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Surgical resection of a rare biatrial giant sarcoma: a case report

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Abstract
Primary cardiac undifferentiated sarcoma is an exceptionally rare entity, marked by nonspecific symptoms that considerably challenge its diagnosis and treatment. Surgical resection generally serves as the primary treatment modality. This article describes the case of a 32-year-old female patient admitted to the hospital with symptoms of abdominal distension and pain. Subsequent echocardiography revealed a cardiac tumor that occupied almost the entire left and right atria. The tumor was surgically removed in an emergency procedure, and subsequent pathological examination confirmed an undifferentiated sarcoma. The patient was successfully discharged 17 days after surgery. She then completed six cycles of chemotherapy. A six-month follow-up showed no signs of tumor recurrence or metastasis.

Keywords Cardiac undifferentiated sarcoma, Malignant cardiac tumor, Tumors of the left and right atria, Cardiac surgery

Background

Primary cardiac tumors are rare in clinical practice, with an incidence of approximately 0.001–0.03%. Of these, 75% are benign and 25% are malignant. Sarcomas are the most prevalent malignant type [1], among which undifferentiated pleomorphic sarcoma exhibiting the highest degree of malignancy [2]. Cardiac sarcomas are typically devoid of specific clinical symptoms, making diagnosis challenging. Surgical resection is the primary treatment strategy, but effective management usually requires a multimodal approach that includes surgery, chemotherapy, and radiotherapy. In this case, the patient initially presented with abdominal distension and pain, which are not typical cardiac symptoms. This presentation delayed the immediate recognition of the cardiac issue. The correct diagnosis was ultimately established during an echocardiographic examination, highlighting the unpredictable nature of the diagnostic process for this condition.

Case presentation

Patient information

A 32-year-old female patient, measuring 150 cm in height and weighing 50 kg (BMI: 22.2 kg/m^2), sought medical attention due to persistent abdominal pain and distension unrelieved by oral medications over the past week. The patient reported experiencing chest tightness and exertional dyspnea for over two weeks. She denied any history of hypertension or cardiovascular diseases, and there was no family history of genetic disorders. The patient had undergone a cesarean Sect. 10 months prior. On physical examination, her body temperature was 36.2 °C, blood

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pressure was 90/70 mmHg (1 mmHg = 0.133 kPa), and oxygen saturation was 93% while receiving oxygen at 2 L/min. Her respiratory rate was 18 breaths per minute, and her heart rate was 100 beats per minute with a regular rhythm. There was No precordial prominence was observed, and the apical impulse was located 0.5 cm lateral to the mid-clavicular line in the left 5th intercostal space. No murmurs were detected in any of the valve areas. Breath sounds were diminished bilaterally. The patient was conscious, exhibited generalized edema, and had experienced a weight gain of 5 kg in the past month.

Preoperative findings

Her echocardiogram showed solid echoes occupying the bilateral atrial cavities and the interatrial septum, measuring approximately 88 × 58 × 82 mm. The inferior margin of the mass extended to the orifices of the mitral and tricuspid valves (Fig. 1A).

Left Heart Chambers and Function: The left atrial diameter was within normal limits, while the left ventricular diameter was slightly reduced. The left ventricular wall thickness was normal, and there were no abnormalities in the left ventricular outflow tract. Segmental systolic function of the left ventricle was normal, with a left ventricular ejection fraction of 62%. Mitral Valve: The mitral valve leaflets were not thickened and exhibited-showed normal mobility. The forward flow velocity was increased, though valve opening was not significantly restricted. Continuous Wave (CW) Doppler measurements revealed a peak velocity of 2.1 m/s, a peak pressure gradient of 18 mmHg, and a mean pressure gradient of 7 mmHg. No mitral regurgitation was detected on color Doppler imaging.

Aortic Valve and Root: The aortic sinus and ascending aorta were of normal dimensions. The aortic valve was tricuspid with normal morphology and an unrestricted opening. No aortic regurgitation was observed on color Doppler. Right Heart Chambers and Function: The inferior vena cava had an inner diameter of 20 mm, showing signs of blood stasis. The right atrial diameter was normal. The right ventricular basal segment diameter and wall thickness were normal, with preserved systolic function. The pulmonary artery was not dilated, and the pulmonary valve was not thickened, maintaining an unrestricted opening. The mean pulmonary artery pressure, measured by CW Doppler, was 18 mmHg. The tricuspid valve was not thickened, and the leaflet mobility was normal. Forward flow velocity was increased, but valve opening was not significantly restricted. CW Doppler measurements recorded a peak velocity of 1.9 m/s, a peak pressure gradient of 15 mmHg, and a mean pressure gradient of 8 mmHg. Valve closure morphology was normal, and color Doppler showed mild tricuspid regurgitation. Pericardial Effusion: A small amount of pericardial fluid was noted, measuring 4 mm posterior to the left ventricle and 8 mm anterior to the right ventricular outflow tract during diastole.

Abdominal and pelvic CT showed ascites. Chest CT revealed bilateral pleural effusion (Fig. 2C) and segmental atelectasis in the lower lung lobes. A chest X-ray indicated a cardiothoracic ratio of 0.51. Her electrocardiogram showed a sinus rhythm of 100 beats/minute without any significant abnormalities.

Whole-body positron Emission Tomography-Computed Tomography (PET-CT) detected a mass with high glucose metabolism in the atria and interatrial septum

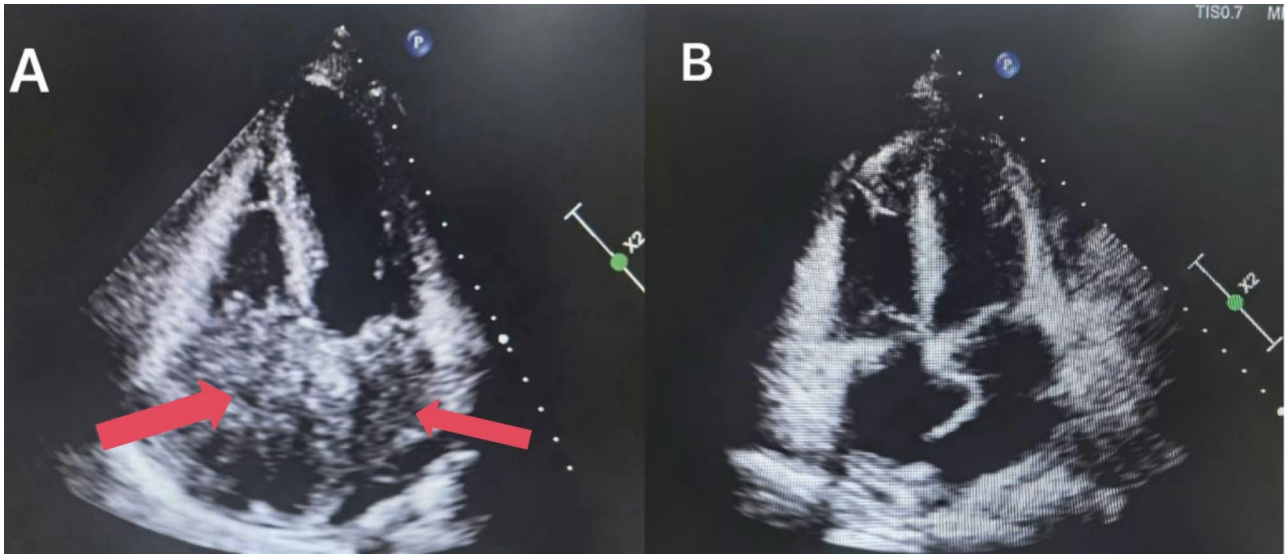

Fig. 1 Transthoracic echocardiography, apical four chamber view showing: (**A**) preoperative huge atrial mass (arrow indicates cardiac mass), (**B**) postoperative

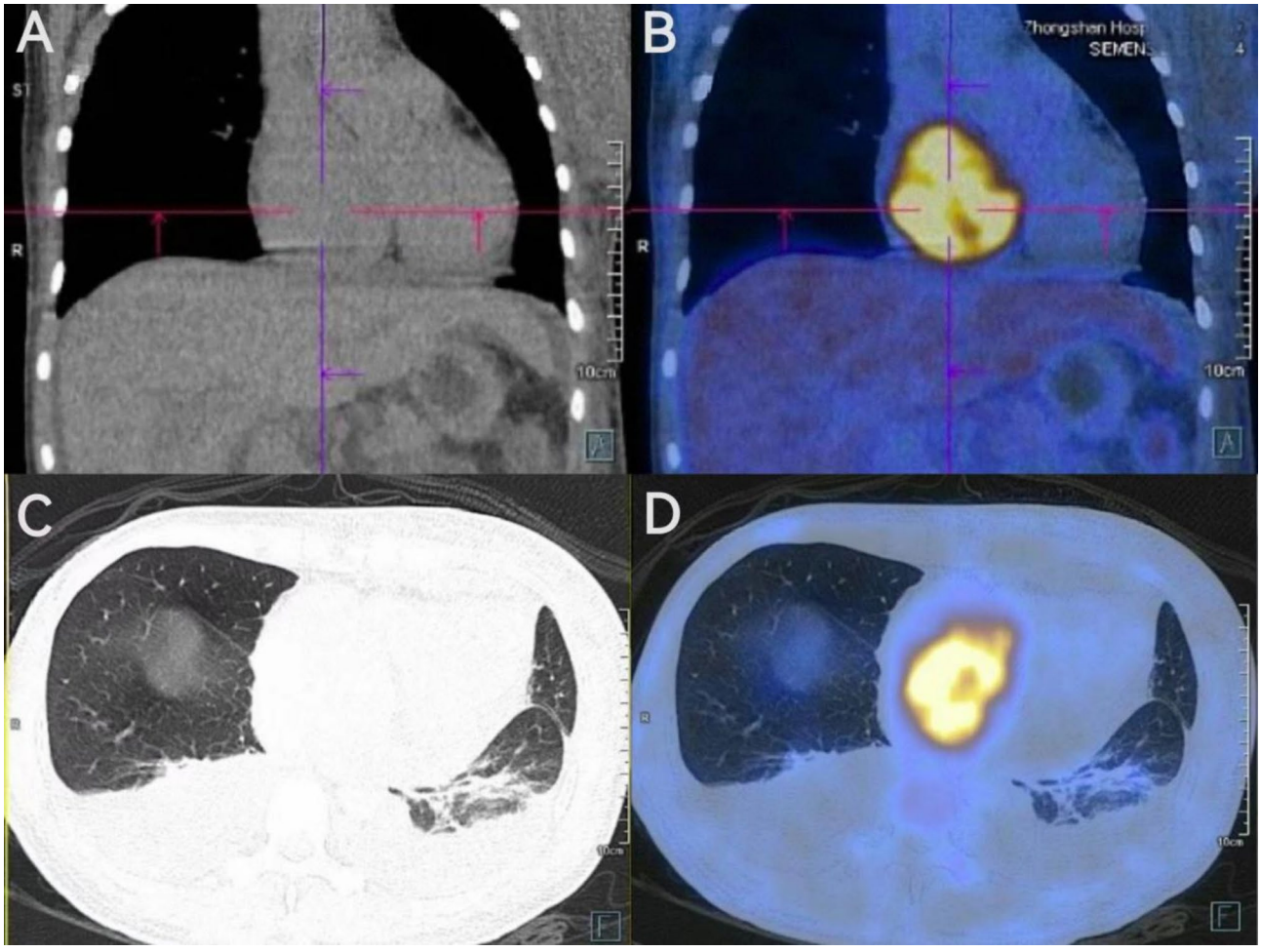

Fig. 2 (**A-D**) PET-CT image demonstrating: (**B**, **D**) A mass with abnormally increased glucose metabolism in the left and right atria and atrial septum is not demarcated from the ventricular wall. (**C**, **D**) Bilateral pleural effusion

(Fig. 2B, D), measuring 69.9 × 66.2 mm, with an unclear boundary with the ventricular wall, CT value of 42.8 HU, and SUVmax of 11.9. A slightly enlarged lymph node was noted at the right cardiophrenic angle, measuring 8.4 × 7.8 mm, with a CT value of 17.0 HU and SUVmax of 0.8. A small amount of pericardial fluid was also present. No regions of increased uptake were observed in the remaining systems, although soft tissue edema was present in multiple areas of the body. PET-CT excluded the presence of malignant tumors in other regions.

Laboratory analysis indicated leukocytosis (18.01 × 10^9/L) and elevated C-reactive protein levels (16.2 mg/L). Abnormal liver function tests showed lactate dehydrogenase at 518 U/L, gamma-glutamyl transferase (γ-GT) at 61 U/L, alanine aminotransferase at 145 U/L, aspartate aminotransferase at 125 U/L, alkaline phosphatase at 124 U/L, and direct bilirubin at 8.5 μmol/L.

Pre - operative discussion

Following a multidisciplinary discussion and considering the large size of the cardiac tumor, which almost entirely occupied the left and right atria and compromised cardiac circulation, it was decided to proceed with emergency surgery.

Anesthesia

Upon the patient's arrival in the operating room, oxygen was administered via a face mask, and continuous monitoring of blood oxygen saturation (SpO2), non-invasive blood pressure, and a five-lead electrocardiogram (ECG) was initiated. Initial readings were as follows: heart rate at 100 beats per minute, blood pressure at 90/69 mmHg, respiratory rate at 20 breaths per minute, and SpO_2 at 94% with oxygen inhalation at a flow rate of 5 L/min. Intravenous access was secured through the median cubital vein. Under local anesthesia with 2% lidocaine, radial artery puncture and catheterization were performed to facilitate invasive blood pressure monitoring,

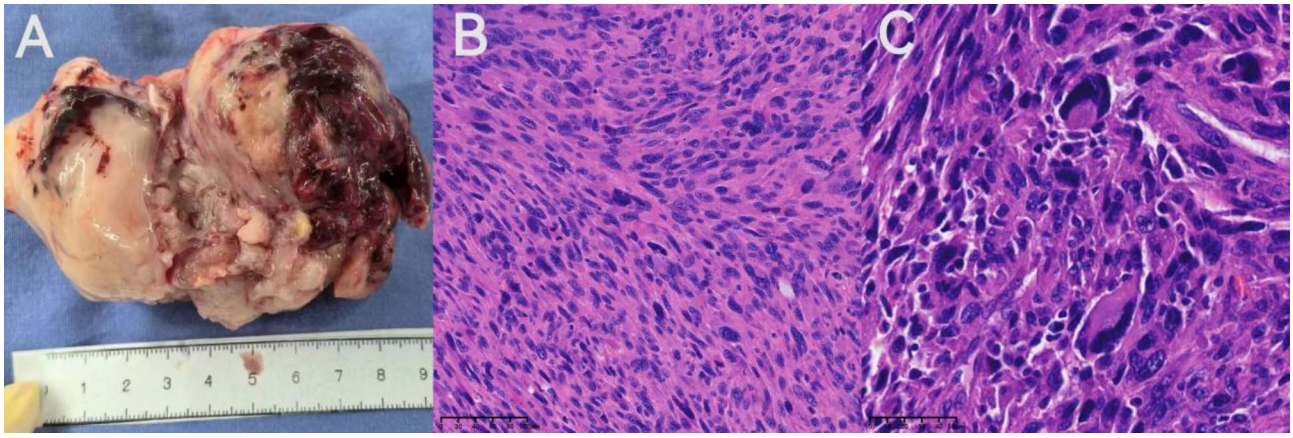

Fig. 3 Macroscopic and microscopic images. **A:** Macroscopic imaging of the tumor. **B:** Tumor cells displayed an ovoid to short-spindle morphology, with focal significant pleomorphism, high proliferative activity, frequent mitotic figures, and eosinophilic cytoplasm (×100). C: Tumor cells exhibited marked pleomorphism and atypia, with prominent nucleoli (×400)

resulting in a reading of 102/78 mmHg. Due to the tumor obstructing the superior vena cava, femoral vein puncture and catheterization were also carried out. The induction of anesthesia was postponed until both the surgical and extracorporeal circulation teams were fully prepared, enabling immediate response to any severe hemodynamic instability or cardiac arrest by performing thoracotomy and initiating extracorporeal circulation.

For the induction of anesthesia, etomidate at a dosage of 0.3 mg/kg, rocuronium at 0.9 mg/kg, and sufentanil at 0.5 μg/kg were administered intravenously in a slow and controlled manner. Tracheal intubation was performed orally under direct visualization. Mechanical ventilation was set to a volume-control mode, with a respiratory rate of 10 to 12 breaths per minute, a tidal volume of 8 to 10 mL/kg, and an oxygen concentration ranging from 50 to 80%. Adjustments to ventilation volume and frequency were made based on blood gas analysis results. Hemodynamic stability was maintained using agents such as norepinephrine, phenylephrine, and dobutamine. Continuous monitoring of invasive arterial pressure, ECG, SpO2, and body temperature was conducted throughout the anesthesia, with arterial blood gas measurements taken intermittently during the procedure. Transesophageal echocardiography revealed solid occupying echoes in the bilateral atrial cavities and the interatrial septum, measuring approximately 88 × 58 × 82 mm. Color Doppler Flow Imaging demonstrated punctate blood flow signals within the mass, with its lower edge extending to the mitral and tricuspid valve orifices.

Intraoperative findings

The patient underwent emergency surgical intervention under cardiopulmonary bypass. Operative exploration revealed that the base of the tumor was located on the lateral wall of the left atrium, invading the entire interatrial septum, part of the roof of the left atrium, and the anterior wall of the right atrium, without involving the mitral and tricuspid valves. The tumor was surgically resected (Fig. 3A). Additionally, due to moderate tricuspid regurgitation, a tricuspid annuloplasty was also performed. The excised tumor was sent to the pathology department for diagnostic confirmation. Transesophageal echocardiography revealed no significant abnormal echoes within the atria. An echo consistent with a patch was observed on the interatrial septum, and color Doppler imaging confirmed the absence of any residual shunt. Following tricuspid valvuloplasty, the tricuspid valve morphology appeared normal, although mild tricuspid regurgitation was detected via color Doppler. The duration of aortic cross-clamping and cardiopulmonary bypass were 77 min and 162 min, respectively. Following the successful completion of the surgical procedure, the patient was transferred to the intensive care unit for further monitoring and care.

Histological findings

After the surgery, the excised atrial mass underwent further histopathological examination. The histological diagnosis confirmed the mass as a mesenchymal-derived malignant tumor. Gross pathological examination showed the tumor as a gray-white to gray-brown mass, measuring 9 × 6.5 × 5.5 cm (Fig. 3A). Microscopic analysis demonstrated that the tumor cells had an ovoid to short-spindle shape, with some cells showing significant pleomorphism (Fig. 3B, C).

Immunohistochemical and molecular testing yielded the following results: Desmin, MyoD1, and Myogenin were negative, ruling out rhabdomyosarcoma. S-100 and SOX10 were negative, excluding neurogenic tumors and malignant melanomas. CD34, CD31, and ERG were negative, ruling out angiosarcoma. TLE1, EMA, and Bcl-2

were negative, and SS18 gene separation was negative (via FISH), excluding synovial sarcoma. Desmin, Calponin, and Caldesmon were negative, ruling out leiomyosarcoma. S-100, CDK4, and MDM2 were negative (assessed through IHC and FISH), with sporadic positivity in P16, excluding liposarcoma. ALK {5A4} was negative, ruling out inflammatory myofibroblastic tumor. Calretinin was negative, excluding cardiac myxoma. The absence of INI-1 deletion ruled out SMARCB1-mutated tumors, and the absence of Brg1 deletion ruled out SMARC4-mutated tumors. CK {pan} was present in small amounts and positive, SMA was partially positive, suggesting exclusion of epithelial tumors. P16 showed weak positivity in scattered areas, Ki-67 showed 30% positivity, and MSA was focally positive [3].

According to the literature, primary cardiac undifferentiated pleomorphic sarcoma is associated with a TP53 mutation [4]. Next-generation sequencing (NGS) results identified a class II TP53p. (W146*) mutation, consistent with previous findings. Based on the histopathological morphology observed in H&E staining, immunohistochemical profiling, FISH, and NGS results, the tumor was diagnosed as primary cardiac pleomorphic undifferentiated sarcoma, classified as pT2N0M0, stage IB.

Postoperative evolution

A postoperative transthoracic echocardiogram showed no masses in the bilateral atria. However, a reduction in right ventricular systolic function and mild tricuspid regurgitation was noted. On the third postoperative day, the patient was transferred from the Intensive Care Unit to the general ward and was discharged on the 17th postoperative day without complications. She was subsequently referred to the oncology department for further management. Between September 6, 2024, and December 26, 2024, the patient underwent six cycles of postoperative adjuvant chemotherapy using the IA regimen. This included ifosfamide at a dosage of 2.4 g (1.8 g/m^2) administered on days 1 to 5, and liposomal doxorubicin at a dosage of 40 mg on day 1, with cycles repeated every three weeks. Six months postoperatively, a follow-up echocardiogram showed no evidence of tumor recurrence.

Discussion

Primary malignant cardiac tumors are exceedingly rare neoplasms that originate from various cellular tissues within the heart. Among these, primary cardiac sarcomas constitute approximately 20% of cases [5]. Sarcomas are a category of tumors that predominantly arise from mesenchymal tissues. Cardiac undifferentiated pleomorphic sarcoma is a highly malignant mesenchymal tumor of the heart, characterized by a lack of specific histological morphology, ultrastructural features, or distinct immunohistochemical markers [6]. This tumor type is notably aggressive. According to the literature, undifferentiated pleomorphic sarcoma is most frequently located in the left atrium with no sex predilection [7]. The clinical presentation is largely determined by the tumor's size, location, and mobility, with symptoms primarily resulting from varying degrees of intracardiac obstruction and embolism due to tumor detachment. Common clinical manifestations include dyspnea, chest pain, heart failure, pericardial effusion, vena cava obstruction, and pulmonary embolism. In the early stages of the disease, the absence of hemodynamic disturbances, such as obstruction, results in a lack of overt symptoms, which can lead to the condition being easily overlooked. Given the extreme rarity of undifferentiated pleomorphic sarcoma, available data are scarce, and the efficacy of current treatments remains uncertain. Extensive surgical resection is the primary therapeutic approach for cardiac tumors, as complete tumor removal can alleviate obstruction and enhance hemodynamic function. Adjuvant radiotherapy or chemotherapy post-surgery may extend survival [8].

The anesthetic risks for patients with undifferentiated pleomorphic sarcoma are predominantly associated with factors including preoperative congestive heart failure, tumor size, the extent of atrioventricular valve obstruction, arrhythmias, and shock. In this case, the cardiac tumor occupied nearly the entire space of the left and right atria, substantially reducing the blood volume within the cardiac chambers. Additionally, the potential movement of the tumor between the atria and ventricles could obstruct the atrioventricular valve orifice, leading to rapid hemodynamic fluctuations and severe outcomes such as heart failure and sudden death. The detachment of the tumor may result in complications like cerebral infarction, pulmonary embolism, pulmonary edema, and infarction of limbs or organs. Consequently, the anesthetic management of such patients necessitates specialized considerations [9].

Prior to surgery, it is imperative to comprehensively understand the patient's condition, with particular emphasis on the correlation between their body position and symptoms, as well as their habitual posture. During the transportation and movement of the patient, abrupt changes in body position should be avoided to prevent the atrial tumor from becoming lodged in the atrioventricular channel due to positional alterations, which could result in significant hemodynamic instability and potentially lead to low cardiac output or sudden death [10]. Preoperatively, the patient effectively alleviated symptoms such as chest tightness and shortness of breath by assuming a semi-Fowler's position. This position was maintained during the transfer from the ward to the operating room and throughout the anesthesia induction phase. In cases where a cardiac cavity tumor

is diagnosed and impacts the patient's hemodynamics, urgent surgical intervention is warranted, necessitating rigorous anesthesia evaluation and meticulous preoperative preparation.

The induction of anesthesia should be conducted with utmost smoothness. It is imperative to select anesthetic agents that possess substantial analgesic effects while minimizing impacts on cardiopulmonary function in order to mitigate adverse reactions such as breath-holding, coughing, muscle fasciculations, and significant fluctuations in heart rate and blood pressure. In this particular patient, the obstruction of the superior vena cava by the tumor has led to congestion in the pulmonary venous system, potentially delaying the effects of intravenous anesthetics. Administering these drugs in small, incremental doses can effectively prevent circulatory disturbances associated with overdose. During the adjustment of the patient's surgical position following anesthesia induction, movements should be executed with care and stability. In the event of an abrupt decline in blood pressure and a marked reduction in heart rate, it should be considered that the tumor may be obstructing the atrioventricular valve orifice. Under such circumstances, the patient should be promptly repositioned into the right lateral decubitus position to alleviate the obstruction, and extracorporeal circulation should be established without delay.

Before initiating extracorporeal circulation, it is imperative to minimize the load on the right heart, keeping the heart rate at or slightly below preoperative levels. In this patient, a combination of intravenous and inhalation general anesthesia was used, primarily utilizing sufentanil. Sufentanil has an analgesic potency 5 to 10 times greater than that of fentanyl and has minimal effects on the cardiovascular system [11]. It effectively stabilizes hemodynamic fluctuations associated with tracheal intubation and surgical interventions. The underlying mechanism may involve the reduction of systemic vascular resistance and the inhibition of baroreceptor sensitivity, thereby reducing cardiovascular impact during general anesthesia [12]. Additionally, given that the tumor has invaded the inter-atrial septum and partial resection was necessary, there is a potential risk of atrioventricular block or severe arrhythmia. Consequently, it is crucial to closely monitor cardiac rhythm alterations following the resumption of cardiac activity.

Conclusions

The diagnosis of undifferentiated pleomorphic sarcoma is typically one of exclusion [13]. When diagnosing a cardiac tumor that affects the patient's hemodynamics, emergency surgical intervention becomes crucial. This requires rigorous anesthesia assessment and meticulous preoperative preparation. During anesthesia induction, maintaining stability, considering the effects of body positioning, and keeping the heart rate constant or slightly reduced are essential to stabilize hemodynamics, thereby ensuring patient safety throughout the perioperative period. Combination therapy, consisting of surgical resection and postoperative chemotherapy, has been associated with improved survival in patients with sarcoma [14].

Abbreviations

CW	Continuous Wave
PET-CT	Positron Emission Tomography-Computed Tomography
NGS	Next-generation sequencing

Supplementary Information

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Supplementary Material 1

Supplementary Material 2

Author contributions

YJ. W, MY. L wrote the main manuscript text and LJ. L, MJ. W prepared Figs. 1, 2 and 3. H. Z, C. L reviewed the manuscript. All authors read and approved the final manuscript.

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Data availability

No datasets were generated or analysed during the current study.

Declarations

Consent for publication

The patient agreed the doctors could use and publish her disease related article with personal information deleted.

Competing interests

The authors declare no competing interests.

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