Incidental Finding of an Aortic Valve Mass on 64-slice Computed Tomographic Coronary Angiography

Quiz

A 41-year-old Chinese man with a background history of hypertension for 8 years underwent a computed tomography coronary angiography for an evaluation of atypical central chest pain. This revealed a coronary artery calcium score of zero with no significant stenosis seen in any of the coronary arteries. There was an incidental finding of a 9 x 6 mm filling defect attached to the right coronary cusp of the aortic valve, projecting into the non-coronary sinus (Figs. 1 and 2).

The patient was afebrile and non-toxic. Transoesophageal echocardiography was performed to elucidate the nature of this mass. This confirmed the presence of a mobile, echodense, papillary mass measuring 2 x 2.4 cm, attached via a stalk to the non-coronary cusp of the aortic valve (Figs. 3 and 4). The aortic valve was otherwise structurally normal without any regurgitation or stenosis.

What is the diagnosis?

a) Infective endocarditis
b) Intracardiac thrombus
c) Cardiac papillary fibroelastoma
d) Cardiac myxoma
e) Cardiac angiosarcoma

(answer below)

Discussion

The diagnosis is that of a cardiac papillary fibroelastoma (CPF) attached to the aortic valve.

The incidence of CPFs in the general population is unknown. In one autopsy series, fibroelastomas constituted 7% of all cardiac tumours detected and primary cardiac tumours have been noted in 0.002% to 0.33% of autopsies.1

Typical echocardiographic features of CPF include a round, oval or irregular appearance, with well-demarcated borders and a homogenous texture. When the image quality is optimal, a “speckled appearance” with “stippling” around the perimeter can be seen.2 Most CPFs are mobile with small stalks and less than 20 mm in the largest dimension.
CPFs may be single or multiple lesions and are most often associated with cardiac valvular disease.\(^2\)

Approximately 90% of reported CPFs were attached to valves, with the majority being on the aortic valve, followed by the mitral valve.\(^2\) When an atrioventricular valve was involved, the tumour was most often on the atrial side of the valve. There are few reports on tricuspid and pulmonic valvular CPFs and non-valvular CPFs.

CPFs are often diagnosed incidentally but have been reported to be associated with neurological events, sudden death, angina, acute myocardial infarction, pulmonary emboli and retinal artery embolism. These manifestations are related to the potential for CPF to cause symptomatic embolisation. In addition, CPFs presenting on the aortic surface of the aortic valve may cause dynamic coronary ostial obstruction leading to myocardial ischaemia.

Treatment of cardiac fibroelastomas is controversial because minimal data is available about therapeutic efficacy. Sun et al\(^3\) proposed that patients with embolic events which are not explained by other cardiovascular or neurological diseases should undergo transthoracic echocardiography and transoesophageal echocardiography if necessary to exclude cardiac sources of emboli, including CPF. A mass seen by echocardiography should be characterised by size, shape, location of attachment, mobility, presence of a stalk and multiplicity. Although the differential diagnosis may still include vegetations, thrombi, degenerative valve tissue and other benign tumours, these lesions can often be differentiated by clinical information, blood cultures and laboratory tests.

Decisions regarding the primary surgical excision of CPF depend on the size, location, mobility and strength of association of the tumour with symptoms.\(^3\) Early surgical excision of lesions close to the coronary ostia and of large lesions attached to the aortic valve may be considered because of the risk of complications, including cardiac ischaemia and sudden death. In cases of recurrent ischaemic events, prompt surgical excision may be necessary. Typically, valve replacement is unnecessary. Small, asymptomatic lesions detected on echocardiography performed for some other reason can usually be treated conservatively. No data exists to evaluate the efficacy of anti-coagulation or anti-platelet therapy for patients with CPF, although it is speculated that CPFs may serve as a nidus for thrombus formation.\(^3\)

REFERENCES


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