. No tumor cells or necrosis were evident in the sections studied. Sections from optic nerve showed extensive perineurialedema. Immunohistochemistry with Melan A, S-100 and HMB-45 were performed on multiple sections and were negative, thus excluding the possibility of melanoma.

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Conclusion: Painful blind eye is a major therapeutic dilemma for ophthalmologist and findings such as avid mibi tracer uptake in the choroid of left eye suggested a high possibility of ocular tumor. We wish to emphasise that inflammatory pathologies of the eye can masquerade as malignancies and to highlight the limitations of the state.

Keywords: Panophthalmitis; melanoma; painful blind eye; masquerade syndrome.

1. INTRODUCTION

Ocular diseases mimicking melanoma are called pseudomelanomas. Melanomas constitute 0.02% of the outpatients in referral institutes in our country. Various lesions that mimic choroidal melanomas are choroidal neoplasms like choroidal nevus, choroidal hemangioma, osteoma, metastasis, melanocytoma, hemorrhagic processes like involutional macular degeneration, extramacular disciform lesion and ruptured arteriolar macroaneurysm, retinal lesions like retinal pigment epithelial hyperplasia or hypertrophy, posterior scleritis and other conditions like hemorrhagic retinal detachment. Various unusual presentations of choroidal melanoma like posterior scleritis, choroidal effusion with secondary glaucoma, posterior staphyloma, offer a diagnosticdilemma to the treating clinician. We report a case which had clinically masqueraded as choroidal melanoma but turned out to be panophthalmitis on histopathology.

2. CASE PRESENTATION

A 45 year-old female presented with sudden painful diminution of vision in left eye for five days. There was no history of trauma. She was not a known diabetic or hypertensive. Visual acuity of the right eye (RE) was 20/20 and left eye (LE) was perception of light with accurate projection of rays. Examination of RE was within normal limits. LE showed an axial proptosis of 3 mm, a shallow anterior chamber and neovascularization of iris at the pupillary border, seclusiopupillae and a fibrinous membrane in the pupillary area. Fundus details could not be made out in the left eye. Intraocular pressure was 14 mm Hg (RE) and 12 mm Hg (LE). Ultrasound B-scan of the LE showed the presence of an echogenic mass projecting into the vitreous cavity with diffuse choroidal thickening and no scleral extension. RE revealed a normal scan. Computerized Tomogram (CT) scan of left orbit revealed an ill-defined enhancing soft tissue density lesion attached to the posterior retina of the left eye. Sclera appeared thickened and hyperenhancing along with mild thickening (1 mm asymmetry) of optic nerve for a distance of 4.6 mm in the retrobulbar portion. Rest of the intraorbital and intracanalicular course and caliber of optic nerve was normal. Intra and extraconal spaces showed normal fat attenuation with no focal lesion or stranding. Bony walls of left orbit, left extraocular muscles and left lacrimal gland were normal. Right orbit, intraorbital contents and brain parenchyma were normal. 99m Tc-mibi whole body scan and SPECT CT showed focal area of increased tracer uptake in anterior and superolateral aspect of choroidal region in LE. Avid MIB 1 tracer uptake in choroid in LE suggested a high possibility of malignant tumor with involvement of retrobulbar portion of the optic nerve. Systemic examination was normal. Laboratory investigations revealed normal hemogram and normal biochemical parameters for sugar, urea, creatinine, Aspartate amino transferase (AST) and Alanine amino transferase (ALT). ELISA for toxoplasmosis serology was negative. Serum for Antinuclear antibody (ANA), Rheumatoid antibody (RA) factor and C-reactive protein (CRP) were negative Poor visual prognosis was explained to the patient, LE enucleation was performed and the tissue was sent for histopathology. The eye ball and optic nerve were sampled entirely in view of the clinical suspicion of melanoma. Histopathology revealed dense

infiltration of lymphocytes, plasma cells and neutrophils in choroid, extending in to the ciliary body. There was dense neovascularization of the choroid and ciliary body. No tumor cells or necrosis were evident in the sections studied. There was no evidence of granulomatous reaction or caseous necrosis. No organisms were identified on histochemical stains.

Sections from optic nerve showed extensive perineural edema. Immunohistochemistry with Melan A, S-100 and HMB-45 were performed on multiple sections and they were negative, thus excluding possibility of melanoma

3. DISCUSSION

Ocular diseases mimicking melanomas are known as pseudomelanomas. Stoffelns et al. report that most frequent ocular diseases suspected for choroidal melanoma were "suspicious nevi" in 31% and "disciform macular degeneration" in 34%, less frequently hyperplasia of pigment epithelium (5%), melanocytoma (10%), choroidalhaemangioma (6%), choroidal detachment (7%) and retinal macroaneurysms with epi-/subretinal haemorrhages (3%) and rare conditions like orbital tumour, posterior scleritis and a combined hamartoma of retina and pigment epithelium (4 %) [1].

According to a study by Biswas et al, uveal melanomas constitute 0.02% of outpatients in a referral eye hospital in India over a 12-year period. Because of the rarity of the uveal melanomas in Asian Indians the possibility of misdiagnosis is also quite high [2].

Blind eyes are suspected to harbor melanomas of the choroid and cause death secondary to distant metastases [3].

Choroidal melanoma has been reported to have highly variable presentations. Some of the manifestations include painful scleritis [4,5] choroidal effusion with secondary glaucoma [6] cataract and staphyloma [3]. The present case had clinical and radiological features suggestive of choroidal melanoma but the histopathology revealed panophthalmitis. In our case the fundus examination could not be done due to presence of seclusion pupillae and presence of pupillary membrane. Ultrasound showed diffuse choroidal thickening with no extension to the sclera and hence the diagnosis of choroidal pathology was made. CT scan of brain and orbit revealed an ill-defined enhancing soft-tissue density seen attached to the posterior retina of the globe of left eye. Sclera was thickened and hyperenhancing with mild thickening (1 mm asymmetry) in the retrobulbar portion of left optic nerve for a distance of 4.6 mm. 99m TC mibi whole body scan and SPECT CT were done. Planar study revealed focal area of increased tracer uptake in superolateral aspect of left eyeball. SPECT CT showed focal area of increased tracer uptake in choroidal region in left eye suggesting high possibility of a malignant pathology. A pre-operative diagnosis of choroidal melanoma was thus made. Pizzuto et al report of a case of choroidal malignant melanoma presenting as panophthalmitis [7].

Coupland et al state that extraocular spread correlated with increased mortality because it was associated with increased tumor malignancy and, in the case of posterior tumors, more advanced disease [8].

Saari et al found that the comparative use of digital color, red-free and red light imaging had 85.7 % (95 % CI; 42.1-99.6) sensitivity, 99.0 % (95% CI; 94.7-99.9) specificity and 98.2% (95% CI; 93.6-99.8) exact agreement versus reference standard in differentiation of small choroidal melanoma from pseudomelanoma They found that the most frequent choroidal

pseudomelanomas were choroid almelanotic and amelanotic nevi, disciform lesions, congenital hypertrophy of the retinal pigment epithelium, and circumscribed choroidal hemangioma [9].

MRI and radioimmunoscintigraphy are sensitive techniques for the diagnosis of choroidal melanomas [10]. 99mTC mibi scanning is an effective imaging modality for whole-body screening of metastatic disease in malignant melanoma patients with the potential to influence treatment planning [11].

Our study highlights the limitations of radiological findings in absence of thorough fundus examination. The possibility of spontaneous resolution of the tumor with extensive necrosis and acute inflammation of the eye ball is highly unlikely in view of the fact that the surgery was performed within 4 days of CT, SPECT CT and 99 TC mibi scans. The enucleated eye ball was sampled entirely in histopathology examination for identifying any focus of residual tumor. In addition to light microscopic examination, immunohistochemical studies were performed to exclude possibility of melanoma.

4. CONCLUSION

Panophthalmitis masquerading as choroidal melanoma is uncommon. This presentation is to highlight the possibility of inflammatory pathology masquerading as a malignancy in spite of an otherwise normal clinical examination, routine investigations and a state of the art radiologic back up.

CONSENT

All authors declare that 'written informed consent was obtained from the patient for publication of this case report and accompanying images.

ETHICAL APPROVAL

The authors obtained ethical approval from Institutional Ethical Committee.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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