DOI: 10.5603/oj.94017

Bilateral uveal tumor — a case report

Karolina Bonińska[1, 2], Sławomir Lis[1], Marzena Szczepaniak[1], Sławomir Cisiecki[1, 2]

[1]Ophthalmology Department, Karol Jonscher Municipal Medical Center in Lodz, Poland
[2]Medical Center "Julianów" in Lodz, Poland

ABSTRACT

BACKGROUND: This case presentation reports a case of bilateral uveal tumor.
CASE PRESENTATION: An 82-year-old patient reported to the ophthalmology department complaining of a limited visual field temporally in his left eye. Visual acuity in the right eye was 0.9, and in the left eye, it was 0.4. Intraocular pressure was 18 mm Hg in each eye. Indirect ophthalmoscopy in the left eye revealed a choroidal tumor. The diagnosis of uveal melanoma was confirmed, and the patient underwent a ^{125}I brachytherapy procedure. During a follow-up visit, a tumor was observed to be spreading beyond the left eyeball, with evidence of metastatic deposits observed on the corneal endothelium. The patient underwent enucleation surgery. Two years later, the patient reported to our Ophthalmology Clinic with a significant deterioration of visual acuity in the right eye (0.2); intraocular pressure was 8 mm Hg. Indirect ophthalmoscopy revealed a choroidal tumor with subretinal fluid.
CONCLUSION: The location of bilateral choroidal melanoma should always be considered. It becomes crucial to carefully and regularly examine both eyes when uveal melanoma is found in one eye. The result of the examination may determine the type of treatment undertaken and, thus the prognosis for long-term survival.

KEY WORDS: uveal melanoma; bilateral tumor

Ophthalmol J 2023; Vol. 8, 138–141

INTRODUCTION

Uveal melanoma accounts for 3-5% of all melanomas in our body and is the most common intraocular neoplasm. It arises from melanocytes of the iris (3–5%), ciliary body (5–8%) and choroid (85–90%) [1–3].

It most often affects patients in the 6th-8th decade of life. The prognosis depends on the tumor's location, size, and histological subtype. Uveal melanomas are usually single, one-sided tumors [1–3].

In this paper, we present a rare case of cooccurrence of ciliary body melanoma and choroidal tumor in one eye and a choroidal tumor in the other eye.

CASE REPORT

An 82-year-old patient reported to the ophthalmology department in October 2017 complaining of a limited visual field in his left eye. Visual acuity with spectacle correction in the right eye was 0.9, and in the left eye, 0.4. Intraocular pressure was 18 mmHg in each eye.

Indirect ophthalmoscopy in the left eye revealed a choroidal tumor peripherally from the nose. An incipient cataract was found in the right eye. Ultrasonography (USG) of the eyeballs was performed, which confirmed a limited choroidal lesion with high internal echogenicity in the left eye. The patient was referred to the Ophthalmic Oncology Department with a suspicion of uveal melanoma to expand the diagnosis and implement an appropriate form of therapy.

The above diagnosis was confirmed. One month later, the patient underwent a ^{125}I brachytherapy procedure. In the following months, he remained under our observation.

CORRESPONDING AUTHOR:
Karolina Bonińska, MD, PhD, Centrum Medyczne "Julianów", ul. Żeglarska 4, 91–321 Łódź; e-mail: karolina.boninska@gmail.com

In February 2018, due to the features of radiation retinopathy with elevated intraocular pressure (left eye = 27.0 mm Hg), panretinal laser coagulation of the left eye was implemented. During several months of observation, features of tumor regression were noted on ultrasound examination.

Due to the deteriorating visual acuity of the right eye (0.4), the patient was referred for cataract surgery. In November 2018. phacoemulsification surgery was performed. Visual acuity in the right eye at discharge was 0.7 and light perception in the left eye.

During a follow-up visit a month later, a tumor was observed to be spreading beyond the left eyeball with evidence of metastatic deposits observed on the corneal endothelium (Fig. 1). The patient was referred to the Ophthalmic Oncology Center for urgent consultation to exclude local recurrence.

After the consultation, the patient was qualified for left eyeball enucleation surgery. This procedure was performed in January 2020. The histopathological examination revealed a mixed type of melanoma tumor of the choroid and ciliary body of the left eye. The patient remained under our observation.

Six months later, the visual acuity in the right eye was 0.8. Intraocular pressure was 9 mm Hg. The anterior and posterior segments of the right eye were within normal limits for his age norm. Topical levofloxacin was prescribed 4 times daily to the left orbit to manage a persistent purulent discharge secondary to the enucleation.

Meanwhile, the oncologist's check-up revealed numerous metastases to the liver, presenting a guarded prognosis without any further treatment options.

In January 2021, the patient reported to our Ophthalmology Clinic with a significant deterioration of visual acuity in the right eye (0.2 cc +1.5 dsph); intraocular pressure was 8 mm Hg. Indirect ophthalmoscopy revealed a choroidal tumor with subretinal fluid. We performed swept source-optical coherence tomography (SS-OCT), which confirmed the presence of a limited, domed choroidal tumor with subretinal neurosensory fluid (Fig. 2). An ultrasound scan showed a tumor with high internal echogenicity within the macula (Fig. 3). The patient was referred to the Ophthalmic Oncology Department for urgent consultation. Even before the scheduled appointment, the patient died.

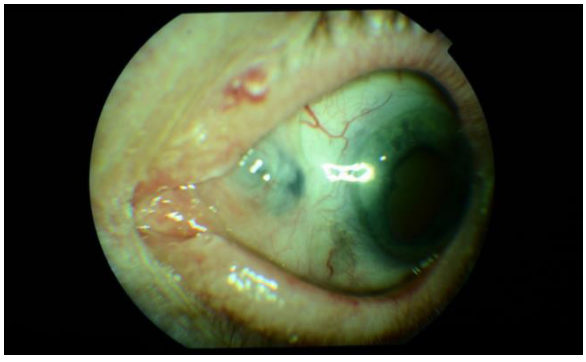

FIGURE 1. Ciliary body melanoma spreading beyond the left eyeball

DISCUSSION

Our case report concerns bilateral uveal melanoma which occurs with frequency of about 0.18%.

Bilateral uveal melanoma presented by us is rare and occurs with a frequency of about 0.18% [4]. Although histopathological diagnosis for the left eye tumor is not available to confirm the diagnosis of a melanoma, the ultrasound studies support this diagnosis. No predisposition to its occurrence has been found in terms of gender, race, mean age at diagnosis, location of the lesion, or presence of distant metastases [4].

Bilateral localization may suggest a genetic predisposition. BAP1 (BRCA-associated protein 1) tumor predisposing syndrome mutation has been identified recently as germline cancer syndrome, which can lead to several cancers, including bilateral uveal melanoma, and follows an autosomal-dominant inheritance pattern [5, 6]. According to Rai et al., analysis of 174 cases of patients with germline BAP1 mutation revealed the most common tumours to be uveal melanoma (31%), mesothelioma (22%), cutaneous melanoma (13%), renal cell carcinoma (10%), breast carcinoma (10%), atypical Spitz tumour (18%) and others [6, 7].

The infiltration of the optic nerve promotes the formation of distant metastases. They are found to occur in 15% at 5 years, 25% at 10 years, and 36% at 20 years [6]. The most common location for them is the liver, affecting 40–60% of patients. In addition, metastases occur in the lungs, bones, skin, and brain [8–11].

The average survival after diagnosis of liver metastasis does not exceed 2–11 months [12–15]. For single metastases, surgical treatment is used, while the primary treatment for multiple metastatic foci is chemotherapy with the use of dacarbazine [16].

The differential diagnosis of choroidal tumors includes primarily choroidal pigmented nevi,

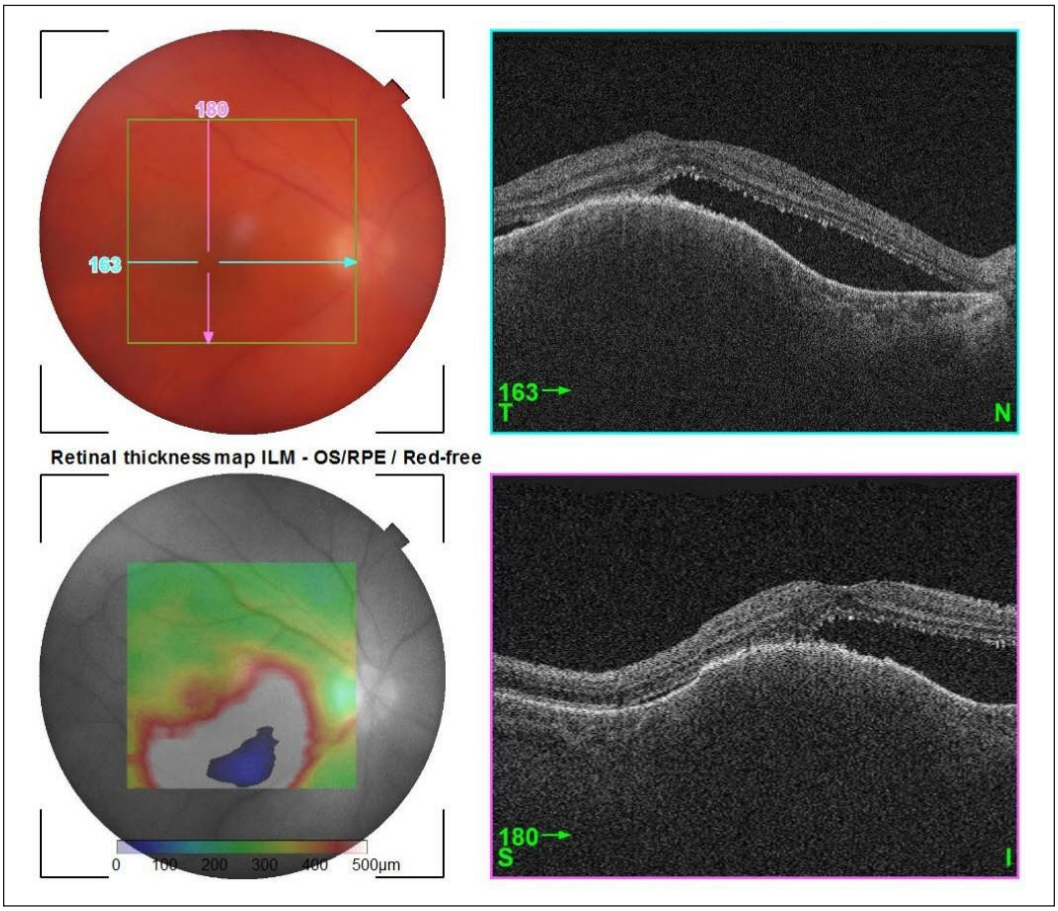

FIGURE 2. Swept source-optical coherence tomography (SS-OCT) of the right eye showing choroidal thickening with subretinal fluid

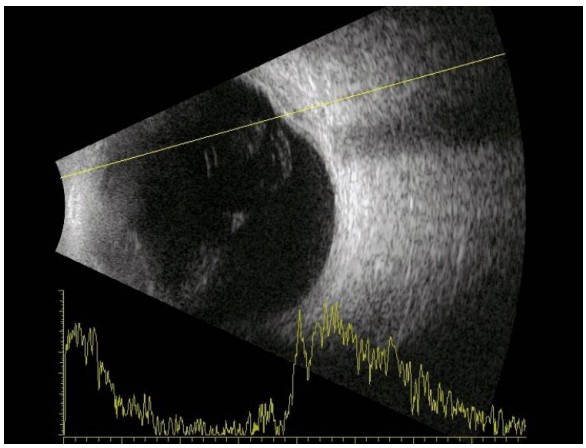

FIGURE 3. A B-scan ultrasonography of the right eye showed a tumor with low internal reflectivity

age-related macular degeneration, choroidal hemangioma, hemorrhagic choroidal detachment, vortex veins distension, subretinal hemorrhage, retinal pigment epithelial hyperplasia, choroidal ossification, toxoplasmic choroiditis, melanocytoma, and retinal astrocytoma [17]. Ultrasonography is extremely helpful in diagnosing and monitoring clinical changes of ocular tumors as the internal reflectivity or the presence of the so-called choroidal incision is pathognomonic, as discussed in the SOCT study [17].

The location of bilateral choroidal melanoma should always be taken into account. It becomes crucial to carefully and regularly examine both eyes when uveal melanoma is found in one eye. The examination result may determine the type of treatment undertaken and, thus, the prognosis for long-term survival. Shields et al. also suggest that all patients with bilateral uveal melanoma and those with history of mesothelioma, cutaneous melanoma, renal cell carcinoma and other cancers should be tested for germline BAP1 tumor predisposition syndrome. Patients identified as positive should have relatives tested and should maintain routine screening for related cancers [6].

Conflict of interest

No conflict of interest.

Funding

No financial support.

REFERENCES

1. McLaughlin CC, Wu XC, Jemal A, et al. Incidence of noncutaneous melanomas in the U.S. Cancer. 2005; 103(5): 1000–1007, doi: 10.1002/cncr.20866, indexed in Pubmed: 15651058.
2. Singh AD, Topham A, Singh AD, et al. Incidence of uveal melanoma in the United States: 1973-1997. Ophthalmology. 2003; 110(5): 956–961, doi: 10.1016/S0161-6420(03)00078-2, indexed in Pubmed: 12750097.
3. Damato B. Progress in the management of patients with uveal melanoma. The 2012 Ashton Lecture. Eye (Lond). 2012; 26(9): 1157–1172, doi: 10.1038/eye.2012.126, indexed in Pubmed: 22744385.
4. Scott JF, Vyas R, Galvin J, et al. Primary bilateral uveal melanoma: a population-based study and systematic review. Clin Exp Ophthalmol. 2018; 46(5): 502–510, doi: 10.1111/ceo.13129, indexed in Pubmed: 29219254.
5. Eide N, Garred Ø, Beiske K, et al. Bilateral uveal melanomas with different gene expression detected with 7 years interval. Acta Ophthalmol. 2016; 94(1): 99–102, doi: 10.1111/aos.12857, indexed in Pubmed: 26496992.
6. Shields CL, Kaliki S, Furuta M, et al. Clinical spectrum and prognosis of uveal melanoma based on age at presentation in 8,033 cases. Retina. 2012; 32(7): 1363–1372, doi: 10.1097/IAE.0b013e31824d09a8, indexed in Pubmed: 22466491.
7. Rai K, Pilarski R, Cebulla CM, et al. Comprehensive review of BAP1 tumor predisposition syndrome with report of two new cases. Clin Genet. 2016; 89(3): 285–294, doi: 10.1111/cge.12630, indexed in Pubmed: 26096145.
8. Żygulska-Mach H. Epidemiology. In: Skowronek J, Mackiewicz A. ed. Czerniak złośliwy. Termedia, Poznan 1998: 145–149.
9. Becker JC, Terheyden P, Kämpgen E, et al. Treatment of disseminated ocular melanoma with sequential fotemustine, interferon α, and interleukin 2. Br J Cancer. 2002; 87(8): 840–845, doi: 10.1038/sj.bjc.6600521, indexed in Pubmed: 12373596.
10. Camp R, Lind DS, Hemming AW. Combined liver and pancreas resection with biochemiother- apy for metastatic ocular melanoma. J Hepatobiliary Pancreat Surg. 2002; 9(4): 519–521, doi: 10.1007/s005340200066, indexed in Pubmed: 12483277.
11. Zygulska-Mach H. [Modern treatment of malignant intraocular melanomas]. Klin Oczna. 1996; 98(1): 61–66, indexed in Pubmed: 9019580.
12. Alexander HR, Libutti SK, Bartlett DL, et al. A phase I-II study of isolated hepatic perfusion using melphalan with or without tumor necrosis factor for patients with ocular melanoma metastatic to liver. Clin Cancer Res. 2000; 6(8): 3062–3070, indexed in Pubmed: 10955785.
13. Kulik A, Olszewska A, Bulski W, et al. Brachytherapy in the conservative treatment of malignant intraocular melanomas. Cancer. 1998; 48: 1077–1088.
14. Stankiewicz A, Figurska M. Melanoma of the eye. Co Oncol. 2003; 7: 589–593.
15. Weinreich DM, Alexander HR. Transarterial perfusion of liver metastases. Semin Oncol. 2002; 29(2): 136–144, doi: 10.1053/sonc.2002.31681, indexed in Pubmed: 11951211.
16. Biedka M, Makarewicz R. Liver metastases in patients with ocular melanoma. Pol Palliat Med. 2005; 4(2).
17. Romanowska-Dixon B, Jakubowska B. Melanoma of the Vascular Membrane. Differential diagnosis of intraocular neoplasms. Krakow 2014.