

CASE REPORT

Mediastinal monophasic synovial sarcoma with pericardial extension causing hemodynamic instability

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Abstract

A 46-year-old man presented with mass on chest x-ray along with a 6-month history of weight loss, dyspnea and cough. He was hypotensive and an echocardiogram showed large extra-cardiac mass compressing the right ventricular outflow tract resulting in features of cardiac tamponade. Chest computed tomography revealed a mediastinal mass invading the pericardium adjacent to right ventricular outflow tract. Biopsy of the mass confirmed primary monophasic synovial sarcoma. Chemotherapy and radiotherapy along with anti-inflammatories were given as surgery was too high risk due to the location of the tumour and pericardial involvement. Patient responded briefly to the treatment with improvement in hemodynamic parameters but over next weeks he became less responsive to treatment with increasing size. He died 2 months after treatment commenced.

INTRODUCTION

Synovial sarcomas are very rare soft tissue tumours derived from primitive pluripotent mesenchyme capable of epithelial differentiation presenting in adolescents and young adults. Primary origin in thorax is extremely rare, accounting for <1% of all malignancies in thorax and more prevalent in adults between 20 and 40 years old. Synovial sarcoma usually occurs in deep soft tissue of the extremities with metastasis to the thorax and abdomen. We report a rare case of mediastinal monophasic synovial sarcoma with pericardial involvement causing compression of right ventricle and right ventricular outflow tract (RVOT) leading to hemodynamic compromise.

CASE REPORT

A 46-year-old man was referred for a chest X-ray following 6-month history of worsening dyspnoea, dry cough, night sweats, intermittent pyrexia and weight loss of 6 kg. The chest X-ray showed a large mass extending from the mediastinum to the left lateral chest. Laboratory blood tests showed a normocytic anaemia picture with a haemoglobin of 12.4 g/dL (13–18), raised ALP at 205 U/L (40–130) and GGT 350 U/L (0–55). Vitals signs recorded heart rate at 104 per minute, blood pressure of 106/70 mmHg, saturation of 94% on room air, and respiratory rate of 24 per minute. Electrocardiogram showed sinus tachycardia with no evidence of electrical alternans and pulsus paradoxus was 24 mmHg.

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The patient subsequently had a trans-thoracic echocardiogram (TTE) which showed localized pericardial effusion in the antero-apical region with maximum dimension of 2.5 cm. A mass was seen within the pericardial space near the RVOT compressing it. Flow turbulence was noted on colour Doppler along with acceleration of 1.5 m/s in the RVOT and in pulmonary trunk.

Urgent computed tomography (CT) scan of the chest showed a solid lobular left hemithorax mass measuring 17.6 × 11.0 × 18 cm causing significant displacement of mediastinal structures to the right. There was invasion into the pericardial cavity, with associated pericardial effusion (Fig. 1).

A biopsy of the mass showed tissue infiltrated with spindle cell tumour of monomorphic appearance consisting of densely packed cells with fine chromatin pattern, partially overlapping nuclei and scanty cytoplasm (Fig. 2). Immunoprofile revealed a strongly positive CD56 and focally positive epithelial membrane antigen (EMA). Fluorescence in situ hybridization studies showed rearrangement of SS18 locus (SYT gene) at 18q:11.2 chromosome within spindle cells pericytomatous blood vessels are present. The above features were morphologically and immunoprofile consistent with monophasic sarcoma of Trojani Grade 2 (Fig. 2). He was reviewed by Oncology specialists who started him on Dexamethasone initially to reduce reactive inflammation to improve his hemodynamics and symptoms. Pulsus paradoxus improved within 48 h allowing for more treatment to commence. Following a case discussion at the multidisciplinary team meeting, he received chemotherapy initially with Liposomal Doxorubicin for cycle one with addition of Iphosphamide for cycle 2. He was to be considered for

surgical resection of the tumour following the second cycle with the expectation of tumour shrink. Unfortunately, the patient died following 2 months of chemotherapy due to severe aggression of the tumour and failure of response to treatment.

DISCUSSION

We describe a rare case of primary monophasic synovial sarcoma in the thorax invading pericardium and RV resulting in extrinsic cardiac compression as evident on echocardiogram and CT of the chest. This case presented with hemodynamic instability similar to cardiac tamponade due to effusion. Pericardiocentesis of the localized moderate effusion was not possible as the approach was obscured by the sarcoma. The reduction in swelling improved his blood pressure and pulsus paradoxus without any invasive treatment.

Synovial sarcomas are very rare soft tissue tumours derived from primitive pluripotent mesenchyme capable of epithelial differentiation presenting in adolescents and young adults. Primary origin in thorax is extremely rare, accounting for <1% of all malignancies in the thorax and more prevalent in adults between 20 and 40 years old [1]. Synovial sarcoma usually occurs in deep soft tissue of the extremities with metastasis to the thorax and abdomen. There are four morphological types of synovial sarcoma—biphasic, monophasic, monophasic epithelial and poorly differentiated tumours [2]. The biomarkers for these tumours include Vimentin, Cytokeratin and EMA positivity with EMA being the most common positive marker. Monophasic synovial sarcoma is a locally invasive tumour and the commonest of all the types constituting spindle cells on histology although epithelial cells might be seen. Genetic identification of t(X:18) translocation involving SYT gene on chromosome 18 and the SSX1 or SSX2 gene on the X chromosome, specific for synovial sarcoma is found in over 90% of patients [1, 3–5].

Treatment involves chemotherapy and or radiotherapy and resection of large tumours. In some cases, complete surgical resection is not possible when the tumours involve pericardium or major vessels.

ACKNOWLEDGEMENTS

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CONFLICT OF INTEREST STATEMENT

There are no conflicts of interest to declare.

FUNDING

No funding was required.

ETHICAL APPROVAL

No ethical approval was needed

CONSENT

Obtained consent from subject when alive. However, the case is sufficiently anonymised to prevent identification.

GUARANTOR

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Richard W. Varcoe.

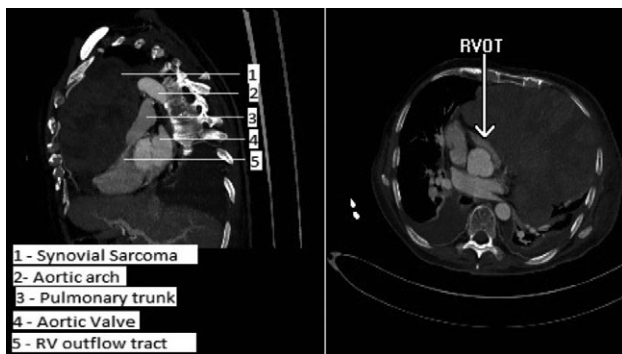


Figure 1: Computerized tomography of the chest: sagittal and transverse views showing marked extrinsic constriction of the RVOT

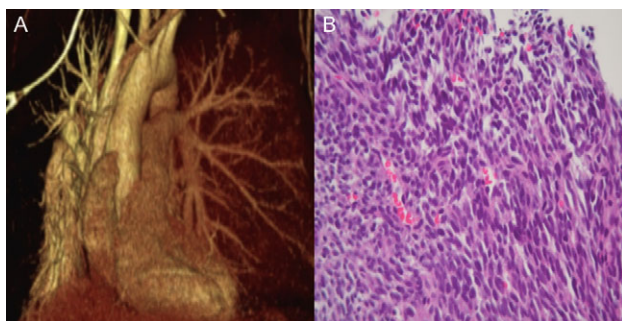


Figure 2: (A) 3D CT chest reconstruction showing marked hypervascularity of the synovial sarcoma and the resulting compression of RV and RVOT. (B) Histology of mass shows densely packed cells of monomorphic appearance with scanty cytoplasm.

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