CASE REPORT

Young Heart, Rare Tumour: A Case Report On Pericardial Synovial Sarcoma

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ABSTRACT

Synovial sarcoma is a rare mesenchymal tumour that typically affects adolescents and adults under 30. It is defined by a translocation between chromosomes X and 18, resulting in SS18:SSX fusion proteins. While it can originate from various soft tissues, the lower limb is most commonly affected. Pericardial synovial sarcoma is an exceptionally rare primary malignant cardiac tumour with an uncertain prognosis, and only a few cases have been reported in India. We present the case of a 15-year-old boy with chest pain and shortness of breath, managed with systemic chemotherapy. This report outlines his diagnosis and management.

Keywords: Pericardial synovial sarcoma, echocardiography, Doxorubicin

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INTRODUCTION

Pericardial synovial sarcoma (PSS) is a rare primary malignant cardiac tumour and a subtype of synovial sarcoma (SS), which accounts for 5-10% of all soft tissue sarcomas and is the fourth most common sarcoma. SS primarily affects individuals aged 20 to 40 years[1] and typically arises near joints in the extremities, though it can rarely occur in other locations such as the thoracic cavity[2]. Patients often present with a deep-seated mass of several years' duration. Most synovial sarcomas exhibit a characteristic chromosomal translocation t(x;18)(p11;q11.2), resulting in SS18-SSX1, SSX2, or SSX4 fusion genes that encode chimeric transcription factors[3]. Histologically, PSS may be monophasic, with uniform spindle cells, or biphasic, containing both spindle and epithelial components. Diagnosis is challenging due to non-specific symptoms, and the rarity of these tumours makes it difficult to establish an optimal chemotherapy regimen[4]. We present such a case here.

CASE PRESENTATION

A 15-year-old male presented in the Department of Paediatrics with sudden onset swelling of bilateral feet and face, accompanied by breathlessness for 2 months, dizziness for 15 days and sweating for 1 week. The symptoms were associated with abdominal pain and distention. His initial baseline investigations were unremarkable. However, echocardiography revealed pericardial effusion with features suggestive of cardiac tamponade. Approximately 250-300ml of haemorrhagic fluid was drained by pericardiocentesis. He was subsequently scheduled for elective pericardiectomy by the Cardiothoracic surgical team. The pericardial fluid analysis was performed, which showed: Protein- 1.6g/dl, Sugar-83mg/dl, Adenosine deaminase (ADA) -50u/L, Lactate dehydrogenase (LDH) - 3856u/L. Pleural fluid analysis shows predominantly neutrophilic exudative effusion (95%), for which chest tube insertion was done.

A computed tomography (CT) of the thorax was also performed to evaluate the mass, which showed thickened

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enhancing pericardium, pleural effusion and left lower lobe consolidation (Image 1). Patient underwent pericardial widening and evacuation of fibromucinous mass, which was sent for biopsy and Immunohistochemistry (IHC) thereafter. Histopathology of the mass revealed pleomorphic spindle cells arranged in fascicles with prominent nucleoli, brisk mitosis, intersecting fascicles, myxoid degeneration & extensive necrosis with areas of haemorrhage in between. (Image 2). IHC was reactive for CK, TLE-1, SSX-SS 18, EMA and was non- reactive for SMA, DESMIN, CD34, S100, with Ki 67 of 40-45% (Image 3). Based on these findings, the patient was diagnosed with a case of Pericardial Synovial Sarcoma.

Subsequently, the patient was shifted to the Department of Medical Oncology, where he received 2 cycles of Doxorubicin, Ifosfamide, and Mesna. He is planned for the 3rd cycle, after which systemic imaging will be repeated to evaluate disease status.

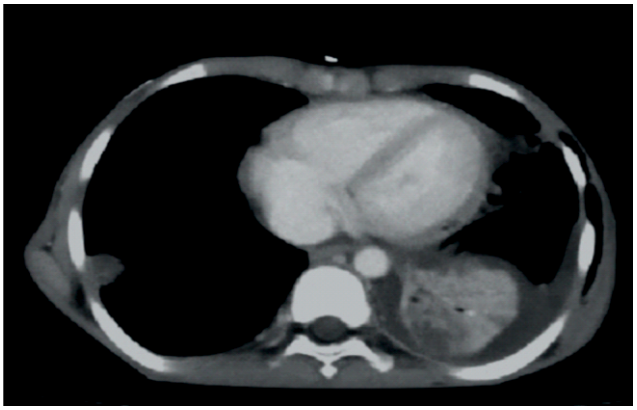

Image 1: Axial section of CECT thorax at the level of heart show enhancing thickening of the pericardium , left lower lobe consolidation and left sided pleural effusion

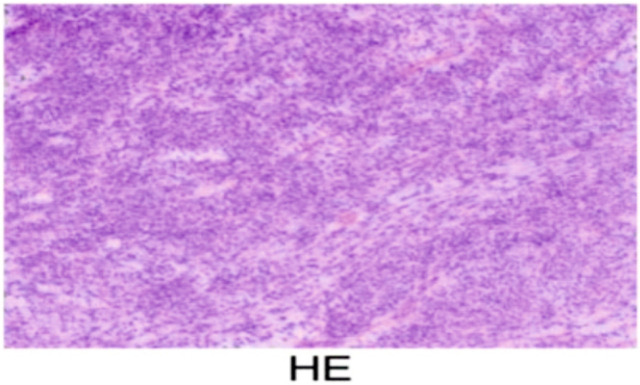

Image 2: Histopathology of the mass showed pleomorphic spindle cells arranged in fascicles with prominent nucleoli, brisk mitosis, intersecting fascicles, myxoid degeneration & extensive necrosis with areas of haemorrhage in between.

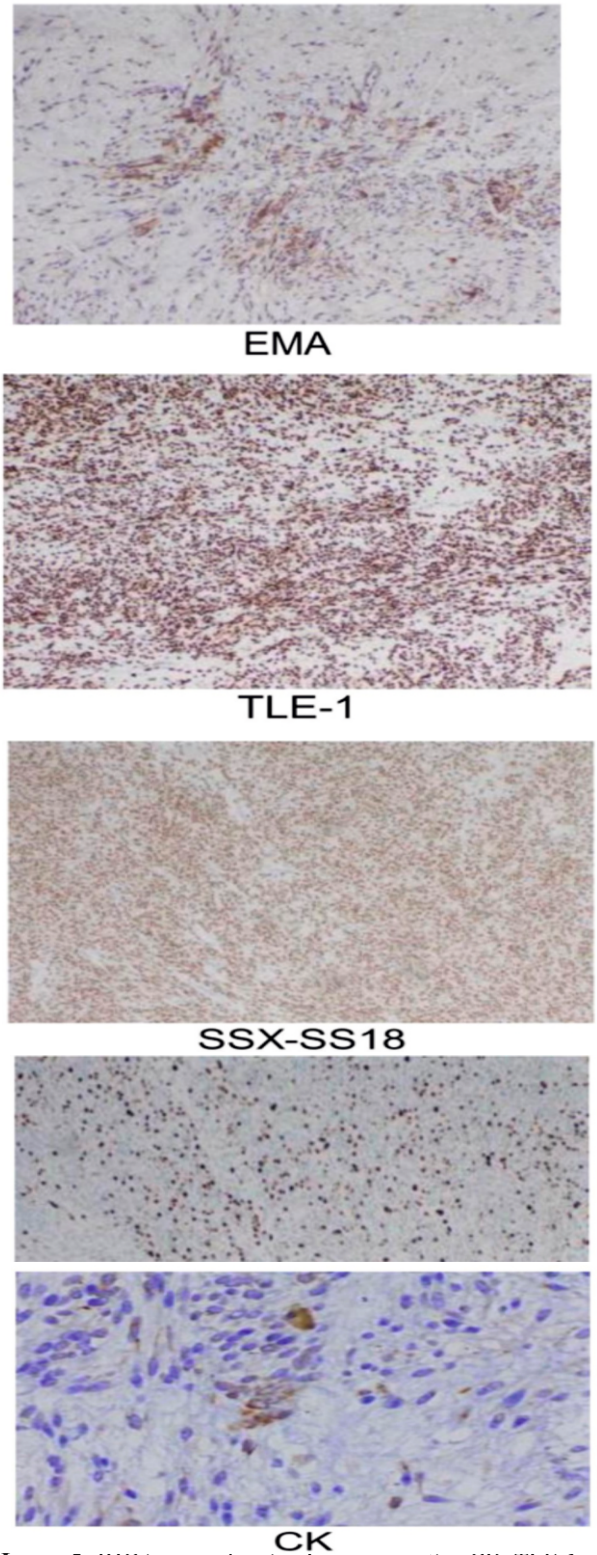

Image 3: IHC images showing Immunoreactive CK, TLE-1, SSX-SS 18, EMA, Ki 67= 40-45%

DISCUSSION

Cardiac tumours are rare, and most of them are accidentally discovered. The incidence ranges from 0.001% to 0.03%[5] [BP2] The overwhelming majority of these are benign, and only a few are malignant. Malignant cardiac tumours, like PSS, are exceedingly rare, with only a few cases reported in the literature so far; thus, limited data prevent further understanding of this condition[6]. This type of tumour is known to have a poor prognosis, with no clarity about the best approach in managing this disease. On average, its 10-year overall survival rate is about 0%–20%[1]. The exact origin of SS is unknown, but a characteristic chromosomal abnormality t(X;18)(p11.2;q11.2) is seen[6]. PSS, although rare in adults, is known for its aggressive nature and challenging management, often requiring a multidisciplinary approach. Even if multimodality treatment based on surgery, chemotherapy and radiotherapy is performed, the prognosis of PSS remains dismal, with an average survival of 27 months[6]. The most common primary tumour seen in the heart is benign tumours such as myxomas that mostly originate from the right atrium[7]. A few soft-tissue sarcomas, such as liposarcoma, rhabdomyosarcoma and angiosarcoma, have also been reported, but SS of the heart, namely the pericardium, is extremely rare.

PSS generally appears as a solid heterogeneous mass with multilocular areas and internal septations[8]. Similarly, in our case, a solid mass showed contrast enhancement on the CT scan. Magnetic Resonance Imaging (MRI), however, was not done due to a lack of resources, although it is the best modality for characterising cardiac tumours[8]. Various other mediastinal and pericardial tumours like thymoma and mesothelioma can also resemble PSS; thus, cytogenetic analysis is necessary for confirmation of diagnosis.

The clinical outcome of PSS is uncertain, but most of these cases are aggressive in nature. The clinical symptoms of the disease are non-specific, with dyspnea and chest pain being the most common presenting symptoms[3]. The patient described here also presented with shortness of breath and chest pain. Gross pericardial effusion was also noted.

Management of SS is challenging and requires a multi-disciplinary approach. Surgical resection is the preferred treatment, but it is often not feasible due to the tumour's proximity to vital structures[9]. In such cases, alternative options like chemotherapy and External Beam Radiotherapy (EBRT) are used to downsize/downstage the tumour and potentially render it resectable.

Due to the rarity of these tumours, it is very difficult to establish an optimal regimen of chemotherapy. In general, these tumours are chemosensitive, particularly to Ifosfamide, with combination regimens like Doxorubicin and Ifosfamide showing higher response rates but with significant toxicity[4, 10]. A case series of 13 patients with SS treated with high-dose Ifosfamide showed a response in all patients, with 4 clinical remissions achieved[10]. Combining doxorubicin with ifosfamide may achieve a higher response rate (58%), but patient tolerability and side effects need to be considered. Similarly, in our patient, there was a marked response seen to Ifosfamide-based chemotherapy.

CONCLUSION

PSS is an extremely rare and aggressive malignancy of the heart. Its presentation is quite vague and non-specific, making it hard to diagnose. Though recognition through advanced imaging and histopathology is available, increased awareness and reporting of such cases are important to improve understanding and therapeutic strategies of the disease.

Conflict of Interest: None

Written Informed Consent: Taken

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