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Patent foramen ovale presenting as visual loss

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Lesson

Retinal artery occlusion in an otherwise healthy, young patient is rare. In this context it is important to consider patent foramen ovale as a differential. Early referral to a cardiology specialist for diagnosis and treatment is important for preventing further ocular and non-ocular events.

Keywords

Cardiovascular medicine, Clinical, Ophthalmology

Case report

A 21-year-old man presented with an eight-day history of a blurry patch of vision in the right eye. He could not identify any precipitants, had no significant medical history and did not take any regular medication. His best corrected visual acuity was 6/4 unaided in both eyes. Intraocular pressure was normal in both eyes and anterior segments appeared quiet. On dilated fundoscopy, a filling defect was apparent in a branch of the inferotemporal retinal artery of the right eye approximately 1–2 disc diameters away from the optic disc, associated with pale areas resembling cotton wool spots. The diagnosis of branch retinal artery occlusion was made. These findings were also visualised with optical coherence tomography – Figure 1. One week later, the patient's symptoms of blurriness had subjectively improved, and the pale areas previously described had reduced in size. Screening for cytomegalovirus, toxoplasma, human immunodeficiency virus, vasculitis and thrombophilia were all negative. Routine blood tests were insignificant with a total cholesterol of 4.6 mmol/L, triglycerides of 1.9 mmol/L, erythrocyte sedimentation rate of 2 urea and normal renal and liver function.

Referral to cardiology was made to investigate for possible cardiac causes of embolism, and the patient was empirically started on aspirin. General cardiological assessments were unremarkable with a blood pressure of 135/83 mmHg, heart rate of 81 beats per minute, normal heart sounds with no murmurs or added sound and no carotid bruits were detected. Magnetic resonance imaging of the head, magnetic resonance angiogram of the head and neck vessels (specifically looking for carotid artery dissection), and carotid doppler scans were all normal. A 72-hour Holter monitoring test showed sinus rhythm throughout, and conventional transthoracic echocardiography was insignificant. However, a transthoracic echocardiographic bubble contrast study, which showed no evidence of bubbles transiting at rest, did show a large right to left shunt at the atrial level within one beat of release of valsalva manoeuvre – Figure 2. This was strongly suggestive of a large patent foramen ovale. No other potential causes for the retinal artery embolus were identified at any point.

In view of the potential consequences of future emboli causing further occlusions of the retinal or cerebral circulations, percutaneous closure of the patent foramen ovale was undertaken, alongside six months of low-dose aspirin and lifelong clopidogrel therapy. This patient has had no further episodes and at 12 months since the episode he is asymptomatic with normal visual acuity (6/6 unaided in each eye).

Discussion

Patent foramen ovale is a persistent cardiac communication between the left and right atria, which normally closes within a year after birth. In utero, this communication allows right-to-left shunting of the circulation. Soon after birth, as the pulmonary circulation is established, the pulmonary pressure drops and the left atrial pressure exceeds that of the right,

Figure 1. Composite demonstrating partial occlusion of a branch retinal arteriole characterised by embolic material within the retinal arteriole and 'cotton wool spots' of the adjacent retina. Spectralis Multicolor™ images comprising multicolour and associated infrared reflectance, green reflectance, blue reflectance images at time of presentation (a) and at six months later (b) with associated retinal thickness heat maps (c) and absolute thickness measurements in microns (d) at the same time points; change in retinal thickness is also shown (e).

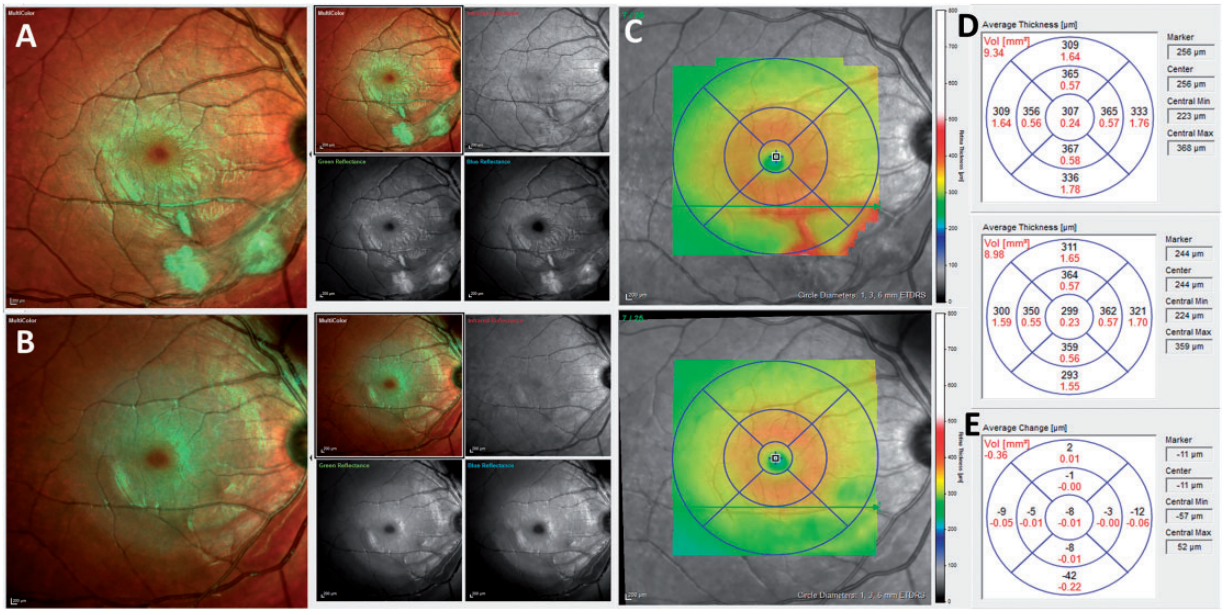
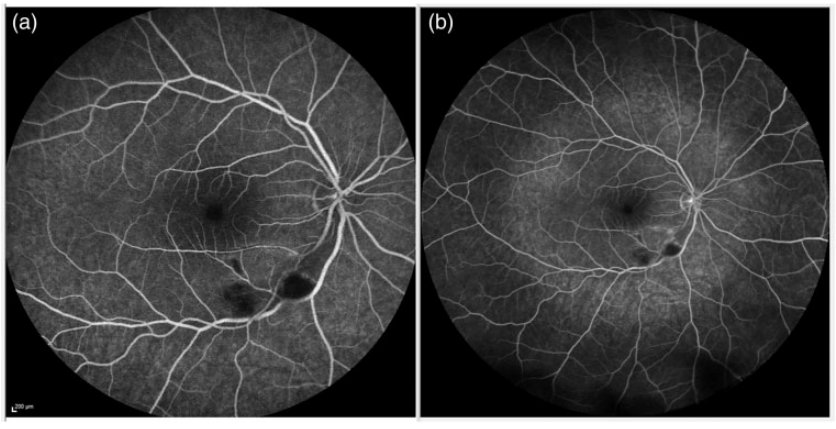


Figure 2. Fundus fluorescein angiogram demonstrating reduced flow through a branch macular arteriole arising from the inferotemporal arcade visualised with 55' lens (a) and ultra-widefield lens (b) of the Heidelberg Spectralis HRA OCT. The cotton wool spots are seen as three hypofluorescent areas.



causing the foramen to close. Patent foramen ovale is prevalent in 20–34% of the population[1] and are generally benign and asymptomatic. Occasionally, they can give rise to systemic emboli, which can cause both ocular and cerebral ischaemic events, such as cryptogenic strokes. Reported ocular manifestations of patent foramen ovale include central and branch retinal arterial occlusion and optic neuropathy.[2,3] Patent foramen ovale has also been associated with decompression sickness in scuba divers and recurrent migraines.[1] In our patient, diagnosis was made using transthoracic echocardiogram bubble study;

however, it has been reported that transoesophageal echocardiogram is more sensitive in the detection of cardiac sources of retinal emboli when compared to transthoracic echocardiogram.[4]

Retinal artery occlusions encompass a group of diseases, whereby sudden obstruction of the arterial circulation causes ischaemic damage to the retina.[5] It is primarily a disease of the older population and is divided into two subtypes: branch retinal artery occlusion and central retinal artery occlusion. Central retinal artery occlusion usually results in profound visual loss in the affected eye, whereas branch retinal artery occlusion may not affect the central vision and instead leave a scotoma, or a visual field defect.

The commonest risk factors for branch retinal artery occlusion are hypertension, atherosclerosis, diabetes mellitus and thrombophilia, but it may also arise from ipsilateral carotid artery atherosclerosis and cardiogenic embolism, such as in atrial fibrillation, cardiac valvular disease and infectious endocarditis. Retinal artery occlusion in young patients with patent foramen ovale is rare and has only been reported in a handful of cases.[2,3,6,7]

This case highlights the importance of considering patent foramen ovale in the context of retinal artery occlusion in an otherwise healthy young patient. Such cases benefit from early referral to a cardiology specialist to enable prompt diagnosis and treatment in order to prevent further serious ocular and non-ocular events.

Declarations

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Guarantor: AD.

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