Available online at www.sciencerepository.org

Science Repository

Case Report and Review of the Literature

Spontaneous and Indentation Subretinal Hemorrhage from Choroidal Melanoma

Pedro A Rebello[1], Melina C Morales[1,2], Juliana HS Mascato[1], Rodrigo B Fernandes[1,2] and Arthur G Fernandes[1,2*]

[1]Ophthal Hospital Especializado, Sao Paulo, Brazil

[2]Department of Ophthalmology and Visual Sciences, Paulista Medical School, Federal University of Sao Paulo, Sao Paulo, Brazil

ARTICLE INFO

Article history:
Received: 27 May, 2022
Accepted: 16 June, 2022
Published: 29 June, 2022

Keywords:
Choroid neoplasms
choroid hemorrhage
differential diagnosis
ophthalmology

ABSTRACT

Choroidal melanoma is the most common primary intraocular malignancy in adults. Herein, the authors report two cases of subretinal hemorrhage associated with it. The first case presented with a large choroidal mass associated with subretinal hemorrhage, no history of trauma and choroidal melanoma diagnosis on ultrasound. The second case was initially diagnosed with retinal detachment but during the surgery a pigmented subretinal mass was observed, starting to bleed after scleral depression. Subretinal hemorrhage can occur due to ruptures in Bruch's membrane spontaneously or precipitated by inadvertent trauma. Despite an unusual manifestation, it should always be considered avoiding misdiagnosis.

Introduction

Choroidal melanoma is the most common primary intraocular malignancy in the adult population. The incidence of uveal melanoma has been reported as 5.1 cases per million people and it has remained stable since the early 1970's [1]. These tumors are generally unilateral and known risk factors include fair skin, light-coloured eyes, 30 age over 60 years, preexisting choroidal nevus, congenital ocular melanocytosis and *BAP1* gene mutation [2, 3].

Most patients are asymptomatic and diagnosis mainly occurs during routine ophthalmic examination. The onset of symptoms is closely related to the location of the tumor and its associated complications. Low visual acuity is the most common symptom, usually related to foveal exudative retinal detachment or direct tumor invasion of the macula [4].

Classical features of choroidal melanoma include dome shaped lesion (75%), total pigmentation or some degree of pigmentation (84%), and exudative retinal detachment (71%). Subretinal hemorrhage is a less common feature reported in 2.9-10% of cases of choroidal melanoma [4-7]. The purpose of this study is to report two cases with subretinal hemorrhage associated with choroidal melanoma.

Case Report

Case 1

A 51-year-old man complaining of worsened visual acuity and loss of inferior visual field in his left eye (OS) for one week, was first examined by a general ophthalmologist that referred him to our service with the diagnosis of rhegmatogenous retinal detachment. He had no relevant ophthalmic history, and his past medical history showed only systemic arterial hypertension. The patient denied any ocular trauma.

Upon examination, the visual acuity was 0.0 LogMAR (20/20) in the right eye (OD) and 1.0 LogMAR (20/200) in OS. Fundus examination of OD was unremarkable. In OS, subretinal fluid was present at the posterior pole, and a pigmented dome-shaped choroidal mass was visible in the periphery of the temporal sector, associated with subretinal hemorrhage and perilesional retinal detachment. On ultrasound examination, the mass proved to be solid, with 5.93mm in high, medium-low internal reflectivity, homogeneous structure, kappa angle sign, choroidal excavation and internal vascularization evident on the kinetic exam. During the fluorescein angiography (FA) exam, leakage was observed, with pinpoints bordered for hypofluorescence (blockage due to subretinal hemorrhage). Figure 1 illustrates the clinical evaluation.

Correspondence to: Arthur Gustavo Fernandes, MPH, Ph.D., Ophthal – Hospital Especializado Ltda, Rua Ministro Gabriel de Rezende Passos, 500, São Paulo, Brazil; Tel: +551150547700; E-mail: arthur_abz@yahoo.com.br

http://dx.doi.org/10.31487/j.JSCR.2022.01.03

Such findings were consistent with the diagnosis of choroidal melanoma. Cranial, chest and abdominal computed tomography revealed no metastases. Brachytherapy was recommended, and the patient is currently waiting for the procedure.

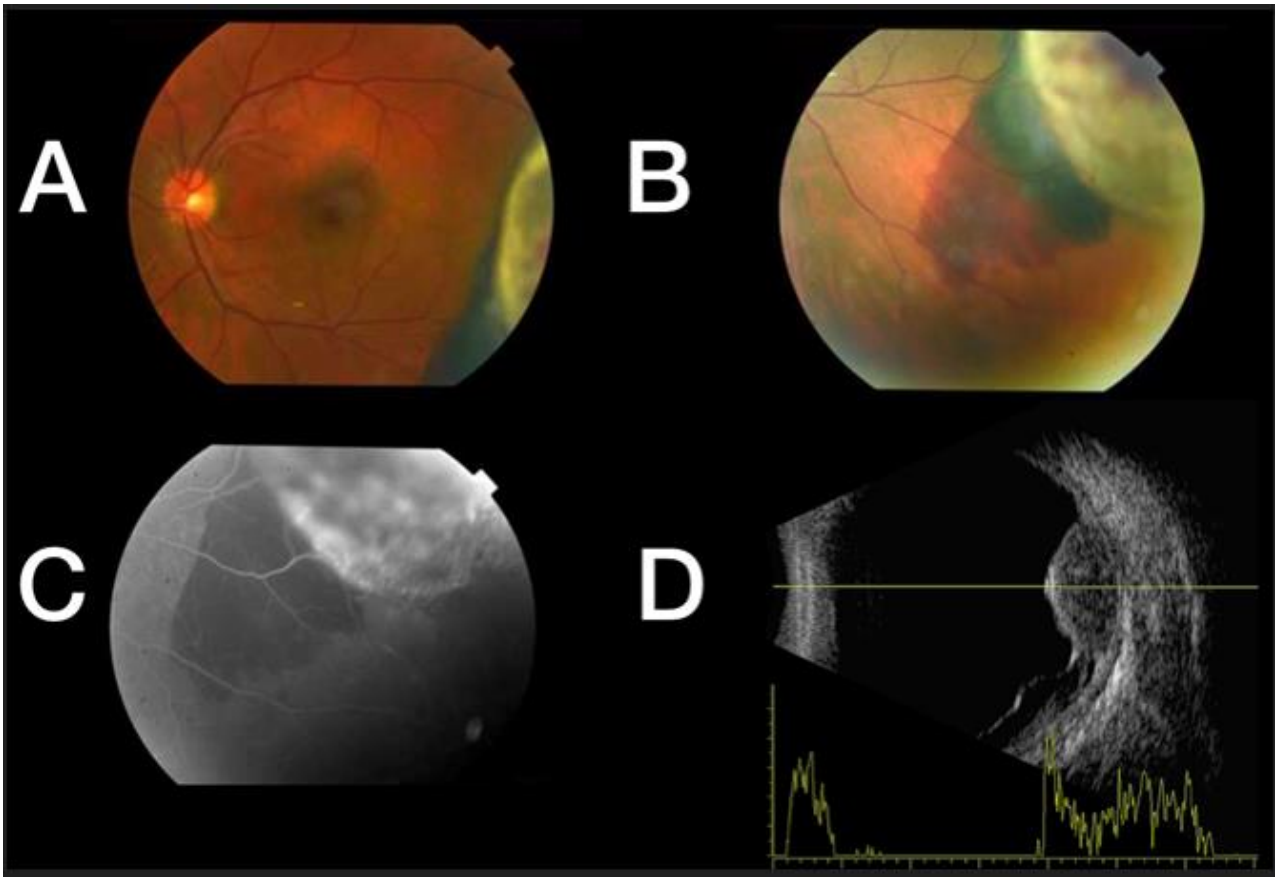

Figure 1: Clinical findings: **A)** Loss of foveal reflex due to subretinal fluid. Choroidal mass at temporal periphery. **B)** Pigmented choroidal mass associated with subretinal hemorrhage. **C)** Angiofluoresceinography: The tumor showed late staining hyperfluorescence and hypofluorescence due to blockage at the site of subretinal hemorrhage. **D)** Transverse ultrasound image shows typical characteristics of choroidal melanoma: solid dome-shaped mass with medium-low internal reflectivity (kappa angle sign) and choroidal excavation. The kinetic examination showed internal vascularization and margins with retinal detachment and subretinal hemorrhage.

Case 2

A 44-year-old female patient with high myopia attended the emergency department, complaining of low visual acuity in the right eye, starting one week after phacoemulsification with intraocular implant in OD at another service. On ophthalmological examination, the best corrected visual acuity was light perception in OD. The anterior segment showed no significant changes. At fundoscopy, a retinal detachment with macula off was present (retinal tear was not found). Primary Pars Plana Vitrectomy (PPV) with C3F8 and endolaser was indicated.

The patient underwent surgery one week later. Intraoperatively a round retinal tear nasal to the optic disc without vitreous traction was discovered in the posterior pole. After hyaloid detachment and vitreous shaving, air fluid exchange was performed to flatten the retina, a pigmented dome-shaped subretinal mass became evident in the upper temporal region. Perfluorooctane was injected, and during vitreous base shaving with scleral indentation, the tumor started to bleed to the subretinal space. Internal drainage of the subretinal fluid was performed through the posterior rupture during the air fluid exchange and silicone oil was implanted as a vitreous substitute.

Ophthalmoscopy and ultrasonography were compromised due to blood overlying the lesion and the presence of silicon oil. Hence, Magnetic Resonance Imaging (MRI) was indicated for initial evaluation of the lesion by the ocular oncology department, it was suggestive of choroidal melanoma. With reabsorption of the subretinal blood over the mass, it was possible to see a typical pigmented choroidal melanoma at fundus ophthalmoscopy, with increased size when compared to intraoperative images. Two months after VVP, the patient underwent enucleation of the OD. Complementary screening systemic examinations did not show metastases. Histopathological findings confirmed the diagnosis of mixed choroidal melanoma, with a predominance of spindle B type fusocellular cells (70%) and epithelioid cells (30%), without extra-scleral extension. Figure 2 shows intraoperative images of tumor indentation and progression of the bleeding, and also tumor rapid growth e histopathological confirmation. The patient continues to be followed up at our retinal and oncology eye service for 2 years now, in addition to regular follow-up with a clinical oncologist.

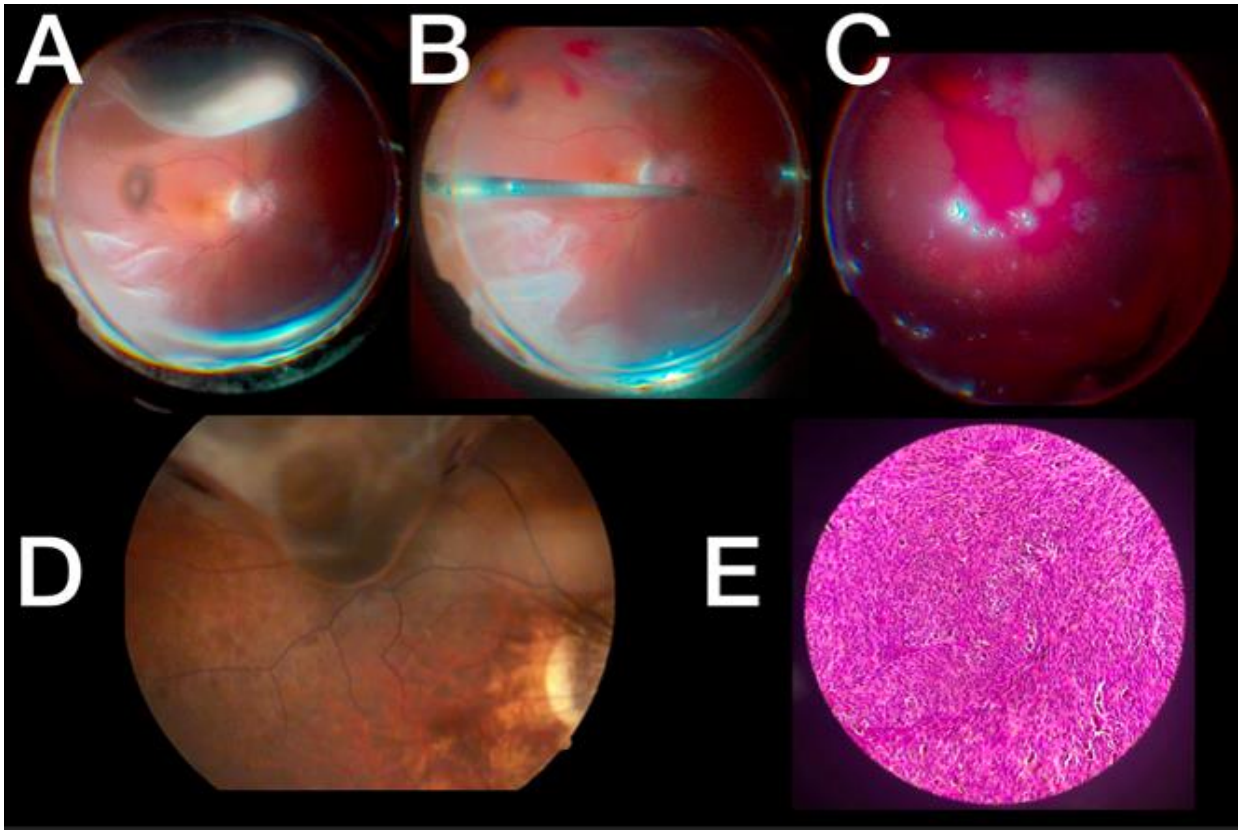

Figure 2: Clinical findings: (A, B, C) Photographs taken intraoperatively. **A)** Rhegmatogenous retinal detachment associated with a pigmented mass bulging the upper retina. Photograph taken during scleral indentation. **B)** Subretinal hemorrhage begins immediately after scleral indentation. **C)** The hemorrhage increased during the surgical procedure, eventually progressing to the posterior pole. **D)** 2 months after surgery, hemorrhage over the tumor reabsorbed, fundus retinography showed a pigmented mushroom-shaped tumor secondary to Bruch's membrane rupture, with increased size when compared to intraoperative images. **E)** After enucleation, histological analysis confirms the diagnosis of mixed choroidal melanoma.

Discussion

Subretinal hemorrhage is an uncommon feature that may be present in choroidal melanoma cases. There are three other case reports in the literature associating subretinal hemorrhage and choroidal melanoma [6-8]. When faced with a picture of extensive subretinal hemorrhage, the main syndromic differential diagnoses that must be remembered are caused by benign conditions divided into 2 main groups: choroidal neovascularization and trauma. It is essential to think and exclude conditions such as: age-related macular degeneration (AMD), polypoid choroidal vasculopathy (PCV), peripheral exudative hemorrhagic chorioretinopathy (PEHCR), ruptured arterial macroaneurysm, angioid streaks and others. A previous study has shown cases of extensive subretinal hemorrhage with ultrasound and magnetic resonance exams wrongly pointing to choroidal melanoma diagnosis when histopathological analysis later showed to be hemorrhages caused by degenerative conditions [9]. Subretinal hemorrhage in cases of intraocular neoplasms, may occur by tumor bleeding itself or secondary to choroidal neovascularization.

The first form is the most common in choroidal melanomas and occurs due to ruptures in the Bruch's membrane by progressive growth in the thickness of the tumor, progressing with tumor prolapse to the subretinal space. Progressive compression of the tumor occurs as if it were a tourniquet, leading to a decrease in venous return (stasis and venous dilation), which can progress to vascular rupture, causing subretinal hemorrhage. The occurrence of vascular rupture and subretinal hemorrhage appears spontaneously or precipitated by inadvertent trauma, such as scleral depression, brachytherapy plaque implantation and transillumination. Therefore, in view of the suspicion of choroidal melanoma, caution is essential when performing maneuvers that generate compressions in the eyeball [6, 7, 10]. In our second case, the patient had subretinal hemorrhage in the presentation and absence of a previous history of eye trauma but in the first case, the subretinal hemorrhage was triggered by surgical scleral depression.

The second mechanism capable of explaining subretinal hemorrhage is secondary to choroidal neovascularization, but there is only one case report of choroidal neovascularization associated with choroidal melanoma [8]. Other choroidal tumors are more associated with this subretinal bleeding mechanism, mainly choroidal osteoma and, more rarely, choroidal nevus.

Diagnostical differentiation of causes of subretinal hemorrhage apart from Posterior Vitreous Detachment and Retinal tears should be considered by ophthalmologists, especially by vitreoretinal specialists. The occurrence of subretinal hemorrhage associated with choroidal

melanoma should be seen as a factor with a probable unfavourable evolution, due to the increased risk of systemic metastasis [5].

Despite the fact that the vast majority of subretinal hemorrhage cases are associated with benign and controllable conditions, less common pathologies, like intraocular tumors, can also be associated with subretinal hemorrhage. In the face of such suspicion, untimely maneuvers of manipulation and ocular compression should be performed with caution.

Conflicts of Interest

None.

Funding

None.

REFERENCES

1. Singh AD, Turell ME, Topham AK (2011) Uveal melanoma: trends in incidence, treatment, and survival. *Ophthalmology* 118: 1881-1885. [Crossref]
2. Carbone M, Ferris LK, Baumann F, Napolitano A, Lum CA et al. (2012) BAP1 cancer syndrome: malignant mesothelioma, uveal and cutaneous melanoma, and MBAITs. *J Transl Med* 10: 179. [Crossref]
3. Chattopadhyay C, Kim DW, Gombos DS, Oba J, Qin Y et al. (2016) Uveal melanoma: From diagnosis to treatment and the science in between. *Cancer* 122: 2299-2312. [Crossref]
4. Singh P, Singh A (2012) Choroidal melanoma. *Oman J Ophthalmol* 5: 3-9. [Crossref]
5. Shields CL, Furuta M, Thangappan A et al. (2009) Metastasis of uveal melanoma millimeter-by-millimeter in 8033 consecutive eyes. *Arch Ophthalmol* 127: 989-998. [Crossref]
6. Choxi H, Sinha N, Shields CL (2004) Choroidal Melanoma Camouflaged by Extensive Subretinal Hemorrhage. *Curr Opin Ophthalmol* 15: 244-251.
7. Oellers P, Wolkow N, Jakobiec FA, Kim IK (2018) Hemorrhagic choroidal melanoma. *Am J Ophthalmol Case Rep* 10: 105-107. [Crossref]
8. Lubin JR, Gragoudas ES, Albert DM (1982) Choroidal neovascularization associated with malignant melanoma: a case report. *Acta Ophthalmol (Copenh)* 60: 412-418. [Crossref]
9. Cong C, Lin J, Wang L, Wang Y et al. (2014) [Clinical and pathological observation of seven cases of spontaneous sub-retinal hemorrhage]. *Zhonghua Yan Ke Za Zhi* 50: 737-742. [Crossref]
10. Shields JA, Shields CL (2008) Intraocular Tumors: An Atlas and Textbook, 2nd ed. Philadelphia, PA: Lippincott Williams & Wilkins 85-175.