Epimacular membrane secondary to an optic nerve head lesion

A 57-year-old woman presented to us with blurred vision in her right eye for approximately a year. She gave no history of any systemic illness, ocular surgery or trauma. Best-corrected visual acuity (BCVA) was 20/70 in the right eye and 20/20 in the left. The anterior segments were unremarkable in both eyes. Fundus examination of right eye showed a tuft of anomalous papillary vessels with adjoining epimacular membrane (EMM) (figure 1A). Fundus of the left eye was normal. There was no evidence of posterior vitreous detachment or any coexistent retinal pathology in either eye. Fluorescein angiography of the right eye showed delayed filling of the papillary aneurysms with characteristic erythrocyte–plasma interface, as well as distortion and leakage of parafoveal capillaries (figure 1B). Optical coherence tomography (OCT; Stratus OCT; Carl Zeiss Meditec, Dublin, California, USA) of the right eye confirmed the EMM, central foveal thickness was 556 μm (figure 1C). A vertical OCT scan also demonstrated the layering of erythrocytes in the aneurysms (figure 1D). Systemic evaluation and neuroimaging revealed no cutaneous or central nervous system involvement. There was no family history of any ocular or systemic disease. The patient’s daughter was also screened; she had unremarkable fundus and systemic examinations. The patient’s parents and siblings were not available for evaluation. Surgery was offered to the patient to remove the EMM. With informed consent of the patient, she underwent pars plana vitrectomy in the right eye. One month postoperatively, BCVA improved in the right eye with a corresponding decrease in macular thickness (figure 1E,F). The visual outcome was maintained over the follow-up. This study was performed in compliance with the tenets of the Declaration of Helsinki.

QUESTIONS
1. What is the differential diagnosis of the fundus lesion?
2. What investigations should be performed to reach a definitive diagnosis?
3. How would you manage this case?
See page 1048 for answers

Figure 1  Diagnosis and management of cavernous haemangioma of the optic disc with epimacular membrane (EMM). (A) Fundus photograph of the right eye shows ‘cluster of grapes’ appearance of cavernous haemangioma of the optic nerve head, with an adjoining EMM extending over the macula. (B) Late-phase fluorescein angiogram shows the delayed filling of the saccular aneurysms with erythrocyte sedimentation highlighted by ‘capping of the dye’. Also note the distortion of parafoveal capillaries that leak into the foveal avascular zone. The arrow shows the direction of the vertical scan. (C) Optical coherence tomography (OCT; horizontal 10 mm scan through fovea) distinctly outlines the EMM, as well as its origin from the optic disc haemangioma. Central macular thickness is 556 μm. (D) The vertical OCT scan (5 mm, inferior retina to the left) corroborates the angiographic layering of the dye (white arrow points at the hyporeflective upper plasma interface, with hyporeflective accumulation of erythrocytes below. (E and F) One month after vitrectomy for removal of EMM, the macula is clear, and visual acuity has improved to 20/25, although macular contours are only partly restored (central macular thickness: 355 μm).
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Patient consent Obtained.

Ethics approval This study was performed in compliance with the tenets of the Declaration of Helsinki.

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The description ‘haemangioma’ is probably a misnomer for this lesion, which is most likely a vascular malformation rather than a neoplastic lesion. Although retinal cavernous haemangiomata are typically asymptomatic and stationary, the ophthalmologist has an important role to play, particularly when the patient is also systemically asymptomatic. Retinal tumours are generally associated with cerebral cavernous malformations as part of an autosomal dominant syndrome. In contrast to retinal lesions, the cerebral aneurysms are frequently associated with seizures and intracranial haemorrhages. It is therefore essential that all patients with retinal cavernous haemangioma and their first-degree relatives are comprehensively evaluated by oculocutaneous and neurological examination, including neuroimaging studies, even if they are asymptomatic.

As most tumours are peripheral and stationary, they are typically asymptomatic. Visual symptoms are more common with large and posterior angiomas, which are rare. Most cases are unilateral, solitary and sporadic. The tumours most frequently present with floaters or visual loss due to vitreous haemorrhage, which typically resolves spontaneously, and rarely warrants vitrectomy. Haller and Knox attributed vitreous haemorrhage to contraction of the fibroglial component on the papillary cavernous haemangioma. Other causes of visual loss include amblyopia and macular location of a haemangioma.

In spite of frequent association with fibroglial tissue, secondary macular pucker has not been reported, probably because of the rarity of the posterior haemangiomata. Messmer et al have described an EMM associated with retinal cavernous haemangioma; however, the haemangioma was located in the inferior mid-periphery, and no direct cause and effect relationship could be established between the hamartoma and macular membrane. The treatment of this membrane was also not mentioned by the authors. We report an EMM in direct continuation with the cavernous haemangioma of the optic nerve head, both clinically and by OCT. There is a possibility of the co-existence of an idiopathic EMM with cavernous haemangioma in this patient in view of her age, ethnicity and the recently reported observation of a high prevalence of EMM in a multi-ethnic population, particularly in Asian individuals and the elderly. This possibility, however, appeared remote in view of the continuation of EMM with haemangioma, the unilaterality of the membrane, and the known association of fibroglial tissue with retinal cavernous haemangioma. Furthermore, symptomatic and visible EMM as seen in this patient (‘pre-macular fibrosis’) is uncommon as an idiopathic entity; risk factors for secondary EMM such as cataetar surgery, posterior vitreous detachment other vitreoretinal diseases were ruled out by history and examination. Removal of EMM led to excellent visual recovery in spite of the chronicity of the membrane.

ANSWERS

From questions on page 1038
1. The clinical picture of dark-red saccular aneurysms in a ‘cluster of grapes’ configuration is so characteristic of retinal cavernous haemangioma that there are few differential diagnoses. Nevertheless, this lesion should be differentiated from other vascular disorders such as Coats’ disease, retinal capillary haemangioma, retinal arteriovenous communications and retinal vasoproliferative tumours. The characteristic absence of capillary telangiectasia, dilated feeder vessels or retinal exudation helps to distinguish cavernous haemangioma from the aforementioned conditions.

2. The main confirmatory investigation is fluorescein angiography, which in this case showed a typical ‘capping of dye’ in the late phase due to sedimentation of the erythrocytes (blocked fluorescence) below, and clear plasma above (hyperfluorescent cap). There was a characteristic absence of fluorescein leakage. Vertical OCT scan showed a similar appearance of clear space above and hyperreflective echoes below in the aneurysms. OCT may therefore be a non-invasive adjunct to fluorescein angiography in such cases.

3. The management of this case is twofold: systemic and ocular. Although most cases are unilateral, solitary, sporadic and stationary, a thorough systemic evaluation with focus on the skin and central nervous system is necessary because of the risk of cutaneous and potentially fatal cerebral aneurysms, particularly in the presence of retinal lesions. The patient and the available family members underwent ocular, dermatological and neurological evaluation, including neuroimaging for the patient. The systemic evaluation of this patient and her family was unremarkable.

As this patient was symptomatic with an EMM secondary to papillary cavernous haemangioma, surgery was offered to the patient to remove the membrane. With informed consent of the patient, she underwent pars plana vitrectomy in the right eye. Posterior vitreous detachment was induced carefully to avoid iatrogenic vitreous haemorrhage from the papillary aneurysms. Peeling of epiretinal and internal limiting membranes was performed after sequential staining with trypan blue 0.15% (Retiblue, Aurolab, Madurai, India) and brilliant blue G 0.05% (Ocublue Plus, Aurolab, Madurai, India) dyes respectively. Sulphur hexafluoride 20% gas was used for tamponade. One month post-operatively, BCVA in the right eye improved to 20/25; OCT showed the decreased macular thickness (365 μm). During an event-free follow-up of 10 months; the visual status was maintained.

REFERENCES
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