



Unilateral Ocular Mass in a Patient with Migraine Headaches

Sayena Jabbehdari, Michael J Heiferman, William F Mieler*

Department of Ophthalmology and Visual Sciences, University of Illinois, USA

Abstract

Purpose: This case report discusses a case of unilateral ocular mass and migraine headaches in a 28-year-old female who was referred to the retina service.

Observations: Funduscopic examination, Spectral Domain Optical Coherence Tomography (SD-OCT) and B-scan echography revealed diffuse choroidal thickening with moderate homogeneity and alteration of choroidal color. Additionally, there was asymmetry in the appearance of the optic nerves. All of these findings accompanied with the history of glaucoma led us to ask the patient specifically if she had a facial hemangioma earlier in childhood. Reviewing photographs from her childhood demonstrated the presence of a prominent facial hemangioma with left upper eyelid involvement, confirming the diagnosis of diffuse choroidal hemangioma in the setting of Sturge-Weber Syndrome (SWS).

Conclusion and Importance: Since the patient had no Cystoid Macular Edema (CME), subretinal fluid, and/or retinal detachment, our management consisted of close observation with serial dilated fundus examinations.

Keywords: Choroidal thickness; Diffuse choroidal hemangioma; Ocular mass; Sturge-weber syndrome

Introduction

Choroidal hemangiomas are an uncommon and benign vascular hamartoma, which are associated with uveal tract abnormality in patients with Sturge-Weber Syndrome (SWS) [1,2]. SWS is a sporadic neurocutaneous disorder presenting with facial nevus flammeus (Port-wine stain), epilepsy, leptomeningeal angiomas, ipsilateral intracranial hemangioma, glaucoma, developmental delay and cognitive impairment [3,4]. Choroidal hemangiomas are classified as either Circumscribed Choroidal Hemangiomas (CCH), which are more common in non-syndromic cases, or Diffuse Choroidal Hemangiomas (DCH) [5], which are often associated with SWS. DCH often times manifests as an asymptomatic ill-defined choroidal mass with unclear borders [3]. The main concern of DCH is vision loss that is often secondary to non-rhegmatogenous retinal detachments, amblyopia, cystoid degeneration and/or CME [6]. Exudative retinal detachment with subretinal fluid is seen in 50% of patients with DCH and SWS [7]. The localized choroidal lesion over diffuse choroidal thickness is often times seen in patients with DCH who develop exudative retinal detachments [7].

Case Presentation

A 28-year-old female was referred to the retina service for evaluation of a left ocular mass lesion. Prior to presentation, the patient developed headaches described as a pressure sensation over the left eye without any associated change in baseline visual function. A CT scan was obtained revealing an ocular mass lesion described as a 1.4 cm × 0.7 cm × 0.5 cm ovoid, well-circumscribed homogenous mass in the left posterior upper aspect of the globe. Past ocular history included mildly decreased vision in the left eye since childhood, without any recent acute vision changes. The patient had a past medical history of migraine headaches and glaucoma. There was no past medical history of trauma, cancer, infectious, inflammatory, or autoimmune conditions. She had no known drug allergies and was not taking any medications except a brimonidine eye drop OS. Reviews of systems as well as family and social history were unremarkable.

On ocular examination, the best-corrected visual acuity was 20/20 OD and 20/60 OS. Extraocular movements were intact, and pupils were round and reactive. There was a relative afferent pupillary

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*Correspondence:

William F Mieler, Department of Ophthalmology and Visual Sciences, University of Illinois, Illinois, 1905 West Taylor Street Chicago, IL 60612, USA, E-mail: wmieler@uic.edu

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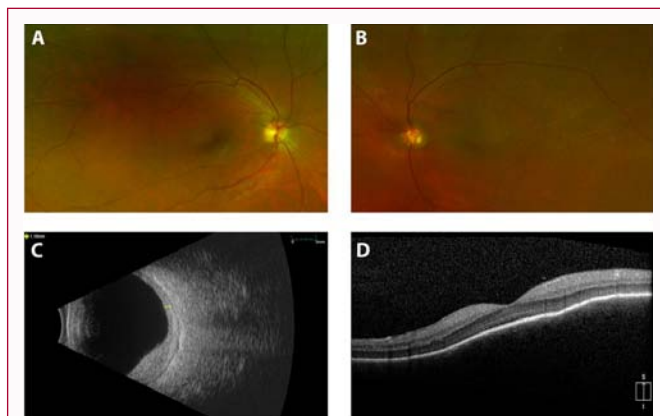


Figure 1: A) Color fundus photograph of the right and B) Left Eye. C) B-scan photograph of the left eye demonstrating diffuse choroidal thickening with moderate homogeneity. D) Spectral-domain optical coherence tomography image of the macula in left eye.



Figure 2: Presence of facial hemangioma with left upper eyelid involvement.

defect in the left eye. Intraocular pressure by applanation was 14 mmHg OD and 24 mmHg OS. Anterior and posterior segment examinations were unremarkable except for dilated episcleral vessels and glaucomatous optic nerve in the left eye, respectively (Figure 1A, 1B). B-scan echography of the left eye was subsequently performed revealing diffuse choroidal thickening with moderate homogeneity (Figure 1C). SD-OCT revealed normal retinal architecture without retinal pigment epithelium atrophy, retinal exudation, or subretinal fluid (Figure 1D). The presence of diffuse choroidal thickening, alteration of choroidal color and asymmetry in the appearance of the optic nerves accompanied with the history of glaucoma led us to ask the patient specifically if she ever had laser treatment of a facial hemangioma. Reviewing her childhood's photographs demonstrated the presence of facial hemangioma with left upper eyelid involvement (Figure 2). The differential diagnosis for ill-defined diffuse choroidal thickening includes choroidal melanoma, choroidal metastasis, choroidal osteoma, choroidal hemangioma, posterior uveitis, central serous chorioretinopathy, posterior scleritis, Vogt-Koyanagi-Harada's syndrome, and hypotony maculopathy/retinopathy. In accordance with our patient's history of left facial hemangioma with left upper eyelid involvement and characteristic "tomato-ketchup" appearance, the diagnosis of unilateral diffuse choroidal hemangioma associated with SWS was made.

Discussion

There have been numerous treatments reported in patient with DCH including low-dose External Beam Radiation Therapy

(EBRT), [3] Photodynamic Therapy (PDT) [7,8] anti-VEGF (reported for CCH), [9] Radioactive Plaque therapy [5,10], Trans-Pupillary Thermotherapy (TTT), oral propranolol and thermal laser photocoagulation [7]. Although DCH with exudative retinal detachment is classically treated with EBRT, this often times requires three to four weeks of treatment [3]. Radioactive plaque therapy has been shown to be superior to photocoagulation in patients with CCH and subretinal fluid [10]. Photodynamic therapy with verteporfin is another approach that works to selectively destroy tumor vasculature and helps to resolve subretinal fluid in DCH [7,8]. Others have suggested performing PDT or thermal laser in combination with a VEGF inhibitor, as a useful alternative in cases with CCH and subretinal fluid [9]. All these options should be considered in cases with exudative retinal detachment or CME. Conversely, cases with no evidence of retinal detachment or macular involvement (our patient) should be monitored closely with periodic observation. Given that the lesion was not proximal to the fovea and had no CME or subretinal fluid, there was no indication for PDT, anti-VEGF antibodies, or EBRT.

Conclusion

Our management consisted of close observation with serial dilated fundus examinations. Our patient was extensively informed about her condition and prognosis. At her most recent follow-up appointment at one-year post-presentation, the patient's visual acuity and examination were stable.

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