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Undifferentiated pleomorphic sarcoma with focal myogenic differentiation mimicking left atrial myxoma

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Abstract

Primary cardiac tumors are very rare. Cardiac sarcomas can be detected at any age, regardless of gender. At diagnosis, these are mostly symptomatic and one-third metastatic. Undifferentiated pleomorphic sarcomas account for 1.7% of all cardiac tumors, are typically localized in the left atrium, and involve the mitral valve. Here we present a case of left atrial undifferentiated pleomorphic sarcoma with myogenic differentiation that clinically and radiologically mimicked myxoma.

Keywords: Left atrium, Myogenic differentiation, Myxoma, Sarcoma

Introduction

Primary cardiac tumors are very rare. Their incidence in the autopsy series has been reported as 0.001-0.28%. Undifferentiated pleomorphic sarcomas (UPSs) account for 1.7% of all cardiac tumors [1]. UPSs are typically localized in the left atrium and tend to involve the mitral valve [2]. Here we present a case of left atrial undifferentiated pleomorphic sarcoma with myogenic differentiation that clinically and radiologically mimicked myxoma.

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Informed Consent
The authors stated that the written consent was obtained from the patient presented with images in the study.

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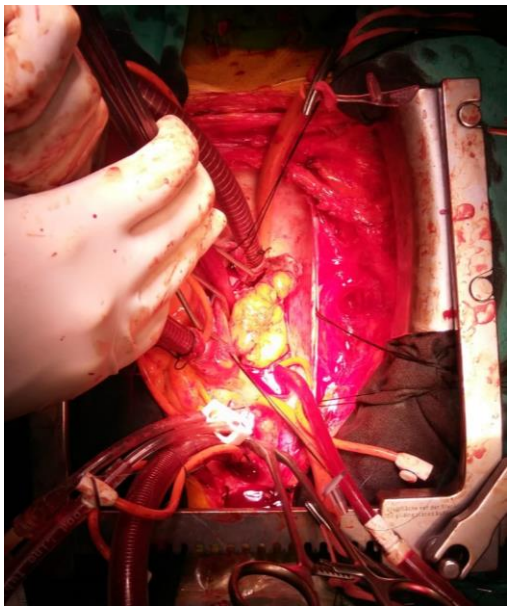
Case presentation

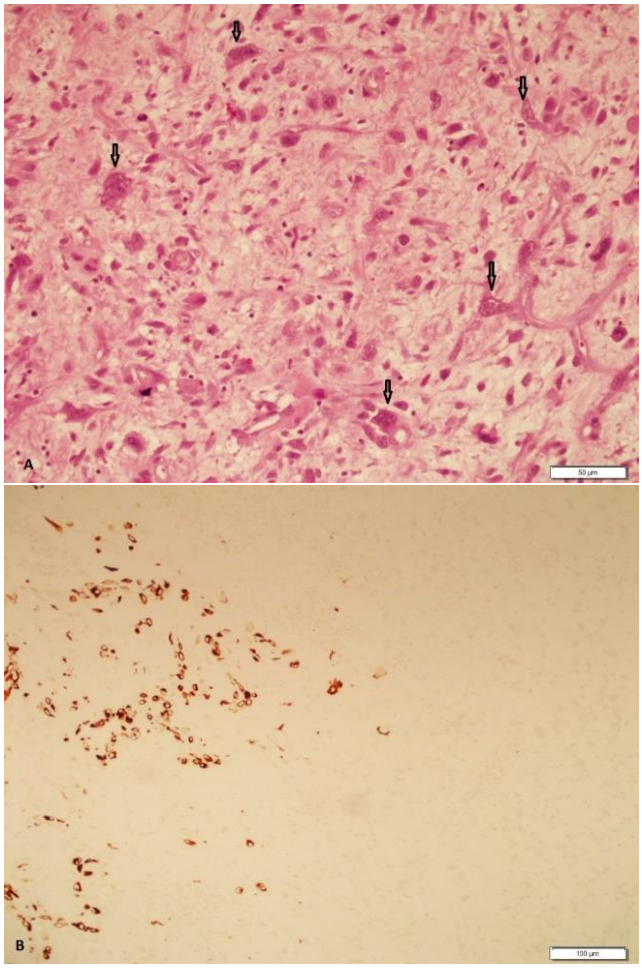
A 37-year-old male patient was admitted due to the complaints of chest pain, palpitation, syncope attacks, and dyspnea that had continued for approximately the past 2 months. His physical examination revealed a 6/5 murmur in the mitral focus. Echocardiography identified a mass (prediagnosed with myxoma) associated with a mitral valve in the left atrium, with a size of 41 × 28 mm, grade 2 mitral regurgitation, and advanced tricuspid regurgitation. The pulmonary arterial pressure was 90 mmHg, and the ejection fraction was 58%. After performing preoperative preparations of the patient and providing required pre-surgical patient instructions, he was submitted to median sternotomy under general anesthesia and right atriotomy under cardiopulmonary bypass following implementation of aorto bicaval cannulation. Left atriotomy was performed because the neighborhood of the mass could not be clearly differentiated when resection of the mass was attempted, and it was observed that the mass elongated from the subendocardial region to the anterolateral commissure of the mitral valve and obstructed mitral orifice (Figure 1). The tumor was subtotally resected. The patient who was postoperatively taken to the CVC intensive care unit was weaned from mechanical ventilation at the postoperative 4 h and externed 6 days later.

Figure 1: Subendocardial cream yellowish mass seen during left atriotomy



Pathological examination, macroscopically, indicated a dirty-cream colored mass with a size of 6 × 4 × 2.5 cm, regular contour, and its crosssection surface with cream-brown color and heterogeneous appearance. Microscopic examination encountered a tumor characterized by diffuse infiltration of pleomorphic, partly multinuclear atypical cells with a large polygonal shape, a large eosinophilic cytoplasm, and intense and atypical mitotic figures in the myxoid stroma (Figure 2A). There were foci of necrosis, and some areas were more cellular. The tumor cells were diffusely immunoreactive with vimentin and focally immunoreactive with SMA and desmin (Figure 2B). Pancytokeratin, S-100, CD34, CD31, myogenin, and myoD1 were non-immunoreactive. The Ki-67 proliferative index was 40%. The patient was diagnosed with undifferentiated pleomorphic sarcoma with myogenic differentiation.

Figure 2: A. Diffuse infiltration of atypical cells (arrows) with hyperchromatic nuclei, a large polygonal shape, large eosinophilic cytoplasm, atypical mitotic figures in the myxoid stroma (Hematoxylin-eosin ×200). B. Focal immunoreactivity in tumor cells with desmin (Desminx100)



No new mass was encountered by echocardiography imaging of the patient at the seventh postoperative month. No residual valvular leak or organic complication was detected by the echocardiographic examination.

Discussion

Cardiac sarcomas can be detected at any age, regardless of gender. At diagnosis, they are mostly symptomatic and one-third metastatic [3]. Dyspnea, pulmonary venous hypertension, and pulmonary edema are the most commonly seen symptoms [4]. Other symptoms are fever, arrhythmia, chest pain, pericardial tamponade, and congestive heart failure [3]. Also, in our case, dyspnea, tachycardia, and pulmonary hypertension were present consistently with the literature. However, no metastasis was detected.

Left atrial primary sarcomas constitute at least 50% of cardiac sarcomas. UPS is the most frequently type seen among those. A small part of UPS may be seen in the other cardiac sites. These tumors can be easily differentiated from myxomas with characteristics of infiltrative growth in the arterial wall and the absence of a connection with atrial septum by cardiac MRI [5, 6]. Cardiac sarcomas are mostly diagnosed by transthoracic echocardiography. The combination of echocardiography, CT, and MRI guides the preoperative evaluation of the tumor in terms of size, localization, its anatomical relationship with cardiac structures and valves, and differentiation between benign and malignant tumors [3]. This differentiation has importance in the determination of the most appropriate surgical procedure.

The presence of the mass can be detected using only echocardiography, whereas its use alone may be deceptive in identifying the real nature of the atrial masses [7]. UPS may be confused with myxoma because of its endocardial growth pattern in echocardiography [6]. Also, in our case, only echocardiography was implemented as the preoperative imaging technique. However, MRI or CT was not performed. Consequently, the tumor with left atrial localization that presented endocardial growth was erroneously prediagnosed with myxoma in the preoperative period.

The definite diagnosis of the cardiac tumors requires histopathological examination. Almost all soft tissue sarcomas may be seen in the heart. However, consideration of the same histological classification is difficult because there is a limited site where they may reveal morphologically specific differentiation. Besides, even detailed immunohistochemical assessment may be inadequate for differential diagnosis due to the frequent lack of tissue-specific antigens [3]. Microscopically, the important diagnostic parameters are structural, cellular, and microvascular patterns with stromal reaction, hemorrhage, and necrosis [3]. The diagnosis of UPS is reached as an elimination diagnosis. Therefore, a detailed immunohistochemical panel is crucial for differential diagnosis. The differential diagnosis involves other pleomorphic sarcomas, melanoma, anaplastic large cell lymphoma, and pleomorphic and metastatic carcinomas. In our case, anaplastic large cell lymphoma, melanoma, pleomorphic and metastatic carcinomas, and rhabdomyosarcoma were eliminated by the negativity of LCA, S-100, pan-cytokeratin, and myogenin, respectively. UPSs may rarely indicate myogenic differentiation, and this entity has been reported as a poor prognostic factor [8]. Cipriani et al. [8] have detected myogenic differentiation in 15 of 38 UPS cases in their study. Focal expression of desmin and SMA was present in our case. Thus, our patient was diagnosed with undifferentiated pleomorphic sarcoma with myogenic differentiation based on the absence of diffuse expression for these two muscle markers (desmin and SMA), myxoid background, and pleomorphic cells.

The treatment of cardiac sarcomas is complete surgical resection. However, it is usually unavailable. Palliative surgery may be performed to relieve valvular or vascular obstruction symptoms in cases where complete surgical resection cannot be performed. However, local recurrence or metastasis is frequently seen in the first year for the cases who underwent palliative surgery [9]. Although additional adjuvant treatment with chemotherapy or radiotherapy has been defined after surgical treatment, they can provide a limited treatment benefit, and there is still no evidence-based recommendation yet [9, 10]. Complete surgical resection could not be performed. Also, in our case, the mass that elongated from the subendocardial field to the mitral valve and caused an obstruction could be subtotally resected. No recurrence and/or metastasis was detected in the patient at the seventh postoperative month.

Conclusion

Primary cardiac sarcomas are rarely seen, presenting a poor prognosis despite surgical and adjuvant treatments. Even though concurrent use of multiple imaging techniques is useful at the stage of prediagnosis to create an appropriate surgical strategy in the establishment of the accurate diagnosis, definite diagnosis requires histopathological examination. It should be kept in mind that, particularly as an elimination diagnosis, that UPS may present myogenic differentiation. The inclusion of such rarely seen subtypes of cardiac sarcoma in the literature will help raise physicians' awareness of this possibility.

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