Right Atrial Fibrosarcoma Mimicking Myxoma with Intermittent Complete Atrioventricular Block in a Young Adult: A Case Report

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Abstract : *Primary cardiac tumors are rare and may be associated with nonspecific symptoms and cardiac conduction disturbances. Preoperative imaging may not reliably distinguish benign from malignant cardiac masses. A 19-year-old man presented with progressive dyspnea, fatigue, and diaphoresis. Electrocardiographic assessment identified first-degree atrioventricular (AV) block with intermittent complete AV block. Transthoracic echocardiography revealed a 2 × 2 cm right atrial mass with a left ventricular ejection fraction of 65%, and cardiac computed tomography raised suspicion of myxoma. While awaiting surgery, the patient received dopamine at 15 µg/kg/min and oxygen at 3 L/min by nasal cannula. He underwent tumor excision with cardiopulmonary bypass and permanent pacemaker implantation. Intraoperative ventricular fibrillation caused cardiac arrest, for which defibrillation and adrenaline were administered. Histopathology was reported as low-grade fibrosarcoma. After three days of intensive care, he was discharged in stable condition on postoperative day four. A right atrial tumor clinically suspected to be myxoma was associated with advanced intermittent AV conduction disease and was subsequently reported as low-grade fibrosarcoma. This case emphasizes cardiac imaging, rhythm surveillance, histopathological assessment, and careful interpretation of perioperative outcomes.*

Keywords : *Right Atrial Tumor; Cardiac Myxoma; Cardiac Fibrosarcoma; Atrioventricular Block; Permanent Pacemaker; Ventricular Fibrillation*

Abstrak : Tumor primer jantung merupakan kondisi langka yang dapat menimbulkan gejala tidak spesifik dan gangguan konduksi jantung. Pemeriksaan pencitraan praoperatif belum tentu dapat membedakan massa jantung jinak dan ganas secara akurat. Seorang laki-laki berusia 19 tahun datang dengan sesak napas progresif, kelelahan, dan diaforesis. Pemeriksaan elektrokardiografi menunjukkan blok atrioventrikular (AV) derajat satu disertai episode intermiten blok AV total. Ekokardiografi transtorakal menunjukkan massa atrium kanan berukuran 2 × 2 cm dengan fraksi ejeksi ventrikel kiri 65%, sedangkan tomografi komputer jantung mengarah pada dugaan miksoma. Selama menunggu operasi, pasien mendapat infus dopamin 15 µg/kg/menit dan oksigen 3 L/menit melalui kanula nasal. Pasien menjalani eksisi tumor menggunakan pintasan jantung-paru serta implantasi alat pacu jantung permanen.

Fibrilasi ventrikel intraoperatif menyebabkan henti jantung yang ditangani dengan defibrilasi dan adrenalin. Pemeriksaan histopatologi melaporkan fibrosarkoma derajat rendah. Setelah tiga hari perawatan intensif, pasien dipulangkan dalam kondisi stabil pada hari keempat pascaoperasi. Tumor atrium kanan yang awalnya diduga miksoma disertai gangguan konduksi AV intermiten derajat lanjut, tetapi secara histopatologis dilaporkan sebagai fibrosarkoma derajat rendah. Kasus ini menekankan pentingnya pencitraan jantung, pemantauan irama, pemeriksaan histopatologi, dan evaluasi luaran perioperatif secara cermat.

Kata Kunci : Tumor Atrium Kanan; Miksoma Jantung; Fibrosarkoma Jantung; Blok Atrioventrikular; Alat Pacu Jantung Permanen; Fibrilasi Ventrikel.

INTRODUCTION

Primary cardiac tumours are relatively uncommon in clinical practice, and their manifestations vary according to tumour location, size, and biological characteristics. Myxomas are among the most common benign primary cardiac tumours, whereas primary malignant mesenchymal tumours are considerably rarer (Reynen, 1995; Silverman, 1980). Clinical manifestations may include intracardiac blood flow obstruction, embolic events, constitutional symptoms, and cardiac electrical conduction disturbances. Non-specific symptoms may complicate early diagnosis, particularly in young patients without a previous history of cardiac disease. Therefore, identifying the clinical characteristics and performing appropriate investigations are essential for establishing an accurate diagnosis and selecting suitable management strategies.

Echocardiography plays a key role in the initial assessment of intracardiac masses by evaluating their location, size, mobility, and haemodynamic effects. Cardiac computed tomography and magnetic resonance imaging provide additional information regarding tumour anatomy and tissue characteristics. However, imaging does not always reliably distinguish benign from malignant tumours, making histopathological examination necessary for definitive diagnosis (Angeli et al., 2024; Kabashi et al., 2013). In addition to structural manifestations, cardiac tumours may be associated with atrioventricular (AV) conduction disturbances, although these are relatively uncommon. Several case reports have described associations between right atrial masses and AV block, including advanced-grade and complete AV block (Davila, 2014; Yamada et al., 2022). A systematic review of right atrial myxomas also identified AV block as one of the less frequently reported electrocardiographic abnormalities (Rabiee Rad et al., 2024). These findings indicate that conduction disturbances should be considered when evaluating patients with intracardiac masses, without assuming a causal relationship in the absence of sufficient clinical evidence.

Diagnostic challenges arise when imaging characteristics are inconsistent with histopathological findings or when the accompanying clinical manifestations are atypical. The combination of a right atrial tumour, first-degree AV block, and intermittent episodes of complete AV block in a young adult represents a clinical presentation requiring comprehensive evaluation. Furthermore, an initial diagnosis of myxoma does not necessarily reflect the actual biological characteristics of the tumour. Although imaging supports diagnostic assessment and surgical planning, definitive tumour classification requires tissue confirmation. Conduction disturbances accompanying a cardiac mass must also be interpreted cautiously, as the anatomical relationship between the tumour and conduction system, reversible causes, and mechanisms underlying rhythm disturbances cannot always be established from initial investigations (Roberts & Ramsdale, 1990; Angeli et al., 2024).

In response to these diagnostic challenges, this case report aims to describe the clinical presentation, electrocardiographic and imaging findings, surgical management, and histopathological results of a young adult with a right atrial mass initially suspected to be a myxoma, accompanied by first-degree AV block and intermittent complete AV block. The novelty of this report lies in

documenting this combination of findings alongside a histopathological diagnosis of low-grade fibrosarcoma, which differed from the preoperative suspicion. This report is expected to enrich the clinical documentation of uncommon presentations of primary cardiac tumours and emphasise the importance of integrating electrocardiography, imaging, histopathology, and postoperative monitoring. Accordingly, this case may inform clinical assessment of cardiac masses accompanied by conduction disturbances, particularly when initial investigations cannot definitively establish the tumour's biological nature.

METHODS

This study employed a descriptive case report design to document the clinical presentation, diagnostic evaluation, management, and histopathological findings of a 19-year-old man presenting with a right atrial mass and atrioventricular (AV) conduction abnormalities. Clinical information was obtained from the available medical records, including the patient's presenting symptoms, physical examination findings, laboratory investigations, electrocardiographic assessment, transthoracic echocardiography, cardiac computed tomography, operative records, and histopathological examination. The diagnostic assessment focused on characterising the intracardiac mass, identifying associated conduction disturbances, and evaluating the discrepancy between the preoperative imaging impression and the final histopathological diagnosis. The patient's clinical course was reviewed from initial hospital presentation through surgical intervention and postoperative care. The report describes the administration of supportive treatment, surgical tumour excision under cardiopulmonary bypass, permanent pacemaker implantation, management of the intraoperative ventricular fibrillation event, and subsequent clinical recovery. Histopathological findings were summarised from the available pathology report. No additional immunohistochemical or molecular findings were available for analysis. The clinical findings were interpreted descriptively, without statistical analysis or causal inference. Verbal consent for publication was obtained directly from the patient; however, written informed consent had not been obtained at the time of reporting. This limitation should be addressed in accordance with the target journal's publication ethics requirements before submission.

RESULTS AND DISCUSSION

Results

A 19-year-old man was referred to the emergency department with a two-day history of progressive shortness of breath, associated with fatigue and diaphoresis. He had no significant reported medical history. On admission, blood pressure was 109/64 mmHg, heart rate 56 beats/min, respiratory rate 22 breaths/min, temperature 36.7°C, and oxygen saturation 98% while receiving oxygen at 3 L/min via nasal cannula. All four extremities were cool. No peripheral edema, cyanosis, clubbing, jugular venous distension, carotid bruit, or cardiac murmur was reported. Breath sounds were vesicular without crackles or wheezing, and abdominal examination was unremarkable. Electrocardiographic assessment revealed first-degree AV block with intermittent episodes of complete AV block from the initial hospital presentation. Transthoracic echocardiography demonstrated a right atrial mass measuring approximately 2 × 2 cm, preserved left ventricular systolic function (ejection fraction 65%), and mild mitral regurgitation. Cardiac multislice computed tomography identified a right atrial tumor suspected to be a myxoma and described tricuspid insufficiency.

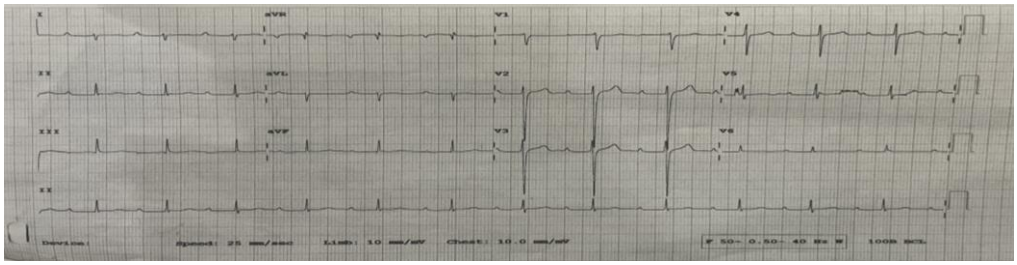


Figure 1. Twelve-lead electrocardiogram obtained during the initial hospital evaluation. Electrocardiographic findings were reported as first-degree atrioventricular block with intermittent episodes of complete atrioventricular block.

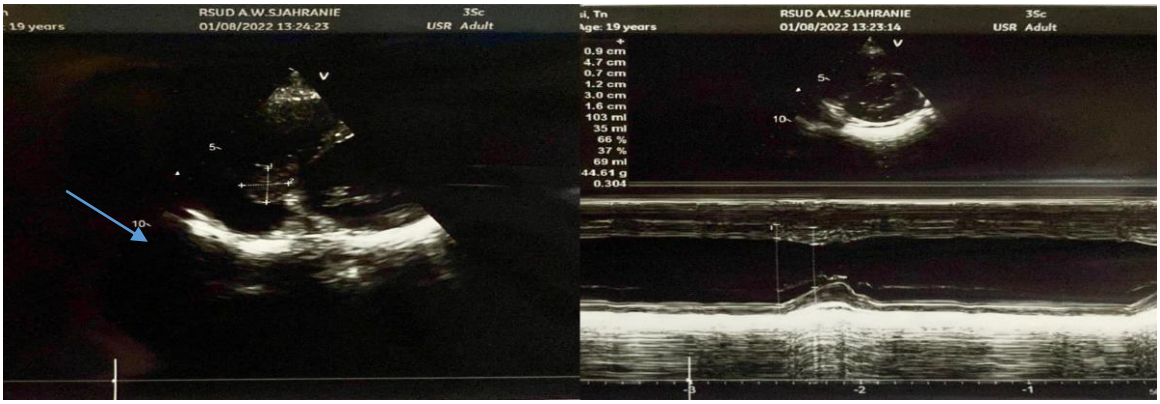


Figure 2. Transthoracic echocardiographic images obtained during the initial diagnostic evaluation. Echocardiography demonstrated a right atrial mass measuring approximately 2 × 2 cm, with preserved left ventricular systolic function (left ventricular ejection fraction, 65%).

Laboratory investigations revealed a leukocyte count of 19.15 × 10^3/µL, hemoglobin 10.4 g/dL, hematocrit 29.5%, and platelet count 194 × 10^3/µL. Sodium was 140 mmol/L, potassium 3.9 mmol/L, chloride 103 mmol/L, urea 34 mg/dL, creatinine 1.04 mg/dL, albumin 3.5 g/dL, calcium 8.8 mg/dL, and magnesium 2.1 mg/dL. The activated partial thromboplastin time was 30.0 seconds, prothrombin time 12.6 seconds, and international normalized ratio 1.17. While awaiting the scheduled operation, the patient received a dopamine infusion at 15 µg/kg/min and supplemental oxygen at 3 L/min through a nasal cannula. Surgical excision of the right atrial tumor was performed using cardiopulmonary bypass. During the operation, ventricular fibrillation resulted in cardiac arrest, and defibrillation and adrenaline were administered. Permanent pacemaker implantation was also performed. The available record did not establish the precise intraoperative phase of the arrhythmia.

Macroscopic examination of the excised tissue showed grayish, elastic fragments measuring 0.5–3 cm. Microscopic examination revealed solid proliferation of atypical fibroblastic mesenchymal cells with nuclear pleomorphism, vesicular nuclei, somewhat coarse chromatin, and inconspicuous nucleoli. The pathology report documented a nuclear atypia score of 1, mitotic activity of 5–10 mitoses per high-power field (reported score 2), and no tumor necrosis. The final pathology report described the lesion as low-grade fibrosarcoma of the right atrium. Immunohistochemical and molecular findings were not provided. The patient was monitored in the intensive care unit for three days. On postoperative day four, he was discharged in stable condition with blood pressure 112/66 mmHg, heart rate 94 beats/min, respiratory rate 20 breaths/min, temperature 36.0°C, and oxygen saturation 97% on room air. Discharge medications were oral paracetamol 500 mg four times daily, furosemide 40 mg once daily, and spironolactone 25 mg once daily. Postoperative ECG recordings and pacemaker interrogation results were unavailable; therefore, postoperative rhythm and device function could not be assessed.

Discussion

This case described a 19-year-old patient with a right atrial tumor who presented with first-degree atrioventricular (AV) block and intermittent complete AV block. Although preoperative imaging suggested a cardiac myxoma, histopathological examination of the resected tissue was reported as low-grade fibrosarcoma. The combination of a right atrial mass, significant conduction abnormalities, and an unexpected histopathological diagnosis made this case clinically noteworthy. Cardiac myxomas are commonly found in the atria and may cause dyspnea through intracardiac obstruction or altered cardiac filling (Muthiah, 2016; Roberts & Ramsdale, 1990; Willey et al., 1980). However, imaging findings alone cannot reliably establish tumor histology. Echocardiography is useful for evaluating the size, location, mobility, and hemodynamic effects of cardiac masses, while computed tomography and magnetic resonance imaging provide additional anatomical and tissue characterization (Angeli et al., 2024). In this patient, echocardiography demonstrated a 2 × 2 cm right atrial mass, and cardiac computed tomography initially suggested myxoma. The subsequent

histopathological diagnosis highlighted the limitations of imaging in distinguishing benign from malignant cardiac tumors.

The presence of first-degree AV block and intermittent complete AV block before surgery was a notable finding. Previous reports have described conduction disturbances associated with right atrial tumors. Roberts and Ramsdale reported first-degree AV block in a patient with right atrial myxoma, while Davila et al. described a right atrial lipoma associated with advanced AV block (Davila, 2014). Similarly, Yamada et al. reported complete AV block in a patient with a cardiac tumor involving the coronary sinus and right atrium (Yamada et al., 2022). A systematic review by Rabiee Rad et al. also identified AV block among the less frequently reported electrocardiographic abnormalities in patients with right atrial myxoma (Rabiee Rad et al., 2024). These findings suggest that the anatomical location of a right atrial mass may contribute to AV conduction disturbances, particularly when the tumor is situated near the interatrial septum or AV nodal region. However, the precise attachment site of the tumor and its relationship to the conduction system were not documented in this patient. Therefore, although tumor-related conduction disturbance was considered a possible explanation, a direct causal relationship could not be established.

The patient received dopamine infusion while awaiting surgery and subsequently underwent permanent pacemaker implantation. According to contemporary bradycardia guidelines, permanent pacing is recommended for acquired high-grade or third-degree AV block when reversible or physiological causes have been excluded (Kusumoto et al., 2019). In this case, intermittent complete AV block had been documented before surgery. However, the available records did not establish whether all potentially reversible causes had been excluded, and postoperative pacemaker interrogation results were unavailable. During surgery, the patient developed ventricular fibrillation resulting in cardiac arrest, which was managed with defibrillation and adrenaline administration. Ventricular arrhythmias during cardiac surgery may occur in association with myocardial manipulation, ischemia, electrolyte disturbances, or cardiopulmonary bypass. Nevertheless, the precise mechanism of ventricular fibrillation in this patient remained unknown. This intraoperative event was considered separately from the AV conduction abnormalities documented before surgery.

Histopathological examination revealed a proliferation of atypical fibroblastic mesenchymal cells with nuclear pleomorphism and mitotic activity, without tumor necrosis. The pathology report concluded that the lesion was low-grade fibrosarcoma. Primary cardiac fibrosarcoma is exceptionally uncommon (Kabashi et al., 2013). Although the diagnosis was established in the available pathology report, immunohistochemical and molecular findings were not documented, limiting further characterization of the tumor. Despite the intraoperative complication, the patient recovered sufficiently to be discharged in stable condition on postoperative day four. Given the reported malignant histology, continued multidisciplinary evaluation and longitudinal cardiac imaging would be important to assess postoperative outcomes and monitor for possible tumor recurrence.

Several limitations should be acknowledged. The original PR interval, rhythm strips documenting intermittent complete AV block, and detailed tumor attachment site were not available for independent review. The operative record did not specify the exact timing or mechanism of ventricular fibrillation beyond its intraoperative occurrence. Postoperative ECG and permanent pacemaker interrogation data were unavailable, precluding assessment of postoperative conduction status and device function. Although the pathology report concluded low-grade fibrosarcoma, immunohistochemical and molecular characterization were not documented. Resection margin status, staging investigations, and longer-term follow-up were also unavailable, limiting conclusions about oncological prognosis and recurrence. Finally, the observations from this single case could not establish a causal relationship between the right atrial tumor and AV block.

CONCLUSION

This case describes a 19-year-old man with a right atrial tumour presenting with first-degree atrioventricular (AV) block and intermittent complete AV block. Although preoperative cardiac imaging suggested a myxoma, histopathological examination identified the lesion as low-grade fibrosarcoma. The patient underwent surgical tumour excision and permanent pacemaker implantation following the documented conduction abnormalities and was discharged in a stable

condition on postoperative day four. This case highlights the limitations of imaging in determining tumour histology and the clinical relevance of investigating structural cardiac abnormalities in young patients presenting with unexplained conduction disturbances. However, a direct causal relationship between the tumour and AV block could not be established. Further clinical assessment should include postoperative electrocardiographic monitoring, pacemaker interrogation, follow-up cardiac imaging, and appropriate oncological surveillance to evaluate cardiac function and the risk of tumour recurrence. Future case reports and clinical studies should incorporate detailed tumour localisation, comprehensive histopathological and immunohistochemical characterisation, and longitudinal follow-up. These measures may improve understanding of the relationship between rare cardiac tumours and conduction abnormalities and support more comprehensive diagnostic and management strategies.

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