

Myxoma of the aortic valve

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Received 23 November 2011; received in revised form 31 January 2012; accepted 18 February 2012

Abstract

Myxomatous tumours can arise from different cardiac structures. They have a special predilection for the left atrium and are an exceedingly uncommon finding in cardiac valves. We report the case of a 28-year old man who presented with a stroke and was found to have a mass arising from his aortic valve. The patient underwent a successful surgical excision of the aortic valve with the implantation of a mechanical prosthesis. The histopathological examination of the aortic valve confirmed the diagnosis of myxoma. Some aspects related to the diagnosis and management of this entity are discussed in this article.

Keywords: Myxoma • Aortic valve

INTRODUCTION

Myxomas are the most common cardiac tumours in adults. They derive from multipotential mesenchymal cells located within the endocardium and can originate from any chamber or cardiac structure [1]. Myxomas arising from heart valves are very infrequent [2], and to the best of our knowledge, only eight cases of isolated aortic valve myxomas have been published previously [2–10]. We report the case of a patient with a stroke who was found to have a mass implanted on his aortic valve. After surgical resection, the histopathological examination confirmed the diagnosis of myxoma.

CASE REPORT

A 28-year old male was admitted to the emergency department with a sudden onset of left hemiplegia and dysarthria. An urgent cerebral CT scan demonstrated an ischaemic stroke involving the territory of the middle cerebral artery. His medical history included epilepsy of unknown aetiology since the age of 18, but the patient had been free from seizures during the last 5 years.

On physical examination, peripheral pulses were present and normal. An early diastolic murmur could be heard in the aortic area. The ECG showed a normal sinus rhythm with signs of mild left ventricular hypertrophy. Transoesophageal echocardiography revealed a grade II/IV aortic incompetence and a mobile mass measuring 15 × 7 mm originating from the ventricular surface of the aortic valve was identified (Fig. 1).

A doppler ultrasound of the carotid arteries was normal. Blood cultures, inflammatory markers and immunological tests were all negative. The patient remained afebrile during the

whole period. Early physiotherapy was initiated with satisfactory progress of the patient's neurological status. Weekly follow-up transthoracic echocardiographic examinations were performed. Four weeks after admission, the mobile mass that was identified in relation to the aortic valve remained unchanged; therefore, surgical excision was scheduled after discussing the risk of new perioperative thromboembolic complications with the patient. An intraoperative inspection of the aortic valve revealed a 15 × 7 mm sessile mass attached to the ventricular surface of the right and left coronary leaflets. The mass was gelatinous soft, friable and yellowish in colour, with multiple haemorrhagic areas (Fig. 2). The patient underwent a standard aortic valve replacement with mechanical prosthesis.

The surgical specimen was fixed in 10% neutral buffered formalin. Routine 5 µm-thick sections were prepared from paraffin-embedded tissue and stained with haematoxylin–eosin. An immunohistochemical stain was performed using EnVision Plus method (Dako Glostrup, Denmark). The antibody used was directed against CD34 (clone QBEND/10, BioGenex; 1:20). Histopathological examination of the mass confirmed the diagnosis of myxoma. There was a sparse population of round and stellate cells mostly concentrating beneath the surface, with some cells forming solid cords and vascular channels (demonstrated with CD34 antibody). Cells were surrounded by abundant myxoid stroma. Mitosis, pleomorphism and necrosis were all absent (Fig. 2).

DISCUSSION

Although myxomas can originate from any cavity or cardiac structure, they originate most frequently from the left atrium,

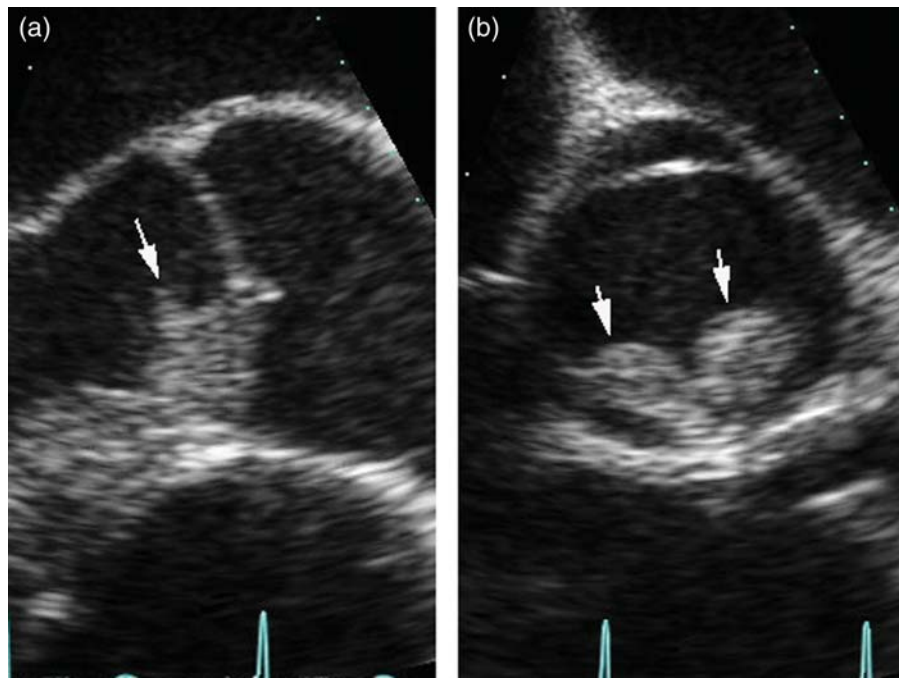


Figure 1: Preoperative transoesophageal echocardiogram. (a) Longitudinal view. The tumour is attached to the ventricular aspect of the aortic valve (arrow). (b) Transversal view. The tumour arises from the right and left aortic leaflets (arrows).

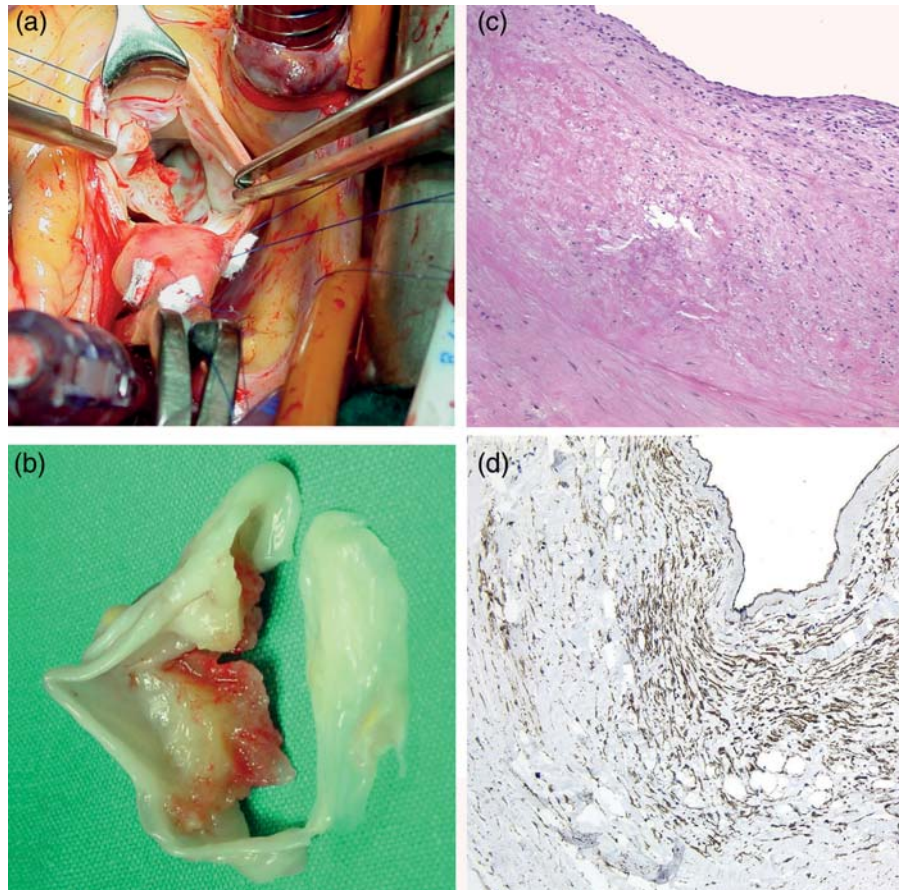


Figure 2: (a) Intraoperative image of the aortic valve showing a mass on the ventricular surface of the right and left cusps of the aortic valve. (b) Macroscopic view of the excised valve. (c) Histological section through the tumour (haematoxylin and eosin $\times 20$). (d) Vascular channels are demonstrated using CD34 immunostaining ($\times 10$).

followed to a lesser extent by the right atrium, then the left and right ventricles [1]. Myxomas of the cardiac valves are very unusual, especially those of the aortic valve [2]. To our knowledge, only eight cases of aortic valve myxomas have been reported. The first was described as a *post mortem* finding [2], while the clinical presentations of the other seven cases included: acute embolic stroke [3, 9], acute embolic myocardial infarction [8], acute embolic lower limb ischaemia [4], aortic stenosis [5] and accidental finding during a routine echocardiogram [6, 7]. This type of tumour has been described as arising from both the ventricular aspect [3, 6, 7] and the margin [8] of the valve cusps. The right [3, 6], left [5, 7] and non-coronary [8] leaflets may be affected either together or individually. In one of the reported cases [4], the right and left cusps were simultaneously affected (as in our patient).

Differential diagnosis of aortic valve myxoma includes vegetations, papillary fibroelastoma and Lamb's excrescences [1, 3, 4, 7]. Microscopic and immunohistochemical characteristics allow the distinction between these entities. As we have observed, aortic valve myxomas are a potential source of emboli; therefore, surgical removal should be indicated as soon as the diagnosis is confirmed. Surgical excision should include not only the tumour but also the implantation site to minimize the risk of local recurrence. Tumour resection with conservation of the native valve should be intended [7, 8], but sometimes due to a big tumour size and/or structural valve degeneration, replacement of the aortic valve may become necessary [4, 6]. Follow-up of these patients is highly recommended since distal tumour growth at the site of previous embolization as well as local recurrence of the tumour have been described [1] in previous reports.

Conflict of interest: none declared.

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eComment. Isolated valvular myxoma

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doi:10.1093/icvts/ivs280

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We read with great interest the report by Fernández *et al* [1]. They presented a case of a 28-year old male patient with a sessile myxomatous tumour localized on the ventricle aspect of the right and left cusps of the aortic valve. Valve-sparing aortic root reconstruction was impossible, therefore a mechanical aortic valve was implanted.

Myxomas are the most common primary cardiac tumours, typically located in the left atrium at the level of the fossa ovalis (three-quarters of these tumours) [2]. Valvular myxomas are extremely rare, atrio-ventricular valves being more involved than the aortic or pulmonary valves. The mitral valve is the most frequently affected structure, followed by the tricuspid, the aortic, and the pulmonary valves [3]. This case provides the possibility to discuss the clinical and pathologic features of valvular myxomas. Myxomas of the aortic valve are usually located at the ventricle side, involving one or two aortic cusps. The mean age at diagnosis is 44 (range 15–72). Ten cases of aortic valve myxomas have been published in the English literature [1, 4, 5], and they all showed a unique feature, that is, an isolated myxoma on the aortic valve. None of these patients presented with Carney's complex, therefore we can conclude that, to date, familial myxomas do not involve aortic valves.

Recently, an additional case report was published on the same subject [4]. A 10x8-mm myxoma attached to the non-coronary cusp of the aortic valve from the ventricle side was incidentally found during an echocardiogram. The 72-year old man was operated under cardiopulmonary bypass, the tumour was resected, and the aortic valve was successfully repaired. The postoperative course was uneventful. This is the oldest patient operated on with an aortic valve myxoma.

We completely concur with the statement that the main differential diagnosis is made with papillary fibroelastoma, which is generally described as arising from the ventricle aspect of the aortic valve, that a surgical approach should be considered in all these patients, and that echocardiogram follow-up must be recommended [5].

Conflict of interest: none declared.

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