

Primary Pericardial Synovial Sarcoma: An Extremely Rare Cardiac Neoplasm

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Abstract

Primary pericardial tumors are an entity that is infrequently encountered and may be a cause of pericardial effusion. Primary synovial sarcomas of the pericardium are even rarer malignant invasive tumors that are a challenge to recognize due to their vague presentation and difficulty in diagnosing non-invasively. Here, we report a case of a 48-year-old gentleman of South Asian descent, who was incidentally found to have pericardial and bilateral pleural effusions and subsequently diagnosed to have primary pericardial synovial sarcoma.

Categories: Cardiac/Thoracic/Vascular Surgery, Cardiology, Oncology

Keywords: pericardial effusion, pericardial synovial sarcoma, cardiac tumor

Introduction

Primary cardiac tumors are an extremely rare entity with a prevalence rate of 0.02% to 0.056% [1-2]. Primary tumors of the pericardium are even rarer, and their prevalence ranges from 0.001% to 0.007% [3]. Primary pericardial tumors may be benign or malignant. Benign tumors include pericardial cysts, lipomas, lipoblastomas, paragangliomas, germ-cell tumors, hemangiomas, fibromas, and inflammatory pseudo-tumors. Pericardial cysts are the most common benign tumors followed by lipomas. Primary malignant pericardial neoplasms include mesotheliomas, a wide variety of sarcomas, lymphomas, and primitive neuroectodermal tumors amongst which mesotheliomas are the most common [4]. In this article, we present a rare case of primary pericardial synovial sarcoma with a review of the literature.

Case Presentation

A 48-year-old gentleman of South Asian descent who had no prior medical illness was referred to our hospital from a peripheral clinic for the evaluation of incidental sonographic finding of pericardial and bilateral pleural effusions while undergoing evaluation for renal colic. On admission, he was primarily complaining of right flank pain associated with dysuria of two days duration. On a detailed review of history, he admitted having shortness of breath associated with a mild nonproductive cough of 10 days duration. Also, he was complaining of anorexia and paroxysmal nocturnal dyspnea (PND). On examination his vital signs on admission were: pulse rate 103/minute, blood pressure 122/80 mmHg, respiratory rate 18/minute, temperature 37°C, and oxygen saturation 99% breathing on room air. Jugular venous pulse was not raised. Systemic examination revealed a comfortably lying patient without any distress. Respiratory system examination showed dullness on percussion with decreased intensity of breath sounds over the left infra-scapular area. Other system examinations, including the cardiovascular system, were unremarkable.

His initial laboratory investigations were unremarkable except for a mildly elevated C-reactive protein (CRP) of 30 mg/L (Table 1).

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| Test | Result | Normal Range |
|---------------------------------|---------------------------|--------------|
| WBC | 10.6 x10 ³ /uL | 4.0-10.0 |
| RBC | 5.7 x10 ⁶ /uL | 4.5-5.5 |
| Hemoglobin | 13.1 gm/dL | 13.0-17.0 |
| Hematocrit | 40.2 % | 40.0-50.0 |
| Platelet | 302 x10 ³ /uL | 150-400 |
| Absolute Neutrophil count (ANC) | 9.4 x10 ³ /uL | 2.0-7.0 |
| Lymphocytes count | 0.66 x10 ³ /uL | 1.00-3.00 |
| Urea | 7.6 mmol/L | 3.2-7.4 |
| Creatinine | 86 umol/L | 64-110 |
| Total Protein | 64 gm/L | 64-83 |
| Albumin | 40 gm/L | 35-50 |
| Alkaline Phosphatase | 64 U/L | 40-150 |
| ALT | 42 U/L | 0-55 |
| AST | 34 U/L | 5-34 |
| LDH | 259 U/L | 125-220 |
| High Sensitivity Troponin-T | 6.05 ng/L | 0.00-14.00 |
| Glucose | 5.0 mmol/L | 3.3-5.5 |
| C-Reactive Protein | 30 mg/L | 0-5 |
| Procalcitonin | 0.14 ng/mL | |

TABLE 1: Showing baseline laboratory results

WBC-White blood cell count, RBC-Red blood cell count, ALT-Alanine aminotransferase, AST-Aspartate aminotransferase, LDH-Lactate dehydrogenase

Electrocardiogram (ECG) on admission showed sinus tachycardia with electrical alternans (Figure 1).

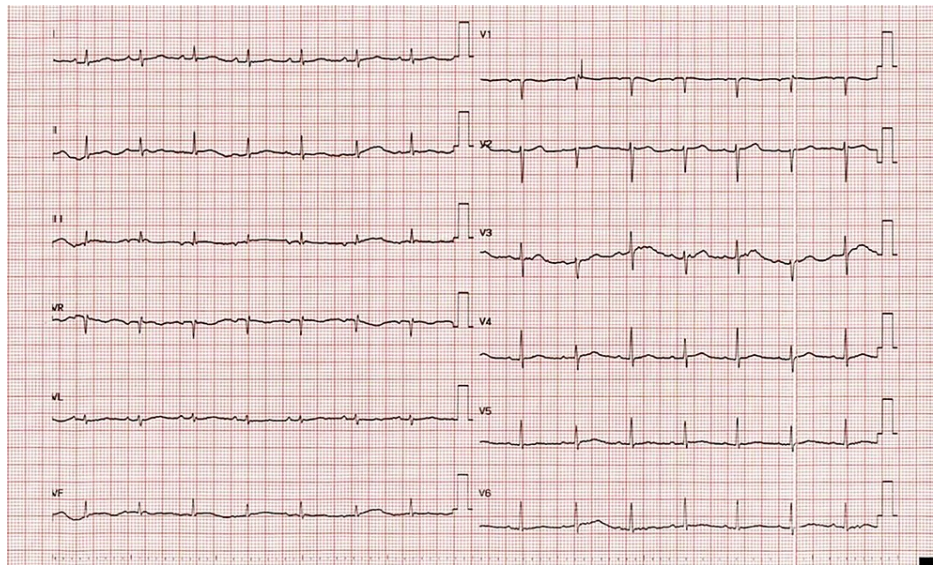


FIGURE 1: Electrocardiogram showing electrical alternans

Chest X-ray demonstrated an enlarged cardiac silhouette with blunting of the left costophrenic angle. Transthoracic echocardiography revealed a large pericardial effusion suggestive of cardiac tamponade (Figure 2).

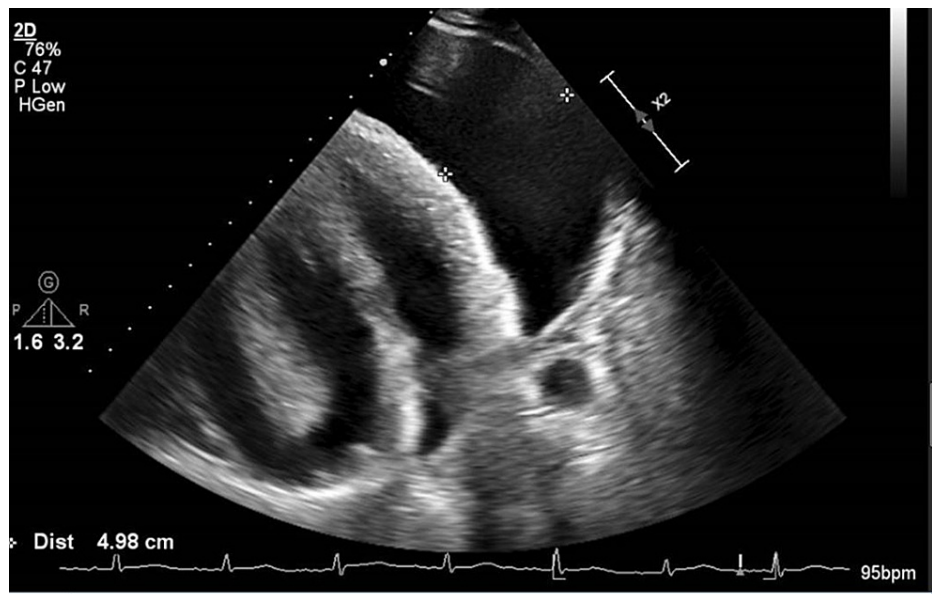


FIGURE 2: Transthoracic echocardiogram showing large pericardial effusion

Abdominal sonography showed a right renal calculus measuring 7.6 mm without hydronephrosis. The patient underwent emergent pericardiocentesis with drainage of 950 ml of sanguineous fluid and a left-sided pericardial drain was subsequently inserted. His shortness of breath improved following pericardiocentesis and flank pain resolved with conservative management. Pericardial fluid analysis revealed an exudative effusion with no bacteria or neoplastic cells (Table 2).

| Pericardial fluid analysis | Result |
|----------------------------|---|
| Color | Red |
| Appearance | Bloody |
| WBC | 6,125 /uL |
| RBC | 646,125 /uL |
| Lymphocytes | 57.0 % |
| Monocytes | 29.0 % |
| Macrophages | 8.0 % |
| Mesothelial cells | 2.0 % |
| Glucose | 6.9 mmol/L |
| LDH | 273.0 U/L |
| pH | 7.471 |
| Protein | 29.0 gm/L |
| Albumin | 18.0 gm/L |
| Cytology | Lymphohistiocytic effusion. No malignant cells seen |
| Fluid Culture | No growth |

TABLE 2: Results of pericardial fluid analysis

WBC-White blood cell count, RBC-Red blood cell, LDH- Lactate dehydrogenase

Initially, a possibility of tuberculous etiology was considered; however, workup was negative for the presence of acid-fast bacilli (AFB) in the smear, tuberculosis polymerase chain reaction (TB-PCR), and culture. Quantiferon-TB Gold (QFT) was also negative. Screening for common autoimmune causes of serositis did not yield any positive results.

His COVID-19 PCR was also negative. Meanwhile, the patient was given symptomatic management with ibuprofen 600 mg every six hours and colchicine 0.5 mg daily.

With the initial workup being unable to delineate the cause of serositis, a computed tomography (CT) scan of the chest, abdomen, and pelvis was performed, which showed a well-defined heterogeneous structure measuring 13.5 x 5 cm on the lower aspect of the pericardium extending into the left side of the thorax, abutting the cardiac border and the anterior part of the aorta without any infiltration (Figures 3-5).

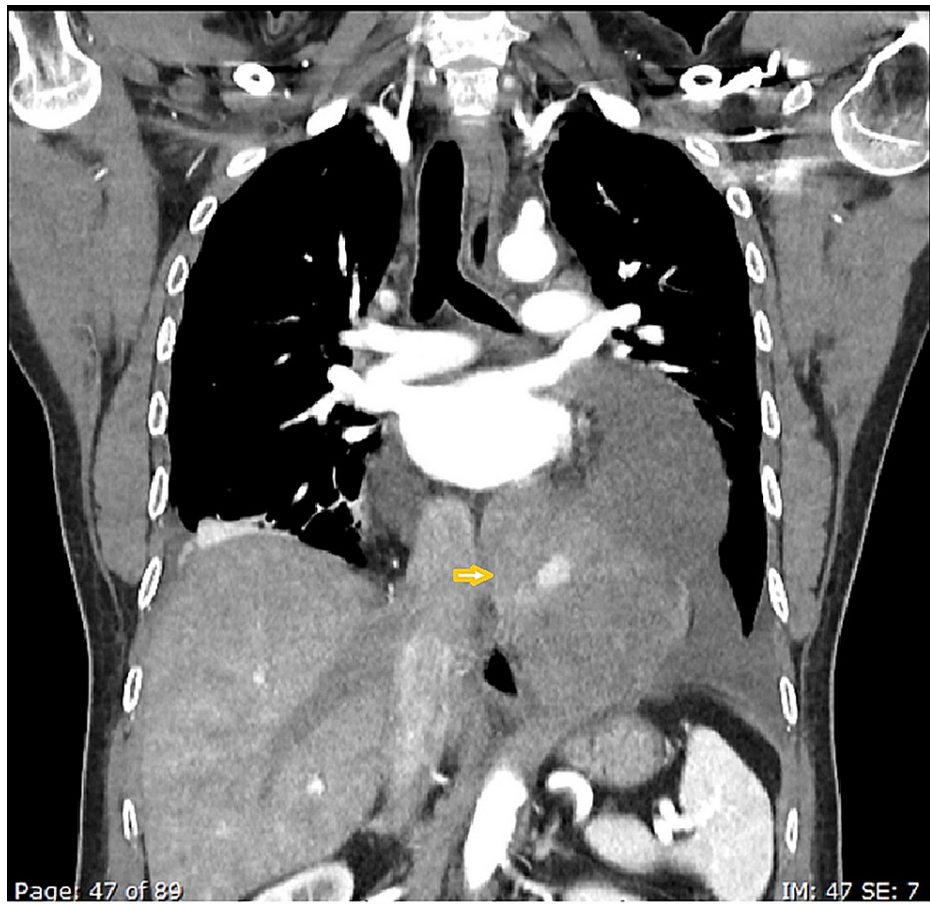


FIGURE 3: Coronal contrast CT (arterial phase) images showing a large, heterogeneously enhancing pericardial mass (arrowhead)

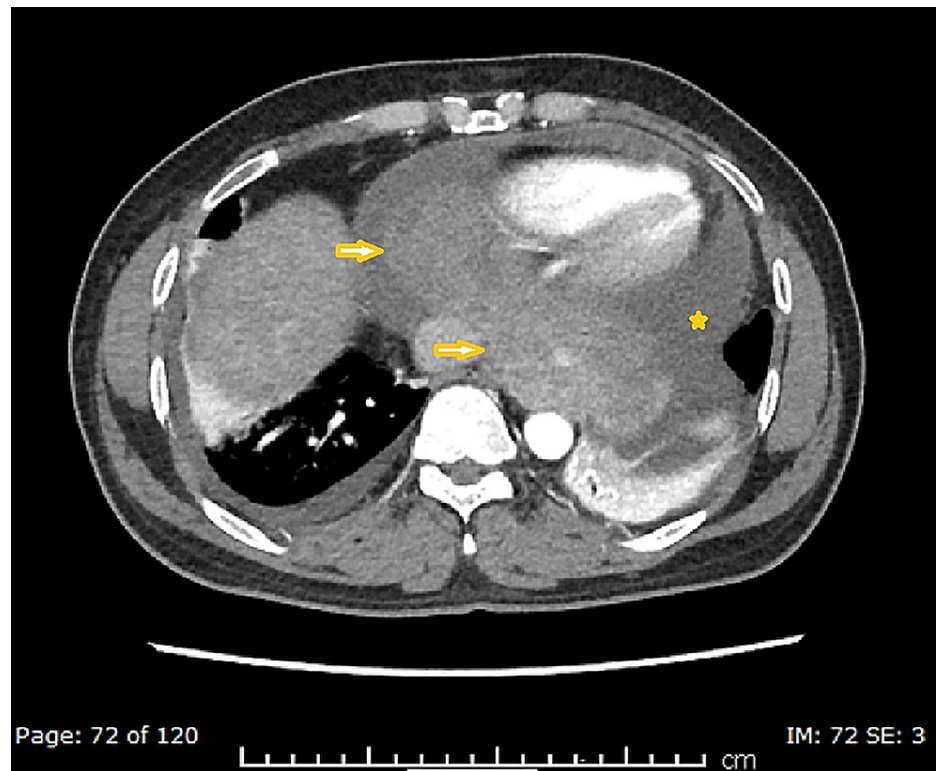


FIGURE 4: Axial contrast CT (arterial phase) images showing a large, heterogeneously enhancing pericardial mass (arrowheads) with pericardial effusion (star)



FIGURE 5: Sagittal contrast CT (arterial phase) images showing a large, heterogeneously enhancing pericardial mass (arrowhead) with pericardial effusion (star)

The radiological findings and the sanguineous nature of the pericardial fluid indicated an organized hematoma versus a neoplastic etiology. The CT also confirmed bilateral pleural effusion, more on the left side. No free fluid was seen in the abdomen. Subsequent diagnostic pleurocentesis from the left pleural effusion yielded 80 ml of sanguineous fluid, which was exudative as well. It was predominantly lymphohistiocytic, without any growth of bacteria, AFB, or neoplastic cells.

The patient underwent further characterization of the suspected hematoma/neoplasm with a cardiac magnetic resonance imaging (MRI), which showed three well-defined pericardial soft tissue masses exhibiting features of local invasiveness on the left inferior pleura and left hemidiaphragm without infiltration, highly suggestive of a neoplastic origin; however, atypical hemangioma was still a consideration (Figures 6-8).

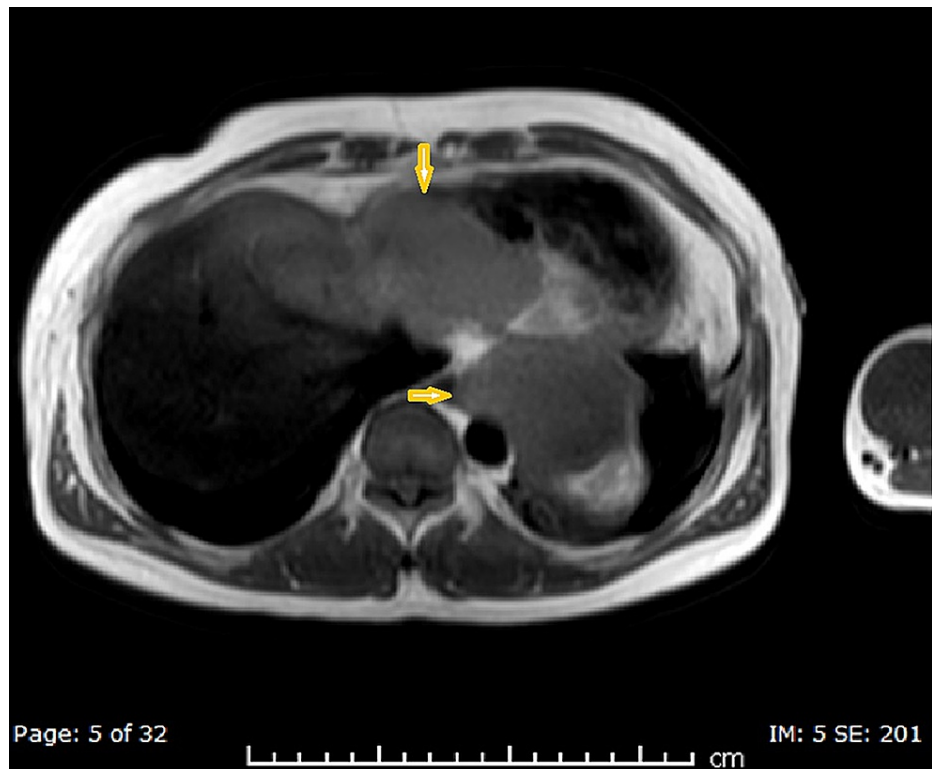


FIGURE 6: MRI T2-weighted axial image showing a pericardial mass lesion (arrowheads)

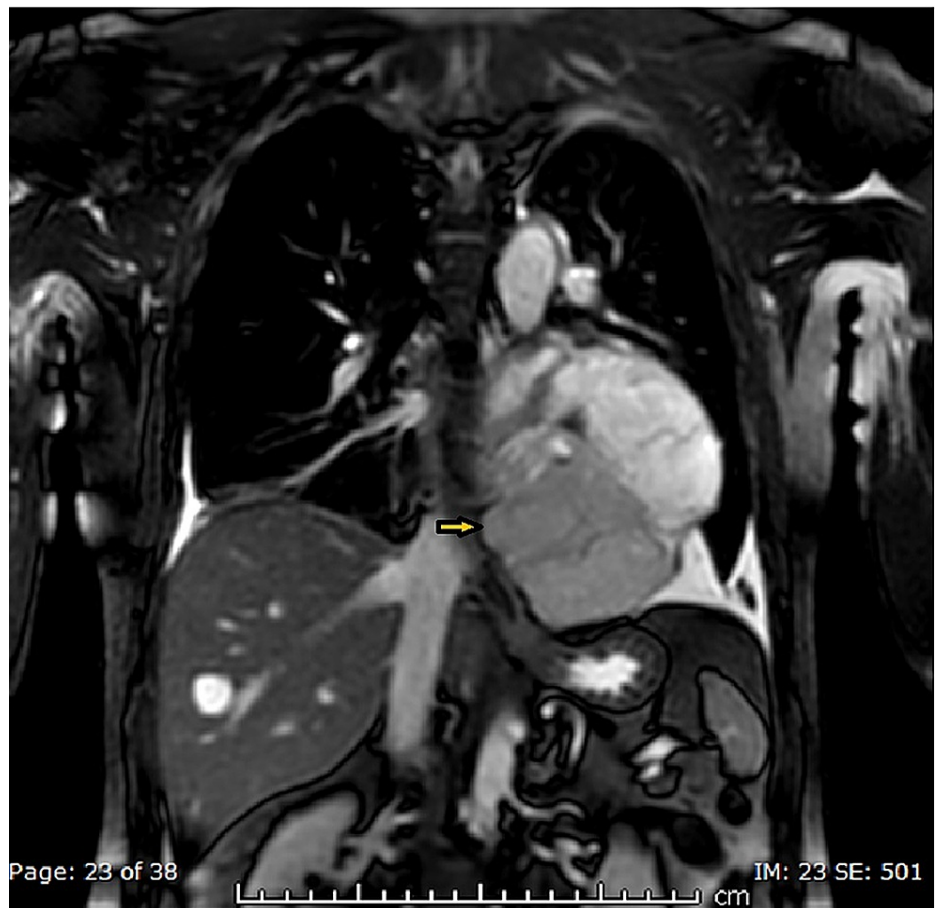


FIGURE 7: MRI-STIR sequence coronal image showing a pericardial mass lesion (arrowhead)

STIR: short-T1 inversion recovery

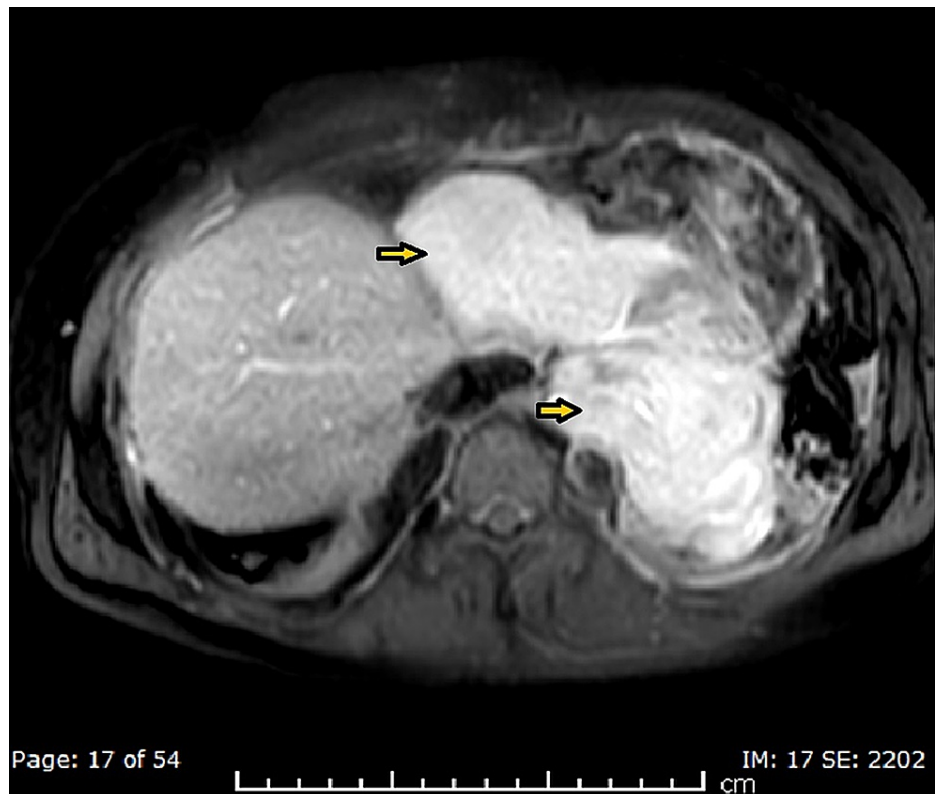


FIGURE 8: MRI post-contrast axial image showing a pericardial mass lesion (arrowheads)

Follow-up echocardiography after three days revealed the recurrence of a large circumferential pericardial effusion without features of tamponade. The patient remained clinically well without any new complaints; however, his hemoglobin was progressively declining from 13.1 gm/dL to 7.5 gm /dL, which was managed with packed red blood cells (PRBC) transfusion. A positron emission tomography (PET) scan confirmed increased fluorodeoxyglucose (FDG) uptake in the pericardial and pleural masses without any uptake elsewhere, indicating a primary pericardial-pleural pathology (Figures 9-11).

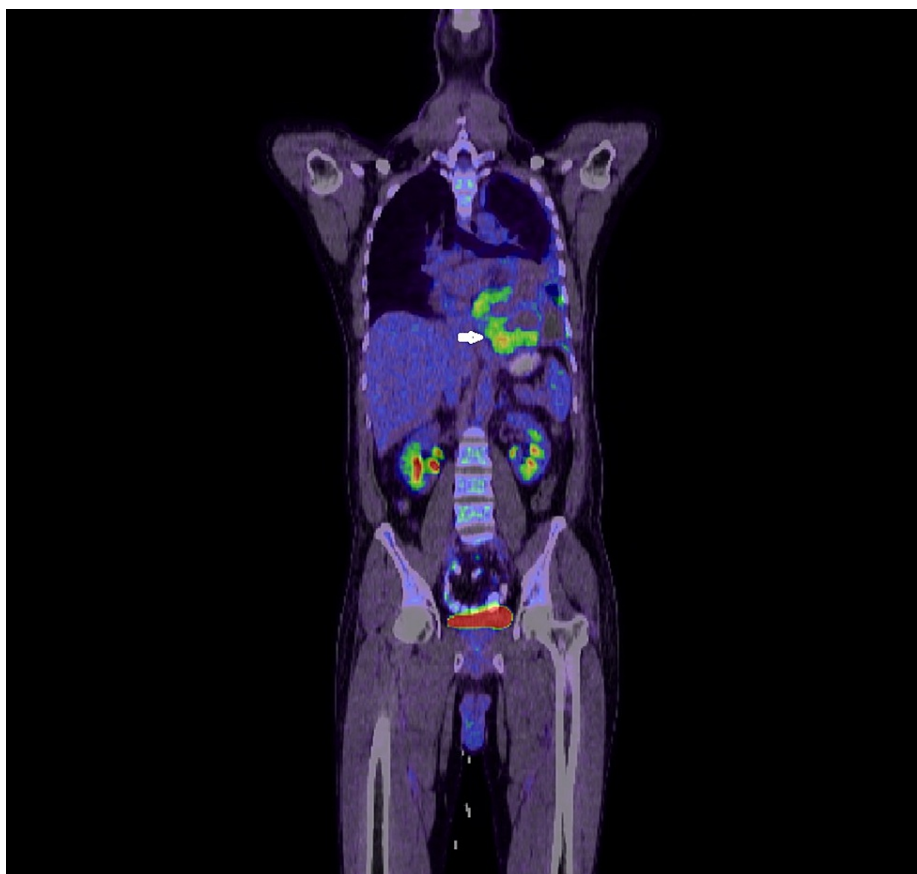


FIGURE 9: PET scan coronal section image showing increased uptake by a pericardial mass lesion (arrowhead)

PET: positron emission tomography

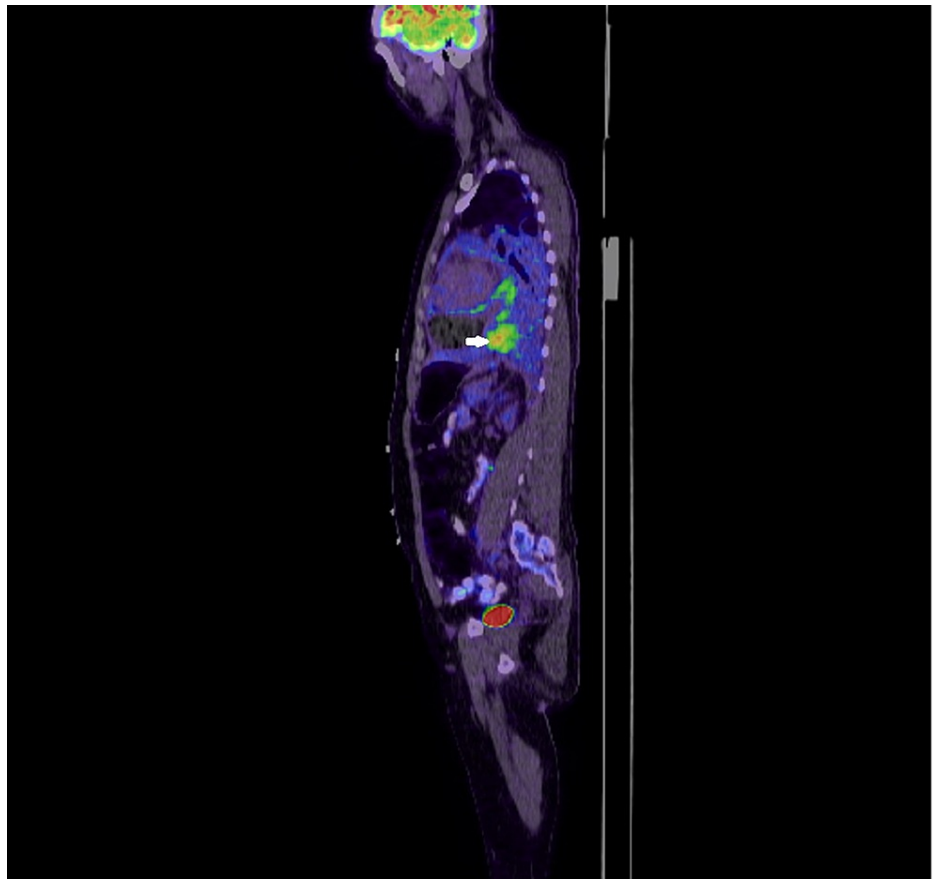


FIGURE 10: PET scan sagittal section image showing increased uptake by a pericardial mass lesion (arrowhead)

PET: positron emission tomography

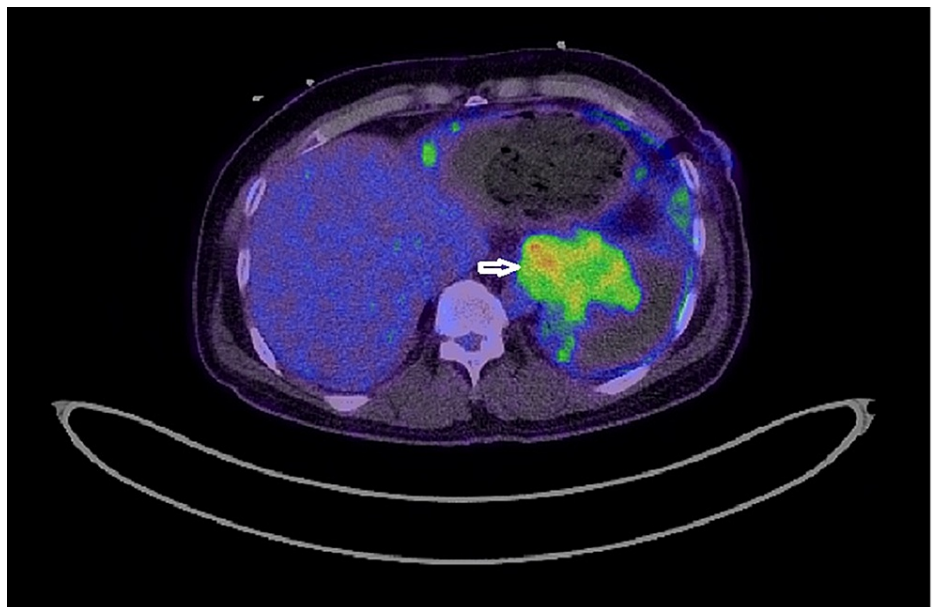


FIGURE 11: PET scan axial section image showing increased uptake by a pericardial mass lesion (arrowhead)

PET: positron emission tomography

The patient subsequently underwent a left video-assisted thoracoscopic (VATS) biopsy of the pericardial mass. Histopathological examination of the tissue samples from the pericardial and pleural segments of the mass showed fascicles of spindle cells with moderate cellular atypia, frequent mitoses, and necrosis most compatible with a diagnosis of synovial sarcoma. By immunohistochemistry, the cells were strongly positive for cytokeratin AE1/AE3, FLI-1, cytokeratin 8/18, TLE-1, BCL-2, CD99, cytokeratin 7, and D2-40 and showed patchy positivity with epithelial membrane antigen, WT-1, cytokeratin 5/6, Gata-3, and ERG. The proliferative index (immunostaining with Ki67) was estimated to be around 60%. Fluorescence in-situ hybridization (FISH) testing was positive for synovial sarcoma translocation on chromosome 18 (SS18) rearrangement and negative for Ewing sarcoma breakpoint region 1 (EWSR1) rearrangement, further confirming the diagnosis of a locally advanced primary synovial sarcoma of the pericardium.

Discussion

Primary pericardial sarcomas are very uncommon and aggressive tumors. They form approximately 13% of all primary cardiac sarcomas [5]. They have been classified into subtypes based on histopathological appearance, which includes angiosarcoma, liposarcoma, rhabdomyosarcoma, synovial sarcoma, and undifferentiated sarcoma [6]. Angiosarcoma is the most common type followed by undifferentiated sarcoma and leiomyosarcoma.

Like other primary pericardial neoplasms, the primary pericardial synovial sarcomas will also have varied clinical presentations. The symptoms include dyspnea, chest pain, cough, palpitation, and fever. The symptoms are mainly due to the presence of pericarditis, pericardial effusion, and cardiac tamponade.

Evaluation of pericardial synovial sarcomas includes chest radiography, which might reveal cardiomegaly or the presence of a widened mediastinum [7]. Echocardiographic findings include pericardial effusion or a thickened pericardium [8]. CT scan will detect the location of the tumor and will show the local invasion or distant metastasis. Magnetic resonance imaging is considered a better imaging tool than a CT scan, as it provides higher resolution and can detect invasion of the myocardium [7-9].

Very few cases of synovial sarcoma have been reported in the past. A literature review by Cheng et al. on 14 published cases of primary pericardial synovial sarcoma showed that the median age at the time of diagnosis was 32 years, with a preponderance in males [7], and the mean size of the tumor was 10.6 cm [10]. The most common symptom was dyspnea followed by chest pain and cough, with most of the symptoms attributed to cardiac tamponade. Our patient was in his late forties and his renal colic may be coincidental, the severity of which might have precluded/distracted the patient from complaining of chest symptoms initially.

Conclusions

Although primary pericardial synovial sarcoma is extremely rare, it should be considered as one of the differential diagnoses in the evaluation of patients presenting with pericardial effusion of an unknown etiology. CT scan can detect the location of the tumor as well as metastasis and local invasion; however, for detecting myocardial invasion, MRI is considered to be a better imaging tool.

Additional Information

Disclosures

Human subjects: Consent was obtained or waived by all participants in this study. Institutional Review Board, Medical Research Center, Hamad Medical Corporation issued approval MRC-04-21-255. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

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