Case report - Cardiac general

Prolonged survival with left atrial spindle cell sarcoma

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Abstract

Primary spindle cell sarcoma of the left atrium is a rare tumour. Optimal treatment is to obtain complete surgical clearance of the tumour. The anatomic location of the tumour, infiltration into vital structures and difficult access provides a surgical challenge for resection of the lesion and reconstruction of the defect. The prognosis of patients with a primary cardiac sarcoma is very poor because of their resistance to treatment with chemotherapy and radiotherapy. Metastases and local recurrences are common despite optimal multimodality treatment. This report describes a 48-year-old gentleman who underwent multiple surgeries to achieve an 11-year survival since the diagnosis. The operative techniques have been described.

Keywords: Cardiac tumour; Spindle cell sarcoma; Left atrium; Survival; Outcome

1. Case report

A 48-year-old man initially presented 11 years ago with a left atrial mass, assumed to be a myxoma. Following surgical resection, histological analysis, however, revealed a sarcoma. He re-presented with a right scapula metastasis and left atrial recurrence, 3 and 7 years following his primary surgery, respectively. Despite tumour debulking and adjuvant chemotherapy, he was labelled as ‘inoperable’ at another centre. Subsequent to this, he re-presented with a tumour obstructing the mitral valve, 8 years following his primary surgery. Magnetic resonance imaging delineated a fat plane surrounding the apex of the tumour projecting into the atrioventricular groove. In view of this, he was treated by radical surgical excision of the tumour at our institution.

Following a redo median sternotomy, hypothermic (25 °C) cardiopulmonary bypass (CPB) was commenced with bicaval venous drainage and ascending aortic return. The heart was arrested and protected with intermittent, antegrade direct ostial cold blood cardioplegia and topical cooling. The tumour was large (LA size = 3.8 cm) and the only area from where we could approach without cutting into it was the roof of the left atrium. The aorta, main pulmonary artery and the superior vena cava were transected as decided preoperatively. The right atrium was opened parallel to the atrioventricular groove and the incision carried over to the atrial appendage and into the atrial septum.

The roof of the left atrium was opened at the base of the left atrial appendage and the incision was redirected into the atrioventricular groove towards A1. The tumour was obstructing the mitral valve (Fig. 1) and it was dissected free from its attachment in the atrioventricular groove and the mitral valve. The left atrial appendage was resected en bloc with the lesion (Fig. 2). The circumflex artery was seen in the atrioventricular groove and a clear view of the left ventricular free wall was obtained due to the absence of atrioventricular attachment and the A1–P1 commissure. The atrioventricular groove was reconstructed with a sandwich of Teflon. A large patch of bovine pericardium was sutured within the ventricular free wall, coming out across the mitral annulus. The A1–P1 commissure was sutured to the inner surface of the patch and the rest of the patch was used to reconstruct the left atrium. The great vessels were re-anastomosed following by atrial closure and weaning from CPB. Mild–moderate central mitral regurgitation was accepted and it was decided against replacing the mitral valve to avoid atrioventricular disruption. Histopathological analysis confirmed cellular spindle cell sarcoma with at least 7 mm clearance from the deep surgical resection margin. Postoperatively, he remained dyspnoeic with severe mitral regurgitation and so a mitral valve replacement was performed at six weeks following this surgery.

In spite of the previous radical surgery, the patient re-presented, 11 years following his primary surgery, with recurrent sarcoma obstructing the mitral valve prosthesis necessitating emergency surgery. Hypothermic CPB (32 °C) was instituted between femoral vessels prior to median sternotomy. A balloon endoclamp was inflated and cardioplegia was delivered into the aortic root. The left atrium

Fig. 1. Intraoperative view of the left atrial tumour with transected major vessels. (Aorta, MPA, main pulmonary artery and PV, pulmonary vein.)

Fig. 2. Excised left atrial spindle cell sarcoma.

was opened anterior to the pulmonary veins. Recurrent tumour was observed both on the native left atrium and on the bovine pericardial tissue from the previous surgery with two projections passing through the mitral prosthesis. The tumour and the mitral prosthesis were excised en bloc. It was decided against attempting to repair the back of the atrium that was left as a raw surface and a 27-mm bileaflet mitral prosthesis was implanted. The left atrium was closed and heart was weaned from bypass with the use of an intra-aortic balloon pump.

Histology confirmed recurrent sarcoma that had progressed to a higher-grade lesion with Mib1 staining up to 17.5%. Postoperative TTE reported well functioning valves without evidence of residual tumour and the patient was discharged on postoperative day 14.

2. Discussion

Primary cardiac neoplasia is rare [1]. Cardiac spindle cell sarcomas have been reported in the great vessels, pulmonary veins and the right atrium, but its occurrence in the left atrium is rare.

They manifest clinically by obstructing forward flow as in our case or with arrhythmias, embolic events and metastases [2]. Complete surgical excision prolongs survival [3] and life expectancy is almost twice as long for patients who undergo complete tumour resection compared to those with incomplete excision [4]. Surgical approach through the Water-son’s sulcus or interatrial septum is not feasible for large tumours as in our case. An alternative technique of autotransplantation was described in 1985 [5]. This method involves transection of both cavae, ascending aorta, main pulmonary artery and the left atrium just anterior to the pulmonary veins. The heart is explanted which gives excellent views and then reimplanted back after tumour resection. For locally advanced primary tumours, orthotropic allotransplantation could be considered. However, this method is limited by the poor availability of donor organs and requires lifelong immunosuppression that would be a risk in itself for neoplasia [6].

The most common cause of death even after complete macroscopic resection is local recurrence of the tumour in half the patients. Whatever the treatment, the prognosis is generally poor with a mean survival of 16.5 months after diagnosis [7].

We were aware of the proximity of the tumour to the atrioventricular groove but considering the age of the patient we considered that repairing the atrioventricular groove would not be an impossible task. Our technique offered an excellent view of the tumour. We were not only able to excise it, but were also able to reconstruct the defect in spite of multiple previous surgeries. Unfortunately, the patient had a recurrence again and needed another resection in just < 3 years. Our patient is alive for 11 years after the preliminary diagnosis of cardiac sarcoma with a good quality life.

In conclusion, we describe an alternative technique of accessing large left atrial tumours. Repeated attempts at resection of neoplasia in the left atrium are possible, although, technically difficult and require careful planning with skilled perioperative support. However, if such support is available intermediate survival is possible with good quality of life between surgical events.

References