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Case Report

Diagnostic dilemma of cardiac synovial sarcoma – Primary or secondary

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ABSTRACT

Cardiac synovial sarcomas are rare tumors. Few cases are reported in literature. They commonly present with heart failure. Surgery is the primary modality of treatment where possible. Radiation and chemotherapy are used for local and systemic control as indicated. We present a rare case of synovial sarcoma with simultaneous cardiac and soft tissue involvement. The case is presented for its rarity and diagnostic difficulty in determining the primary site of origin.

1. Introduction

Synovial sarcoma is a rare malignancy, comprising approximately 10% of all soft tissue sarcomas (STS).[1] It occurs most often in children and young adults and is an aggressive tumor with 10-year-overall survival rates as low as 0–20%.[2,3] A characteristic chromosomal abnormality t(X;18) is used for the confirmation of synovial sarcoma.[4]

Primary or metastatic cardiac synovial sarcomas are very rare. We present a case of metastatic monophasic synovial sarcoma to heart.

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2. Case report

A 34-year-old male presented with severe pain in the left side of the chest with restricted neck movements and multiple painless subcutaneous swellings over the upper and lower limbs of 3 months duration associated with anorexia and fatigability.

Contrast enhanced computed tomography (CECT) scan of the chest showed a large irregular, heterogeneously enhancing lobulated, hypodense lesion involving the pericardium posteriorly with loss of fat planes between the lesion and wall of the left atrium and both the ventricles. The lesion was causing pressure effect over the left pulmonary artery extending close to the left hila suggesting a pericardial infiltrative pathology (Fig. 1).

CECT scan of the abdomen with section of the upper thigh showed a small, well defined soft tissue lesion in the right side of the pelvis, lateral to the greater trochanter involving the gluteus medius muscle. Similar hypodense lesion was also noted in the upper third of the left thigh medially involving the abductor muscle. A bone scan showed no evidence of metastasis. Echocardiogram was suggestive of multiple cardiac masses attached to the inter ventricular septum (IVS), interatrial septum, pulmonary artery wall and pericardium, a hypo kinetic IVS with normal LV systolic function (Fig. 2).

Histopathological examination from the limb lesions revealed uniform and relatively small spindle cells arranged in fascicle and sheets with ovoid pale staining nuclei and inconspicuous nucleoli. Cytoplasm was sparse with indistinct cell bodies. There were 2–3 mitoses per 10 high power field (monophasic synovial sarcoma). Immunohistochemistry showed strong positivity for BCL-2 and S-100, and focal positivity for cytokeratin (Figs. 3 and 4).

Which lesion, was the primary amongst the two was impossible to determine. The possibility that both were secondary lesions with an unrecognized primary source cannot be ruled out.

In view of the metastatic presentation, ifosfamide-based chemotherapy was started. Ifosfamide was given at a dose of 2 g/m^2 day 1–5 and doxorubicin was administered at 75 mg/m^2 day 1 q 3 weekly. After 3 cycles of chemotherapy the patient had partial response and the remaining 3 cycles of chemotherapy were completed. End of therapy evaluation showed stable disease. After a DFS of 2 months he presented with exertional dyspnea, extreme fatiguability and bilateral pedal edema. 2 D- Echo showed an increase in size of the swelling, thrombus in left ventricle and severe left ventricular dysfunction. He was treated symptomatically but succumbed due to cardiac dysfunction despite best possible efforts.

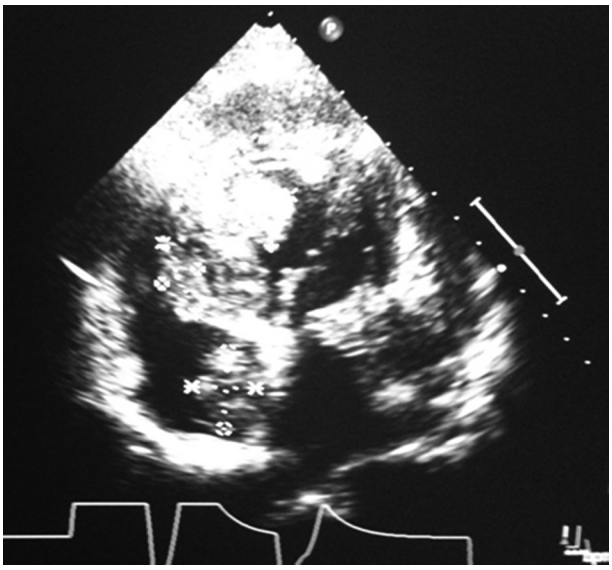

Fig. 2 – 2D Echocardiogram showing multiple cardiac masses attached to the IVS.

3. Discussion

The term “synovial” sarcoma is a misnomer as it rarely occur in joint capsules. The tissue of origin is unknown. It occurs primarily in the extremities arising from bursae and tendon

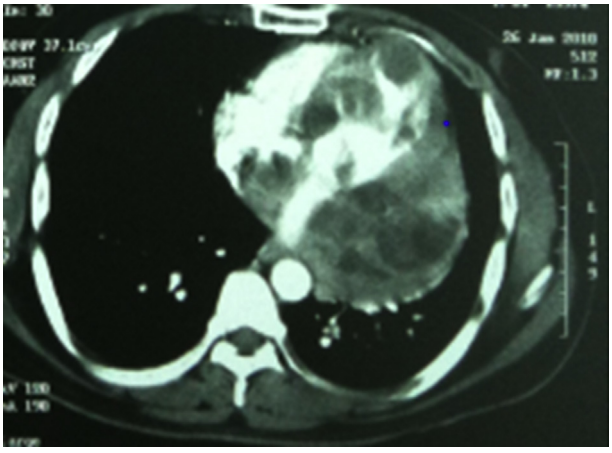

Fig. 1 – CECT Chest showing a large hypodense lesion involving the pericardium.

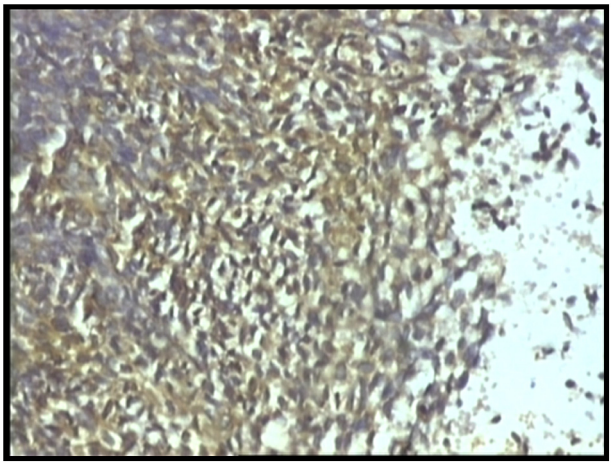

Fig. 3 – BCL2 positive.