A rare but important differential diagnosis in transient monocular blindness

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DESCRIPTION

A 45-year-old man presented with transient episodes of monocular blindness. Physical examination, including full neurology and funduscopy examination was unremarkable. Head CT was negative for intracranial bleed. Carotid ultrasound with Doppler was unremarkable. A two-dimensional echocardiogram was performed and showed a mobile mass attached to the aortic valve. To further evaluate the mass, a transesophageal echocardiography (TEE) was performed which showed a multilobulated, mobile, pedunculated mass attached to the aortic surface of the non-coronary aortic cusp, measuring 1.5 cm×0.4 cm without any valvular involvement ([figure 1](#) and [video 1](#)). Via thoracotomy, the mass was surgically excised. Aortic valve competence was confirmed intraoperatively before the aortotomy was closed. Gross examination showed a pedunculated mass with frond-like projections and filamentous endings, appearing like a 'sea anemone' ([figure 2](#)). Histopathology revealed branching avascular papillae with single layer of endothelium, confirming the diagnosis of papillary fibroelastoma (PFE; [figure 3](#)). No symptomatic recurrence or evidence of regurgitation was seen on follow-up at 6 months.

PFEs are rare cardiac tumours, but should be differentiated from thrombus, vegetation, myxoma and Lambl's excrescence.¹ Although patients are typically asymptomatic, PFEs can potentially lead to severe complications, primarily due to their embolic potential. A TEE most often helps in making a presumptive diagnosis.² Surgical excision is curative and is indicated in symptomatic patients, lesions larger than 1 cm, and in patients with left-sided lesions.³ Our case underscores the

importance of PFE in the differential diagnosis of transient blindness, especially in young patients without significant medical histories.

Video 1 TEE showing a multilobulated, mobile, pedunculated mass attached to the aortic surface of the noncoronary aortic cusp.

Figure 2 Gross examination showing pedunculated mass with frond like projections and filamentous endings appearing as 'sea anemone'.

Figure 1 Transesophageal echocardiographic image of aortic valve.

Figure 3 Histopathological examination showing avascular branching papillae covered by single layer of endothelium in a mucopolysaccharide matrix.

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Learning points

- ▶ Papillary fibroelastoma are rare cardiac tumours, typically found on the heart valves, that has the embolic potential.
- ▶ A transesophageal echocardiography will help diagnose them and they should be differentiated from thrombus, vegetation, myxoma and Lambl's excrescence.
- ▶ Surgical excision is curative and is indicated in symptomatic patients, lesions larger than 1 cm, and in patients with left-sided lesions.

Contributors Both authors reviewed the scientific literature, interpreted the data, and wrote the manuscript.

Competing interests None.

Patient consent Obtained.

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