

Case Report

Cardiac leiomyosarcoma, a case report

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Abstract: In this case report we present the history of a patient admitted with recurrent pulmonary edema. Trans-oesophageal echocardiography showed a tumour in the left atrium, occluding the ostium of the mitral valve and mimicking intermittent mitral stenosis. Cardiac surgery followed by pathological examination revealed that the tumour was a leiomyosarcoma. Images from the echocardiography as well as the pathological findings are shown and discussed. The present case report illustrates that atrial tumors comprise also sarcomas, suggesting the use of careful, rapid diagnostic procedures and treatment to prevent dissemination of malignancy.

Keywords: Leiomyosarcoma, cardiac tumours

Introduction

Primary cardiac neoplasm is a rare condition with a prevalence of 0.001% to 0.03% in autopsy series. More common are metastases to or direct invasion of the heart, particular from neoplasm's with primary origin in the lungs or breast. About 75% of primary neoplasms are benign and most often atrial myxomas. Among the malignant primary tumours, angiosarcomas are most frequent, followed by rhabdomyosarcoma, mesothelioma and fibrosarcoma. Leiomyosarcoma occur in less than 1% of the malignant cases.

A leiomyosarcoma is a mesenchymal tumour that arises in smooth muscle cell tissue. In the heart, it is mostly localised in left atrium and often involves pulmonary veins causing dyspnoea, pericardial effusions, chest pain, atrial arrhythmias, peripheral embolism or heart failure. The tumour is growing rapidly with a high rate of distant metastases and local recurrence after removal. Therefore, prognosis is poor with a mean survival of 6 months after diagnosis [1].

Case presentation

We present a case of an 86-year old woman, hospitalised due to severe pulmonary oedema. Transthoracic echocardiography gave suspi-

cion of stenosis of the mitral valve. A trans-oesophageal echocardiography revealed a large tumour in the left atrium. The tumour was connected to the top of the atrium as well as to the muscular part of the atrial septum, consisting of both solid and cystic tissue. It was very mobile and was oscillating into the mitral valve ostium during diastole causing a moderate mitral stenosis (**Figure 1A-C**). Because of the morphology of the tumour, Positron Emission Tomography - Computed Tomography was performed preoperatively, showing activity solely in the left atrium, not revealing any metastases. The patient was unstable with several incidences of severe pulmonary oedema and low blood pressure despite medical treatment. Surgical removal of the tumour was uncomplicated. The 4 cm tumour involved both the muscular septum and the right upper pulmonary vein. After surgery the patient was discussed with oncologists at a specialised sarcoma centre and chemo- or radiation therapy was not recommended. The patient died 15 months after surgery.

Pathological findings

The tissue consisted of several firm and more soft pieces of tissue with colors varying from white to grey. In the biggest piece of tissue there was a cyst measuring 3 cm in diameter.

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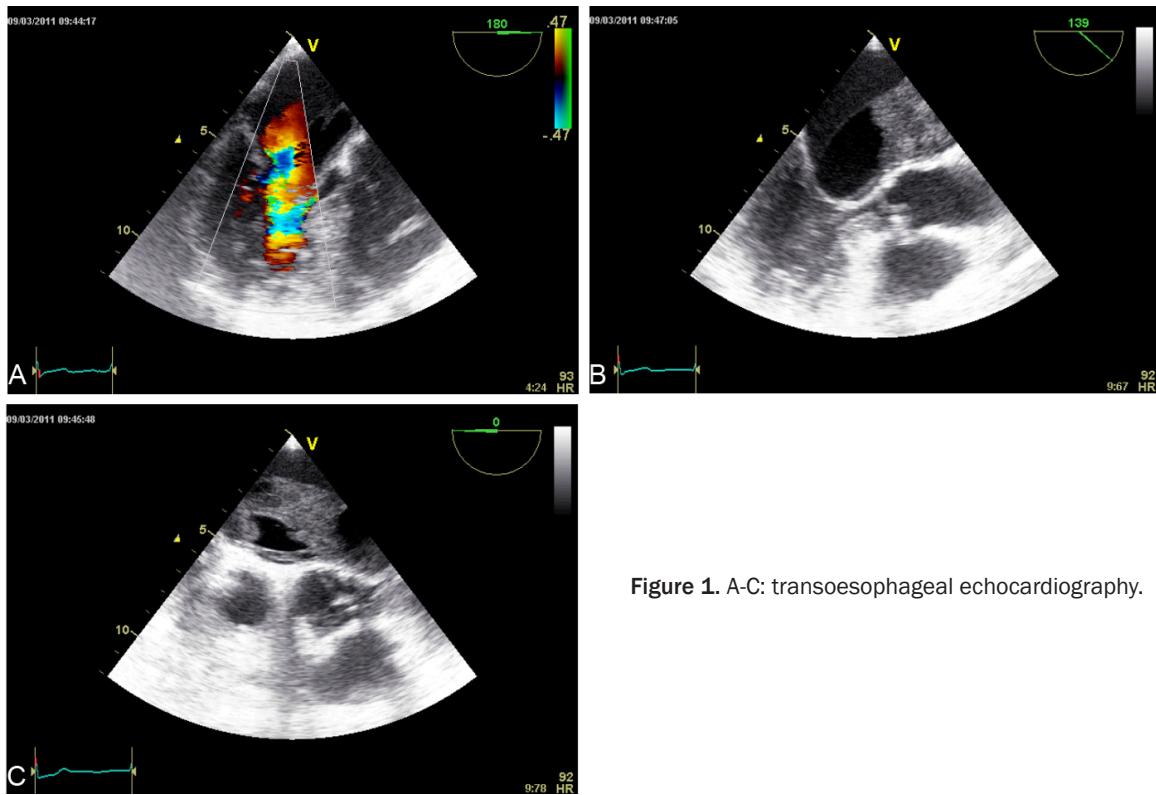


Figure 1. A-C: transoesophageal echocardiography.

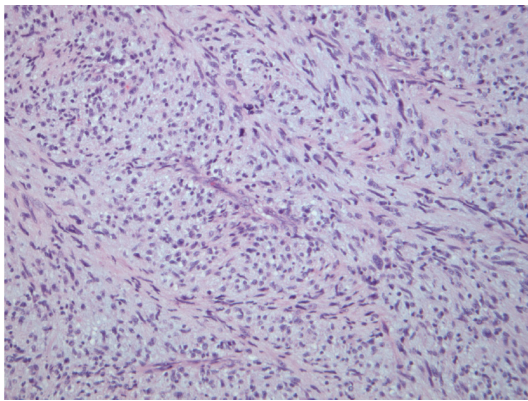


Figure 2. Hematoxylin-eosin staining of histological section of the tumour.

The histological sections had both vital and necrotic areas. About 30% of the tissue was necrotic. The vital areas consisted both of well-differentiated areas having a fascicular growth pattern but also of areas with a more reticular pattern. Some areas had high cellularity, but myxoid areas were also present. The tumour cell nuclei were characteristically elongated with a variable degree of pleomorphism. Mitotic figures were found with a frequency of 3 per 10 high power fields (**Figure 2**). Immunohistoche-

mical staining with the proliferation marker ki-67 revealed a proliferation index of 20%. Other immunohistochemical stainings showed positive areas for alpha-smooth-muscle actin and desmin. Based on these investigations, it was concluded that the tumour was a grade II leiomyosarcoma.

Discussion

We present a rare case of a large cardiac leiomyosarcoma involving the characteristic areas of both the smooth muscle cells of the left atrium and pulmonary veins. The clinical distinction between benign atrial myxoma and the malignant sarcoma may be difficult, and diagnosis must therefore be confirmed with both histopathological and immunohistochemical examination, where a strong positive reaction in tumour cells to alpha-smooth-muscle actin and desmin is diagnostic. Leiomyosarcoma has a high rate of local recurrences, and metastases are also described after radical surgical extirpation. Our case did not have detectable metastases at the time of diagnosis, but large areas with necrosis and a high mitotic activity were found, which may have worsened the prognosis. Other authors reported up to 6-12

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months of survival without surgery, and in cases with surgery the survival was extended to 24 months. Regarding adjuvant therapy, leiomyosarcoma has low radiosensitivity, and the risk of developing myocarditis and pericarditis exceed the benefits of radiotherapy. Concerning chemotherapy, the efficacy is still unknown [2-4]. As for all malignant diseases, early histopathological diagnosis is important for choice of treatment and prognosis. Therefore, any atypical morphology of something thought to be a typical benign atrial myxoma, should result in rapid diagnosis and treatment before dissemination of malignancy.

Disclosures

None.

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