Case Reports in Ophthalmology

Case Rep Ophthalmol 2023;14:421–425
DOI: 10.1159/000527087
Received: March 16, 2022
Accepted: September 14, 2022
Published online: September 6, 2023

Case Report

Presumed Melanocytoma-Associated Choroidal Neovascular Membrane with Hemorrhage Successfully Treated with Intravitreal Aflibercept Injections

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Keywords
Aflibercept · Anti-VEGF · Choroidal neovascular membrane · Eylea · Melanocytoma

Abstract
A patient presented with melanocytoma and associated choroidal neovascular membrane with hemorrhage involving the macula. The patient was treated with monthly aflibercept (Eylea) injections with significant improvement of best corrected visual acuity. In this report, we explore the development of a choroidal neovascular membrane (CNVM) formation in a patient with melanocytoma and the effect of intravitreal aflibercept (Eylea) on disease course. Case report study used patient data obtained from examination and imaging. The patient was treated with monthly intravitreal aflibercept injections leading to complete resolution of CNVM and hemorrhage, with significant improvement of best corrected visual acuity. Awareness and proper monitoring for the sequelae of melanocytoma are important for early detection and prevention of visually threatening outcomes. In cases of melanocytoma-associated CNVM formation with large subretinal hemorrhage, intravitreal aflibercept can be an effective tool for inducing CNVM regression and allowing improvement of visual acuity.

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Introduction

Melanocytoma is a densely pigmented nevus classically found unilaterally at the optic disc. This benign lesion is derived from uveal melanocytes and is thought to be a congenital lesion that becomes clinically apparent in adulthood. Pathogenesis of melanocytoma is unknown at this time. Mean age of diagnosis of this lesion is 50 years, and some studies have demonstrated a predilection for Caucasian patients.

Although largely classified as a benign and visually insignificant lesion, rare associations between melanocytomas and local complications such as optic disc edema, intraretinal/subretinal fluid, retinal vein occlusion, and focal hemorrhage have been reported [1]. Upon review of available peer-reviewed literature, only 8 cases of melanocytoma-associated choroidal neovascular membrane (CNVM) have been reported. In this report, we document a rare case of an optic disc melanocytoma with secondary CNVM formation and resultant large subretinal hemorrhage, managed by a series of intravitreal aflibercept (Eylea) injections.

Case Report

A 75-year-old Caucasian male presented with new onset metamorphopsia and paracentral scotoma of the left eye (OS). Past ocular history was significant for peripapillary melanocytoma (shown in Fig. 1a, b) which had been stable for 6 years prior to this presentation and epiretinal membrane OS (not pictured). Past medical history was significant for use of apixaban at the time of presentation. On the examination, Snellen best corrected visual acuity was 20/25 right eye (OD) and 20/150 OS. Visual acuity was noted to be 20/70 OS 3 months prior. Intraocular pressure was 12 in the OD and 13 OS. Anterior examination was unremarkable in both eyes (OU). Fundus examination OD was significant only for postsurgical changes following membrane peeling for epiretinal membrane several years prior. Fundus examination of the OS revealed an inferotemporal peripapillary optic disc melanocytoma measuring 1 mm in greatest basal diameter with an associated large area of subretinal hemorrhage extending into the macula and abutting the fovea (shown in Fig. 1c).

OCT over the optic nerve demonstrated irregularities in Bruch's membrane, which are consistent with the diagnosis of a presumed CNVM. Unfortunately, fluorescein angiography was not available at presentation (Fig. 1d).

The patient was diagnosed with melanocytoma with presumed associated CNVM. Anti-VEGF therapy was initiated. Aflibercept was chosen due to treating provider preference in managing CNVM given the presence of a large fovea-threatening subretinal hemorrhage. He underwent a total of 6 aflibercept injections (2 mg/0.05 mL) over the course of 10 months. During this time, the subretinal fluid resolved and the subretinal hemorrhage largely resolved. Treatment ceased once there was no longer clinically evident fresh hemorrhage and subretinal fluid resolved on OCT (Fig. 1f). His vision improved to 20/60 OS eccentrically with residual paracentral scotoma. He also noted significant subjective improvement with resolution of metamorphopsia and decrease in the size and density of the perceived scotoma. After resolution of the presumed CNVM and macular hemorrhage, the patient underwent pars plana vitrectomy with epiretinal membrane peel with further improvement of vision to 20/30 at postoperative month 1.

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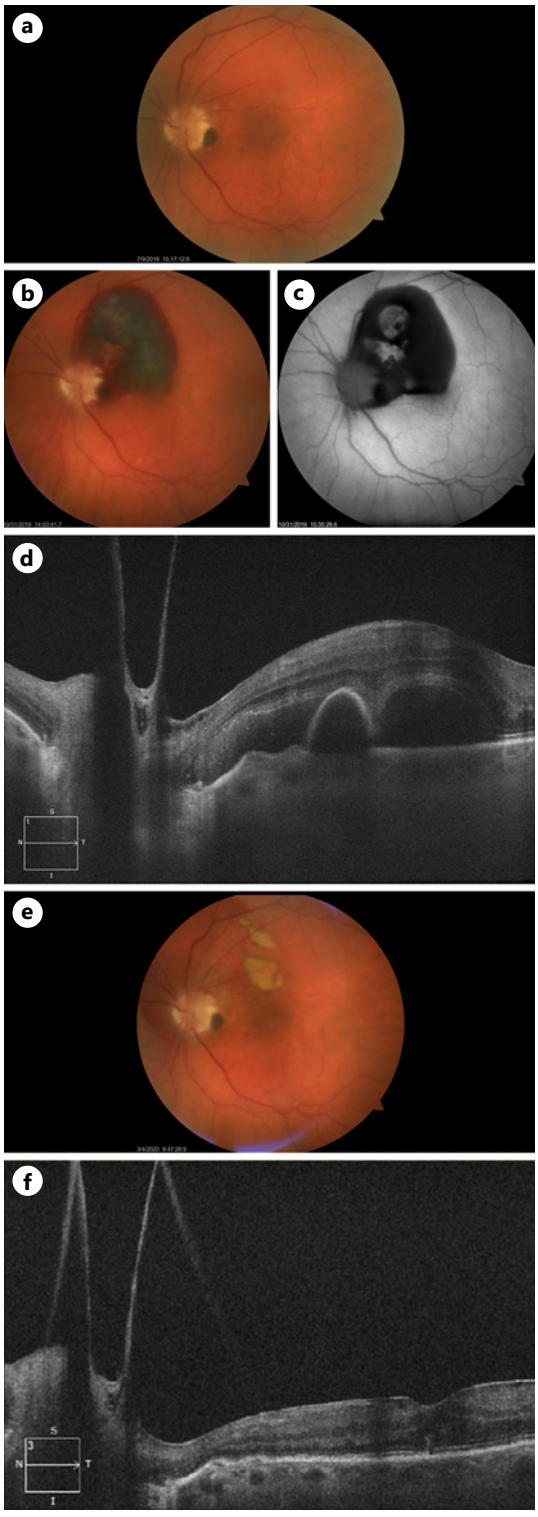

Fig. 1. Fundus photo from before the subretinal hemorrhage (**a**) demonstrated an optic disc melanocytoma in the 4 o'clock position. Fundus photo (**b**) and fundus autofluorescence (**c**) after presumed CNVM formation demonstrated subretinal hemorrhage emanating from the optic disc melanocytoma and abutting the fovea. OCT (**d**) demonstrated subretinal and sub-RPE hemorrhage consistent with the fundus examination. Fundus photo from after resolution of the presumed CNVM and hemorrhage (**e**) demonstrated 2 distinct areas of residual subretinal fibrin. OCT (**f**) from after aflibercept treatment course demonstrated resolution of hemorrhage and subretinal fluid consistent with presumed regressed CNVM.

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Discussion

Optic disc melanocytoma is a benign pigmented variant of a melanocytic nevus that typically remains stable and is not associated with visually significant complications. In a study of 115 eyes with melanocytoma, Shields et al. [1] found the most commonly associated complications to be optic disc edema (25%), retinal edema (16%), localized subretinal fluid (14%), retinal exudation (12%), retinal hemorrhage (5%), vitreous seeds (4%), and retinal vein occlusion (3%). Resultant decrease in visual acuity secondary to melanocytoma is estimated to occur in about 26% of cases [1]. The most common mechanism for visual compromise is mild retinal exudation involving the fovea or neuroretinitis from tumor necrosis [2, 3]. Choroidal neovascular membrane in association with optic nerve melanocytoma is an exceedingly rare complication. Though uncommon, melanocytoma-associated CNVM has the potential to result in significant long-term visual compromise. The majority of those cases reported have been associated with small subretinal hemorrhages [4–9].

In rare cases of melanocytoma-associated CNVM, a variety of different treatments have been reported including photodynamic therapy and intravitreal anti-vascular endothelial growth factor (anti-VEGF) injections. Chalam et al. [10] describe a papillomacular CNVM secondary to optic disc melanocytoma. In that case, treatment with photodynamic therapy resulted in scarring of the CNVM and improvement in visual acuity from 20/200 to 20/30. Three cases were treated with intravitreal bevacizumab and one with intravitreal ranibizumab [4–7]. Two of the three cases utilizing bevacizumab therapy required only one injection for resolution of fluid and prolonged stability of the CNVM. One reported case of melanocytoma-associated CNVM treated with a series of intravitreal aflibercept injections has been reported by Hamza et al. [8]. This case presented with macular edema without hemorrhage, unlike the case presented in the current report. The previously reported case demonstrated aflibercept to be effective in producing long-term CNVM regression after three monthly injections with 13 months of follow-up. Prolonged anti-VEGF treatment may be required for more severe pathology including persistent retinal hemorrhage, persistent subretinal or intraretinal fluid, or progression of CNVM. Both our case and that described by Hamza and colleagues required multiple injections. This highlights the fact that CNVM associated with melanocytoma may lead to visual compromise and in some cases require an extended treatment course.

In summary, although many cases of melanocytoma remain stable and isolated without associated complications, these lesions can be associated with choroidal neovascular membrane formation which may lead to visual compromise through accumulation of subretinal fluid or hemorrhage. Awareness and proper monitoring for the sequelae of melanocytoma are important for early detection and prevention of visually threatening outcomes. In cases of melanocytoma-associated CNVM formation with large subretinal hemorrhage, intravitreal aflibercept can be an effective tool for inducing CNVM regression and allowing improvement of visual acuity.

Statement of Ethics

This study protocol was reviewed and need for approval was waived by VA Healthcare (VISN 4) Multi-Site Institutional Review Board. This included the waiver of the need of written informed consent from the patient to publish the case report and any accompanying images.

Conflict of Interest Statement

All authors have no financial disclosures or conflicts of interest.

Funding Sources

No funding support.

Author Contributions

Austin S. Yu and Jordan A. Sugarman: manuscript drafting and analysis. Stephanie J. Weiss: manuscript final review and data acquisition.

Data Availability Statement

The authors confirm that the data supporting the findings of this study are available within the article. Further inquiries can be directed to the corresponding author.

References

1 Shields JA, Demirci H, Mashayekhi A, Eagle RC Jr, Shields CL. Melanocytoma of the optic disk: a review. Indian J Ophthalmol. 2019;67(12):1949–58.
2 Shields JA, Demirci H, Mashayekhi A, Shields CL. Melanocytoma of optic disc in 115 cases: the 2004 samuel johnson memorial lecture, part 1. Ophthalmology. 2004;111(9):1739–46.
3 Archdale TW, Magnus DE. Melanocytoma of the optic disc. J Am Optom Assoc. 1993;64(2):98–103.
4 Kamisasanuk T, Uchino E, Sakamoto T. Choroidal neovascularization of optic disk melanocytoma treated with bevacizumab. Eur J Ophthalmol. 2012;22(3):503–5.
5 Batmanabane V, Soman M, Nair U. Management of optic disc melanocytoma associated with choroidal neovascularisation and macular detachment. J Ophthalmol Clin Res. 2015;2(3):1–4.
6 Al-Halafi AM. Successful treatment of melanocytoma associated choroidal neovascular membrane with intravitreal bevacizumab. Saudi J Ophthalmol. 2013;27(2):117–9.
7 Urrets-Zavalia JA, Crim N, Esposito E, Correa L, Gonzalez-Castellanos ME, Martinez D. Bevacizumab for the treatment of a complicated posterior melanocytoma. Clin Ophthalmol. 2015;9:455–9.
8 Hamza HS, Moussa M, Elhusseiny AM. Management of choroidal neovascularization associated with optic disc melanocytoma with intravitreal aflibercept: a case report and review of the literature. Case Rep Ophthalmol Med. 2019;2019:2672798.
9 Garcia-Arumi J, Salvador F, Corcostegui B, Mateo C. Neuroretinitis associated with melanocytoma of the optic disk. Retina. 1994;14(2):173–6.
10 Chalam KV, Gupta SK, Shah GY, Agarwal S. Successful management of melanocytoma-associated choroidal neovascularization with photodynamic therapy. Eur J Ophthalmol. 2006;16(5):776–8.