Case Report

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Coexistence of choroidal melanoma and rhegmatogenous retinal detachment: a case report and literature review

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Introduction and importance: The simultaneous occurrence of rhegmatogenous retinal detachment (RRD) and choroidal melanoma is extremely rare, and diagnosis of choroidal melanoma in RRD patients is challenging. As a result, choroidal masses in RRD patients tend to be overlooked, resulting in delayed treatment. The authors report a rare case presenting with simultaneous choroidal melanoma and RRD, and the authors review the related literature.

Case presentation: A 45-year-old Thai man who presented with inferior RRD and choroidal elevation in the left eye was examined using fundoscopy and ultrasonography. The presumptive diagnosis was simultaneous RRD and haemorrhagic choroidal detachment (CD). Vitrectomy and external drainage were attempted but were unsuccessful in draining fluid from the presumed haemorrhagic CD. Although the retina was reattached, the choroidal elevation remained unchanged. After being lost to follow-up, the patient returned later with severe proptosis in the left eye. MRI findings suggested a presumptive diagnosis including choroidal melanoma and choroidal metastasis, which choroidal melanoma was later confirmed through histopathological examination after exenteration.

Clinical discussion: Important clinical clues were provided for distinguishing between choroidal melanoma and haemorrhagic CD in cases of coexisting RRD.

Conclusions: In the differential diagnosis of RRD with suspicious choroidal elevation, the possibility of the presence of choroidal melanoma should be considered. In particular, in cases where haemorrhagic CD fails to drain during surgery, the possibility of underlying choroidal melanoma should be investigated.

Keywords: choroidal melanoma, magnetic resonance imaging, rhegmatogenous retinal detachment, ultrasonography

Introduction

Choroidal melanoma (CM) is the most common primary intraocular malignancy found in adult Caucasian populations, yet it is uncommon in Asian and African nations[1]. Clinical manifestations of CM encompass blurred vision, photopsia, floaters, visual field loss, visible tumour, pain, and metamorphopsia; however, around 30% of CM cases are asymptomatic at the time of diagnosis, and the rate of misdiagnosis in patients with CM has previously been reported to be as high as 23%[2]. Commonly presenting as a dome-shaped mass or with a mushroom configuration, the lesion can be pigmented or non-pigmented, and it can be associated with retinal detachment, intraocular haemorrhage, or extraocular extension[3].

HIGHLIGHTS

- Simultaneous occurrence of choroidal melanoma and rhegmatogenous retinal detachment highlights the importance of considering the possibility of underlying choroidal melanoma in cases with suspected choroidal detachment or haemorrhagic choroidal detachment.
- Preoperative ocular imaging, along with biopsy during retinal detachment repair surgery, can effectively diagnose and prevent tumour invasion and metastasis.

The simultaneous occurrence of rhegmatogenous retinal detachment (RRD) and CM is extremely rare, with less than 1% of CM coexisting with RRD[4]. RRD with choroidal elevation can be caused by choroidal detachment (CD), which is more common than haemorrhagic CD[5,6], or by choroidal tumour, which includes CM[4,7–12]. Diagnosis of CM in RRD patients is challenging, and the choroidal mass in RRD patients tends to be overlooked, resulting in delayed treatment. For CM, early diagnosis and treatment not only preserves vision but also reduce rates of metastasis and can prolong patients' survival time. At present, there is a lack of data related to cases presenting with simultaneous RRD and pathologically proven CM, especially in Asia[4,7–10,12]. One such case is presented in this report. This case report has been reported in line with the SCARE Criteria[13].

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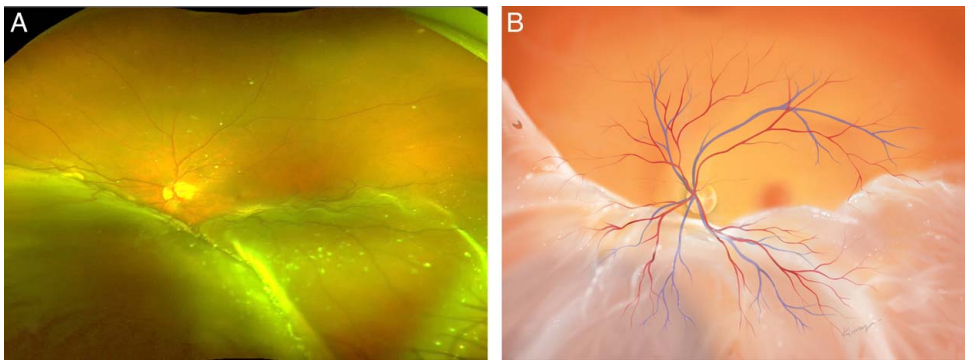

Figure 1. Ultra-widefield fundus photo (A) and schematic diagram (B) of the left eye showing inferior bullous retinal detachment overlying a large choroidal elevation at the inferonasal retina.

Case presentation

A 45-year-old Thai man was referred to our retina clinic due to RRD which had been causing hazy vision in his left eye for 1 month. He denied any history of flashes or floaters, myopia, or trauma in either eye, and he had otherwise been healthy. At initial eye examination, his best-corrected visual acuity was 20/20 in the right eye and 20/250 in the left. The intraocular pressure was 14 mmHg in the right eye and 16 mmHg in the left. There was no proptosis and no abnormality in eyelids or ocular adnexa. Slit-lamp examination demonstrated normal anterior segment and early senile cataracts in both eyes. Posterior segment examination was unremarkable in his right eye, while fundus examination of the left eye showed macula-off inferior bullous retinal detachment with small horseshoe tear at the 10 o'clock region (Fig. 1). At the inferonasal retina, a large non-pigmented choroidal elevation was discovered underneath the detached retina. B-scan ultrasonography (US) showed a dome-shaped heterogeneous hyperechoic mass and subretinal fluid, and A-scan ultrasonography revealed moderate to high internal reflectivity. The lesion measured 12.15 mm thick by 15.05 mm in the largest basal diameter (Fig. 2). Diagnosis of RRD with haemorrhagic CD in the left eye was made.

Phacoemulsification with intraocular lens implantation and pars plana vitrectomy, as well as endolaser and silicone oil tamponade, were performed by an experienced vitreoretinal surgeon Intraoperative findings revealed total retinal detachment with a peripheral retinal break at the superonasal retina and large choroidal elevation at the inferonasal retina extending to the ora serrata (Fig. 1). External drainage of haemorrhagic CD was also performed but failed to drain any fluid or pigment granules. After the surgery, the retina was reattached with unchanged choroidal elevation. The patient was not informed about the ongoing issue of choroidal elevation, as the possibility of choroidal melanoma had not been considered at that point. However, due to a concerning choroidal elevation, he was scheduled for a follow-up appointment for 1 week after his discharge from the hospital. Unfortunately, he was lost to follow-up for 3 months during the COVID-19 pandemic but subsequently revisited the clinic with progressive painful proptosis in the left eye. His best-corrected visual acuity was 20/20 in the right eye and poor light projection in the left. Anterior segment was obscured due to marked corneal oedema precluding fundus examination. Detailed assessment of the posterior segment in the left eye was not possible due to image artifacts seen during ocular US in the silicone-filled eye. MRI was therefore performed and demonstrated moderate hyperintensity on fat-saturated T1-weighted image and homogeneous hyperintensity on fat-saturated T2-weighted image (Fig. 3).

The radiologist's presumptive diagnosis included choroidal melanoma, choroidal metastasis, and choroidal haemorrhage. The results of systemic workup, including computed tomography

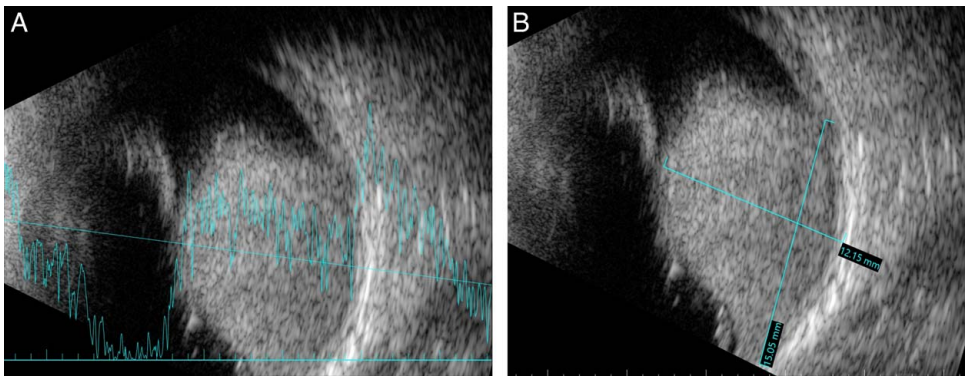

Figure 2. Preoperative ultrasonography of the choroidal elevation revealing a dome-shaped heterogeneous hyperechoic mass and subretinal fluid in the B-scan ultrasonogram, and moderate to high internal reflectivity in the A-scan ultrasonogram (A). The largest basal diameter of the lesion was 15.05 mm by 12.15 mm thick (B).

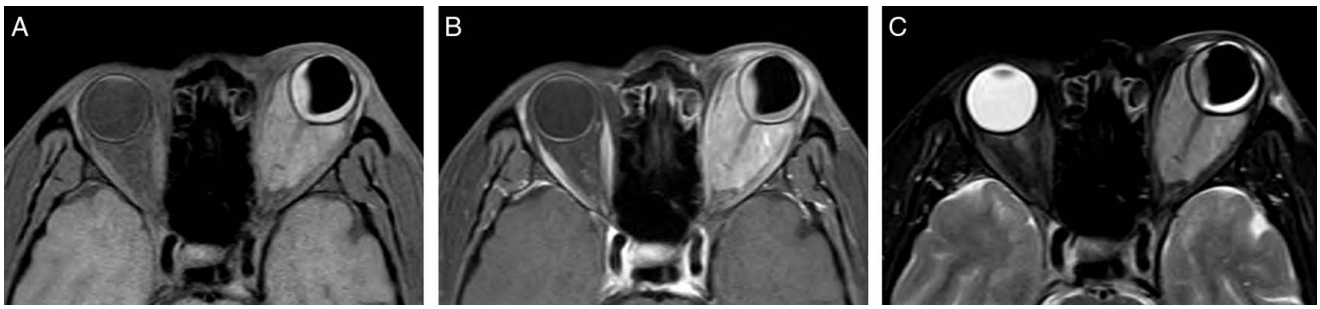

Figure 3. Axial magnetic resonance image of the orbits shows thickening of the posterior choroid of the left eye globe and a lobulated extraocular retrobulbar mass. The retrobulbar mass totally occupies retroorbital fat, encasing the left optic nerve and causing left eye proptosis. These lesions exhibit moderate hyperintensity on fat-saturated T1-weighted image (A), homogeneous enhancement on contrast-enhanced fat-saturated T1-weighted image (B), and homogeneous hyperintensity on fat-saturated T2-weighted image (C).

(CT) of the thorax, US of the upper abdomen, liver function test, alpha-fetoprotein, carcinoembryonic antigen, and cancer antigen 19-9 were unremarkable. Primary diagnosis of CM was considered at this point. The left eye was exenterated after comprehensive discussion with the patient. The histopathological examination confirmed diagnosis of epithelioid type choroidal melanoma (Fig. 4).

Discussion

Our case highlights the importance of being aware of the possibility of coexistence of RRD and CM, as CM presents with choroidal elevation similar to CD, which is more common. For patients with RRD with choroidal elevation, the B-scan US is an essential tool for ruling out CD from CM and haemorrhagic CD. However, although the ultrasonogram in our case was able to exclude CD due to its higher echogenicity, it could not exclude haemorrhagic CD from CM. Although extremely rare, the coexistence of RRD and CM, or RRD with haemorrhagic CD, may be encountered, and it poses a dilemma for clinical practice because the management of each is completely different; therefore, both early detection and early treatment are critical for a positive long-term survival outcome in CM. Table 1 displays a literature review regarding patients with simultaneous CM and RRD. Most previous reports have included Caucasians and clinical manifestation revealed CM and subretinal fluid communication, and normal or near normal intraocular pressure. There have been only a few cases, along with ours, which have yielded confirmatory biopsy results. Table 2 demonstrates clinical indicators for distinguishing between CM and haemorrhagic CD in coexisting RRD. Aside from hypertension and systemic anticoagulation, which were previously identified as risk factors for spontaneous haemorrhagic CD, high myopia was discovered to be an important risk factor for a patient with simultaneous RRD and haemorrhagic CD[5,6]. Imaging studies could also play a vital role in differentiating between these two conditions. For ultrasonography, despite the similarity of the lesion echogenicity, the distinguishing power of the test will be evident if haemorrhagic CD coexists with CM[14]. Orbital imaging with MRI is not commonly used in the evaluation of CM because clinical examination features and B-scan US are usually enough to make the diagnosis[2]. However, in our case, concomitant RRD and suspicious choroidal elevation made diagnosis more difficult, necessitating MRI. In spite of the similarity of signal intensity of both entities demonstrated by MRI, vivid contrast enhancement after gadolinium injection can help differentiate CM from CD and haemorrhagic CD[15–18]. This imaging method also has an important role in the evaluation of extraocular extension of CM[14,15].

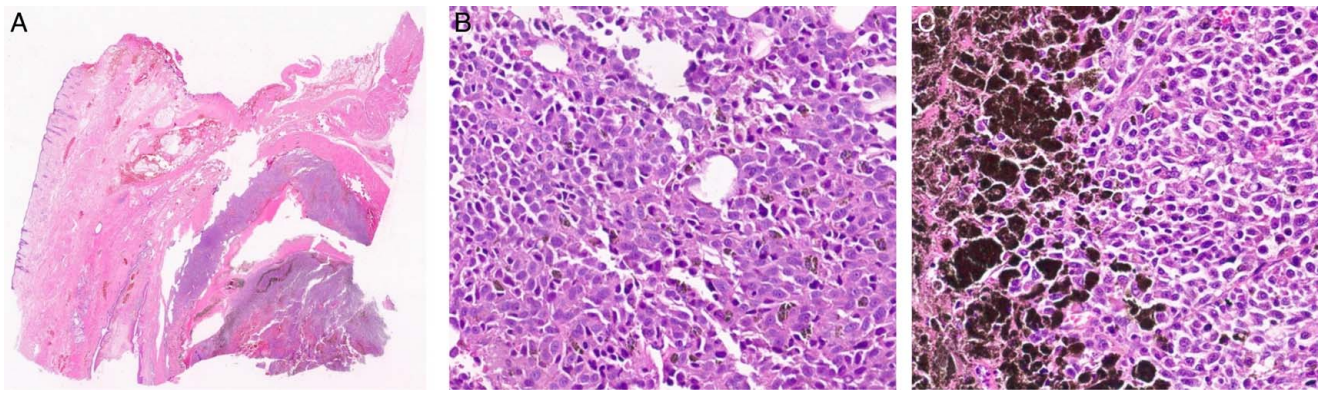

Figure 4. Histopathological sections of the left orbital content revealing epithelioid cell type uveal melanoma (A). The characteristic mitotic count: 15 mitoses/1 sq. mm (B), ciliary body involvement (C). (magnification, ×40).

Table 1

A literature review relating to patients with simultaneous choroidal melanoma and rhegmatogenous retinal detachment

Author (year)	Age/ sex/ eye	Country	Character of choroidal abnormality	Communication between SRF and CM	IOP of affected eye (and other eye)	Confirmatory imaging at initial presentation or referral to hospital	Methods of diagnosis of CM before treatment	Delayed diagnosis after RRD diagnosis and the cause	Treatment	Type of CM and outcome
Present study (2023)	45/M/L	Thailand	Haemorrhagic CD anterior to the equator	Yes	16 (14)	US	Clinical	3 months, loss to follow-up	PPV, external drainage, endolaser, silicone oil tamponade for RRD Exenteration	Epithelioid cell type No evidence of tumour metastasis
Robertson *et al.* (1971)[7]	62/F/R[a]	USA	Large solid-appearing mass located at posterior pole to oral serrata	Yes	20 (20)	NA	Clinical	Not clear (referred case)	Enucleation	Mixed-cell type
Haimovici *et al.* (1996)[8] (only 3 cases associated with CM were selected)	66/M/R	USA	Haemorrhagic CD	No	NA	US	Clinical	6 months, postoperative vitreous haemorrhage	Scleral buckle with external drainage for RRD Proton beam irradiation	Reattached retina No pathological examination Tumour regression with no evidence of tumour growth
	54/M/R	USA	Undifferentiated elevated mushroom-shaped pigmented mass	No	NA	US	Clinical	3 months, postoperative vitreous haemorrhage	Pneumatic retinopexy (C_2F_6) for RRD Iodine-125 brachytherapy	Reattached retina No pathological examination Tumour regression with no evidence of tumour growth
	69/M/L	USA	Pigmented choroidal tumour at temporal periphery	Yes	NA	US	Clinical	No	Scleral buckle for RRD Iodine-125 brachytherapy	Reattached retina No pathological examination Tumour regression with no evidence of tumour growth
Lakosha *et al.* (1999)[4]	74/F/L[a]	Canada	Pigmented mass at the posterior pole	Yes	18 (18)	US, FA	Clinical	Not clear (referred case)	Iodine-125 brachytherapy	No pathological examination Decrease in the height of the tumour No evidence of tumour metastasis
	50/M/L[a]	Canada	Pigmented tumour	Yes	20 (20)	US	Clinical	No	Iodine-125 brachytherapy	No pathological examination Decrease in the height of the tumour No evidence of tumour metastasis
Wilson *et al.* (2001)[9]	30/M/L	New Zealand	Elevated pigmented mass anterior to the equator	No	NA	US, FA	Clinical	No	Scleral buckle without external drainage but with external cryotherapy for RRD Iodine-125 brachytherapy	Reattached retina No pathological examination Radiation papillopathy

Kase *et al.* (2018)[10]	78/F/R[a]	Japan	A yellowish flat choroidal elevation	Yes	12 (16)	US, FA, ICGA, MRI	Clinical	Not clear (referred case)	Enucleation	and radiation retinopathy Mixed-cell type Liver metastasis developed about 1.5 years after the enucleation

[a]Referred case.
CD, choroidal detachment; CM, choroidal melanoma; F, female; FA, fundus fluorescein angiography; ICGA, indocyanine green angiography; IOP, intraocular pressure; L, left eye; M, male; NA, not available; PPV, pars plana vitrectomy; R, right eye; RRD, rhegmatogenous retinal detachment; SRF, subretinal fluid; US, ultrasonography.

Initially, the presumptive diagnosis of RRD with haemorrhagic CD prompted us to perform external drainage, which failed. Unfortunately, the possibility of RRD and CM coexistence did not cross our minds, and we did not instantly conduct a transscleral biopsy for further pathological examination. In general, due to the high accuracy of clinical diagnosis and the risk of procedure-related complications, biopsy of the lesion is not recommended for CM[15,19]. Nevertheless, it provides an opportunity to determine the prognosis for metastasis while also providing tissue resources for further research in order to develop molecular-based targeted therapies. Because of the problematic nature of presumptive diagnosis, as exemplified in our case, we emphasize the importance of tumour biopsy. Several intraocular biopsy techniques for CM, such as transscleral incisional and excisional biopsy, fine needle aspiration biopsy, and pars plana vitrectomy-assisted approaches using a vitreous cutter, have been described[20]. Use of either the transvitreal or transscleral route involves only a low risk of intraocular and extrascleral tumour dissemination after surgery[20–22]. However, our review of the literature found that there was no recommended technique for tumour biopsy in the event of coexistence of CM and RRD (Table 1). As a result, in our case scenario presenting with RRD and thick mass at presentation, we suggest two management options: (1) transscleral incisional and excisional biopsy, together with scleral buckling procedure; and (2) transretinal biopsy using a 23-gauge or 25-gauge vitreous cutter, together with RRD repair with intraocular tamponade[20,22]. The decision on which options to use is influenced by the location and thickness of the mass and the surgeons' experience.

Unfortunately, the patient was lost to follow-up before further investigations, such as an MRI, were made. Although orbital MRI with contrast did not strongly point to the diagnosis of CM or choroidal metastasis, we decided to exenterate the affected eye due to the significant proptosis which we felt could have resulted from extrascleral extension following external drainage. Histopathological examination confirmed the diagnosis of epithelioid cell type choroidal melanoma in atypical presentation for a CM case.

In conclusion, we report a case of simultaneous RRD with CM, which is an extremely rare presentation. In the differential diagnosis of RRD with presumed CD or haemorrhagic CD, the possibility of underlying CM should be considered. Adjunct imaging methods, such as ultrasonography and MRI, may play a vital role in the confirmatory diagnosis and appropriate management of such atypical manifestations. In addition, tumour biopsy in combination with RRD repair surgery can be a helpful diagnostic tool for preventing further tumour invasion and metastasis. In particular, a CD that does not drain in the operating room should prompt consideration of diagnosis of CM.

Method of literature search

A systematic literature search was performed using the PubMed database for articles in English with the following search terms: rhegmatogenous retinal detachment and choroidal melanoma; and rhegmatogenous retinal detachment and uveal melanoma. Out of the eleven that were found, seven papers (10 cases) with concurrent RRD and CM were chosen. There are no copies of the two papers from 1983 and 1984; as a result, the review included five articles (eight cases).

Table 2

Clinical indicators for distinguishing between choroidal melanoma and haemorrhagic choroidal detachment in coexisting rhegmatogenous retinal detachment

	Simultaneous RRD and CM[1,4,7–10,14,15]	Simultaneous RRD and haemorrhagic CD[5,6,14,16,17]
Age (years)	59–62 (Caucasian) 45–55 (Asian)	Variable
Intraocular pressure	Near normal/normal	Hypotony
Susceptibility factors	Fair skin, light eye colour, inability to tan, ocular or oculodermal melanocytosis, cutaneous or iris or choroidal naevus	High myopia[a], hypertension, systemic anticoagulation
Characteristics of choroidal elevation	Pigmented/non-pigmented	Non-pigmented
Ultrasonography features	Dome-shaped or mushroom-shaped, homogeneous hyperechoic mass with acoustic hollowness, and choroidal excavation	Dome-shaped mass with changeable echogenicity:
	If haemorrhagic CD is the presenting sign of choroidal melanoma, it may feature large haemorrhagic choroidal detachments that are separable from the underlying mass	Acute stage: hyperechogenicity and variable degree of heterogenicity Late stage: hypoechogenicity and homogenicity
MRI finding	Typically displays high signal intensity on T1-weighted images and low signal intensity on T2-weighted images.	High signal intensity on T1-weighted images and low signal intensity on T2-weighted images.
	Vivid contrast enhancement after gadolinium injection	Slight peripheral contrast enhancement after gadolinium injection
	Imaging appearance may be variable based on the degree of pigmentation and the presence of areas of necrosis or cavitation	
Management	Resection Radiation therapy Enucleation	External drainage ± Vitrectomy

[a]Common risk factor for simultaneous RRD and haemorrhagic CD.

CD, choroidal detachment; CM, choroidal melanoma; RRD, rhegmatogenous retinal detachment.

Ethical approval

The study followed the tenets of the Declaration of Helsinki and was approved by the Ethics Committee of Rajavithi Hospital (approval No. 096/2565).

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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None.

Author contributions

N.P. and S.S. conceived the study, participated in design and coordination, data interpretation, drafted and edited the article. N.P., W.P., and S.S. participated in coordination and data collection. N.P., W.P., and S.S. edited the article. All authors read and approved the final version of the article.

Conflicts of interest disclosure

There are no conflicts of interest.

Research registration unique identifying number (UIN)

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