Case Report

Primary pericardial synovial sarcoma presenting as cardiac tamponade

Jaideep Dey[1], Ratna Sud[2], Kumaresh Vaidya[3], Ranjan Kachru[4], Tapan Ghose[5]

From [1]Associate Director and Chief, Department of Non-invasive Cardiology, BLK Max Superspeciality Hospital, [2]Consultant, Department of Non-invasive Cardiology, Fortis Flt. Lt. Rajan Dhall Hospital, [3]Consultant, Department of Non-invasive Cardiology, Ayushman Hospital and Health Services, [4]Additional Director and [5]Senior Director and Head, Department of Cardiology, Fortis Flt. Lt. Rajan Dhall Hospital, New Delhi, India

ABSTRACT

Primary malignant cardiac neoplasms are a rare entity. Malignant pericardial tumors may present with pericardial effusion or non-specific symptoms. We report about a middle-aged male patient who presented with cardiac tamponade and was diagnosed with biphasic phenotype of primary pericardial synovial sarcoma, using histopathology and immunohistochemistry. Diagnosis of pericardial synovial sarcoma requires a multipronged approach involving clinical, imaging, histopathological and cytogenetic evaluation. Demonstration of a specific cytogenetic alteration (chromosomal translocation t(X;18)(p11.2;q11.2) SYT-SSX) remains the ideal confirmatory test. However, in resource constrained settings, a detailed histopathological examination combined with immunohistochemistry, specifically nuclear staining for Transducin like enhancer of slit-1 in tumor cells is useful for accurate diagnosis, due to its high sensitivity and specificity. Early diagnosis followed by prompt initiation of therapy may lead to improved outcomes despite the dismal prognosis associated with this neoplasm.

Keywords: Cardiac tamponade, Histopathology, Pericardial effusion, Synovial sarcoma

Primary pericardial malignancies are extremely rare, with an estimated prevalence of 0.001–0.007% [1]. A significant minority of newly diagnosed large pericardial effusions (PEf) may have a malignant etiology. The most common primary pericardial malignancies include mesothelioma, sarcoma, and lymphoma [1]. Synovial sarcoma (SS) is a soft-tissue sarcoma arising from pluripotent mesenchymal cells, having structural homology to developing synovial tissue [2]. Although most commonly seen in the joints of the lower extremities, SS can arise from multiple locations.

We report an unusual case of primary pericardial SS (PSS) presenting as cardiac tamponade (CT). Significant PEf are often ascribed to infectious and secondary malignant causes among patients in clinical practice. This case highlights the importance of keeping primary pericardial malignancy as a differential diagnosis to prevent diagnostic delay and ensure early initiation of appropriate therapy.

Access this article online	
Received - 27 October 2025 Initial Review - 14 November 2025 Accepted - 22 December 2025	Quick Response code
DOI: 10.32677/ijcr.v12i1.7920	

CASE REPORT

A 46-year-old male patient with no prior medical ailments presented with vomiting and uneasiness for the last 2 days. He gave a history of weight loss and anorexia since the preceding 2 months.

On examination, his respiratory rate was 40/min, with a heart rate of 130/min and blood pressure of 110/60 mmHg. Pulsus paradoxus was present. Jugular venous pressure was raised with absent "y" descent. There was no pallor, digital clubbing, cyanosis, or lymphadenopathy. Cardiovascular examination revealed muffled heart sounds, with no audible murmurs or rub. Other systemic examination was normal.

Electrocardiogram (ECG) showed sinus tachycardia, PR segment depression, and QRS electrical alternans (Fig. 1a). Chest roentgenogram (CR) demonstrated cardiomegaly with bilateral pleural effusion (PlEf) and a prominent bulge on the left cardiac border (Fig. 1b).

Transthoracic echocardiography revealed a large circumferential PEf, with an ill-defined echogenic mass abutting the left cardiac border (Fig. 2). A swinging motion of the heart in the pericardial cavity was noted along with significant respiratory variation across the

Correspondence to: Dr. Jaideep Dey, Associate Director and Chief, Department of Non-invasive Cardiology, BLK Max Super Speciality Hospital, New Delhi - 110005, India. E-mail: drdey1978@gmail.com

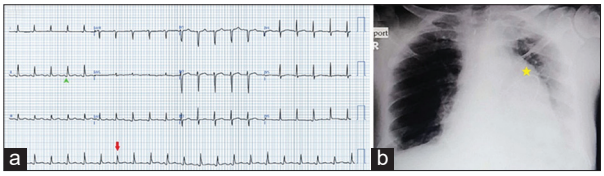

Figure 1: 12-lead electrocardiogram (a) showing sinus tachycardia, PR segment depression (green arrowhead), and electrical alternans (red arrow). Chest roentgenogram (b) showing cardiomegaly, bulging along the left border of the cardiac silhouette (yellow star), and bilateral pleural effusion. Pericardial drainage catheter also noted *in situ*

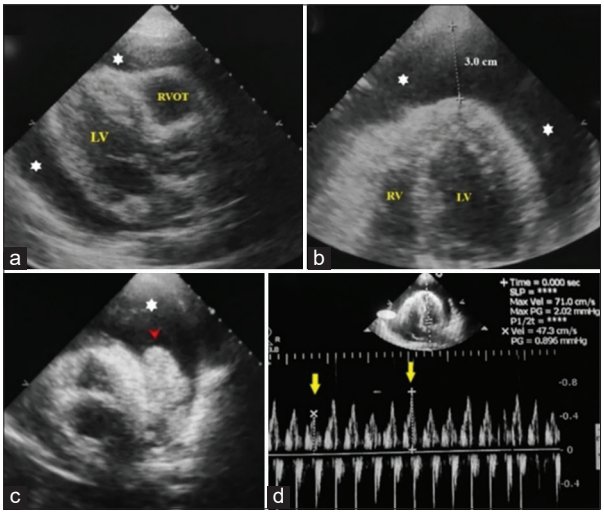

Figure 2: Transthoracic echocardiography in parasternal long axis and apical 4-chamber views (a-c), demonstrating large circumferential pericardial effusion (white stars). An echogenic mass lesion noted in pericardial cavity (c), lateral to the left ventricle (red arrowhead). Significant respiratory variation (33%) noted along mitral valve inflow (yellow arrows), suggestive of tamponade physiology (d). LV: Left ventricle, RV: Right ventricle, RVOT: Right ventricular outflow tract

mitral and tricuspid valve inflow. Right ventricular diastolic collapse was present. The inferior vena cava was dilated (2.3 cm) with blunted respiratory variation. The echocardiographic features were consistent with CT.

The patient subsequently underwent emergency pericardiocentesis. 600 mL of hemorrhagic pericardial fluid was drained immediately, followed by 80 mL over the next 24 h. Pericardial fluid examination revealed a hemorrhagic, exudative fluid having 3000 cells/cu mm (85% neutrophils, 15% lymphocytes), with no coagulum formation. Bacterial, fungal, and mycobacterial cultures were negative, and adenosine deaminase was not elevated. Cytological examination revealed scattered, degenerated, and reactive mesothelial cells in a background of red blood cells, polymorphonuclear leukocytes, and a few lymphocytes. No atypical/malignant cells were noted.

Hematological investigations revealed hemoglobin – 11.5 g/dL, with total leukocyte count – 14,890/cu mm (neutrophilic predominance). The liver and kidney function tests were normal. Ultrasound abdomen was significant only for features of cholelithiasis with chronic cholecystitis.

Chest computed tomography showed PEf, PlEf with a large, heterogeneously enhancing mass (8.1 × 7.5 × 4.4 cm) in the anterior mediastinum, insinuating the aortic and pulmonary root with infiltration into the pericardium (Fig. 3). Findings were consistent with neoplastic etiology (? thymic origin/lymphoma/pericardial lesion). There were no additional lesions in the chest, abdomen, or significant lymphadenopathy.

Patient underwent thoracotomy with debulking of the pericardial lesion. Intraoperatively, the anterior mediastinum was devoid of any tumorous growth. On pericardiotomy, 200 mL of hemorrhagic pericardial fluid was drained. A large, fleshy lobulated pericardial mass was noted along the left cardiac border (Fig. 4). This was resected with some remnant tissue left near the base, due to invasion of the aorta and main pulmonary artery. Frozen section analysis of the mass was provisionally diagnosed as a round cell tumor.

Histopathological examination revealed tumor cells in a biphasic distribution consistent with SS (Fig. 5). Morphologically, epithelial cells were columnar/cuboidal, arranged in glandular, cord-like, or rounded clump patterns. Cells had a moderate to large amount of cytoplasm, ovoid or round vesicular nuclei, and non-prominent nucleoli. Some glandular elements demonstrated intraluminal homogeneous eosinophilic secretions. Spindle-shaped cells were arranged in a fascicular pattern, admixed with epithelial cells. Connective tissue fibers separated adjacent layers of cells. Spindle cells had scant cytoplasm, oval hyperchromatic nuclei, and indistinct cell margins. Nuclei of adjacent cells appeared to have an overlapping pattern. There were numerous interspersed cleft-like hemangiopericytic spaces, lined by epithelial cells. Mitotic figures were rarely noted, while there were no areas of calcification or necrosis.

On immunohistochemistry, tumor cells stained positively for Transducin-like enhancer of slit-1 (TLE 1), showing a diffuse nuclear staining (3+) pattern, of both the spindle and epithelial elements (Fig. 6).

Post-procedure, the patient had an uneventful immediate post-operative course, and further course of management was being planned in consultation with the medical oncology and thoracic surgery teams.

DISCUSSION

CT is most common with pericarditis due to infection, hemorrhage, idiopathic, neoplastic, and connective tissue diseases [3]. Approx. 20% cases of undiagnosed, non-resolving, large symptomatic PEf may be the initial presentation of malignancy [4]. Primary cardiac neoplasms are rare, with an incidence of 0.001–0.03%; benign tumors being the most common (>80%) [5]. Most malignant primary tumors are sarcomas, with angiosarcoma most frequent [5]. Primary cardiac SS (PCSS) has been infrequently reported. A search of the PubMed database (www.pubmed.ncbi.nlm.nih.gov) with keywords “primary cardiac synovial sarcoma” and “pericardial synovial sarcoma with cardiac tamponade” case reports revealed 163 and 13 results, respectively, as of October 2025. PCSS has a 3:1 male-to-female

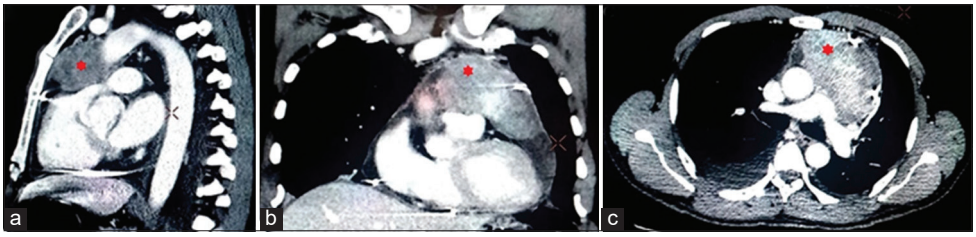

Figure 3: Computed tomography images of thorax in sagittal (a), coronal (b), and cross section (c) views, showing heterogeneously enhancing mass lesion (red stars) in anterior mediastinum in relation to origin of great vessels and extending into pericardial cavity. Pericardial and pleural effusion, along with pericardial drainage catheter noted

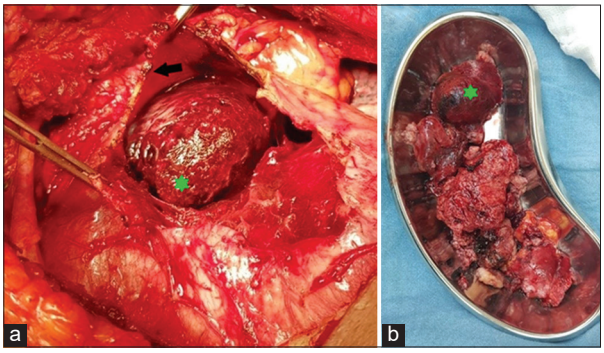

Figure 4: Intraoperative findings (a) on incision of pericardium (black arrow) showing the polypoidal pericardial mass lesion (green star). Final resected mass (green star) (b)

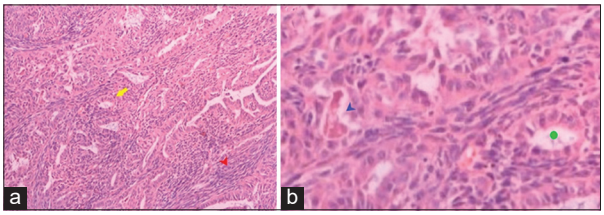

Figure 5: Histopathological photomicrograph (hematoxylin-eosin stain, ×100 magnification) showing biphasic spindle cells in fascicles (red arrowhead) and epithelial cells (yellow arrow) in a glandular pattern of growth (a). Zoomed image of figure (a), clearly showing tumor cells in fascicular arrangement and glandular pattern of cells (b), containing intraluminal eosinophilic secretions (green dot and blue arrowhead)

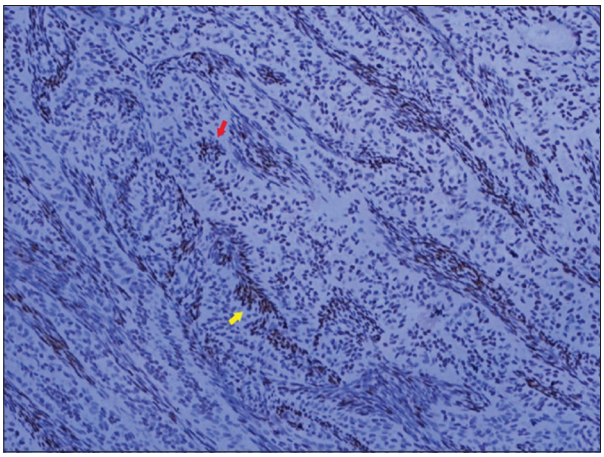

Figure 6: Immunohistochemistry depicting the neoplastic cells positivity for Transducin like enhancer of slit-1 (avidin biotin, ×100 magnification) of both, epithelial (red arrow) and spindle elements (yellow arrow)

prevalence [6]. The majority arise in pericardium, followed by the right atrium and other locations [6]. Dyspnea is the most frequent presenting symptom, though non-specific symptoms may sometimes be the sole manifestation.

CT is the most common presentation of PSS; it may also present with only systemic manifestations and PEf, without any pericardial mass [7]. ECG in PSS may show low voltage complexes and non-specific ST-T changes. CR may be normal in some patients of PCSS, while it may demonstrate cardiomegaly, PlEf, or pulmonary congestion in others [6]. Echocardiography is more sensitive in intracardiac SS, while in PSS with large PEf, tumor mass may not be detected [6]. It can also help in diagnosing CT. Computed tomography demonstrates a heterogeneously enhancing mass, with evidence of local infiltration and/or presence of PEf. There may be foci of calcification or cystic areas within. However, it is important to differentiate it from other tumors, such as mesothelioma [6,7]. Cardiac magnetic resonance imaging is also a good imaging modality for diagnosis, but may sometimes miss the lesion in favor of pericarditis [6].

Definitive diagnosis is made by histopathological examination, immunohistochemistry, and demonstrating cytogenetic alterations. Three subtypes of PSS are identified histopathologically: (i) Monophasic – spindle/epithelial cells in a monomorphic pattern, (ii) Biphasic – spindle and epithelial cells present in varying proportions. Epithelial cells sometimes show a glandular pattern of growth, and (iii) Poorly differentiated type, with a spherical/polygonal cell pattern [8]. Histopathology and immunohistochemistry confirmed the diagnosis in our case. TLE 1 is one of four TLE genes involved in embryogenesis, hematopoiesis, neurogenesis, and body-patterning [9]. TLE 1 is overexpressed in SS. TLE 1 is also involved with the *Wnt* signaling pathway, which is linked to the development of SS [9,10]. TLE 1 positivity is a reliable marker for the diagnosis of SS [8]. In a meta-analysis of 13 studies using TLE 1 staining for the diagnosis of SS, the overall sensitivity was 94% with a specificity of 81% [10]. An antibody panel immunopositivity may not be able to differentiate SS from other similar tumors comprising the close differential diagnoses- sarcomatoid mesothelioma, malignant peripheral nerve sheath tumors, fibrosarcoma, sarcomatoid carcinoma, and hemangiopericytoma [2]. The characteristic biphasic phenotype with strong TLE 1 positivity narrowed the diagnosis of SS in our patient. The gold standard test for SS diagnosis remains the demonstration of highly specific chromosomal translocation t(X;18)(p11.2;q11.2) SYT-SSX [11]. This may be detected in >95% cases of

SS. Cytogenetic evaluation would have been the ideal confirmatory test in our patient; however, in view of logistical considerations, we performed a histopathological and immunohistochemical analysis to confirm the diagnosis.

Prognosis of SS is poor with an estimated 5-year survival in adults between 50% and 60%, while in the case of PCSS, outcomes are worse; with reported survival rates between a few months to 1 year [11,12]. Favorable prognostic factors specific for PCSS include age at diagnosis <30 years, absence of complex chromosomal abnormalities, chemotherapy, and pericardial origin of the tumor [12,13].

There is a paucity of established guidelines on the ideal treatment regimen. Wide surgical excision is the primary treatment modality. Radical surgery is often impossible, due to infiltration of adjacent vital structures [14,15]. External beam radiation therapy combined with neo-adjuvant chemotherapy (ifosfamide + doxorubicin) has been employed in non-resectable tumors before surgery [14]. Intraoperative radiotherapy has also been used in these patients. Recurrent lesions have been treated with surgical resection and docetaxel, especially in ifosfamide-resistant cases [14]. In advanced metastatic disease, treatment with anthracycline/ ifosfamide alone or in combination has been shown to have 25–60% response rates [11]. The reader is referred to an excellent review on treatment regimens and clinical trials for the management of advanced SS [11].

CONCLUSION

We report a case of primary PSS of the biphasic phenotype, presenting with CT. Despite being uncommon, PSS can be diagnosed conclusively after analyzing clinical presentation with appropriate laboratory techniques and cytogenetic evaluation. Meticulous histopathological and immunohistochemical analysis helps in clinching the diagnosis, while ruling out other similar tumors, in settings where cytogenetic studies may not be feasible. Despite having a grim prognosis, early diagnosis and prompt initiation of a multi-pronged treatment plan are important. This may lead to better outcomes and improved overall disease-free survival, especially in patients with favorable prognostic factors.

ACKNOWLEDGMENT

We would like to express our sincere gratitude to Dr. Sabyasachi Bal (Chairman, Thoracic surgery, Sir Ganga Ram Hospital, New Delhi), Dr. Sanjay Gupta (Director, Cardio thoracic vascular surgery, Fortis Flt. Lt. Rajan Dhall Hospital, Vasant Kunj, New Delhi), and Dr. Naorem Gopendro Singh for providing invaluable guidance and assistance in preparation of this manuscript.

AUTHOR'S CONTRIBUTIONS

JD: Conception of study, patient care, drafting of manuscript; RS and KV: Patient care, imaging; RK: Patient management, guidance, and supervision; TG: Guidance and supervision.

REFERENCES

1. Restrepo CS, Vargas D, Ocazionez D, Martínez-Jiménez S, Betancourt Cuellar SL, Gutierrez FR. Primary pericardial tumors. Radiographics 2013;33:1613-30.
2. Arun Priya S, Vasudevan S, Anu Priya S, Aishwarya S. Synovial sarcoma- a review. Int J Contemp Med Res 2017;4:1344-6.
3. Collins D. Aetiology and management of acute cardiac tamponade. Crit Care Resusc 2004;6:54-8.
4. Lewinter MM, Cremer PC, Klein AL. Pericardial diseases. In: Libby P, Bonow RO, Mann DL, Tomaselli GF, Bhatt DL, Solomon SD, *et al*, editors. Braunwald's Heart Disease a Textbook of Cardiovascular Medicine. 12th ed. Philadelphia, PA: Elsevier; 2022. p. 1615-34.
5. Lenihan DJ, Reardon MJ, Hundley WG. Tumors affecting the cardiovascular system. In: Libby P, Bonow RO, Mann DL, Tomaselli GF, Bhatt DL, Solomon SD, *et al,* editors. Braunwald's Heart Disease a Textbook of Cardiovascular Medicine. 12th ed. Philadelphia, PA: Elsevier; 2022. p. 1829-40.
6. Wang JG, Li NN. Primary cardiac synovial sarcoma. Ann Thorac Surg 2013;95:2202-9.
7. Youn HC, Lee Y, Kim SC. Pericardial synovial sarcoma presenting with large recurrent pericardial effusion. J Thorac Dis 2016;8:E412-6.
8. Kaur H, Kumar T, Singh S, Vemulakonda LA, Bhargava R. Primary pericardial synovial sarcoma: A case series and review of literature. Cureus 2025;17:e86731.
9. Li Y, Zhang X, Wang A, Ding S, Li Y, Wang H, *et al*. Transducer-like enhancer of split 1 (TLE1) as a novel biomarker for diagnosis of synovial sarcoma correlates with translocation t(X;18): A study of 155 cases in China. Int J Clin Exp Pathol 2019;12:251-8.
10. El Beaino M, Jupiter DC, Assi T, Rassy E, Lazar AJ, Araujo DM, *et al*. Diagnostic value of TLE1 in synovial sarcoma: A systematic review and meta-analysis. Sarcoma 2020;2020:7192347.
11. Stacchiotti S, Van Tine BA. Synovial sarcoma: Current concepts and future perspectives. J Clin Oncol 2018;36:180-7.
12. Varma T, Adegboyega P. Primary cardiac synovial sarcoma. Arch Pathol Lab Med 2012;136:454-8.
13. Coli A, Chiariello GA, Novello M, Colizzi C, Massetti M. Treatment of cardiac synovial sarcoma: Experience of two cases. J Cardiothorac Surg 2018;13:84.
14. Phatak P, Khanagavi J, Aronow WS, Puri S, Yusuf Y, Puccio C. Pericardial synovial sarcoma: Challenges in diagnosis and management. F1000Res 2014;3:15.
15. Sharma A, Dixit S, Sharma M, Sharma N, Sharma O. Primary synovial cell sarcoma of the heart: A rare case. Heart Views 2015;16:62-4.

Funding: Nil; Conflicts of interest: Nil.

How to cite this article: Dey J, Sud R, Vaidya K, Kachru R, Ghose T. Primary pericardial synovial sarcoma presenting as cardiac tamponade. Indian J Case Reports. 2026; 12(1):32-35.