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Multicentric Cardiac Myxoma Treated with Extended Surgery

M Yilmaz, O Gurbuz and M Cengiz

Department of Cardiovascular Surgery, Faculty of Medicine, Uludag University, Bursa, Turkey

We present the case of a 37-year-old male patient with a multicentric myxoma admitted to hospital with dyspnoea, syncope and chest pain. Physical examination revealed a grade 3/6 systolic murmur at the left lower sternal border and diffuse bilateral lung rales. Transthoracic echocardiography and thoraco-abdominal computed tomography revealed a right atrial mass and a right ventricular mass obstructing the outflow tract. The patient underwent urgent surgical treatment. At operation, a solitary right atrial myxoma and a right ventricular myxoma originating from the tricuspid valve and attached to the free wall of the right ventricle were seen. The tumours were successfully excised and the tricuspid valve was replaced with a No. 33 Omnicarbon® metallic valve. The right ventricular free wall was repaired with a glutaraldehyde-treated pericardial patch. Histopathological examination of the tumours confirmed the diagnosis of myxoma. Post-operative recovery was uneventful and there was no recurrence after 1 year's follow-up.

KEY WORDS: Myxoma; Multicentric; Tricuspid valve

Introduction

Myxomas are the most common benign cardiac tumours, accounting for 24% of all cardiac tumours.[1] Most occur in the left atrium (75 – 80%).[2] Right ventricular myxomas are rare, representing 5% of all myxomas, and 15% of right ventricular myxomas are associated with other cardiac myxomas.[3] The proportion of myxomas that are multicentric was reported to be less than 5% in a large series.[4] Valvar myxomas are extremely rare; only 22 myxomas originating from the tricuspid valve have been reported in the English literature and none of these was multicentric.[5] We present here the case of a 37-year-old male with multicentric (right atrial and right ventricular) myxoma that was successfully treated with extended surgery.

Case report

A 37-year-old male with a history of dyspnoea on exertion over a period of 1 year presented with worsening of symptoms, syncope and chest pain. He had severe dyspnoea with a heart rate of 110 beats/min and a blood pressure of 110/80 mmHg. Physical examination revealed a grade 3/6 systolic murmur at the left lower sternal border and diffuse bilateral lung rales. The liver was enlarged and palpable 7 cm from the right costal margin. There was diffuse peripheral oedema of the lower limbs. Electrocardiography showed sinus rhythm

with no signs of myocardial ischaemia. A chest X-ray revealed cardiomegaly with moderate pulmonary congestion. Transthoracic echocardiography showed a right atrial mass and another mass in the right ventricular cavity that was obstructing the right ventricular outflow tract. Thoracoabdominal computed tomography revealed right atrial and right ventricular masses. The presenting symptoms were attributed to right crural venous thrombosis leading to pulmonary embolism. The patient was scheduled for urgent operation.

OPERATIVE TECHNIQUE

Surgery was performed by standard median sternotomy. Extracorporeal circulation was instituted using bicaval and ascending aortic cannulation. Moderate systemic hypothermia (32 °C) was used and antegrade cardioplegia was administered for myocardial protection. The right atrium was then opened. An oval-shaped mass (4 cm in diameter) was found at the inferior edge of the coronary sinus; the pedicle of the mass was attached to the free wall of the right atrium. The mass was excised with the surrounding atrial wall and the right atrial wall was closed with a polypropylene 4/0 suture. The interatrial septum was then incised at the level of the fossa ovalis and the left atrium examined; no pathology was seen. The right atrial approach did not allow sufficient exposure of the second mass originating from the anterior leaflet of the tricuspid valve. Thus, a right ventriculotomy was performed and the large mass (7×4 cm) was excised together with the surrounding ventricular free wall and the anterior leaflet of the tricuspid valve (Fig. 1). The right ventricular free wall was repaired with an autologous glutaraldehyde-treated pericardial patch and a No. 33 Omnicarbon® metallic valve (MedicalCV Inc., Minneapolis, Minnesota, USA) was inserted at the tricuspid position. After direct closure of the atrial septum and right atrium, the patient was weaned off the extracorporeal circulation.

PATHOLOGICAL EXAMINATION

The microscopic appearance of frozen sections of the masses was consistent with the diagnosis of myxoma. This was subsequently confirmed by histopathological examination.

POST-OPERATIVE COURSE AND FOLLOW-UP

The patient had a fairly uncomplicated post-operative period. The constitutional symptoms resolved immediately after surgery. After the operation, the patient was given anticoagulation treatment using warfarin; there were no further episodes of pulmonary

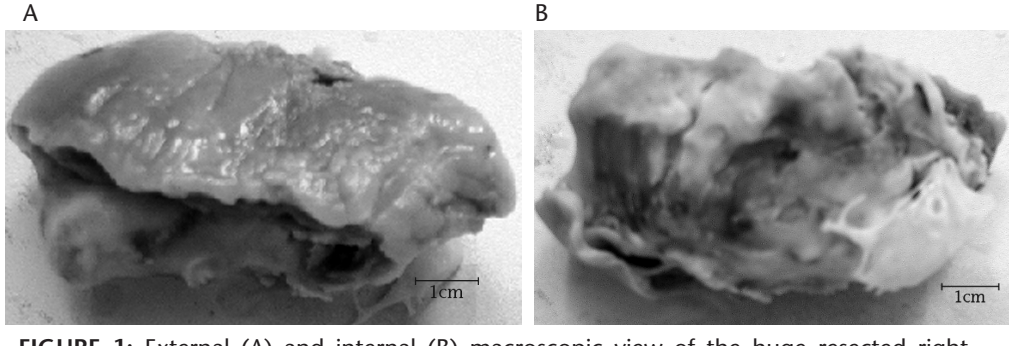

FIGURE 1: External (A) and internal (B) macroscopic view of the huge resected right ventricular mass, including the anterior leaflet of the tricuspid valve

embolism or any anticoagulation-related problems. At echocardiographic follow-up after 1 year, no recurrences were observed.

Discussion

Primary cardiac valve tumours are very uncommon and the involvement of any part of the tricuspid valve apparatus is extremely rare. Atypical locations and multiple myxomas occur most frequently in cases with familial myxoma.[6] Fewer than 1% of these tumours are part of Carney's complex – an autosomal dominant syndrome characterized by spotty pigmentation, cardiac and cutaneous myxomas, myxoid fibroadenomas of the breast, Cushing syndrome, acromegaly, bilateral sex cord tumours of the testicles and schwannomas.[7] Under these circumstances, screening of first-degree relatives should be undertaken. In the case presented here, there was no familial history and other signs of Carney's complex were absent.

The clinical features of myxomas are determined by their location, size, nature and mobility. Myxomas in the right ventricle can cause the clinical triad of intracardiac obstruction (30%), embolism (10%) and systemic symptoms such as syncope, exertional dyspnoea, chest pain and cyanosis.[8] Pulmonary embolism is seen in fewer than 10% of cases, but is the most dangerous complication of right-sided myxoma. Constitutional symptoms that resolve rapidly after surgical excision are seen in a number of patients.[8]

Once the diagnosis of a cardiac myxoma has been made, surgical excision should be performed without delay. Pulmonary embolism is the most critical complication of right-sided myxoma during surgery, but embolic complications can also occur during induction of anaesthesia, sternotomy or insertion of the venous cannula into the right atrium before the establishment of extracorporeal circulation. Caval or femoral vein cannulation, avoidance of excessive manipulation of the heart during surgery and use of a cross-clamp on the main pulmonary artery may help to reduce the risk of embolism. In the present case, cardiopulmonary bypass was instituted using direct caval cannulation in order to avoid pulmonary embolism and pulmonary hypertension during or after the operation.

Removal of adequate margins has been advocated to eliminate the risk of recurrence. Longitudinal right ventriculotomy has also been advocated as the approach of choice in right ventricular myxomas, although tumours have been excised through the right atrium.[3] There is still controversy concerning the most appropriate surgical approach to achieve complete excision of intracardiac myxomas. The best way to eliminate the risk of residual tumour in ventricular myxomas is to choose an approach that gives good exposure and the possibility of tumour-free resection margins. Although the use of a wide right-ventricular free wall resection may seem excessive, most of the wall requiring resection is not contractile before the operation because of tumour involvement. The glutaraldehyde-treated pericardial patch gives excellent handling and allows remodelling of the right ventricle, as in congenital pathologies.

Myxomas of the tricuspid valve are exceptional; only 22 cases have been reported to date.[5] Most tricuspid valve myxomas arise from the septal leaflet and can be excised without valve replacement. Kinugasa *et al.*[9] reported a myxoma arising from the anterior papillary muscle of the tricuspid valve that was excised and the valve was repaired by chordoplasty and annuloplasty. Follow-up revealed no recurrence or tricuspid regurgitation. Karagounis and Sarsam[10] reported a similar case in a 30-year-old

female patient who, again, had a satisfactory outcome. In the case presented here, the ventricular myxoma originated from the anterior leaflet of the tricuspid valve and the anterior leaflet was totally attached to the tumour. The anterior leaflet and tumour were resected together. Tricuspid annuloplasty has a risk of valve stenosis when the annular diameter of the tricuspid valve is normal. In the present case, a previous history of pulmonary embolism (requiring anti-coagulation) and the risk of tricuspid valve stenosis made metallic tricuspid valve replacement preferable to annuloplasty.

Today, primary cardiac tumours are a potentially curable form of heart disease. Many of the recurrences that do occur can be attributed to multicentric tumours or inadequate resection margins.[11] Some authors have suggested that structural and haemodynamic abnormalities of the cardiac valves may even occur with tumours that are not located on the valve because of the traumatic effect of friction between the leaflets and the tumour mass.[12,13] In the case presented here, nearly all the free wall of the right ventricle was resected and the wall was successfully repaired with an autologous glutaraldehyde-treated pericardial patch. We suggest that the resection margin should be free of tumour, including that of any involved valve. Repair of the valve is then the procedure of choice, but valve replacement should also be considered.

Conflicts of interest

No conflicts of interest were declared in relation to this article.

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Address for correspondence
Dr M Yilmaz
Department of Cardiovascular Surgery, Faculty of Medicine, Uludag University, Bursa 16059, Turkey.
E-mail: mertyilmaz@uludag.edu.tr or cvitr@yahoo.com