Aortic valve mass and sudden blindness

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A 63-year-old man presented with a sudden and painless loss of vision in his left eye. He had no history of heart disease or arrhythmias, diabetes mellitus, hyperlipidaemia, hypertension or smoking. On admission his blood pressure was 160/90 mmHg, and his physical examination was unremarkable. Electrocardiography demonstrated normal sinus rhythm, with no signs of ischaemia or left ventricular hypertrophy. Ophthalmologic examination revealed no light perception in his left eye, and a pale and oedematous retina. The findings suggested a high probability of an embolic central retinal artery occlusion. Therefore 1.5 million units of streptokinase were given, with only a slight improvement of his vision. A transthoracic echocardiography (TTE) and carotid ultrasound performed the next day were interpreted as normal. The patient was discharged on warfarin therapy. While at home, he continued to have episodes of lightheadedness, dizziness and visual hallucinations once or twice a week. Transoesophageal echocardiography (TEE) was performed a few weeks after discharge (figure).

Questions
1 What abnormality is shown in the aorta on the echocardiogram?
2 What is the differential diagnosis?
3 What is the most probable diagnosis, considering the echocardiographic findings and the patient’s history and physical findings?
4 What is the treatment of choice?

Figure Transoesophageal echocardiography

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Answers

QUESTION 1
A 7 x 7 mm papillary mass can be seen attached
to the aortic face of the non-coronary aortic
cusp.

QUESTION 2
The differential diagnosis is cardiac tumour,
aortic valve vegetation, thrombus, or calcified
atherosclerotic mass.

QUESTION 3
The most probable diagnosis is a papillary
fibroelastoma.

QUESTION 4
The recommended treatment is immediate
surgical excision of the mass.

Discussion

Papillary fibroelastomas are rare, benign and
avascular cardiac tumours, derived from normal
components of the endocardium. They
were found to be the most common primary
cardiac valve tumour in a literature review of
128 case reports. Most tumours are attached
to either side of the cardiac valves, but may also
be located on the surface of papillary muscles,
chordae tendinae, ventricular septum and
endocardium of all cardiac chambers. They are
mostly identified on autopsy, but rarely during
life. Most of the patients who were diagnosed
by echocardiography, had prior symptoms –
stroke, myocardial infarction, or sudden
death.

The possibility of a cardiac vegetation, a
thrombus, or an atherosclerotic aortic calcified
mass were considered unlikely in our patient,
since these were not supported by the clinical
and echocardiographic findings. TTE per-
formed one day after his presentation was
interpreted as normal, and the tumour was
only discovered a few weeks later on TEE. The
patient was sent for immediate surgery and the
tumour was excised successfully. Histological
examination confirmed the diagnosis of papil-
lar fibroelastoma. He has had no further
embolic events in 5 years of follow-up.

It has been suggested that TEE should be
performed in every patient with signs and
symptoms of systemic embolisation and a nor-
mal cardiac history, physical findings, and nor-
mal non-invasive studies or TTE. We suggest
that cardiac valve tumours, mainly papillary
fibroelastoma, should be included in the differ-
ential diagnosis of ocular or systemic embolic
events. Once diagnosed, even in an asympto-
matic patient, they should be excised immedi-
ately.

Final diagnosis

Papillary fibroelastoma of the aortic valve,
causing an embolic central retinal artery occlu-
sion.

Keywords: papillary fibroelastoma; blindness; trans-
oesophageal echocardiography

Learning points

- papillary fibroelastomas are rare, but are the
  most prevalent primary cardiac valve tumours
- sudden unilateral blindness may be of an
  embolic source
- TEE should be performed whenever systemic
  embolisation is suspected and other non-invasive
tests are unrevealing
- most papillary fibroelastomas are asymptomatic,
  but once diagnosed they should be excised
  immediately

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