A case of retinal cavernous haemangioma – don't let it slip through the grapevine

BY MARY HENRY AND JERALD WILLIAM

We present a case of a seven-year-old male who presented to the emergency eye referral clinic with red eye. He was diagnosed and treated for allergic conjunctivitis, however, on clinic review, wide field retinal imaging was performed (as has been trust protocol since 2015 for children over the age of three). A suspicious lesion on the retina was seen, obscured by the inferior lashes, in the inferonasal periphery (Figure 1). Therefore, a further image focussed on the area of interest was taken (Figure 2). This revealed a deep red, cluster lesion which appeared vascular in nature, measuring five-disc diameters in length and approximately one- to two-disc diameters in width, with normal surrounding retina and no subretinal fluid, exudates or haemorrhage.

Classically described as a 'cluster of grapes', this was diagnosed by the typical appearance as a retinal cavernous haemangioma; a benign, vascular hamartoma, seen as clumps of dark intraretinal aneurysms. They are usually incidental findings unless there is macular involvement and rarely present with self-limiting vitreous haemorrhage as a result of vitreous and epiretinal membrane traction [1]. Very rarely they can present with hyphaema [2].

Cavernous haemangiomas can be single or multiple but do not progress and can even spontaneously regress. Most cases are sporadic but some are inherited in an autosomal dominant inheritance and can be associated with skin, hepatic and cerebral haemangiomas. This is often associated with a splice mutation in CCM1 gene or KRIT protein [3].

Cavernous haemangiomas are usually a clinical diagnosis made on the typical cluster of grapes appearance but if diagnosis is in doubt, then fundus fluorescein angiography can be performed, which would display dilated saccular lesions that fill slowly. Plasma-erythrocyte layering can occur as a result of sluggish blood flow. This appearance with an absence of subretinal fluid and leakage is suggestive of cavernous haemangioma instead of other differentials such as coats disease, that can show capillary dropout and leakage. Coats, along with other differentials of retinal capillary haemangiomas and racemose haemangiomas, can result in exudative retinal detachments. Racemose haemangiomas are arteriovenous malformations with no intervening capillary bed, where larger retinal vessels are dilated than those in cavernous haemangiomas [4,5].

Cerebral cavernous haemangiomas can have fatal consequences if left undiagnosed, including seizures and stroke, and it is important to consider these in patients with incidental retinal

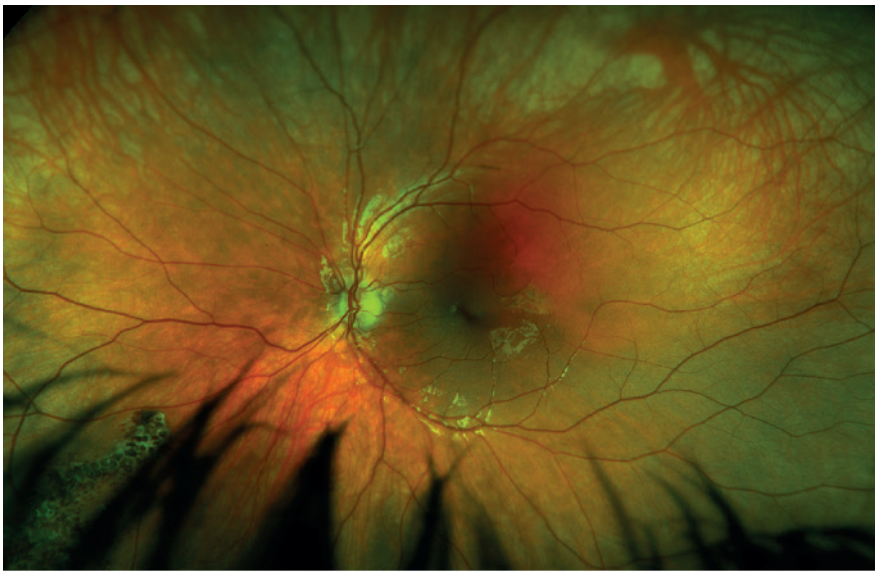

Figure 1: Widefield fundal image of a left eye suggesting the presence of a lesion in the inferonasal periphery obscured by the inferior lashes.

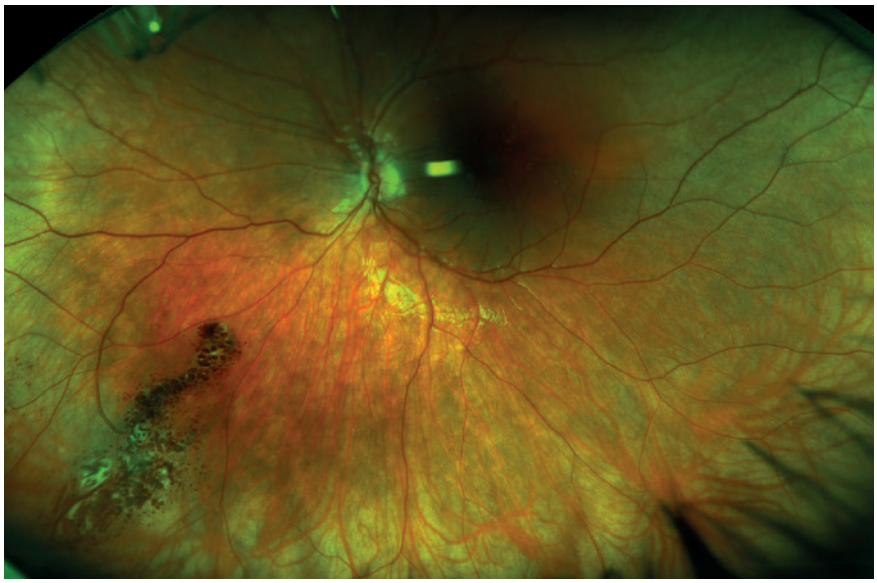

Figure 2: Widefield fundal image of a left eye showing a dilated vascular lesion in the inferonasal periphery, resembling a 'cluster of grapes'.

cavernous haemangiomas. Though in this case neuroimaging was normal, it is recommended that patients with cavernous haemangiomas have neuroimaging in the form of MRI, with contrast, to rule out or diagnose any such lesions that may require surgical intervention or specialist observation [2]. Ophthalmologists play a key role in diagnosing and coordinating long-term care with specialists in the field.

Moreover, as cavernous haemangiomas can often be incidental, it is important to ensure any fundal imaging requested is reviewed by a clinician, even if patients are only being reviewed for anterior segment pathology.

References

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TAKE HOME MESSAGE

- Retinal cavernous haemangiomas can be solitary or numerous, sporadic or syndromic.
- They are often asymptomatic and incidental findings on fundal examination but can rarely present with vitreous haemorrhage or hyphaema.
- Retinal cavernous haemangiomas have a typical cluste of grapes appearance on fundoscopy but can be differentiated from other differentials on fundus fluorescein angiography.
- We recommend neuroimaging on patients with incidentally noted cavernous haemangiomas to rule out or identify any associated cerebral haemangiomas.
- It is important to ensure all imaging that is taken to be reviewed by a trained clinician in the field.

AUTHORS

Mary Henry,
ST6 Trainee, University Hospitals Coventry & Warwickshire, UK.

Jerald William,
Consultant, University Hospitals Coventry & Warwickshire, UK.

Declaration of competing interests:
None declared.