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Case Report

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Primary Cardiac Synovial Sarcoma of the Left Ventricular Outflow Tract Mimicking Cardiac Myxoma

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Abstract

Background: Primary cardiac synovial sarcoma (PCSS) is exceedingly rare and typically presents with imaging features characteristic of malignancy, including aggressive growth, poorly defined margins, and limited mobility. However, a subset of cases may closely mimic a myxoma with a benign imaging phenotype, thereby posing a significant preoperative diagnostic pitfall and compromising treatment planning.

Case summary: A 34-year-old man was admitted with acute cerebral infarction. Echocardiography and coronary CTA demonstrated a solid, pedunculated subaortic mass attached to the left ventricular outflow tract, with marked mobility and imaging features highly consistent with a myxoma. Given the acute phase of cerebral infarction and high perioperative risk, tumor resection was deferred following multidisciplinary team discussion. At 4-month follow-up, the mass showed a mild interval increase in size, and open-heart surgery with mass excision was performed. Intraoperatively, the tumor exhibited a botryoid, semi-translucent appearance but was soft externally and firm internally on palpation. Histopathology and

immunohistochemistry revealed positivity for TLE1 and SS18-SSX, and FISH confirmed SS18 gene rearrangement, establishing the diagnosis of PCSS. Adjuvant chemotherapy was administered postoperatively, with no evidence of residual tumor on follow-up imaging.

Discussion: This case highlights that PCSS may completely mimic the growth pattern of a myxoma in its early phase, including a pedunculated attachment, marked mobility, and well-circumscribed borders—imaging features that readily lead to a benign misdiagnosis. For masses located in atypical locations with benign imaging features but complicated by systemic embolization, a high index of suspicion for malignancy should be maintained.

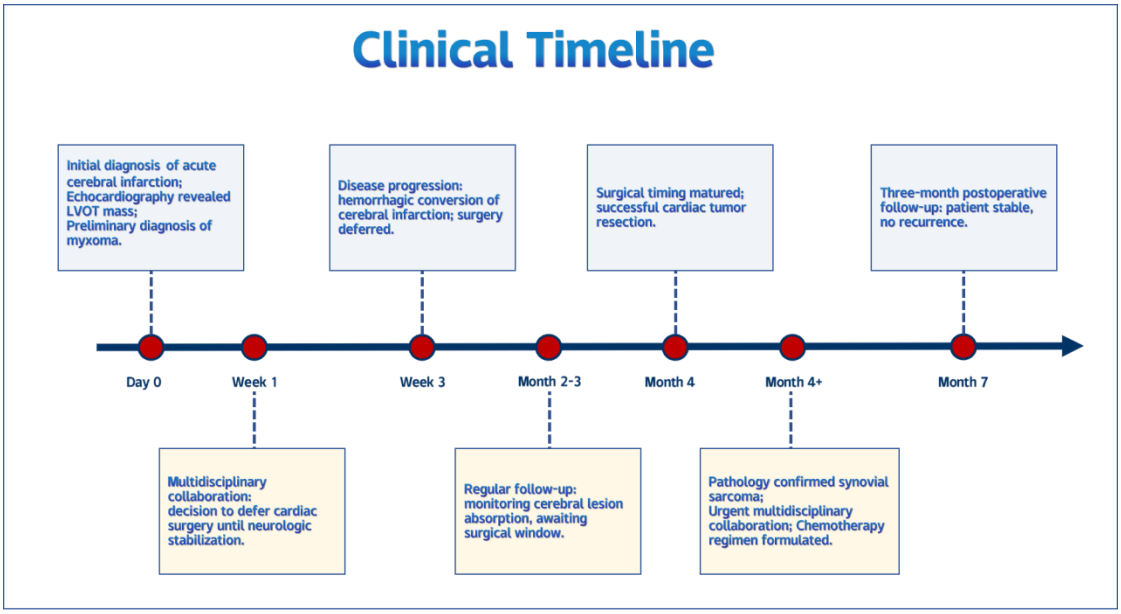
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Learning points

1. PCSS may present with imaging features closely mimicking a myxoma in its early phase, including a pedunculated attachment, marked mobility, and well-circumscribed borders, thereby readily leading to benign misdiagnosis. A high index of suspicion for malignancy should be maintained for masses in atypical locations.

2. Systemic embolization (e.g., cerebral infarction) may represent the initial manifestation of a malignant cardiac tumor. Even in the presence of imaging features highly suggestive of a myxoma, malignancy cannot be definitively excluded.

Summary figure



Introduction

Primary cardiac tumors are exceedingly rare. Most are benign, with myxomas representing the predominant histotype in adults [1]. Sarcomas constitute the majority of malignant cases, accounting for approximately 88.5% [2], whereas primary cardiac synovial sarcoma (PCSS) represents less than 4% of cardiac sarcomas [3]. Synovial

sarcoma typically demonstrates malignant characteristics, including infiltrative growth, a broad-based attachment, irregular margins, and relative immobility. It frequently involves the pericardium or right-sided cardiac chambers, thereby conferring substantial malignant predictive value on imaging assessment[3]. However, some cardiac synovial sarcomas may initially demonstrate imaging morphology closely resembling that of benign lesions, such as myxomas, characterized by a pedunculated attachment, high mobility, regular margins, and absence of myocardial infiltration, thereby posing a substantial risk of preoperative misdiagnosis.Such deceptive imaging manifestations have been rarely reported in the existing literature, often resulting in delayed diagnosis and inadequate treatment preparation.The present case describes a primary cardiac synovial sarcoma originating from the subaortic region, with acute cerebral infarction as the initial clinical manifestation. Preoperative imaging features were highly concordant with those of a myxoma, and the definitive diagnosis was ultimately established by histopathology and the identification of the SS18-SSX fusion gene.This case underscores that cardiac synovial sarcoma may completely mimic myxoma on imaging, particularly when the tumor is located in an atypical site and accompanied by systemic embolization. Clinicians and radiologists should maintain heightened vigilance to avert misdiagnosis and delayed treatment.

Case description

The patient was a 34-year-old man who presented with sudden onset of left-sided weakness and numbness of 1-day duration. Cranial CT performed at an outside hospital revealed right cerebral infarction, and echocardiography demonstrated a subaortic mass. He had no prior history of cardiac disease or family history of malignancy.

On admission, physical examination revealed leftward deviation of the tongue, decreased muscle strength in the left upper extremity, and a positive Babinski sign on the left. Laboratory investigations, electrocardiography, chest CT, and abdominal ultrasonography were all unremarkable.Cranial MRI revealed a large acute infarct in the right cerebral hemisphere (Figure 1a-b). Coronary CTA showed no coronary artery embolism but demonstrated a subaortic mass (Figure 1c-d).

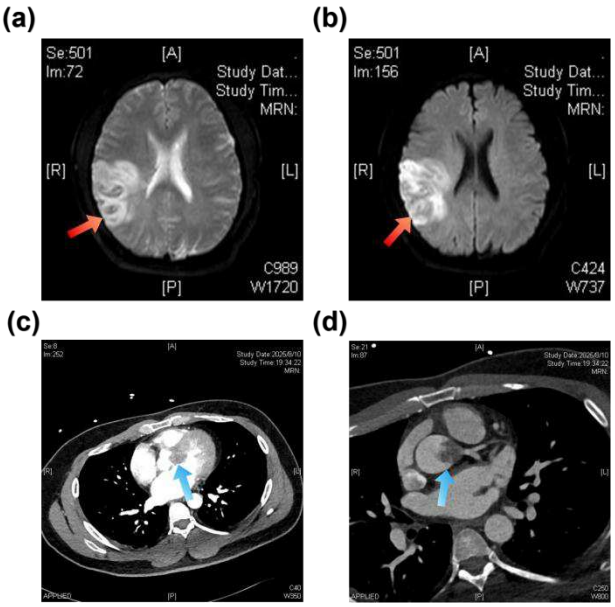


Figure 1. Cranial MRI revealing a large acute infarct in the right cerebral hemisphere. Coronary CTA demonstrating a subaortic mass.

(a-b) Cranial MRI demonstrates patchy areas hypointense on T1-weighted images and hyperintense on T2-weighted images in the right frontoparietotemporal and insular regions, with associated gyriform swelling. Diffusion-weighted imaging (DWI) reveals hyperintensity, and apparent diffusion coefficient (ADC) maps demonstrate hypointensity (red arrows). (c-d) Coronary CTA demonstrates a mobile solid subaortic mass (blue arrows) situated between the left ventricular outflow tract (LVOT) and the aortic sinuses, without involvement of the left coronary artery.

TTE and TEE demonstrated a solid, lobulated, and pedunculated subaortic mass attached inferior to the commissure between the left and right coronary cusps. The mass exhibited heterogeneous echogenicity with focal areas of cystic necrosis. It was highly mobile, with marked excursion throughout the cardiac cycle, and caused dynamic mechanical obstruction of the aortic valve orifice during systole (Figure 2a-f and Supplementary Video 1).

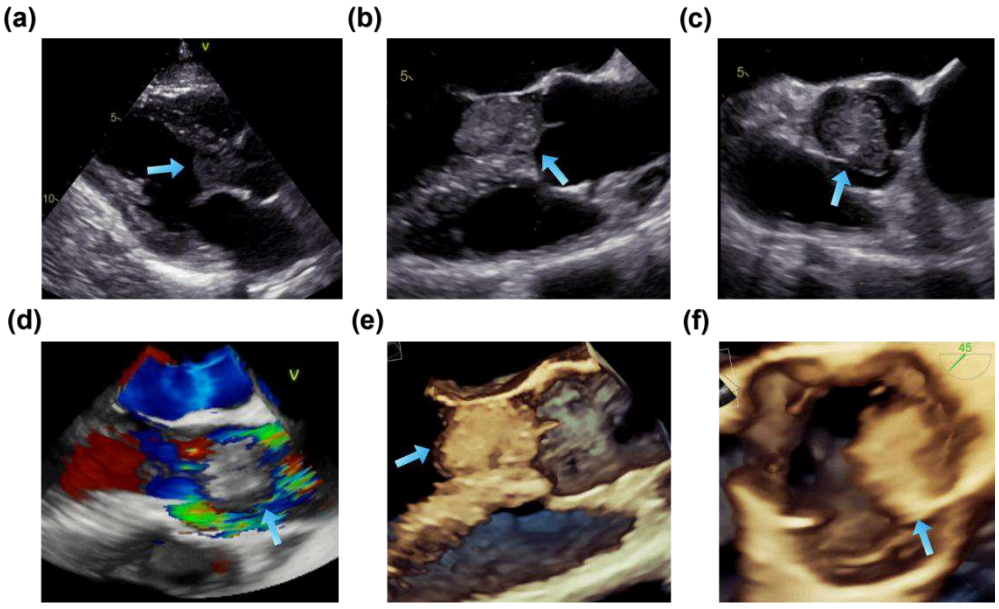


Figure 2. Transthoracic and transesophageal echocardiography demonstrating a pedunculated solid

subaortic mass and its structural characteristics.

(a) TTE demonstrating an irregular hypoechoic solid subaortic mass.(b-c) Two-dimensional TEE providing superior delineation of the heterogeneous echotexture within the mass, with punctate hyperechoic foci and focal hypoechoic to anechoic areas. The mass was highly mobile throughout the cardiac cycle, without restriction of aortic leaflet excursion.(d) Color Doppler flow imaging (CDFI) on TEE demonstrating a narrowed, high-velocity flow stream across the aortic valve orifice during systole.(e) Three-dimensional TEE en face view at 135° demonstrating an irregular mass surface with granular projections.(f) Three-dimensional TEE en face view at 45° demonstrating the stalk of the mass attached inferior to the commissure between the left and right coronary cusps.

The patient was admitted with cerebral infarction as the initial manifestation. In conjunction with the CT and echocardiographic findings, there was a high clinical suspicion that the subaortic mass represented a myxoma. Multidisciplinary evaluation by neurology and cardiac surgery teams led to the decision to defer surgery due to the high perioperative risk of intracranial hemorrhage in the setting of acute cerebral infarction with significant edema, and the anticoagulation-hemostasis dilemma during the perioperative period. Surgical intervention was planned after neurologic stabilization.

The patient subsequently underwent monthly imaging follow-up of the brain and heart. At 4 months, cranial CT demonstrated the cerebral infarct with partial encephalomalacia (Figure 3a-b). TTE revealed a mild interval increase in the size of the subaortic mass (Figure 3c, Supplementary Video 2). Following a multidisciplinary collaboration (MDT) discussion involving cardiac surgery, anesthesiology, and neurology, open-heart surgery for resection of the cardiac mass was ultimately undertaken.

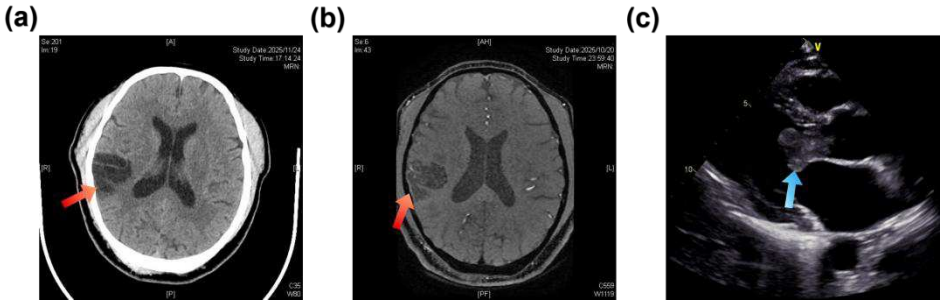


Figure 3. Four-month imaging follow-up.

(a) Cranial CT demonstrating patchy hypodense areas in the right frontoparietotemporal and insular regions, with a few strip-like slightly hyperdense foci internally, consistent with post-infarct encephalomalacic changes.(b) Cranial MRI demonstrating patchy areas hypointense on T1-weighted images and hyperintense on T2-weighted images in the right frontoparietotemporal and insular regions. A few linear hyperintense foci were noted within the lesion, with mild gyriform swelling.(c) TTE in the parasternal long-axis view demonstrating the subaortic mass (blue arrows) with a mild interval increase in its longitudinal diameter. During diastole, the mass extended beyond the level of the anterior mitral leaflet tip.

Intraoperative inspection revealed a subaortic mass measuring approximately 31 × 26 × 29 mm, with a botryoid appearance and semi-translucent surface. The stalk was well-defined, and the mass was sharply demarcated from the surrounding tissue without invasion of the aortic valve leaflets. On palpation, the tumor was soft externally but firm internally, a consistency inconsistent with the loose gelatinous

texture characteristic of a typical myxoma. An intraoperative frozen section was therefore sent for immediate pathological examination.

Gross examination revealed grayish-white to yellow soft tissue. Microscopically, diffuse proliferation of short spindle cells was observed (Figure 4b-c). Immunohistochemistry showed positivity for TLE1, CD99, BCL-2, SS18-SSX, and Vimentin; the Ki-67 proliferation index was approximately 40%. Staining for S-100, Desmin, and SMA was negative. Fluorescence in situ hybridization (FISH) confirmed a positive SS18 gene rearrangement. The final diagnosis was PCSS. Following multidisciplinary discussion by the oncology team, adjuvant chemotherapy was initiated. At 3-month postoperative follow-up, echocardiography showed no evidence of residual tumor. The patient remains in good general condition with ongoing close surveillance.

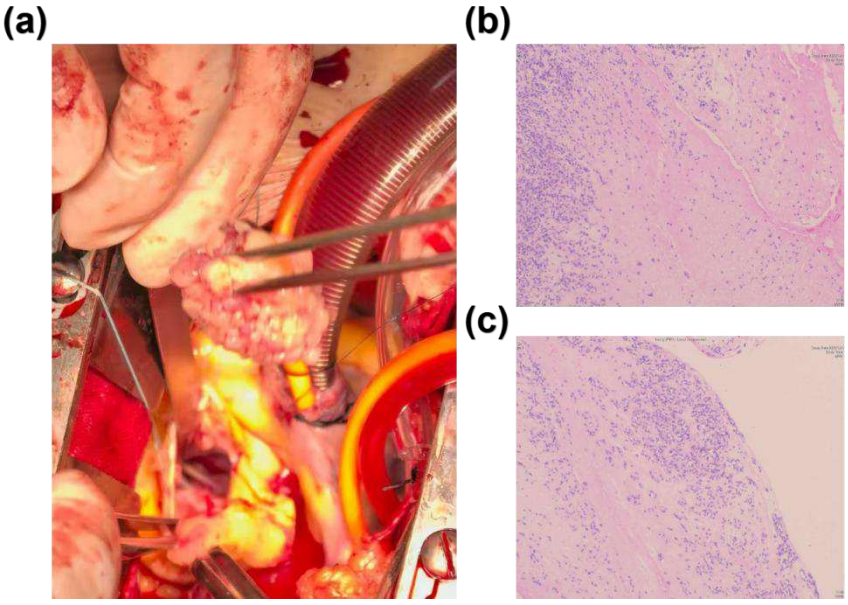


Figure 4. Intraoperative gross appearance and histopathological features.

(a) Intraoperative photograph demonstrating a pedunculated subaortic mass with a botryoid appearance and semi-translucent, irregular surface. The mass is well-circumscribed with a soft but elastic consistency (stalk grasped by forceps).(b-c) Histopathology showing diffuse proliferation of short spindle cells with a diffuse growth pattern, consistent with a mesenchymal neoplasm.

Discussion

1. The Mimic Challenge

The diagnostic challenge in this case was that the malignant cardiac tumor exhibited imaging features characteristic of a typical myxoma, thereby posing a significant preoperative diagnostic pitfall. Typically, cardiac myxomas present as pedunculated masses attached to the fossa ovalis of the interatrial septum, appearing as a solid hypoechoic mass with well-circumscribed borders and a smooth or villous surface, and oscillate on a stalk in a regular pattern throughout the cardiac cycle [4]. Given these characteristic echocardiographic features, the diagnosis of myxoma is generally straightforward.

In contrast, PCSS is a malignant mesenchymal neoplasm that commonly arises

from the pericardium or right-sided cardiac chambers. Imaging typically reveals a large mass with a broad base, extensive contact with the atrial or ventricular wall, poorly demarcated borders with the myocardium, frequent valvular involvement, irregular morphology, indistinct margins, an irregular surface, heterogeneous internal echogenicity, and limited mobility. These typical findings are usually suggestive of malignancy.

However, the synovial sarcoma in the present case closely mimicked a myxoma in both clinical presentation and imaging morphology: a pedunculated attachment, marked mobility, well-circumscribed borders, and an absence of obvious infiltration were all consistent with features of benignancy, thereby enabling the tumor to completely masquerade as a myxoma preoperatively. Such masquerading has been infrequently reported in the literature [5–9], further compounding the difficulty of preoperative differential diagnosis.

2.Key Differential Diagnosis

In this case, PCSS was located in the subaortic region, with cerebral infarction as the sole initial manifestation—a clinical pattern that is exceedingly rare in the existing literature. Although the imaging features closely mimicked a myxoma, posing a significant diagnostic pitfall, several clues suggestive of malignancy were evident on retrospective review.

1) Epidemiologic features: Myxoma predominantly occurs in women aged 40 to 60 years, whereas cardiac synovial sarcoma more frequently affects young males, with a mean age of approximately 35 years [5,10]. The patient in this case was a 34-year-old man; epidemiologically, this profile is more consistent with synovial sarcoma and suggests that myxoma-like imaging findings should be interpreted with caution.

2) Imaging features:

① Although myxomas can arise in any cardiac chamber, they are most commonly located in the left atrium along the interatrial septum [11]. A subaortic location is exceedingly rare for myxomas and should raise concern for an underlying malignant neoplasm.

② Myxomas are composed predominantly of myxoid stroma and typically exhibit a soft, translucent, gelatinous consistency. Synovial sarcoma, in contrast, is a mesenchymal neoplasm with a firmer texture. Intraoperative palpation in this case revealed a mass that was soft externally but firm internally—a consistency inconsistent with the uniformly loose gelatinous texture of a typical myxoma and an important diagnostic clue. Preoperatively, the marked mobility of the tumor had been misinterpreted as indicative of a friable architecture; however, mobility does not directly reflect tissue histology.

③ Calcification characteristics: Calcification is exceedingly rare in myxomas; when present, it is typically coarse. In contrast, synovial sarcoma may demonstrate fine calcifications in 28% to 30% of cases [12,13]. In this case, TEE revealed fine punctate hyperechoic foci within the mass, corresponding to microcalcifications—a finding suggestive of malignancy.

3) Histopathology: The definitive diagnosis relies on postoperative

histopathological examination, including immunohistochemistry and testing for the SS18 gene rearrangement. In this case, positivity for TLE1 and SS18-SSX, together with FISH confirmation of the SS18 rearrangement, established the diagnosis of primary cardiac synovial sarcoma.

3. Treatment Strategy and Lessons Learned

Previous studies have demonstrated that cardiac synovial sarcoma is associated with a poor overall prognosis, with a median survival of approximately 27 months. Radiotherapy and chemotherapy may improve survival to some extent, regardless of whether complete resection is achieved [14]. In this case, the patient was at high risk in the setting of acute cerebral infarction, and preoperative imaging was highly suggestive of benignancy. Tumor-directed therapy was therefore deferred. No evidence of obvious local infiltration or rapid progression was observed during the follow-up period, thereby preserving surgical feasibility. No evidence of obvious local infiltration or rapid progression was observed during the follow-up period, thereby preserving surgical feasibility.

This case suggests that in patients with cardiac tumors and concomitant acute cerebral infarction, treatment strategies must strike a careful balance among surgical timing, neurological risk, and potential tumor progression. If preoperative multimodality imaging had identified potential clues suggestive of malignancy earlier—such as focal infiltration, abnormal tissue characteristics, or an atypical location—it might have prompted earlier initiation of oncological evaluation, thereby optimizing the overall treatment pathway. This experience underscores the critical role of MDT in the management of complex cardiac tumors.

Conclusions

PCSS may present with imaging features closely mimicking a myxoma in its early phase, particularly when the tumor is located in an atypical location or is complicated by systemic embolization, thereby posing a significant risk of misdiagnosis. This case underscores that the diagnosis of cardiac tumors requires an integrated approach incorporating epidemiology, imaging findings, and histopathology. Multimodality imaging plays a critical role in identifying potential clues suggestive of malignancy. For masses with atypical features despite suspected benignancy, a high index of suspicion should be maintained, with early planning for histopathological confirmation. Timely surgical excision and appropriate oncological management are crucial for improving prognosis.

Consent for publication

Written informed consent for publication of the anonymized echocardiographic images and related clinical data was obtained separately from the patient involved in this study.

Competing Interests:

The authors declare no competing interests.

Data Availability Statement

The data supporting the findings of this study are available upon reasonable request to the corresponding author. Access to the data may be subject to certain conditions to ensure the privacy and confidentiality of participants.

Permissions statement:

The authors do hereby declare that all illustrations and figures in the manuscript are original and not require reprint permission.

Author notes

Na Ouyanga and Hao Zuo contributed equally to the study.

Ethical Statement

Patient consent was obtained for the publication of this case report. The study adhered to ethical standards, and all data were managed confidentially and anonymized to protect the patient's privacy.

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