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Case Report

Primary Pulmonary Vein Leiomyosarcoma: A Case Report

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ABSTRACT

Primary leiomyosarcoma of the heart is extremely rare and found in about 0.2% of all cardiac tumors. Here in a 26-year-old man with progressive dyspnea, which had started since 2 months ago, is presented. Echocardiography revealed a left atrial mass, which was suggestive for a atrial myxoma.On the surgery, a tumoral tissue in the left atrium and pulmonary veins with attachment to peripheral soft tissue, was seen and incompletely resected. Histologic examination exhibited a hypercellular,necrotic and mitotically active spindle-celled tumor with fascicular arrangement. Immunohistochemistry showed a positive reaction to SMA in tumoral cells. The patient was advised to refer for postoperative chemotherapy, which was rejected. One year later, the patient was brought to hospital with tumor recurrence. Chemotherapy was initiated for the patient immediately, but the patient was expired 3 days later.

Key words: Heart, Leiomyosarcoma, Iran

Introduction

Primary cardiac neoplasms are unusual and found in only about 0.0017% of autopsies (1, 2). Approximately 25% of cardiac neoplasms are malignant (3) and mostly represented by sarcomas (4, 5). Leiomyosarcoma of vascular origin comprises a seemingly rare group of tumors illustrated by the fact that a few hundred cases have been reported in the literature since the initial report by Pearl in 1871 (2, 4). Primary leiomyosarcoma of the heart is also extremely rare (6) and found in less than 0.2% of all cardiac tumors (3). At the time of primary diagnosis, leiomyosarcoma often shows advanced local invasion or even metastasis and the prognosis is poor (1, 7).

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Case report

A 26 year-old man was referred with the history of progressive dyspnea since 2 months ago. Complete clinical and paraclinical evaluation was performed. Systolic murmur was detected in physical exam. A left atrial mass, suggestive of atrial myxoma, was noted at echocardiography while chest CT scan showed a large heterogeneous mass involving pulmonary vein and left atrium. All lab data, expect for a high creatinin level, were within normal limits prior to surgery.

A large mass was detected in operation room, which could not be resected totally due to firm adhesion to adjacent structures. No metastasis was noted .The specimen in formalin sent to the Pathology Department consisted of multiple irregular elastic masses with the dimensions 10x6x4cm. Cross sections showed creamy-yellow non-homogeneous cut surfaces with dilated vascular channels slit appearance were seen as well. Microscopic examination showed a hypercellular neoplasm composed of spindle cells with myoid features arranged in a fascicular pattern and occasionally surrounding large vessels. In some areas, there were individual cells with elongated eosinophilic cytoplasm and central nuclei embedded in a loose myxoid stroma containing chronic inflammatory cells. Extensive necrosis and hemorrhage were also evident with 1-3 mitotic figures in 10 high power field (Fig. 1, 2). Immunohistochemically, the tumor cells were positive for smooth muscle actin and desmin (Fig. 3) and were negative for myogenin, myoD1and cytokeratin .Percentage of Ki67 positivity was about 25%. Histologic and IHC findings are in favor of a leiomyosarcoma.

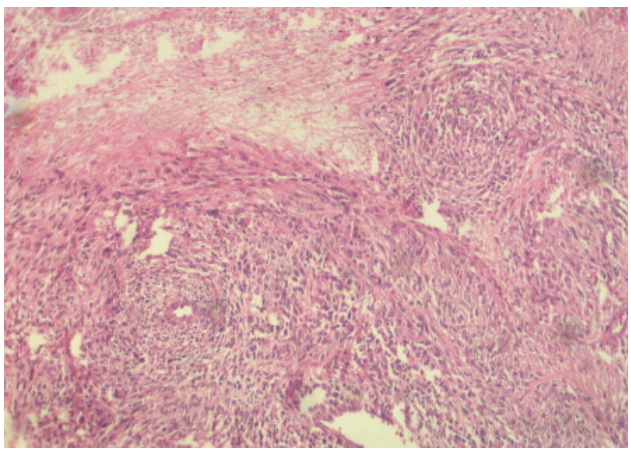

Fig. 1: Hypercellular neoplasm of spindle cells with extensive necrosis and hemorrhage (H&E x100)

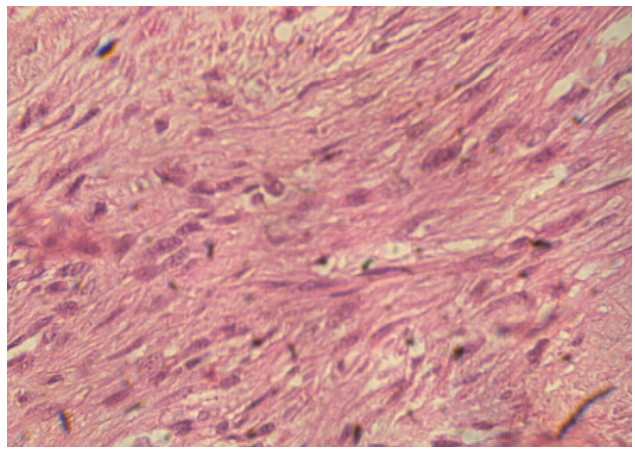

Fig. 2: Highly atypical spindle cells with myoid feature arranged in fascicular pattern(H&E x400)

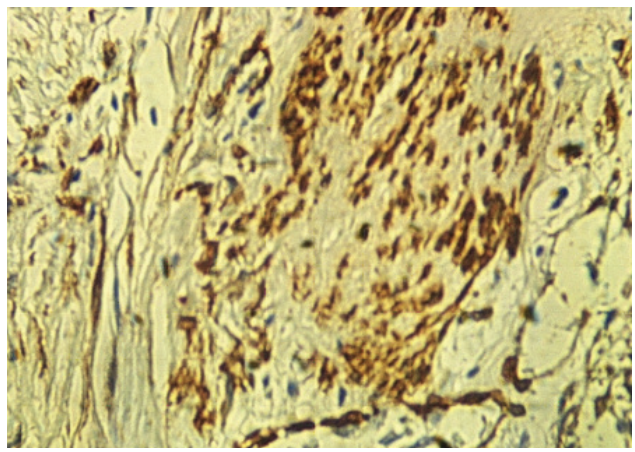

Fig. 3: Positivity of tumoral celle for smooth muscle actin (SMA) marker

The patient developed ATN after the surgery but was finally discharged from hospital with good general condition and was advised to refer for postoperative chemotherapy, which was rejected.

One year later the patient was brought to hospital with servere dyspnea (FC II – III), cough, hemoptysis , weight loss and hoarsness . Physical exam revealed a holosystolic murmur of III / VI of severity. All lab data were within normal limits except for a serum uric acid of 9.4 gr/dl. Chest X Ray showed left atrial enlargement. No evidence of metastasis was detected. Chemotherapy was initiated for the patient immediately, but the patient suffered from decreased level of consciousness and respiratory failure. Eventually he was expired 3 days later.

Discussion

Cardiac malignant tumor is a rare disease. Most of these tumors are sarcomas including angiosarcoma,

rhabdomyosarcoma, fibrosarcoma and liposarcoma. Primary cardiac leiomyosarcoma is very rare (6).

Clinically the patient most often presents with dyspnea, as did the reported case, but have different clinical, morphologic or radiologic features (2, 6).

Cardiac malignant tumors occur preferentially in the right side of the heart. An exception is leiomyosarcoma that occurs predominantly in the left atrium and tends to invade pulmonary veins and mitral valve (8-10).

Some authors reported cases of left atrial leiomyosarcoma originating from the wall of pulmonary vein (2).This also possible for our case because both left atrium and base of pulmonary vein were involved by tumor.

The preferential left atrial location and frequently myxoid appearance of carciac leiomyosarcoma make it difficult to differentiate them preoperatively from atrial myxomas, so intraoperative frozen section diagnosis and wide surgical margin are recommended for all atrial myxoid tumors (10).

Diagnosis of left atrial leiomyosarcoma is frequently delayed and may result in poor prognosis. Complete surgical resection is still primary therapy and postoperative chemotherapy should be considered because of possible incomplete resection, as was in this case (1, 4, 8).

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