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Case Report

CASE REPORT

Primary intimal sarcoma of the left atrium presenting with constitutional symptoms

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Abstract

Intimal (spindle-cell) sarcomas are exceptionally rare and are highly aggressive cardiac tumors. The authors describe a case of a 43-year-old female, presenting with a 3-month history of constitutional symptoms with fever, night sweats, anorexia and weight loss, associated with productive cough and pleural effusion that was admitted with clinical suspicion of pulmonary tuberculosis. The patient developed sudden acute heart failure symptoms during hospitalization, leading to mechanical ventilation. Computed tomography scan with contrast showed a cardiac tumor filling the left atrium causing compression of pulmonary veins. Surgical resection was performed and histologic examination revealed an intimal sarcoma. Although commenced on adjuvant chemotherapy, local tumor recurrence occurred with pericardium invasion. The patient died within 4 months of initial diagnosis. This report aims to describe an unusual presentation of this rare disease entity, and to discuss its highly aggressive clinical course.

INTRODUCTION

Primary cardiac tumors are rare with an incidence based on autopsy findings ranging from 0.001 to 0.030% [1, 2]. Malignant tumors account for 25% of primary heart tumors, and among those, sarcomas are the most prevalent [3]. Intimal sarcoma is a rare tumor, more commonly encountered in large arterial blood vessels but extremely rare in the heart [4–9]. The aim of this report is to describe an unusual presentation of this rare disease entity, and to underline its highly aggressive clinical course.

CASE REPORT

A healthy 43-year-old female presented in the emergency department with a 3-month history of constitutional symptoms with fever, night sweats, anorexia, weight loss and productive cough. She had been medicated with several antibiotics (co-amoxiclav and clarithromycin) for respiratory infections, without clinical improvement. There were no associated symptoms such as dyspnea, thoracalgia or syncope.

On physical examination, the patient had a regular heart rate of 100 beats per minute; blood pressure was 110/88 mmHg at supine position; body temperature was 36.6°C; cardiovascular system evaluation revealed a normal first and second heart sound, without murmurs; respiratory, abdominal and neurological examinations were also unremarkable. Laboratory analysis revealed a hemoglobin of 11.1 g/dl (normal: 11.8–15.6 g/dl). The remainder of complete blood count, urinalysis, and results of hepatic and renal-function tests were normal, as were electrolytes and glucose blood levels. Electrocardiogram showed

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sinus tachycardia. A chest X-ray was performed revealing bilateral pleural effusion and pulmonary edema. Non-contrast computed tomography (CT) scan confirmed bilateral pleural effusion associated with mediastinal lymphadenopathy and bilateral ground-glass opacification (Fig. 1). The patient was subsequently hospitalized for further management, with clinical suspicion of pulmonary tuberculosis.

Diagnostic thoracentesis revealed an exudate with lymphocytosis and mononuclear cell predominance. Cytologic examination showed acute inflammatory cells with no evidence of malignancy. Bronchoscopy with bronchoalveolar lavage revealed normal findings. Bacteriological and mycobacteriological studies, including blood cultures, sputum and bronchoalveolar lavage culture, were negative.

During hospitalization she showed clinical worsening with development of acute respiratory failure, interpreted as acute pulmonary edema. Non-invasive ventilation was initiated without clinical improvement, leading to orothracheal intubation and mechanical ventilation. In order to rule out pulmonary embolus a CT thorax with contrast was done revealing a 6 cm cardiac tumor that occupied two-third of the left atrium and compressed the pulmonary veins (Fig. 2). A transthoracic echocardiogram confirmed the left atrial mass compressing both pulmonary veins, with associated dilation of the right ventricle and signs of right ventricle overload. There was no evidence of distant metastatic disease on any of the initial scans performed as part of her staging work-up. Surgical resection was performed the following day and two fragments were removed: one of 2.6 × 1.4 × 1 cm and other of 1.6 × 0.5 × 0.6 cm. The bulk of the tumor was removed at that time although with surgical positive margins. Histologically, the specimen revealed a hypercellular malignant spindle-cell neoplasm with fascicular growth pattern and areas of necrosis, with associated lymphocytic focal infiltrate. Immunohistochemistry showed focal positivity staining for CD31 and multifocal positivity for murine double minute 2 (MDM2). Cytokeratin (Antigen E1/Antigen E3), CD34 and desmin expression were negative, compatible with a diagnosis of intimal sarcoma (Fig. 3).

Although submitted to adjuvant chemotherapy with paclitaxel, there was clinical worsening within 1 month period and the patient reported experiencing recurrent shortness of breath and fatigue. A transthoracic echocardiogram was performed, and revealed a new left atrial mass, with compression of pulmonary veins and pericardium invasion, associated with right ventricle dysfunction, suggesting local recurrence with invasion of adjacent structures (Fig. 4). Ultimately, within 4 months of diagnosis, the patient died due to acute heart failure.

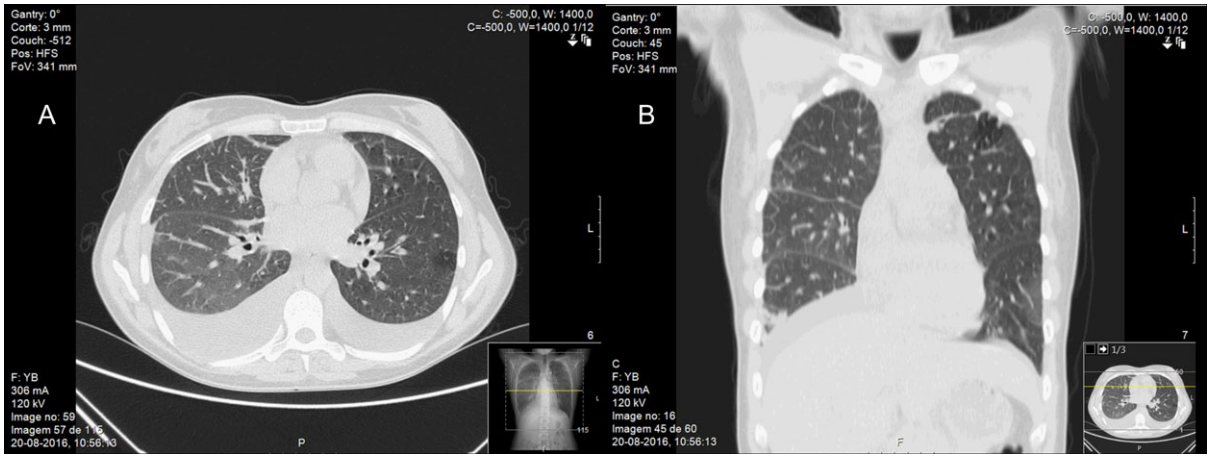

Figure 1: Non-contrast thorax CT scan showing bilateral pleural effusion associated with multiple mediastinal adenopathies and bilateral ground-glass opacification. (**A**) Transverse section and (**B**) Coronal section.

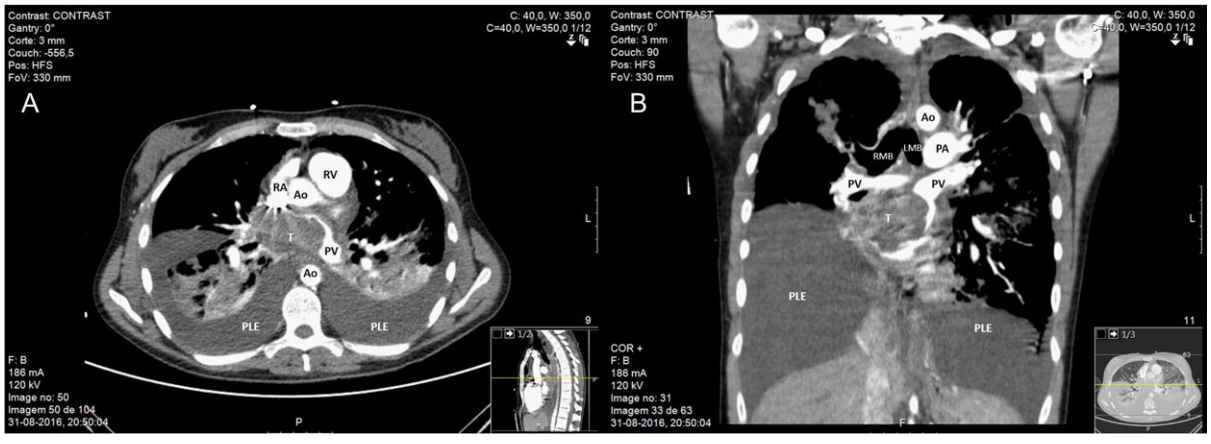

Figure 2: Thorax CT scan with contrast showing tumor in the left atrium compressing both pulmonary veins; (**A**) Transverse section, (**B**) Coronal section. PLE, pleural effusion; RMB, right mainstem bronchus; LMB, left mainstem bronchus; Ao, aorta; RV, right ventricle; RA, right atrium; PV, pulmonary vein; PA, pulmonary artery; T, tumor.

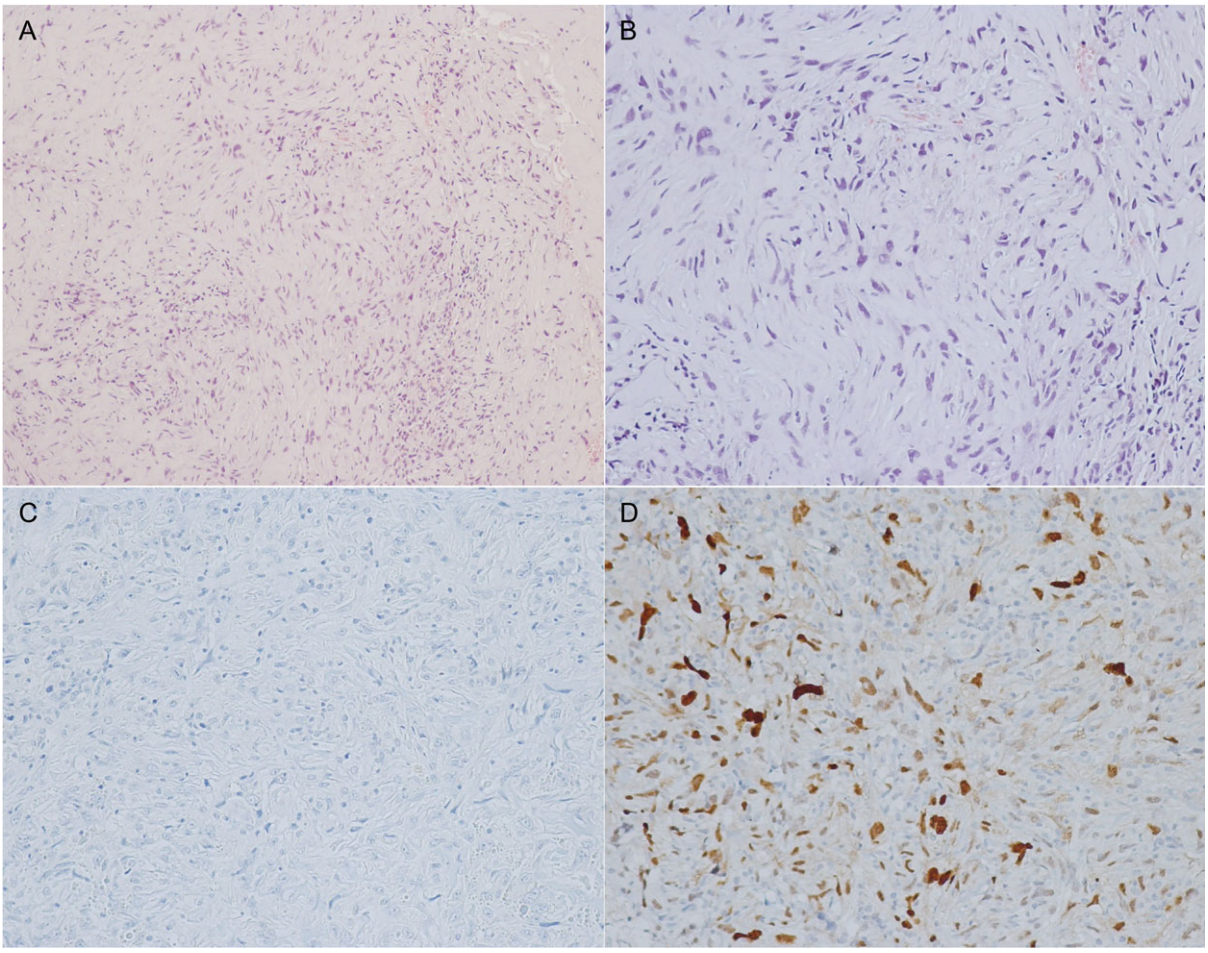

Figure 3: Histologically, the neoplasm showed highly packed spindle cells, with fascicular growth pattern and areas of necrosis, with associated lymphocytic focal infiltrate. (**A**) Hematoxylin-eosin stain, magnification: ×100; (**B**) hematoxylin-eosin stain, magnification: ×200. (**C**) Immunohistochemistry profile shows negativity for desmin and (**D**) positivity for MDM2.

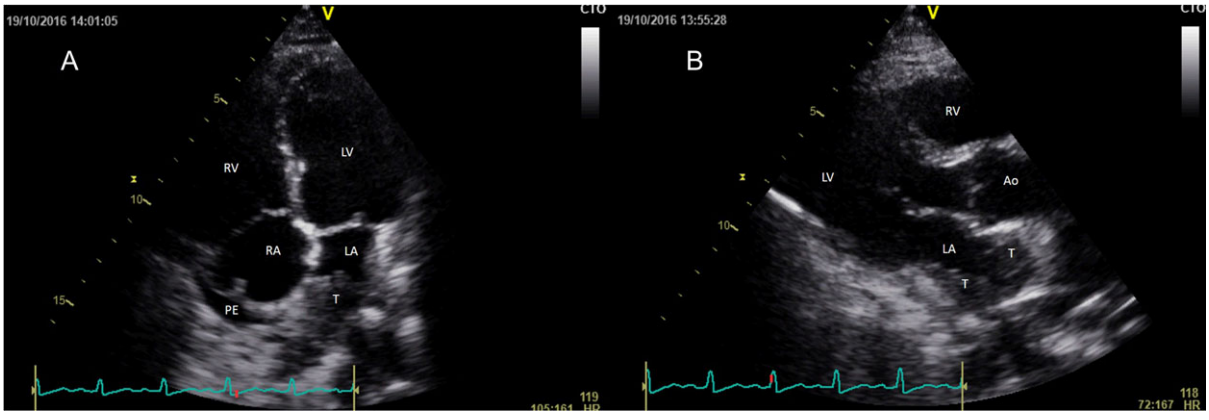

Figure 4: Transthoracic echocardiogram showing the recurrent left atrium tumor. (**A**) Subcostal four chamber view and (**B**) parasternal long axis view. Ao, aorta; LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle; T, tumor; PE, pericardial effusion.

DISCUSSION

Mesenchymal origin sarcomas are the largest group of primary cardiac malignant neoplasms. The most commonly occurring types are angiosarcomas, leiomyosarcomas, liposarcomas and malignant fibrous histiocytomas [2]. Cardiac intimal sarcomas are classified as undifferentiated sarcomas. These tumors are composed by atypical and tightly packed spindle-shaped cells arranged in long fascicles, but can also show areas of necrosis [9]. Since these tumors are extremely rare, initial pathological and radiological features might be confused with those of more common

malignant sarcomas types or even benign cardiac masses. In addition, a prompt diagnosis of this kind of tumors may be particularly challenging as they share common staining features with other sarcomas, such as angiosarcomas.

Cardiac tumors have many clinical presentations. A classic triad often characterizes them: cardiac symptoms and signs resulting from intracardiac obstruction, signs of systemic embolization, and systemic or constitutional symptoms [3]. Furthermore, manifestations due to metastases and other cardiac symptoms can also occur, such as atrial fibrillation, other conduction abnormalities or pericardial effusion. These tumors are diagnosed by transthoracic and transoesophageal echocardiograms, magnetic resonance imaging and CT scan. Intimal sarcomas usually show positive immunoreactivity for vimentin, osteopontin and MDM2, and variable positivity can be observed for desmin [9].

Although elective cardiac sarcoma therapy includes when possible complete surgical excision, followed by radio- and chemotherapy, its prognosis remains poor, as proven in our patient. Achievement of negative resection margins is a significant factor to prolonged patient survival, although complete surgical excision is often not possible due to the highly aggressive nature of the tumor and its frequent involvement of contiguous critical structures. Surgery can offer dramatic palliation of symptoms in cases of valvular and/or vascular obstruction, but local recurrence and metastasis occur frequently and early, usually within the first year [10]. In conclusion, it remains clear that early diagnosis and treatment are of the utmost importance as they have prognostic and therapeutic implications. Diagnostic suspicion must be high, as the symptoms may be non-specific and non-contrast CT scan may be non-diagnostic, such as reported in this case. These features contributed to delayed diagnosis given that there were other more common causes for the clinical findings. Final diagnosis was made upon a sudden clinical worsening during hospitalization by performing a CT scan with contrast. Thus, the report of these cases is of paramount importance in order to reinforce clinical awareness regarding such aggressive tumors.

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CONFLICT OF INTEREST STATEMENT

None declared.

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ETHICAL APPROVAL

No ethical approval was required in this work.

GUARANTOR

Edite Marques Mendes.

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