CASE REPORT

Aortic valve myxoma at the extreme age: a review of literature

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SUMMARY

Primary cardiac tumours are a rare finding, with cardiac myxoma and fibroelastoma representing the majority of these tumours. Cardiac myxomas are most commonly found in the left atrium but are rarely found with attachment to the cardiac valves. The authors describe a case of aortic myxoma found in an 81-year-old man presented with peripheral arterial disease. CT angiogram of the thorax was performed to find the source of emboli and it showed a mass attached to the aortic valve and protruding into the aorta. Details of the location and texture were studied on transoesophageal echocardiography. Preoperative coronary angiography showed coronary artery disease and the patient underwent successful coronary artery bypass grafting and simultaneous resection of the mass. Histopathology revealed the mass as a myxoma.

BACKGROUND

About 80% of cardiac tumours are benign and myxoma comprises 50% of these benign tumours. Most cardiac myxomas have attachments to the cardiac chambers, usually in the left atrium with attachment to the interatrial septum at the fossa ovalis, followed by the right atrium, the left ventricle and the right ventricle, in descending order of frequency. Myxomas attached to the cardiac valves are very rare. Majority of cardiac myxomas occurred in the third to sixth decade of life. Oldest age reported is 72 and 95 years for valvular myxoma and atrial myxoma, respectively. Aortic myxoma can embolise leading to myocardial infarction, stroke and limb ischaemia. This is the first ever case reported of aortic myxoma at 81 years of age.

CASE PRESENTATION

An 81-year-old man with a history of hypertension, chronic kidney disease, peripheral arterial disease and carotid stenosis presented with bilateral superficial femoral artery occlusion, and we suspected the presence of an embolism.

INVESTIGATIONS

A CT angiogram of the thorax (figure 1) indicated the presence of a 2.4×1.3 cm mass just above the aortic root, which was noted as a hypodense region within a column of contrast. The patient then underwent transoesophageal echocardiography (TEE), which indicated the presence of a 1.8×1.2 cm mobile density with broad base attachment to the margin of the left coronary leaflet on the aortic side of the valve (figure 2). Otherwise,

Figure 1 Contrast-enhanced CT of the thorax showing hypodense lesion bulging into the aorta.

the aortic valve was focally thickened without stenosis or insufficiency. Following the administration of an echocardiographic contrast agent, the brightness of the mass increased, which is consistent with tumour vascularity (video 1).

DIFFERENTIAL DIAGNOSIS

The differential diagnoses included haemangioma, angiosarcoma, myxoma and papillary fibroelastoma.

TREATMENT

Owing to the risk of recurrent embolisation of the tumour, we decided to surgically resect the mass. Preoperative coronary angiography demonstrated double-vessel coronary artery disease, and, therefore, simultaneous resection of the myxoma and coronary artery bypass grafting were performed. Transverse aortotomy indicated the presence of a 1.5×1.3 cm yellow-tan-coloured mass attached to the left leaflet of the competent aortic valve. The mass was successfully resected, without the need for additional aortic valve repair. Histological examination of the tumour specimen indicated a myxoid matrix with ovoid myxoma cells (figure 3).

OUTCOME AND FOLLOW-UP

Postoperative course was complicated by volume overload requiring diuresis. Then the patient was transferred to the inpatient rehabilitation unit and was subsequently discharged home in stable condition. No recurrence of the tumour has been identified after 2 years.

DISCUSSION

Primary tumours of the heart are rarely observed, with an incidence of 0.0017–0.19% at autopsy^{1 2}; the tumours are benign in 75% of these cases.^{1–3}

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Figure 2 Echocardiography showing measurements of myxma.

Cardiac myxoma is the most common benign tumour of the heart in adults. The left atrium is the predominant location for this tumour, followed by the right atrium and left and right ventricles. Valvular myxomas are very rare, particularly those of the aortic valve. To the best of our knowledge, only nine cases of aortic myxoma have been reported in the literature (table 1). However, the patient in the present case is the oldest among these cases; the reported age of the cases of valvular myxoma ranged between 15⁴ and 72 years.⁵

TEE enables the detection and further examination of aortic myxomas. Histopathological examination is the gold standard method for diagnosis. At diagnosis, most of the tumours are >1 cm in size. On clinical examination, one of the patients presented with embolic myocardial infarction,⁴ three patients had embolic stroke,^{6–8} one had embolic occlusion of the abdominal aorta, one patient demonstrated embolic occlusion of the common femoral artery⁹ and three patients exhibited an incidental finding on TTE.^{5 10 11} Furthermore, the myxoma was attached to the right coronary cusp in three patients,^{6 7 11} to the left coronary cusp in two patients (including the present case),¹⁰ and to the non-coronary cusp in two patients^{4 5}; two patients exhibited fusion of the left and right coronary cusps.^{8 9} Valvular myxomas tend to have a smaller cellular component, fewer giant cells and less perivascular cuffing of tumour cells.¹²

Recurrences

Recurrences are frequent in cases of cardiac myxoma. The efficacy of surgical resection with clear margins, while preserving the valve architecture^{4 10} and valve repair or replacement,^{9–11} is dependent on the size of the tumour, in order to prevent recurrence. In cases without complex myxoma syndrome, recurrence of tumour is rare if the resection is appropriately performed.¹³ In the present case, the tumour size was appropriate for surgical resection of the mass and implantation of the base. However, rigorous follow-up and surveillance should be the cornerstone of management of such cases.

Learning points

- ▶ Cardiac myxoma is a rare finding and valvular myxoma is extremely rare.
- ▶ Cardiac myxomas usually manifest as distal embolic phenomenon and are detected serendipitously on CT/MRI of the chest.
- ▶ If found, they should be resected to prevent distal emboli.

Video 1 Trans-esophageal echocardiography showing freely mobile myxoma.

Figure 3 Myxoid matrix with scattered ovoid myxoma cells.

Table 1 Characteristics of patients having aortic myxoma

Reference	Age at diagnosis	Presentation/incidental finding	Size	Complications	Surgery	Location	Comorbidity
Javed <i>et al</i>	81	Leg pain	1.8×1.2 cm	Myocardial infarction	CABG and resection	LCC	HTN, HLD, Aortic aneurysm
Kim <i>et al</i> ⁵	72	Shortness of breath	1.5×0.8 cm	None	AVR	NCC	HTN, A fib
Okamoto <i>et al</i> ¹⁰	61	Suspicion of endocarditis/Incidental	1×1 cm	None	Resection	LCC	HTN, DM
Koyalakonda <i>et al</i> ⁶	60	Paroxysmal A fib	1×1 cm	Stroke	Resection	RCC	A fib, HTN
Watarida <i>et al</i> ¹¹	58	Heart murmur	1.1×1 cm	None	AVR	RCC	HTN
Ramsheyi <i>et al</i> ⁷	32	Facial hemiparesis	1 cm	Stroke	AVR	RCC	None
Fernandez <i>et al</i> ⁸	28	Hemiparesis	1.5×0.7 cm	Stroke	AVR	RCC and LCC	Epilepsy
Kennedy <i>et al</i> ⁹	23	Leg pain	1.5 cm	Peripheral vascular disease	AVR	RCC and LCC	None
Dyk <i>et al</i> ⁴	15	Chest pain	4×1 cm	STEMI	Resection	NCC	None

AVR, aortic valve repair/replacement; CABG, coronary artery bypass grafting; DM, diabetes mellitus; HLD, hyperlipidaemia; HTN, hypertension; LCC, left coronary cusp; NCC, non-coronary cusp; RCC, right coronary cusp.

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Competing interests None.

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