Unusual Presentation of Unilateral Choroidal Melanoma with Bilateral Vasculitis in Young Individual: A Case Report and Review of Literature

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ABSTRACT

Ocular melanoma stands as the predominant primary intraocular malignancy, albeit infrequently exhibiting ipsilateral inflammatory manifestations. In this article, we present an exceptional case involving a middle-aged male who presented with unilateral ocular choroidal melanoma alongside bilateral retinal vasculitis. The patient initially received temporary steroid treatment, followed by brachytherapy, which contributed to the resolution of vasculitis symptoms. The study aims to document the atypical occurrence of bilateral retinal vasculitis, which could potentially masquerade as melanoma, emphasizing the need for heightened vigilance and further investigations when encountering choroidal masses in its presence. Future research endeavors are warranted to better understand the incidence of such occurrences in this context.

Keywords*: Melanoma, Vasculitis, Uveal Melanoma, Choroidal Melanoma, Macular Lesion, Uveitis, Ocular melanoma, masquerade.*

INTRODUCTION

Uveal melanoma is the most common primary intraocular malignancy.[1] There are approximately 7095 new cases of uveal melanoma annually, over half of this number in white non-Hispanic populations. This incidence has been stable over a decade.[2] Choroidal melanoma, especially the small lesions, can be difficult to diagnose initially due to the wide variety of mimickers. Diagnosis of choroidal melanoma is usually reached with the aid of ancillary tests including imaging, in addition to the use of indirect ophthalmoscopy.

A masquerade syndrome is an ophthalmological entity where a neoplasm mimics an inflammatory condition. Ocular melanoma may rarely present with ipsilateral inflammatory signs or vitreous hemorrhage and has most frequently been noted in those with tumor necrosis and mixed or epithelioid cell types.[3,4] In this case, we present an unusual case of unilateral macular choroidal melanoma with bilateral vasculitis in a middle-aged patient.

CASE ILLUSTRATION

This is a case of a 37-year-old Saudi male patient, medically free, who presented to the emergency department at Dhahran Eye Specialist Hospital (DESH) complaining of distorted central vision, in the form of metamorphopsia, with minimal decrease of vision of the left eye throughout a couple of months. There was no associated history of trauma, headache, nausea, vomiting, flashes of light, fever, weight loss, night sweating, neck pain, joint pain, or rashes. There was a positive history of left Ramsay Hunt syndrome (*herpes zoster oticus*), which

was treated with steroids and resolved without residual complications. The past ocular, surgical, and family histories were unremarkable. The patient provided a positive history of smoking.

On examination, the visual acuity and intraocular pressure were 20/20 and 20/50, 19 mm Hg and 17 mm Hg in the right and left eyes, respectively. The anterior segment examination was within normal limits in both eyes. Dilated fundus examination, (**Figure 1**), showing the left eye with a pigmented choroidal lesion under the macula with overlying orange pigmentation demonstrated hyperautofluorescence on Fundus Autofluorescence and perivascular sheathing in the periphery in both eyes.

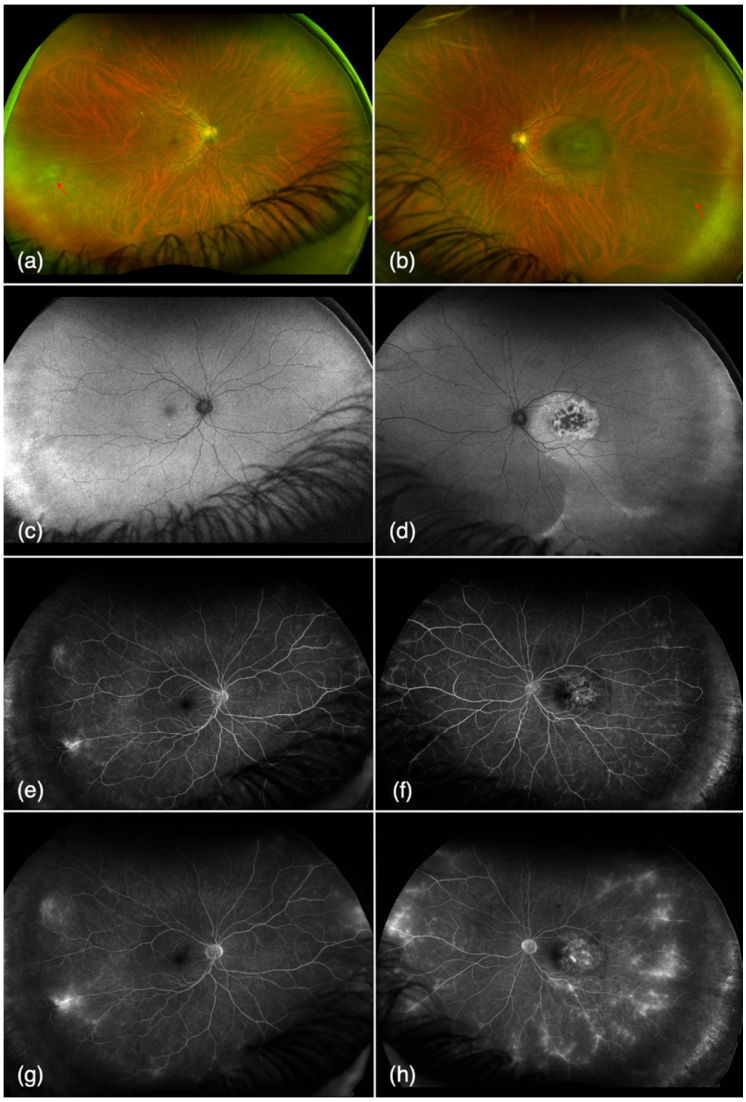

Figure 1. (a,b) Color Fundus Photo of both eyes showing (left eye) with pigmented choroidal lesion under the macula with overlying orange pigmentation and areas of peripheral retinal vasculitis 'red arrows'. (c,d) Fundus Autofluorescence Photo showing hyperautofluorescence of the choroidal lesion (left eye). (e.f) Fundus fluorescein angiography early frames showed patches of hypo and hyperfluorescence in the macula of the left eye, and the peripheries of both eyes showed diffuse capillary leakage with vasculitis. (g,h) late frame Fundus fluorescein angiography photos showing staining and leakage of the mass in the left eye with bilateral peripheral diffuse leakage and staining of the blood vessels which is more in the left eye.

Fundus fluorescein angiography showed patches of hypo and hyperfluorescence in the macula of the left eye. The retinal periphery of both eyes showed diffuse capillary leakage with vasculitis in retinal venules and capillary nonperfusion in late frames (**Figure 1**). B-Scan showed a choroidal lesion sized 7.06*8.54 mm and 3.17 mm thickness with low to medium internal reflectivity (**Figure 2**). Optical Coherence Tomography, (**Figure 3**), showed a choroidal large mass with subretinal fluid and hyperreflective subretinal (lipofuscin) deposits.

Complete blood count with differential (CBC), Liver function tests (LFTs), and Renal function tests all were within normal limits. The most common infectious etiology was ruled out as we found out that Tuberculin skin (TST) or purified-protein derivative (PPD) was negative, The rapid plasma reagin (RPR) test was negative and Toxoplasma latex was negative. ANA (Antinuclear Antibody) Test was positive. The patient was unlikely to have sarcoidosis as the Angiotensin Converting Enzyme (Blood) was within normal limits as well. Magnetic resonance

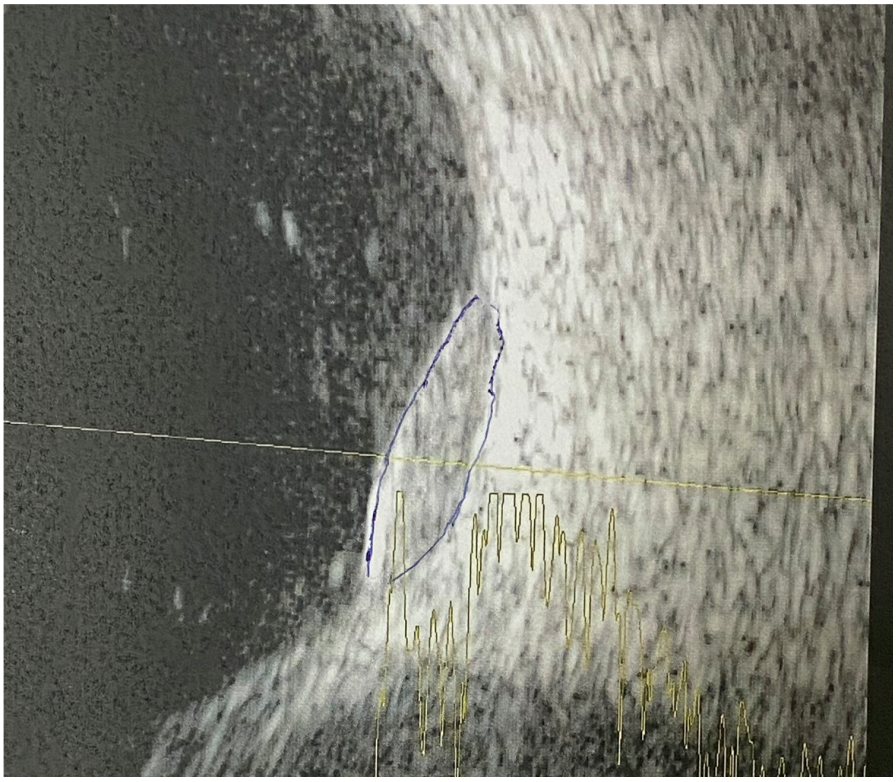

Figure 2. B-scan ultrasonography of the left eye showing choroidal mass with low to medium internal reflectivity.

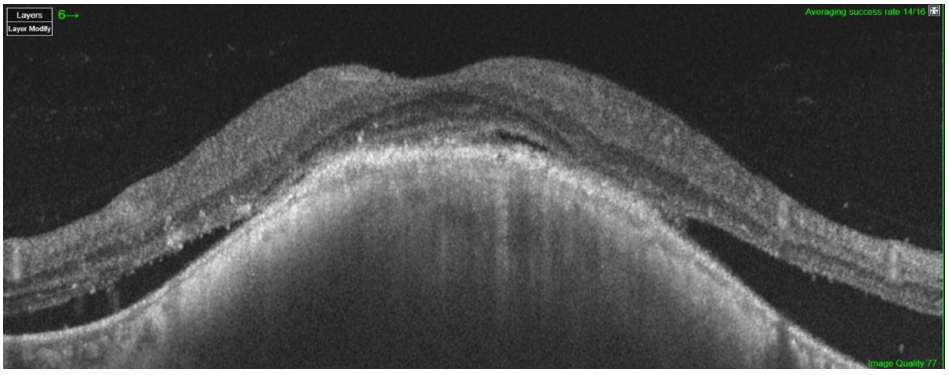

Figure 3. Optical Coherence Tomography showing large choroidal mass with subretinal fluid and hyperreflective subretinal (lipofuscin) deposits.

imaging (MRI) of the brain and spine was done as well as a computerized tomography (CT) scan of chest, abdomen, and pelvis done with no signs of malignancy or any other abnormalities.

The case was diagnosed as a left-eye choroidal melanoma with bilateral ocular paraneoplastic inflammation in the form of vasculitis. The patient was treated with oral prednisolone 60 mg daily tapering weekly by 5 mg for the vasculitis, and the patient was referred for plaque brachytherapy with close serial follow-up.

Before the brachytherapy in the serial follow-up, the mass documented an increase in size in the b-scan which reached a thickness of 4.8 mm, but the vasculitis was controlled on the oral prednisolone. (**Figure 4**)

After the brachytherapy, the patient's visual acuity dropped to 20/400 and he showed a decrease in the size of the mass in serial follow-ups documented by the b-scan, which reached 2.96 mm thickness with omit radiation retinopathy surrounding the area of the mass corresponding to the area of the brachytherapy as well as regression of vasculitis despite the stoppage of oral prednisolone as seen in (**Figure 4**). The patient was planned to regularly visit the clinic and he will be managed accordingly if there are any signs of inflammation or regrowth.

DISCUSSION

Uveal melanoma is the most common primary intraocular tumor with an incidence of 4 to 5 cases per year in the United States and slightly higher in Europe.[5,6] The majority of uveal melanoma patients are between 50 to 80 years of age. Multiple factors play a role in the pathogenesis of uveal melanoma including ethnicity and multiple intrinsic predisposing factors like lighter color of skin and conditions like oculodermal melanosis.[7] However, in this case, the patient was young and of Middle Eastern ethnicity, which we think contributed to the atypical presentation with bilateral vasculitis because a younger patient has a stronger immune system.[8]

To the best of our knowledge, this case is the second macular choroidal melanoma reported with bilateral vasculitis in the literature. The only similar previous case was reported in 1995 by Steel et al,[4] concerning a young male patient with left anterior unilateral choroidal melanoma and bilateral retinal vasculitis. In contrast to our case, the oral corticosteroid did

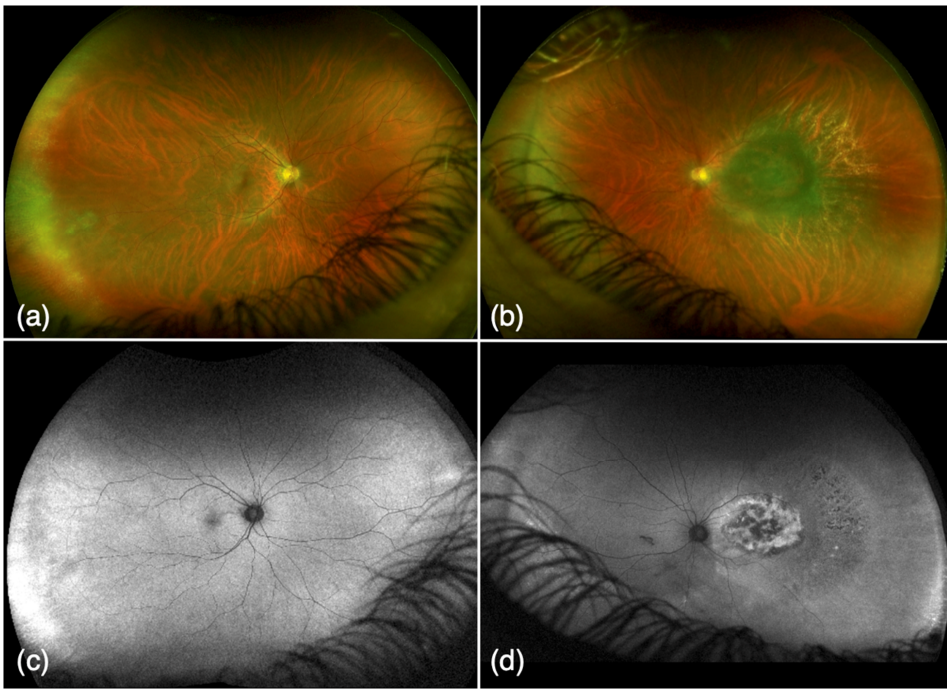

Figure 4. (a,b) Color Fundus Photo of both eyes showing (left eye) with pigmented choroidal lesion under the macula with areas of omit radiation retinopathy surrounding the mass. (c,d) Fundus Autofluorescence Photo showing hyperautofluorescence of the choroidal lesion (left eye).

not affect the retinal vasculitis in their case, but it decreased dramatically after the patient underwent enucleation. That was supported by the finding in our patient who had resolved vasculitis after brachytherapy and the shrinkage of the melanoma size despite the stoppage in oral corticosteroids.

Pathogenesis of bilateral vasculitis is unknown in both cases. However, it is speculated that there is a cross-reactivity between the melanoma and various retinal antigens resulting in altered antiretinal autoimmunity and subsequent retinal vasculitis.[4] This was reinforced by the fact that vasculitis has improved after enucleation in their patient. On the other hand, the association of unilateral uveitis with uveal melanoma is not infrequent. These cases are usually masquerading as panuveitis and are associated with anterior choroidal melanoma or tumor necrosis.[9] Sympathetic ophthalmia is also known as secondary to necrotic choroidal melanomas, often with extraskeletal extension.[10]

CONCLUSION

This article presents a noteworthy instance of choroidal melanoma accompanied by bilateral vasculitis. This report underscores the concealment potential of masquerade syndrome and intraocular hemorrhage for neoplastic origins, with uveal melanoma emerging as the predominant primary intraocular malignancy. Particularly in the context of small lesions, the diagnostic challenge arises from the diverse array of conditions that can mimic choroidal melanoma. When confronted with bilateral intraocular vasculitis and suspected neoplastic pathology, a heightened consideration for biopsy becomes imperative to ensure an accurate diagnosis. Further investigations are warranted to elucidate the association of bilateral vasculitis as an indicative marker of melanoma and to assess its overall incidence within this context.

REFERENCES

1. Seregard S. Posterior uveal melanoma: the Swedish perspective. Acta Ophthalmol Scand. 1996;74:315-29.
2. Kivela T. The epidemiologic challenge of the most frequent eye cancer: retinoblastoma, an issue of birth and death. Br J Ophthalmol 2009;93:1129–31.
3. Fraser DJ, Font RL. Ocular inflammation and hemorrhage as initial manifestations of uveal malignant melanoma: Incidence and prognosis. Arch Ophthalmol. 1979;97(7):1311–4.
4. Steel DH, Mahomed I, Sheffield E. Unilateral choroidal melanoma with bilateral retinal vasculitis. Br J Ophthalmol. 1996;80(9):850–1.
5. Field, Matthew G, Harbour, William J. National library of medicine. Current Opinion in Ophthalmology. 2014; 25(3):234-9.
6. Schoenfield, Lynn. Advances in anatomic pathology. National Library of Medicine. 2014;21(2):138-43.
7. Chen X, Wu Q, Tan L, et al. Methods in molecular biology. Oncogene. 2014;33(39):4724-34.
8. Hirokawa K, Matsuyama M, Kasai M, Kurashima C. Aging and immunity. Acta Pathol Jpn. 1992;42(8):537-48. doi: 10.1111/j.1440-1827.1992.tb03103.x.
9. Feng L, Zhu J, Gao T, et al. Uveal melanoma in the peripheral choroid masquerading as chronic uveitis. Optom Vis Sci. 2014;91:e222Ye225.
10. Duke-Elder S. Uveitis of unknown etiology. System of ophthalmology. Vol. IX. Diseases of the uveal tract. London: Henry Kimpton; 1966. p. 558-602.