

Interactive CardioVascular and Thoracic Surgery

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Interact CardioVasc Thorac Surg 2008;7:945-946; originally published online Jul 17, 2008;

DOI: 10.1510/icvts.2008.184085

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Case report - Cardiac general

Primary left atrial haemangioendothelioma

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Received 19 May 2008; received in revised form 23 June 2008; accepted 27 June 2008

Abstract

We describe the case of a 21-year-old female patient with epithelioid haemangioendothelioma of the roof of the left atrium. The patient underwent radical resection of the tumour with large disease-free margins, which required reconstruction of left and right atrial superior walls. The prognosis is unpredictable, but life expectancies ranging from 1 to 20 years have been described. There is no single treatment that can be prescribed but, because of their biologic behaviour, epithelioid haemangioendotheliomas must be regarded as fully malignant neoplasms and ought to be resected radically to prevent metastatic disease, and to improve life expectancy and quality of life.

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Keywords: Cardiac tumour; Cancer; Haemangioendothelioma

1. Introduction

Primary cardiac tumours are rare, with incidences range from 0.0017% to 0.2% in autopsy series [1, 2]. More than three-quarters of primary tumours are benign, and about half of these benign tumours are myxomas. Among malignant tumours the haemangioendothelioma is a very uncommon vascular tumour present in the soft tissues, characterized by capillary-sized vessels lined by rounded and multilayered endothelial cells.

In this report, we describe a patient with a haemangioendothelioma of the left atrium, with no previous cardiac history.

2. Case report

A 21-year-old female who complained of non-trauma related anterior chest pain for one year consulted her general practitioner in December 2006. She was medicated with anti-inflammatory drugs, with no remission of symptoms. She had previously been healthy and no special medical history was noted. On physical examination, no heart murmurs or cyanosis were found. The liver and spleen were not palpable. Her blood tests showed no relevant alterations. The tumour-related antigens carcinoembryonic antigen (CEA) and alpha-fetal protein (AFP) were in the normal range. Chest X-ray films showed a mildly enlarged anterior mediastinum. Computed tomography and MRI scan revealed a 3.9×2.7 cm mass located between the ascending aorta and the right branch of the pulmonary artery (Fig. 1). A transoesophageal echocardiogram (TEE) revealed

an extra-cardiac mass 'seating' in the roof of the left atrium, behind the aorta and between the pulmonary trunk and its right branch.

The patient was referred to our Centre where she was submitted to a small right exploratory thoracotomy and biopsy of the mass because a lymphoma was presumed initially and a tissue diagnosis was necessary for chemotherapy. Pathological examination revealed that the mass was of vascular origin, angioma or haemangioendothelioma. She was recommended for curative surgery.

A mediansternotomy was performed and cardiopulmonary bypass established with aortic and bicaval cannulation. Cold crystalloid cardioplegia was used. A retro-aortic tumour was found which extended from the intra-pericardiac portion of the right superior pulmonary vein, involved part of the wall of the right atrium and continued along the roof of the left atrium, reaching the base of the left atrial appendage and trunk of the pulmonary artery. There was no endo-cardiac involvement. The tumoural mass was resected en block with the roof of the left atrium, from the base of the left atrial appendage to the right pulmonary vein. Part of the wall of the right atrium including the distal portion of the superior vena cava was also resected. The defect created was reconstructed with bovine pericardium.

Pathological examination revealed capillary-sized vessels lined by rounded and multilayered endothelial cells. Cardiac muscle cells appeared compressed between proliferating vascular channels. Vascular endothelial markers, factor VIII-related antigens CD31 and CD34 were positive, while cytokeratin, desmin and CD117 were negative. There was no evidence of tumour either in the margins of the resected block or in the biopsies taken in the edges of the remaining atrial wall.

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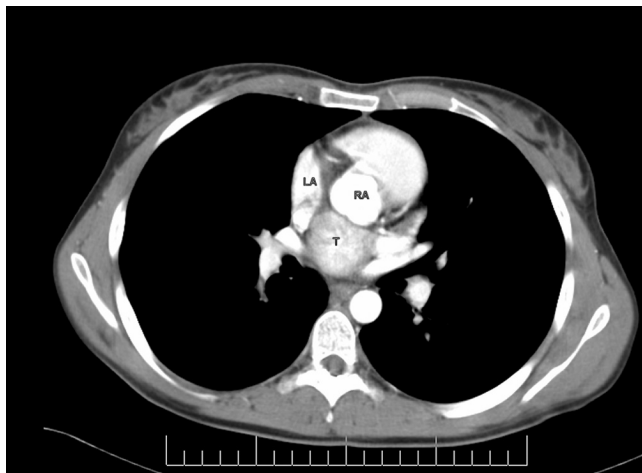


Fig. 1. CT-scan showing the tumour (T) located in roof of left atrium (LA), reaching the right atrium (RA).

During the postoperative course, a mild to moderate mitral regurgitation was detected by routine TEE. A small iatrogenic perforation of the base of the anterior leaflet of the mitral valve was corrected surgically. The patient was discharged from the hospital 15 days after the first intervention. She was asymptomatic, with no signs of recurrence at the 9-month follow-up.

3. Discussion

Primary cardiac tumours are very rare, with only a few cases of primary cardiac haemangioendotheliomas reported. In a review of the literature, we found 20 cases of cardiac haemangioendothelioma, of which only one was located in the left atrium [3–6].

Primary haemangioendothelioma was previously considered a low-grade or borderline malignant vascular lesion. According to the recent World Health Organization classification, it is classified as a malignant tumour, along with angiosarcomas, due to its local aggressiveness and metastasizing potential [7].

The epithelioid haemangiomas represent the benign end of the spectrum whereas the epithelioid angiosarcoma represents the malignant end. The epithelioid haemangioendothelioma is an intermediate variant of the family of vascular tumours. Nonetheless, the occurrence of systemic metastases in 21% of cases described in the medical literature [6], leads to the conclusion that this tumour must be regarded and treated as a fully malignant rather than

borderline neoplasm. Accordingly, it must be resected as radically as possible.

The pathogenesis of these rare cardiac tumours is unknown. They originate from the subendocardium and may occur at any part of the heart. Histologically, they are characterized by capillary-sized vessels lined by proliferating round, often multilayered, epithelial-like endothelial cells with cytoplasmic vacuoles. The expression of vascular endothelial markers, such as von Willebrand factor, CD31 and CD34, rules out metastatic adenocarcinoma or melanoma.

The prognosis is unpredictable but life expectancy ranging from 1 to 20 years was described [5]. In most reports, patients with haemangioendotheliomas have a favourable outcome after surgical resection. Chemotherapy and radiation therapy have a limited effect on this tumour [8]. In our present case, the tumour required extensive resection of the superior and posterior wall of the left atrium, involving also the left superior pulmonary vein and the distal portion of the superior vena cava and adjacent area of the right atrium, and subsequent reconstruction of the cardiac structures. As the resection of the tumour appeared complete, no additional therapy was considered necessary.

Although it is relatively premature, there is no sign of local recurrence or metastasis in our patient. In virtue of its malignant potential and infiltrative growth pattern, a close follow-up is recommended.

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