Left atrial spindle cell sarcoma: a case report

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Background
Primary cardiac spindle cell sarcomas are extremely rare with only a few cases reported. They are frequently misdiagnosed on cardiac magnetic resonance (CMR) imaging as benign myxoma or thrombi and the suspicion of a malignant sarcoma arises only during surgery. This case report describes a case of cardiac spindle cell sarcoma diagnosed after surgery, where the initial diagnostic possibilities included an intramural thrombus and a cardiac myxoma.

Case summary
A 57-year-old woman was referred to our hospital for evaluation of a possible recurrent myxoma in the left atrium on echocardiography. Cardiac magnetic resonance imaging confirmed these masses as mural thrombotic masses, with a possible remnant of myxoma. After 2 months of anticoagulation therapy, the masses did not decrease in size on CMR imaging, and surgical removal was indicated. The atrial masses were surgically resected together with a large part of the left atrium. Histological examination showed spindle cell sarcoma. Unfortunately, the resection margins were positive and it was not possible to remove more atrium. PET-CT revealed metastasis in the right femur. The patient passed away 1 year after surgery.

Discussion
The rarity of spindle cell sarcoma and its similarities to benign cardiac myxoma and thrombi on echocardiography and CMR imaging present a diagnostic challenge when evaluating patients pre-operatively. Therefore, a malignant spindle cell sarcoma may only be diagnosed during surgery, after histological examination.

Keywords
Cardiac spindle cell sarcoma • Left atrial sarcoma • Myxoma • Case report

Learning points
- Benign mobile masses in the left atrium can be hard to distinguish from malignant mobile masses in the diagnostic phase.
- The suspicion of a malignant spindle cell sarcoma can only be confirmed during surgery, by histological examination.

Introduction
Primary cardiac spindle cell sarcomas are extremely rare with only a few cases reported.⁴⁻⁶ On cardiac magnetic resonance (CMR) imaging, they are often falsely diagnosed as benign myxomas or thrombi. A suspicion of malignant sarcoma often arises at a later stage (i.e. during surgery). The prognosis is poor, with a mean survival between 3 months and 1 year. Since the disease is rare, there is no consensus...
on optimal treatment. Some case reports describe improved patient survival when lesions are radically resected. However, evidence-based support is lacking. This case report describes a case of cardiac spindle cell sarcoma diagnosed after surgery, where the initial diagnostic possibilities included an intramural thrombus and a cardiac myxoma.

Timeline

| Date              | Event                                                                 |
|-------------------|------------------------------------------------------------------------|
| November 2014     | Minor stroke — echocardiography: mobile masses in left atrium suspected for primary cardiac myxoma |
| January 2015      | Resection of the left atrial myxoma, pathological confirmation in a foreign country |
| August 2015       | Follow-up echocardiography — recurrence of the mobile masses in the left atrium |
| September 2015    | Cardiac magnetic resonance imaging — mural thrombotic masses in the left atrium, myxoma could not be excluded |
| November 2015     | Cardiac magnetic resonance imaging — unchanged condition of the left atrial masses |
| February 2016     | Resection of the left atrial mass together with a large part of the wall of the left atrium |
| March 2016        | PET-CT — metastasis right femur, histologically confirmed with a CT-guided biopsy |
| July 2016         | Echocardiography — recurrence of the mobile masses in the left atrium |
| January 2017      | Patient passed away |

A 57-year-old woman was referred to our hospital for evaluation of a possibly recurrent lesion in the left atrium. In 2014, after a minor stroke, she was evaluated for a possible cardiac embolism origin. Echocardiography showed two small mobile masses attached to the base of the left atrium, which floated freely and occasionally passed through the mitral valve into the left ventricle, suspected for primary cardiac myxoma.

In January 2015, a resection was performed using minimally invasive surgery; 4 cm² of endothelium around the left pulmonary veins was removed together with two small masses of about 0.8 cm². After expert pathology consultation, a diagnosis of myxoma could be confirmed. The patient was dismissed from the hospital.

Case presentation

In August 2015, a routine follow-up echocardiography showed recurrence, and the patient was referred to the cardiology department at our hospital. She did not experience any (cardiac) complaints. On physical examination, she had normal heart sounds without heart murmurs. Transoesophageal echocardiography (TOE) showed two mobile masses in the posterior of the left atrium. Cardiac magnetic resonance imaging confirmed the presence of multiple mobile masses at the site of the septum, coumadin-ridge, right upper pulmonary vein, and left lower pulmonary vein and were interpreted as mural thrombotic masses (Figure 1). Radiological CMR imaging features best fitted the diagnosis of mural thrombotic masses and were interpreted by both a cardiologist and a cardiac radiologist (Figure 2). Furthermore, removal of endothelium during initial surgery increases the possibility of the formation of thrombotic masses. Nevertheless, myxoma could not yet be excluded (Figure 2D). Anticoagulation treatment in order to decrease the thrombotic masses was started. Despite adequate anticoagulation levels for 2 months, the left atrial masses did not decrease in size and surgical removal was indicated.

A transsternal thoracotomy was performed in February 2016. The large tumour seen on TOE was resected, together with a large part of the wall of the left atrium (Figure 3). A margin near the mitral valve was kept to place sutures for reconstruction. Another small tumour-like structure was removed from the atrial septum at the location of the oval foramen and the atrial septum was repaired with a pericardial patch (3 × 6 cm). Perioperative pathological frozen section analysis suggested malignancy. After inspecting the remaining left atrium, it was concluded that no more tissue could be resected. A total reconstruction of the left atrium through an autologous pericardial patch (10 × 15 cm) was performed. The surgery proceeded uncomplicated and the patient recovered well, aside from developing atrial fibrillation. Post-operatively, an echocardiography was performed and showed no masses in the left atrium.

Histological examination showed a cellular tumour with bundles and storiform fascicles of spindle-shaped tumour cells and some myxoid areas (Figure 4). There were no areas of necrosis. Extensive immunohistochemical workup was performed, showing strong and uniform expression of CD56 and sparse and heterogeneous positivity for pan-keratin, smooth muscle actin, and CD99. Other smooth muscle, lipomatous, vascular, melanocytic, and epithelial markers were negative. The resection margins were positive.
The morphological features were consistent with primary cardiac intimal spindle cell sarcoma. Based on immunophenotypical and molecular (MDM2-FISH, SS18-FISH) characteristics, other classes of sarcoma, including synovial sarcoma and (dedifferentiated) liposarcoma were excluded. However, expression of CD56 leaves a differential diagnostic possibility of malignant peripheral nerve sheath tumour, although morphological features argue against this classification. Unfortunately it was not possible to re-evaluate the first tumour resected in January 2015, since this was performed in another country.

A PET-CT was performed to evaluate possible metastasis. High FDG uptake, suggestive of metastasis, was found in the right femur. This was histologically confirmed with a CT-guided biopsy.

The patient was treated with five sessions of radiotherapy to the right femur. Due to the risk of tumour embolism as side effect of radiation, radiation therapy was not performed at the site of the cardiac mass, despite the positive resection margins after surgery. After treatment, patient returned to her country of origin. Several months later she returned for follow-up. Clinical condition of the patient had deteriorated over these months and she reported increasing fatigue and weight loss. An echocardiography was performed and showed a recurrence as a 6.5 × 2.2 cm large mass in the left atrium, passing through the mitral valve. Best supportive care was started, and the patient passed away in January 2017.

**Discussion**

In this case report, we describe a very rare malignant tumour in the left atrium, with the initial suspected diagnosis of a cardiac myxoma with thrombus but which after subsequent resection was diagnosed as a primary cardiac intimal spindle cell sarcoma. The rarity of this tumour and its radiological similarities to benign cardiac myxoma and thrombi on both CMR imaging and echocardiography present a
The diagnostic challenge when evaluating patients pre-operatively. Pathological examination of the tumour tissue is essential in correctly identifying this unique tumour. Therefore, a malignant spindle cell sarcoma may only be diagnosed after surgical resection. Performing the PET-CT before the surgery would not have changed the decision to perform surgery, since there still would be doubts about the mass in the left atrium. In retrospect, the first suspicion and confirmation of a malignant origin of the mass was made during surgery. Unfortunately, the tumour could not be radically resected and recurrence presented 5 months after surgery with a fatal outcome 11 months after surgery.

**Supplementary material**

Supplementary material is available at *European Heart Journal - Case Reports* online.

**Slide sets:** A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

**Consent:** The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

**Conflict of interest:** none declared.

**References**

1. Muturi A, Kotecha V, Ruturi J, Muhinga M, Waweru W. High-grade spindle cell sarcoma of the heart: a case report and review of literature. *J Cardiothorac Surg* 2015;10:46.
2. Mehta N, Desai A, Shivdasani B, Suryawanshi S, Mehta AB, Behranwala A, Dhabhar B. Left atrial spindle cell sarcoma—case report. *Indian Heart J* 2012;64:416–419.
3. Li Z, Hsieh T, Salehi A. Recurrent cardiac intimal (spindle cell) sarcoma of the left atrium. *J Cardiothorac Vasc Anesth* 2013;27:103–107.
4. Kholaf N, O’Neill BJ. Spindle cell sarcoma of the left atrium: an extremely rare and challenging tumour often masquerading as left atrial myxoma. *Can J Cardiol* 2015;31:104.e5–104.e6.
5. Ibrahim A, Luk A, Singhal P, War B, Zavodni A, Cusimano RJ, Butany J. Primary intimal (spindle cell) sarcoma of the heart: a case report and review of the literature. *Case Rep Med* 2013;2013:461815.
6. Fu B, Yu H, Yang J. Primary intimal (spindle cell) sarcoma of the left atrium. *Echocardiography* 2015;32:192–194.