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CASE REPORT

Choroidal malignant melanoma: the importance of ultrasonography

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Abstract

A 51-year-old Eastern Mediterranean male without known systemic medical illness referred to our clinic with a painless loss of vision for 2 months in the left eye. The diagnosis of choroidal malignant melanoma was suspected when fundus examination and ocular ultrasonography were done after the continuous deterioration in visual acuity. Orbital magnetic resonance imaging evaluation was consisted with choroidal melanoma and enucleation was performed. The post-operative histopathologic report confirmed the choroidal malignant melanoma. This report highlights the importance of B-scan ocular ultrasonography in evaluating an eye with deteriorating visual complaints.

Keywords: Intraocular tumor; malignant melanoma; uveal melanoma.

Uveal melanoma is the most common adulthood primary intraocular malignant tumor. The incidence is between 2 and 11/million every year. It roots from the melanin-producing melanocytes originating from neural crest that migrated to uvea.[1] In melanoma originating from the eye and adnexa, (82.5%) of cases were primary uveal melanoma. Around 85–90% of uveal melanoma cases arise from the choroid, 5–8% from the ciliary body, and 3–5% from the iris. Melanoma can also arise from conjunctiva, cornea, and other parts of the orbit.[1] Patients may come to the clinic with different signs and symptoms such as metamorphopsia, floaters, flashing of light or flickering, decreased vision, pain due to increased intraocular pressure, or even be asymptomatic. Iris melanoma can manifest as small or diffuse lesion, transparent or deeply pigmented.[2] Ciliary body melanoma presents with prominent episcleral vessels, secondary glaucoma, focal cataract, or lens subluxation that might cause asymmetric astigmatism. Choroidal melanoma presents as mushroom-shaped mass with varying size and overlying orange pigment (lipofuscin) with or without exudative retinal detachment that can be seen in B-scan ocular ultrasonography.[3] In this paper, we aim to present a choroidal malignant melanoma case with a unique clinical scenario.

Case Report

A 51-year-old Eastern Mediterranean male without medical illness presented to our hospital with a painless vision loss for 2 months in the left eye. The complaints started 4 months before with minimally decreased vision in the left eye. After that the patient started complaining of meta-

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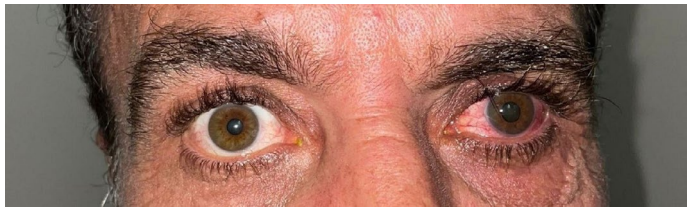

Fig. 1. Slit-lamp examination was unremarkable for the right eye while there was dilatation of episcleral vessels, rubeosis iridis, and opacifications in the lens in the left eye.

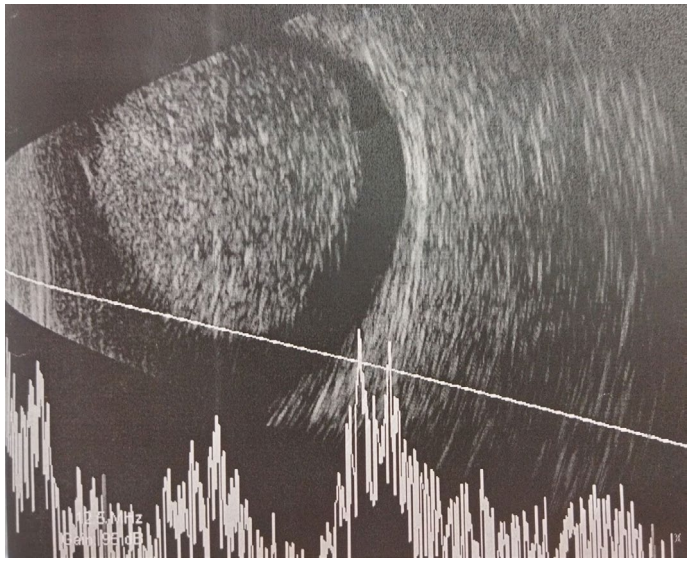

Fig. 2. Standardized B-scan ultrasound revealed a dome-shaped mass filling the vitreous in the left eye.

morphopsia and micropsia in addition to feeling of heaviness sensation in his eye, the visual acuity was getting worse progressively. Due to the war circumstances in Syria, no management was done, the patient was later examined by another ophthalmologist who suspected retinal detachment and performed B-scan ultrasound to the patient. The suspicion of an intraocular masses on ocular ultrasonography led to a referral to a tertiary care hospital. When the patient presented to us, his visual acuity was 20/20 in the right eye with no light perception in the left eye. Intraocular pressure was 15 mmHg and 44 mmHg in the right and left eye, respectively, using Goldmann applanation tonometry. Slit-lamp examination was unremarkable for the right eye while there was dilatation of episcleral vessels, rubeosis iridis, and lens opacifications in the left eye (Fig. 1). Fundus examination was normal in the right while in the left eye, there was a total retinal detachment appearance. Standardized B-scan ocular ultrasonography was performed and it was revealed a dome-shaped mass filling the vitreous in the left eye. The size of the lesion was 25 mm with moderate reflectivity (Fig. 2). After that, orbital magnetic resonance imaging (MRI) with

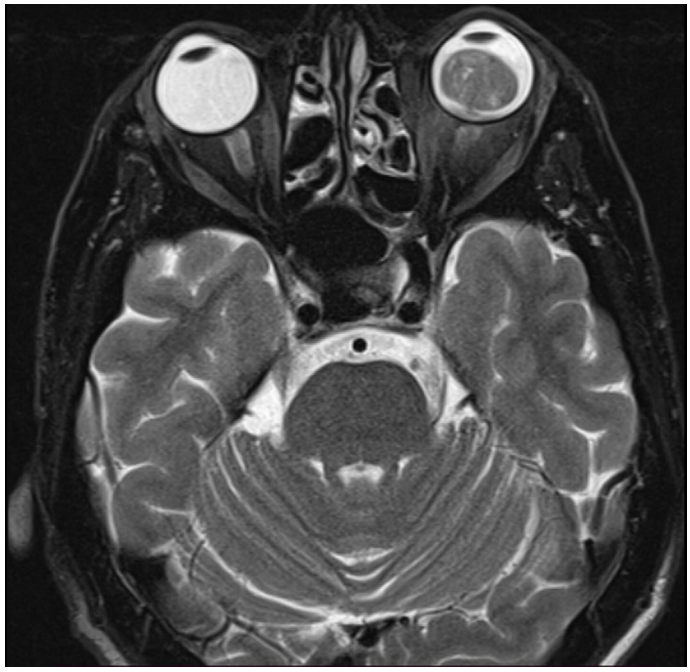

Fig. 3. The orbital magnetic resonance imaging (MRI) reported a little decrease in the size of the left globe compared with the right and a 25 mm well-circumscribed lesion inside the vitreous. T2-weighted axial section MRI of the orbit.

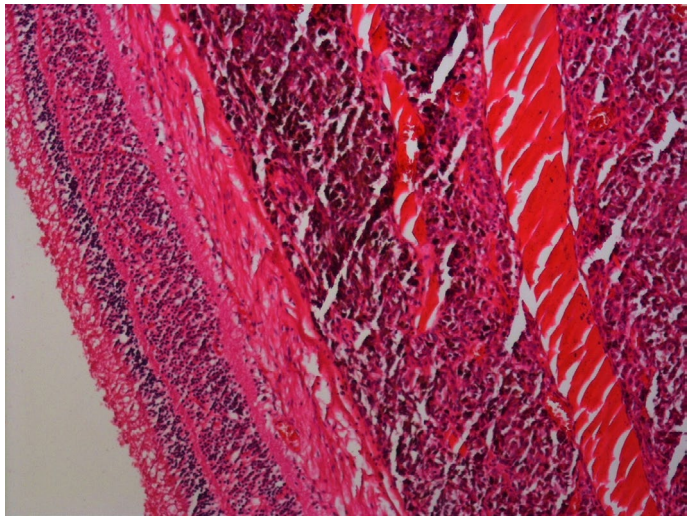

Fig. 4. H&E ×20 magnification showing tumor arising from choroid with densely hypercellular, intensely pigmented tumor pleomorphic epithelioid cells.

contrast was planned and the patient also was referred to oncology clinic. The orbital MRI reported a little decrease in the size of the left globe and a 25 mm hyperintense well-circumscribed lesion inside the vitreous (Fig. 3). Furthermore, positron emission tomography was done which reported a 20 mm × 18 mm the left intraocular lesion with increased metabolic activity, no other focus was detected in the body. The recommended treatment was enucleation of the left

eye based on the size of the lesion, the increased intraocular pressure, and the absence of visual potential. The specimen was sent for histopathologic evaluation. The histopathologic report showed increased mitotic activity with pleomorphic epithelioid cells confirming the diagnosis of malignant melanoma originating from the choroid (Fig. 4).

Discussion

Malignant uveal melanoma represents a challenge for clinicians especially when there is a lack of previous medical records or in case of atypical presentation such in our case. Diagnosis is mainly based on physical examination supported by B-scan ultrasonography.[1] In cases where the posterior segment cannot be clearly distinguished with opaque media, ocular ultrasonography should always be done with suspicion.[1,2] As seen in our case, ocular ultrasonography should be performed for possible malignancy in cases of increased intraocular pressure and rubeosis iridis. Choroidal melanoma can rarely simulate ocular inflammation; therefore, ophthalmologists should be careful in the differential diagnosis.[1-3] The mean age at diagnosis is around 60 years with approximately equal gender distribution (males: 52.3%, females: 47.7%).[3] In a study, the vast majority of cases were from Caucasian origin (98%) compared to Middle Eastern (<1%).[4] Various risk factors exist including fair skin and light eye color. Pre-existing hyperpigmented choroidal nevus, congenital ocular melanocytosis, or primary acquired melanosis were also risk factors. Furthermore, welding was identified as a risk factor.[5] In this report, the atypical presentation and lack of medical facilities due to the Syrian war led to relatively late diagnosis of the tumor. Furthermore, the aforementioned risk factors were absent. According to a multicentered, international registry-based study, around 50% of patients with uveal melanoma will suffer from metastasis and the most common site is the liver.[6] Among multiple prognostic factors such as the patient's age, local invasion to extraocular tissue, lymphatic infiltration and cell type, and others, the tumor size at the time of presentation is very important for metastasis risk.[7] As the size of primary tumor increases, the risk of metastasis increases. In our patient, the tumor size measured 25 mm by ocular ultrasonography and MRI which is considered to be large by the American Joint Committee on Cancer.[5] In general, uveal melanoma treatment is divided into enucleation or eye rescue treatment which is divided into either local surgical resection, radiotherapy, photodynamic therapy, or laser therapy.[8] In cases of choroidal melanoma, the treatment depends on multiple factors such as visual acuity, the tumor size and site, and presence of extraocular spread.[9] For large (more than 10 mm thickness and more than 16 mm in basal diameter) sized tumors, enucleation remains one of the best treatment options with no need for neoadjuvant radiotherapy.[10]

Conclusion

Although uveal melanoma is a rare cause of decreased vision, the survival of the patient and the vision can be preserved with a complete ophthalmologic examination. It is essential that ophthalmologists approach cases of deteriorating and unresolved visual complaints with a high sense of suspicion and perform ocular ultrasonography.

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