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Primary Malignant Mesenchymal Sarcoma of the Left Atrium Complicated with Mitral Valve Disease: A Case Report

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Case Report

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Abstract

Primary cardiac malignant tumors are extremely rare, accounting for less than 0.05% of all cardiac tumors, with malignant mesenchymal sarcomas being one of the most aggressive subtypes. Herein, we report a case of a 56-year-old female patient admitted with acute chest distress and dyspnea, initially suspected of left atrial myxoma combined with moderate mitral stenosis. The patient underwent minimally invasive cardiac surgery under cardiopulmonary bypass, including mitral valve replacement and tumor resection, but converted to thoracotomy due to intraoperative hemorrhage. Postoperative pathological and immunohistochemical examinations confirmed the diagnosis of malignant mesenchymal sarcoma. Severe complications including low cardiac output syndrome, acute respiratory distress syndrome (ARDS), and acute kidney injury occurred postoperatively, which were successfully managed with comprehensive supportive treatments. This case highlights the challenges in preoperative diagnosis, surgical management, and postoperative complication control of primary cardiac malignant sarcomas, and emphasizes the importance of multidisciplinary team (MDT) collaboration in improving patient outcomes.

Introduction

Primary cardiac malignant tumors are uncommon neoplasms, with an incidence of approximately 0.0017% to 0.03% in the general population, which is far lower than that of benign cardiac tumors (such as myxoma) [1]. Among primary cardiac malignant tumors, sarcomas account for about 90%, including angiosarcoma, leiomyosarcoma, and undifferentiated pleomorphic sarcoma [2]. Malignant mesenchymal sarcoma involving the left atrium is particularly rare, and its clinical manifestations are often nonspecific, easily misdiagnosed as benign tumors or other cardiac diseases. Due to the aggressive biological behavior and high recurrence rate, the prognosis of cardiac sarcomas remains poor, with a median survival time of only 6 to 12 months [3]. This case reports the diagnosis, surgical treatment, and postoperative management of a left atrial malignant mesenchymal sarcoma complicated with mitral valve disease, aiming to provide reference for clinical practice.

Case Presentation

A 56-year-old female patient was admitted to our hospital as an emergency on July 14, 2025, with "acute chest distress and dyspnea for 1 hour". She had a medical history of postoperative hypothyroidism (status post thyroidectomy) and hyperlipidemia for 3 years, and chronic bronchitis for 5 years.

Preoperative examinations: Echocardiography at a local hospital revealed a left atrial mass, with possible vegetation on the anterior leaflet of the mitral valve. On July 6, computed tomography (CT) at Xiaolan People's Hospital suggested a left atrial space-occupying lesion, considering myxoma. (Fig. 1A,B,C) Echocardiography at Guangdong Provincial People's Hospital confirmed a left atrial mass (approximately 5.4cm×5cm×2cm) with unclear nature and moderate mitral stenosis. Upon admission, physical examination showed: blood pressure 110/70 mmHg, heart rate 105 beats/min, respiratory rate

24 times/min, oxygen saturation 92% (room air). Cardiac auscultation revealed a diastolic rumble murmur at the mitral valve area. Laboratory tests showed elevated N-terminal pro-brain natriuretic peptide (NT-proBNP) (1860 pg/mL) and mild leukocytosis (11.2×10⁹/L).

Preoperative MDT evaluation: The MDT consisting of cardiothoracic surgeons, anesthesiologists, radiologists, and pathologists determined that the surgical indication was clear, with no absolute contraindications. The surgical plan was formulated as thoracoscopic mitral valve replacement combined with cardiac tumor resection under cardiopulmonary bypass.

Surgical and Postoperative Course

The patient underwent surgery. Intraoperatively, a large mass was found in the left atrium, severely adherent to the atrial wall and mitral valve leaflets. Thoracoscopic mitral valve replacement with a mechanical valve and tumor resection were performed. However, due to massive intraoperative hemorrhage (total blood loss approximately 1000 mL), the operation was converted to thoracotomy for hemostasis. Intraoperative frozen pathology suggested spindle cell proliferation with local infarction and myxoid stroma, considering myxoma with infarction.

Postoperatively, the patient was transferred to the intensive care unit (ICU) for monitoring. She developed low cardiac output syndrome, manifested by hypotension (85/50 mmHg), oliguria, and elevated lactate (3.8 mmol/L). Intra-aortic balloon pump (IABP) support was initiated immediately, combined with inotropic agents (dopamine and dobutamine). Meanwhile, ARDS and acute kidney injury were diagnosed, and non-invasive ventilator support, anti-infection treatment (piperacillin/tazobactam), anti-inflammatory therapy, and myocardial nutrition support were administered.

During the ICU stay, the patient's condition gradually improved. IABP was removed, and non-invasive ventilation was switched to nasal cannula oxygen therapy. Postoperative paraffin pathology showed spindle cells arranged in a storiform and fascicular pattern, accompanied by hemorrhage, geographic necrosis, and myxoid degeneration, with frequent mitotic figures. Consultation with Guangzhou KingMed Diagnostics suggested a malignant tumor. Pathological consultation with Sun Yat-sen University Cancer Center confirmed the diagnosis of malignant mesenchymal sarcoma, with a Ki-67 proliferation index of approximately 20%.

The patient's vital signs were stable (blood pressure 120/80 mmHg, heart rate 88 beats/min, oxygen saturation 98% on room air), and the surgical incision healed well. She was discharged with recommendations to complete immunohistochemical staining and next-generation sequencing (NGS) for further subtyping, followed by oncological consultation for adjuvant treatment.

Pathology and Immunohistochemical Findings

Gross specimen: The resected tumor was 5.4cm×5.0cm×2.0cm in size, with an irregular shape, grayish-white cut surface, and focal hemorrhage and necrosis.

Immunohistochemical results: The tumor cells were positive for vimentin, partially positive for SMA, and negative for CD34, CD117, DOG1, S-100, desmin, and CK. The Ki-67 proliferation index was approximately 20%.(Fig. 2A)

Microscopic examination: Spindle cells were arranged in a storiform and fascicular pattern, with pleomorphic nuclei, prominent nucleoli, and frequent mitotic figures (≥ 5/10 high-power fields). There were extensive hemorrhage, geographic necrosis, and myxoid stromal degeneration.(Fig. 2B,C)

Discussion

Primary cardiac malignant mesenchymal sarcoma is an extremely rare and aggressive tumor. This case presents several clinical challenges: first, preoperative diagnosis is difficult due to nonspecific symptoms and imaging features similar to benign myxoma. In this patient, the initial imaging findings suggested left atrial myxoma, which delayed the suspicion of malignancy. Second, surgical resection is the primary treatment, but the extensive adhesion of the tumor to cardiac structures increases the risk of intraoperative hemorrhage and incomplete resection. Third, postoperative complications are common and severe, including low cardiac output syndrome, ARDS, and acute kidney injury, which require timely and comprehensive intervention.

The key to improving the prognosis of cardiac sarcomas lies in early diagnosis, complete surgical resection, and appropriate adjuvant treatment. Preoperative imaging examinations such as enhanced cardiac CT and magnetic resonance imaging (MRI) can help evaluate the tumor size, location, and invasion range [4]. Intraoperative frozen pathology is important for guiding surgical decision-making, but its accuracy is limited, and final diagnosis depends on paraffin pathology and immunohistochemistry. For unresectable or metastatic tumors, adjuvant treatments such as chemotherapy, radiotherapy, and targeted therapy may be considered, but their efficacy remains controversial [5].

MDT collaboration plays a crucial role in the entire process. In this case, preoperative MDT evaluation ensured the rationality of the surgical plan, while postoperative collaboration between ICU physicians, cardiologists, and pathologists facilitated the timely management of complications and accurate diagnosis.

In conclusion, primary left atrial malignant mesenchymal sarcoma is a rare disease with high misdiagnosis rate and poor prognosis. Comprehensive preoperative evaluation, meticulous surgical operation, effective postoperative complication management, and MDT collaboration are essential for improving patient outcomes. Further studies are needed to explore the optimal treatment strategies for this rare tumor.

Declarations

Patient consent

Written informed consent for the publication of this case report was obtained from the patient.

An ethics statement

The study was approved by the Institutional Review Board of Zhongshan Xiaolan People's Hospital & Zhongshan Fifth People's Hospital.

Funding section

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Competing Interest declaration

None of competing Interest.

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Figures

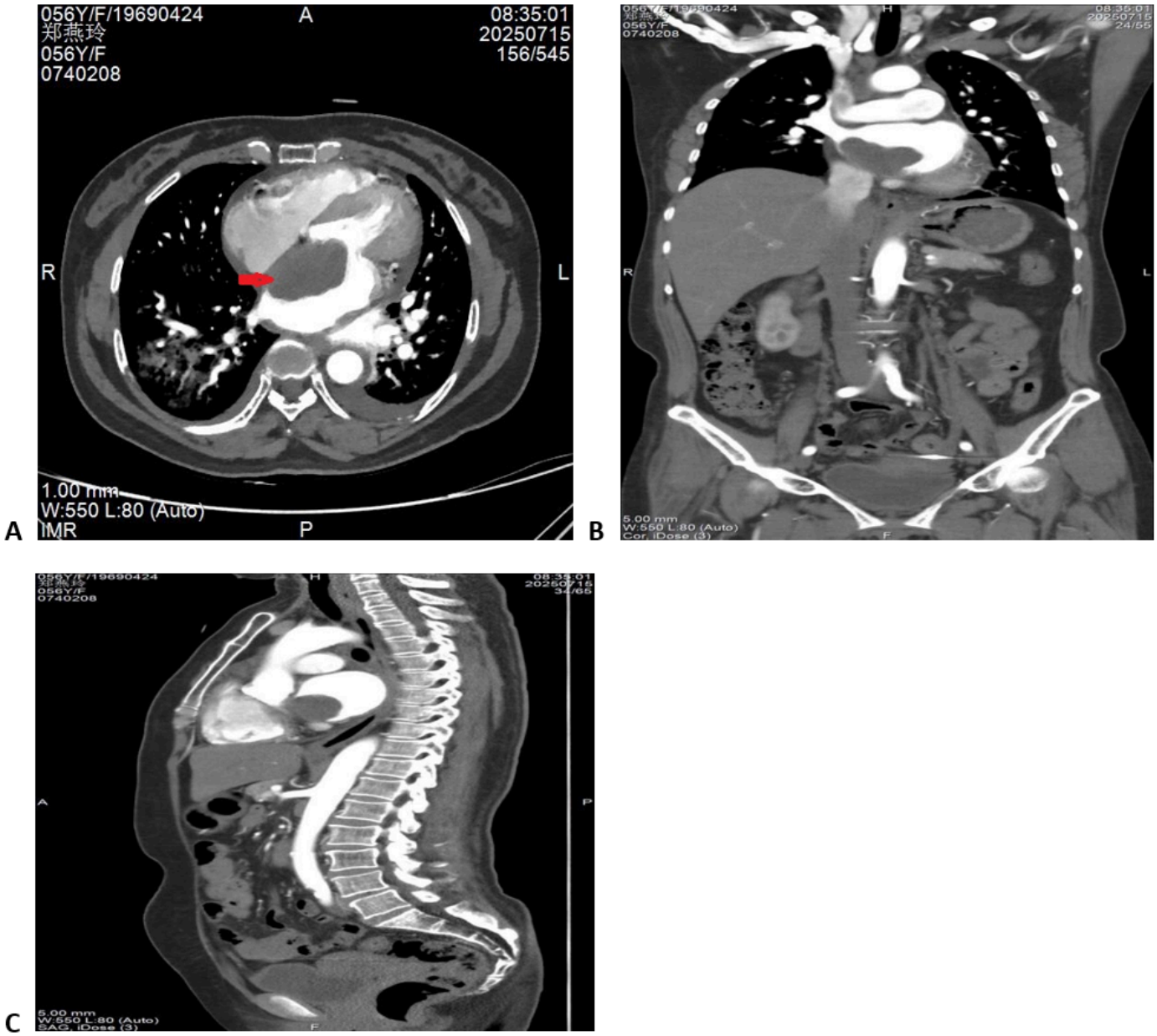


Figure 1

Contrast-enhanced computed tomography (CT) images of the chest and abdomen in a 56-year-old female patient.

Panel A (Sagittal view): A sagittal reformatted image demonstrating the entire thoracic and lumbar spine, with contrast filling the thoracic aorta and visualization of the mediastinal and intra-abdominal organs.Panel B (Axial view, at the level of the heart): An axial image at the level of the heart, showing the contrast-enhanced cardiac chambers. The red arrow points to an area of the right atrial wall. Multiple bilateral pulmonary nodules and ground-glass opacities are also visible, suggestive of metastatic lung disease.Panel C (Coronal view): A coronal reformatted image showing the contrast-enhanced thoracic aorta, bilateral lungs, liver, kidneys, and pelvic bones, providing an overview of the thoracic and abdominal visceral structures.

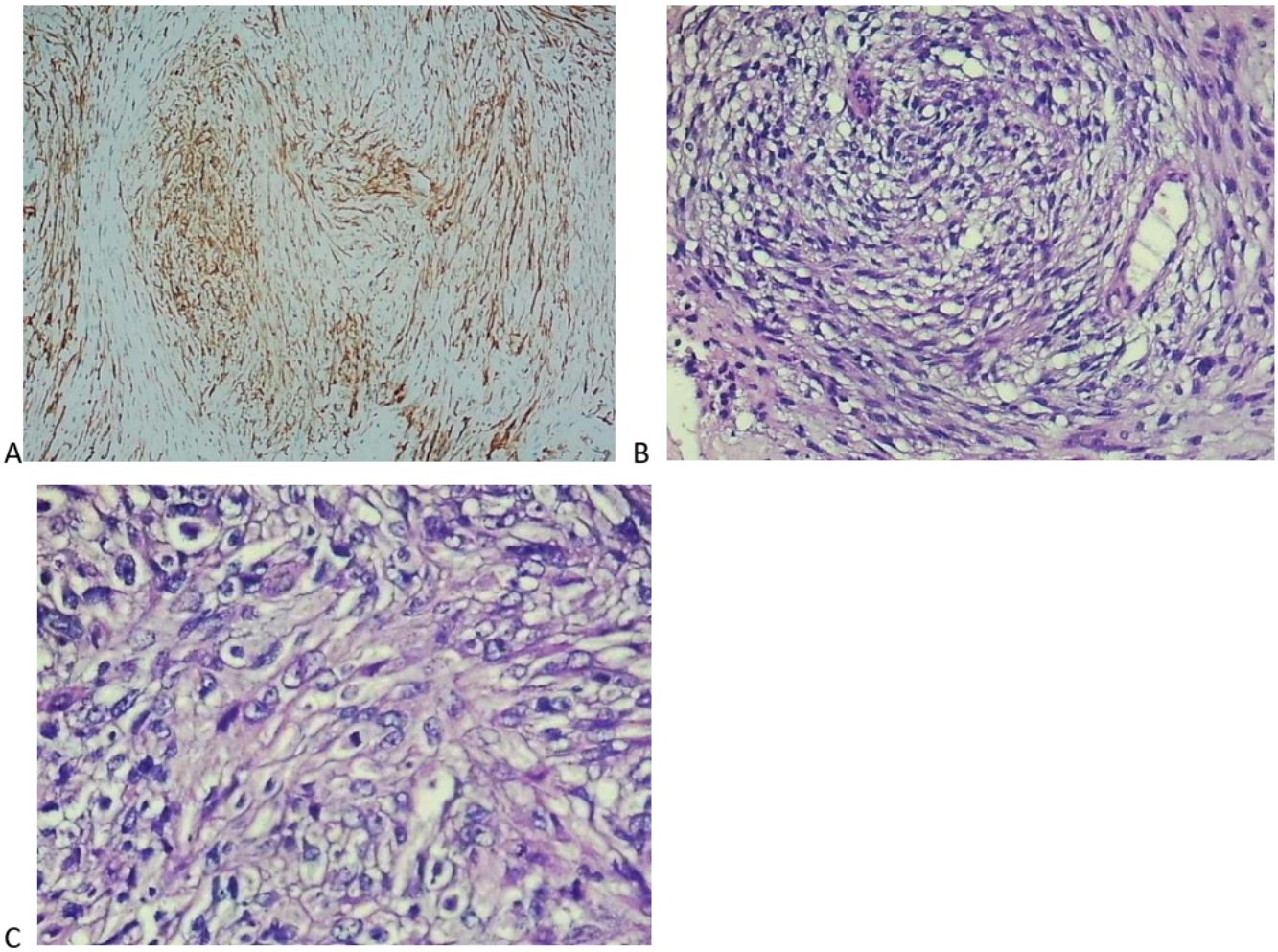


Figure 2

Histopathological and immunohistochemical findings of a soft tissue lesion.

Panel A (Immunohistochemistry stain): Immunohistochemical staining shows diffuse, moderate-to-strong brown cytoplasmic positivity in the spindle cell population, indicating expression of the target antigen.Panel B (Hematoxylin and eosin, H&E stain, low magnification): A low-power view reveals a whorled and storiform growth pattern of spindle cells, with a background of loose myxoid stroma and scattered inflammatory cells.Panel C (Hematoxylin and eosin, H&E stain, high magnification): At higher magnification, the lesion is composed of bland, elongated spindle cells with indistinct cytoplasmic borders, vesicular nuclei, and small nucleoli, arranged in a haphazard pattern within a myxoid matrix.