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Persistent Intraschisis Hemorrhage Simulating Choroidal Melanoma

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Background: Intraschisis hemorrhage of dark green color without accompanying vitreous hemorrhage is a rare complication of degenerative retinoschisis-detachment.

Case: A 37-year-old male patient with unilateral intraschisis hemorrhage closely mimicking a choroidal melanoma is described.

Observations: Ultrasonography and intravenous fluorescein angiography suggested intraschisis hemorrhage. T_1-weighted magnetic resonance imaging demonstrated a hyperintense lesion compared to the vitreous that did not enhance with contrast agent. Managed by observation, the color of the lesion started to change at the 6th month of follow-up. The hemorrhagic lesion regressed to half size in 40 months following the diagnosis, and disappeared in 62 months.

Conclusion: Very rarely may an intraschisis hemorrhage secondary to degenerative retinoschisis-detachment simulate a choroidal melanoma. In our patient, careful interpretation of the conventional methods was adequate for the differential diagnosis. The unusual feature of this patient was that the hemorrhage resolved in 5 years, much slower than expected. **Jpn J Ophthalmol 2002;46:222–225**

Key Words: Intraschisis hemorrhage, retinal detachment, retinoschisis, uveal melanoma.

Introduction

Retinoschisis or splitting of the retina is classified into hereditary, degenerative, and secondary types. Degenerative retinoschisis is mostly age-dependent and can be found in up to 1.5% of patients older than 40 years of age.[1,2] Retinal detachment and posterior extension towards the macula are the most important complications of retinoschisis, which is usually located in the inferotemporal quadrant.[1] The retinal detachment associated with retinoschisis can be either symptomatic and rapidly progressive or more commonly, localized and nonprogressive.[1] Intraschisis hemorrhage alone or in association with vitreous hemorrhage is a fairly rare condition and can be one of the protean manifestations of hereditary retinoschisis.[3] In the past, there have been instances in which eyes with retinoschisis were enucleated for the presumed diagnosis of choroidal melanoma.[4] In this report, a patient with clinically unilateral degenerative retinoschisis and nonprogressive retinal detachment that was further complicated by localized intraschisis hemorrhage simulating a choroidal melanoma is described. It took 5 years for the hemorrhage to clear completely.

Case Report

A 37-year-old man presented with a 2-week history of blurred vision in the right eye. His initial physician referred him to this center because of the possibility of uveal melanoma. He had been otherwise healthy and denied any history of trauma. His best-corrected visual acuity was 20/20 in both eyes. The anterior segments and intraocular pressure were within normal limits bilaterally. There was a well-circumscribed retinoschisis-detachment with surrounding demarcation line formed by mild hyperplasia of the retinal pigment epithelia (RPE) in the inferonasal quadrant of the right fundus. There was a dark green solid lesion 6 mm na-

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sal to the optic disc within the schisis cavity. The mass measured 9 × 6 mm in basal dimensions with an ultrasonic thickness of 4.7 mm. A-mode ultrasonography revealed highly reflective spikes from the cyst walls and low internal reflectivity from the part of the lesion containing the hemorrhage. B-mode ultrasonography displayed a dome-shaped lesion presumably under the outer retinal layer (Figure 1A). The fovea and other parts of the retina were normal. No inner and/or outer retinal holes were found. Fluorescein angiography showed blocked choroidal fluorescence throughout the angiography period with no evidence of neovascularization. No intrinsic vessel within the lesion could be detected. The mass did not transilluminate through the transpupillary route. Magnetic resonance imaging (MRI) done elsewhere demonstrated a hyperintense lesion compared to the vitreous on T_1-weighted sequences (Figure 1B) that did not enhance following contrast agent administration. On T_2-weighted images, the lesion was isointense with respect to the vitreous. The left ocular fundus was normal. Electroretinography (ERG) of the right eye showed a general reduction of amplitude and a relatively normal waveform pattern according to our laboratory normal standards. Dark-adapted scotopic ERG revealed decreased b-wave amplitude and prolonged b-wave implicit time. On maximum combined response recordings, both a- and b-wave amplitudes were decreased. Light-adapted cone response ERG suggested a slight negative shift. Although to a much lesser degree, similar results were recorded on the ERG of the left eye. The results of a systemic work-up were within normal limits.

Six months after the initial visit, the color of the lesion had turned to yellow-tan (Figure 2A) and its ultrasonographic thickness decreased to 3 mm. After 40 months of follow-up, the lesion was reduced to half its original size. At the 62nd month, total clearance of hemorrhage with residual RPE changes and stiff retinal folds were observed (Figure 2B). Repeat fluorescein angiograms failed to show any source of neovascularization or leakage. The patient maintained a visual acuity of 20/20 OU. The retinoschisis-detachment showed a subtle progression to the posterior pole while the macula still remained intact.

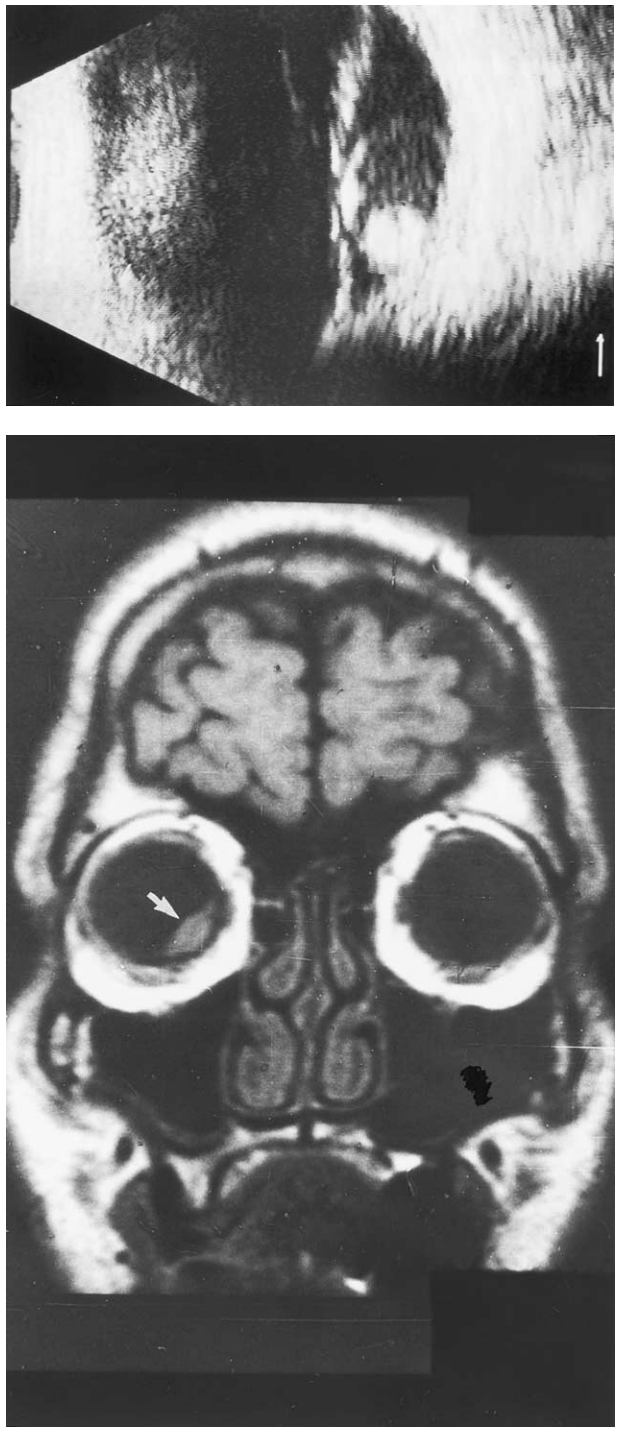

Figure 1. (**Top**) B-mode ultrasonography showing the intraschisis hemorrhage. The vitreous and the choroid are seemingly not involved. (**Bottom**) T_1-weighted coronal cranial magnetic resonance imaging showing a hyperintense lesion in the inferonasal quadrant of the right eye (arrow). As such, the mass is not distinguishable from a melanoma.

Discussion

Hemorrhage into the vitreous or schisis cavity is a highly unusual but well-recognized complication of degenerative retinoschisis.[1,5] Greven et al[3] also observed this complication in 4 of 14 eyes followed for X-linked retinoschisis. The inner layer of the retina is thin with often occluded or thickened vascular channels. Rup-

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ture of these poorly supported fragile vessels, sometimes precipitated by posterior vitreous detachment or even minor trauma, may cause intravitreal or intraschisis hemorrhage.[1,5] Leakage of fluorescein dye can be observed from the attenuated inner layer vessels.[1] However, retinal surface neovascularization induced by the chronic retinal detachment can also account for the hemorrhage.[5] Campo et al[5] reported 3 patients with

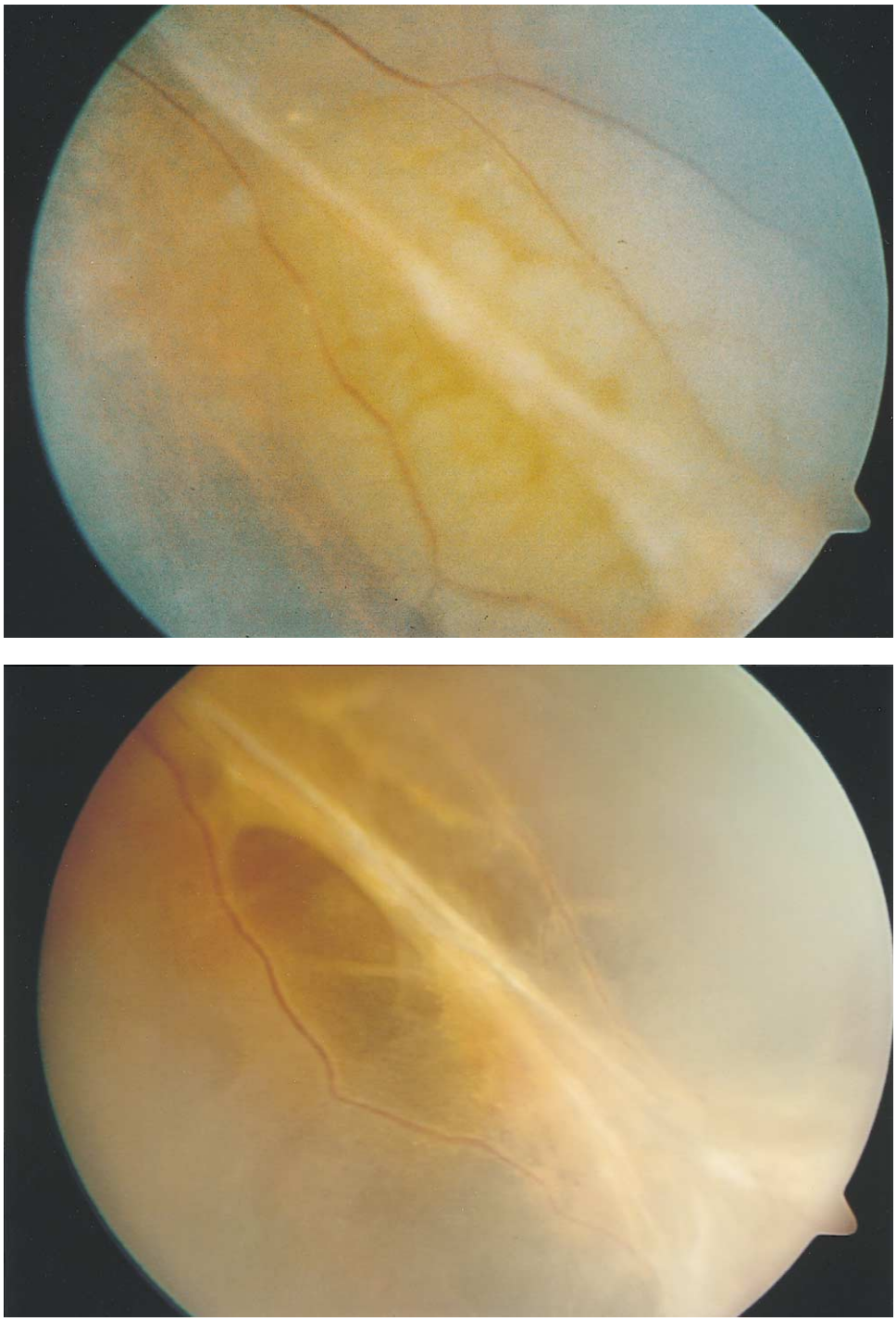

Figure 2. (**A**) Due to dehemoglobination of the hemorrhage, the color of the mass turned to yellow-tan within 6 months. A retinal fold extends parallel to the long axis of the hemorrhagic mass. (**B**) Five years after the diagnosis, the hemorrhage regressed totally with a significant retinal folding and vascular sheathing.

bullous retinoschisis and intravitreal hemorrhage all resulting from neovascularization at the dome of the cyst. The findings and clinical course of this current case tend to suggest possible rupture of some unsupported retinal vessel as cause of hemorrhage rather than choroidal or retinal neovascularization, although neither could be documented or refuted unequivocally.

A large, elevated subretinal hemorrhage with a green hue can sometimes be confused with a choroidal melanoma. On rare occasions, the distinction of subretinal hemorrhage from a melanoma may impose extreme difficulties with standard techniques. It is believed that the greenish hue originates from the retinal nerve fiber layer, which becomes stained by the breakdown products of hemoglobin.[3] In some cases a pigmented line may develop at the posterior margin of the schisis cavity containing hemorrhage.[3] This finding, which was also observed in this patient, can be attributed either to the irritation of the RPE caused by the chronic traction at the posterior border of the schisis or to the migration of the blood breakdown products followed by the staining of the outer layers.[3] It is also possible that chronic detachment of the outer retinal layer, a feature that is found in the present case, might have caused a demarcation line.[3]

There were several distinguishing features in this patient that would favor hemorrhage rather than a melanoma. The margins of the lesion rose abruptly and did not blend with the normal-appearing adjacent choroid, a finding that was also confirmed by B-mode ultrasonography. As in this case, the retina overlying a subretinal hemorrhage may develop stiff folds, which are rarely seen in posterior uveal melanoma. Precontrast MRI scans can be identical in a choroidal melanoma and a subretinal hemorrhage. Due to the presence of methemoglobin, the hemorrhage may give a high signal intensity on T_1-weighted images and a low to high signal intensity on T_2-weighted frames. However, unlike a uveal melanoma, the hemorrhage does not enhance with contrast agent.

Localized, large intraschisis hemorrhage without vitreal involvement is an uncommon complication of degenerative retinoschisis-detachment and may mimic choroidal melanoma. Careful interpretation of clinical observations and the results of conventional ancillary tests made it possible to establish the correct diagnosis in this patient.

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