รายงานกรณีศึกษา: เนื้องอก Atrial Myxoma ขนาดใหญ่ในช่องหัวใจบนขวาเป็นผลให้เกิดลิ้นหัวใจไตรคัสปิดรั่วอย่างรุนแรง

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Case Report: A Huge Atrial Myxoma Located in the Right Atrial Chamber Caused Severe Tricuspid Valve Regurgitation

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Abstract

This case report documents a rare presentation of a primary cardiac tumor, known as myxoma, in the right atrium of a 32-year-old Thai woman with no underlying disease. Myxomas are typically found in the left atrium, making this case unusual. The patient presented with progressive chest pain and dyspnea on exertion. The investigation confirmed the presence of a large myxoma in the right atrium that was obstructive and caused severe tricuspid valve regurgitation and annulus dilation.

The patient underwent surgical treatment, where the mass was excised, and tricuspid valve replacement was carried out using a tissue valve due to the difficulty in repairing the damage caused by the myxoma. Postoperative follow-up showed a successful outcome, with good ejection fraction, tricuspid value function, and no chamber enlargement.

This case report also highlights the importance of prompt diagnosis and surgical intervention in cases of myxomas, especially when they are located in rare locations like the right atrium and are obstructive, causing severe valve damage. It also emphasized the significance of careful monitoring and follow-up after surgery to ensure the patient's full recovery. Overall, this report provides valuable insights into diagnosing and managing rare presentations of primary cardiac tumors like myxomas.

Keywords: Myxomas, Tricuspid valve replacement

บทคัดย่อ

รานงานกรณีศึกษาผู้ป่วยโรคเนื้องอกหัวใจ Cardiac Myxoma ซึ่งพบได้น้อยที่ห้องหัวใจด้านบนขวาของผู้ป่วยหญิงไทย อายุ 32 ปี ไม่มีโรคประจำตัว เพราะปกติโรคเนื้องอหัวใจ Cardiac Myxoma มักพบได้บ่อยที่ห้องหัวใจด้านบนซ้าย โดยผู้ป่วยมาพบแพทย์ด้วยอาการแน่นหน้าอกและหายใจหอบเหนื่อยมากขึ้น ผลการตรวจวินิจฉัยพบก้อนขนาดใหญ่ที่บริเวณห้องหัวใจด้านบนขวาที่ปิดกั้นทางเดินเลือดระหว่างห้องหัวใจด้านบนขวาและห้องหัวใจด้านล่างขวาและทำให้ลิ้นหัวใจไตรคัสปิดรั่วอย่างรุนแรงเนื่องจากทำให้เกิดขอบของลิ้นหัวใจไตรคัสปิดยืดออกมากกว่าปกติ

ผู้ป่วยได้รับการรักษาโดยการผ่าตัดเปิดกลางหน้าอก (open heart surgery) ด้วยเครื่องปอดและหัวใจเทียม (conventional cardiopulmonary bypass) ได้ทำการตัดก้อนออก การซ่อมลิ้นหัวใจในเคสนี้ยากลำบาก จึงทำให้ต้องผ่าตัดเพื่อเปลี่ยนลิ้นหัวใจไตรคัสปิดด้วยลิ้นเนื้อเยื่อหัวใจเทียม ผลการรักษาในผู้ป่วยรายนี้เป็นที่น่าพอใจ การติดตามการรักษาพบการบีบตัวของหัวใจอยู่ในเกณฑ์ปกติ ไม่พบก้อนในห้องหัวใจและการทำงานของลิ้นไตรคัสปิดที่เปลี่ยนดี และไม่มีห้องหัวใจใดโต

ในรายงานกรณีศึกษาผู้ป่วยในเคสนี้เน้นถึงความสำคัญของการวินิจฉัยที่รวดเร็ว และการผ่าตัดในผู้ป่วยที่วินิจฉัยว่าเป็นโรคเนื้องอกหัวใจ Cardiac Myxoma โดยเฉพาะอย่างยิ่งเมื่ออยู่ในตำแหน่งที่พบได้น้อย เช่น ห้องหัวใจด้านบนขวาและปิดกั้นทางเดินเลือดระหว่างห้องหัวใจด้านบนขวาและห้องหัวใจด้านล่างขวา และทำให้ลิ้นหัวใจไตรคัสปิดรั่วอย่างรุนแรง นอกจากนี้ยังเน้นย้ำถึงความสำคัญของการดูแลและติดตามการรักษาโดยการผ่าตัดและหลังผ่าตัดเพื่อให้ผู้ป่วยกลับมามีคุณภาพชีวิตได้ปกติและปลอดภัย ซึ่งโดยภาพรวมแล้ว รายงานกรณีศึกษานี้ให้ข้อมูลเชิงลึกที่มีคุณค่าในการวินิจฉัยและจัดการการนำเสนอที่เคสผู้ป่วยโรคเนื้องอกหัวใจ Cardiac Myxoma

Keywords: เนื้องอก cardiac myxoma, การเปลี่ยนลิ้นหัวใจไตรคัสปิด

Introduction

A 32-year-old Thai woman with no medical conditions presented with progressive chest pain and dyspnea on exertion over the past three months. She was not taking any medication. A cardiovascular examination revealed regular heart rhythm with pansystolic murmurs at the left parasternal border, and examination of other systems was normal.

Case presentation

On chest X-ray, pre-operative investigations showed clear lung fields with cardiomegaly (cardiothoracic ratio 0.73), globular heart shape, and biatrial enlargement. An electrocardiogram (ECG) showed sinus rhythm with 1st- degree atrioventricular (AV) block and right axis deviation with right bundle branch block and prolonged QT. A transthoracic echocardiogram revealed a large, heterogenous mass in the right atrium (RA) with a stalk attached to the lateral wall protruding into right ventricule (RV). There was increased RA and RV volume overload and a dilated inferior vena cava. Left ventricular ejection fraction (LVEF) was 71% (by 2D-Teichholz), without regional wall motion abnormalities. **(Figure 1)**

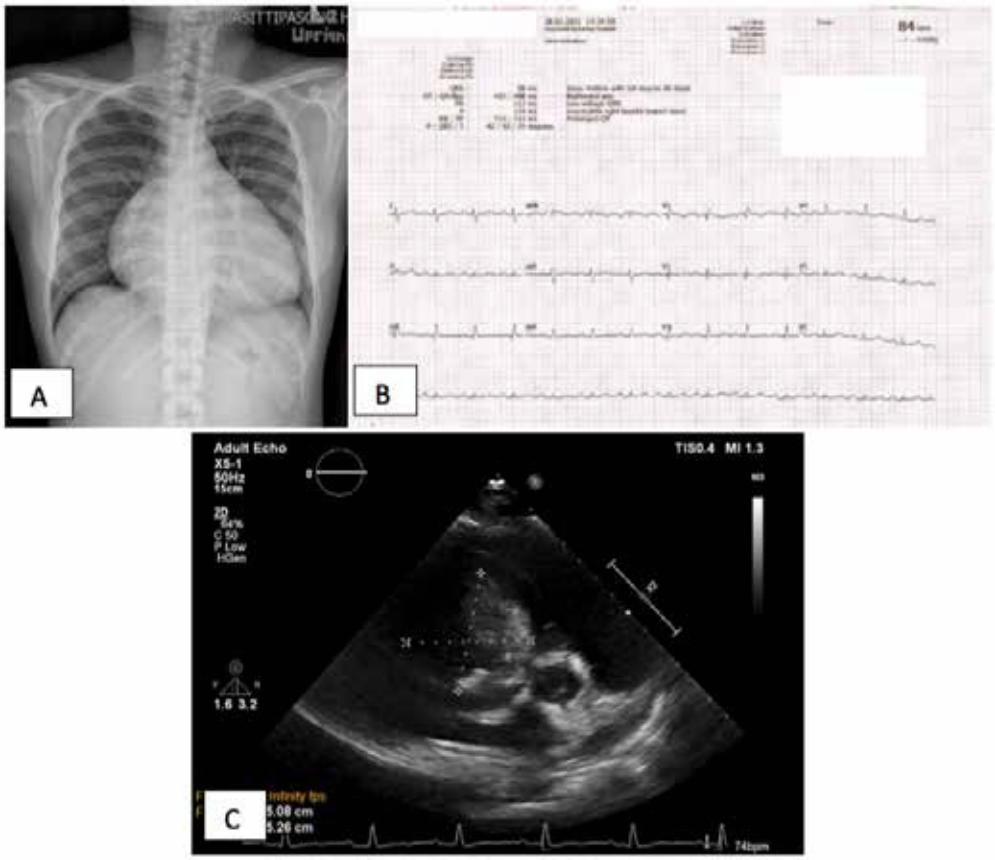

Figure 1: A: Chest X-ray, B: Electrocardiogram, C: Echocardiogram (Pre-Operative)

After providing written informed consent and undergoing pre-operative investigation, the patient underwent median sternotomy under general anesthesia with intra-operative transesophageal echocardiography. Aortic cannulation using EOPA® cannula, Bicaval venous cannulation was performed with an angle-tip cannula in the superior vena cava and a straight tip cannula in the inferior vena cava. The aorta was cross-clamped, then an antegrade cold cardioplegic solution, Custodiol®, was given intermittently with mild hypothermia (28.0°C). Cardiomegaly was observed due to biatrial and RV enlargement. Cardiopulmonary bypass (CPB) was established, and the RA mass was approached via the right longitudinal atriotomy.

The tumor was mobile, pedunculated, lobulated, transparent, and gelatinous liked, with implantation nearly at the opening of the inferior vena cava **(Figure 2)** and protruding into the tricuspid valve (TV). The mass was resected and sent it for histological assessment. The patient underwent TV replacement with a tissue valve due to severe tricuspid regurgitation and severe annulus dilatation. The RA was closed during rewarming, and de-airing was done routinely. The heart returned to sinus rhythm, CPB was discontinued without any problems, and the patient remained hemodynamically stable. The procedure was concluded in the usual fashion. Our patient was transferred to the intensive care unit (ICU) in good hemodynamic condition. Postoperative complications included atrial fibrillation, which was reversed with intravenous amiodarone, and the patient was discharged from the ICU on the third postoperative day and from the ward on the seventh postoperative day. Follow-up in the out patient units showed normal blood tests and imaging exams.

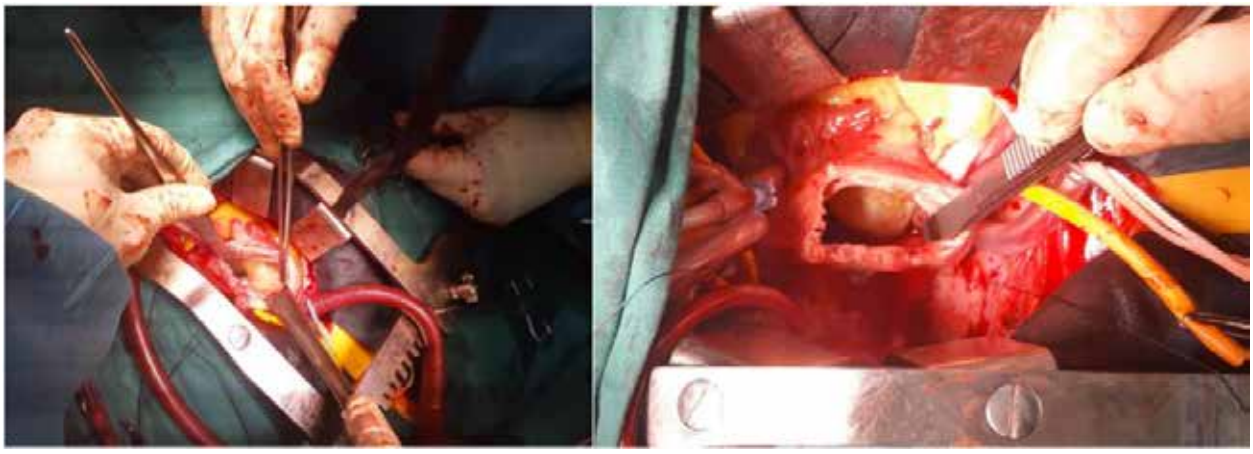

Figure 2: Intra-operative Right Atrial Mass

Macroscopically, the tumor presented as a 7x5x9 cm rubbery glistening tan dark gray mass weighing 120 gm **(Figure 3)**. Microscopically, the tumor was composed of elongated and fusiform myxoma cells with bland nuclei and slightly eosinophilic cytoplasm admixed with plasma cells and lymphocytes within the pale myxoid background. Some myxoma cells are arranged in single or multiple layers around vascular channels, forming a ring structure. There were few mitoses identified. A pathological examination confirmed the diagnosis of myxoma **(Figure 4)**.

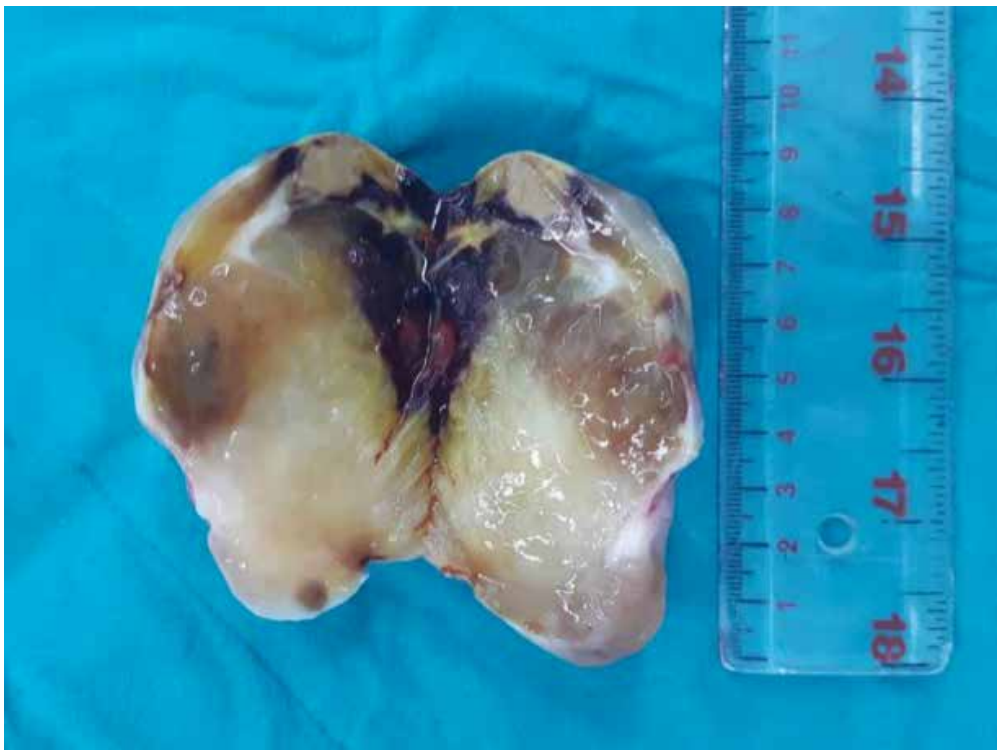

Figure 3: Macroscopic Tumor

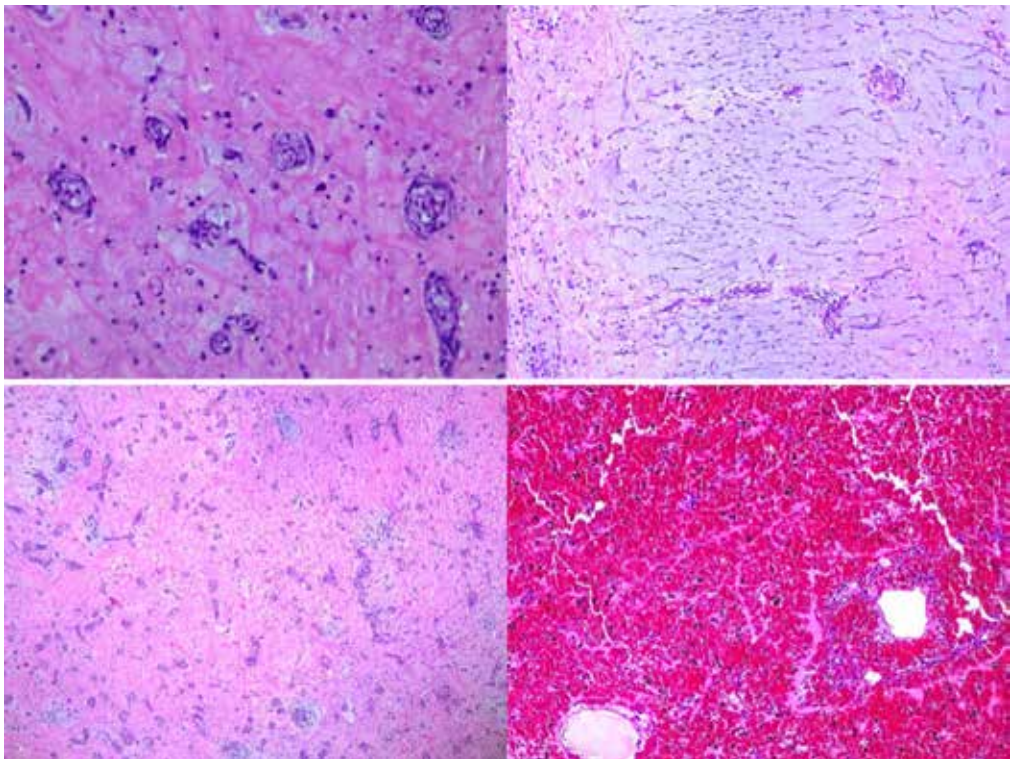

Figure 4: Microscopic Tumor

Six-month post-operative follow-up showed sinus rhythm with 1st degree AV block on ECG, clear lung fields with normal heart size (cardiothoracic ratio 0.48) on chest X-ray, and normal LV size, good LV contraction with LVEF of 80%, slightly dilated RA & RV, and normal RV systolic function on transthoracic echocardiography. There was no evidence of tumor recurrence or any mass or clot in the RA. The tricuspid bioprosthetic valve functioned well **(Figure 5)**.

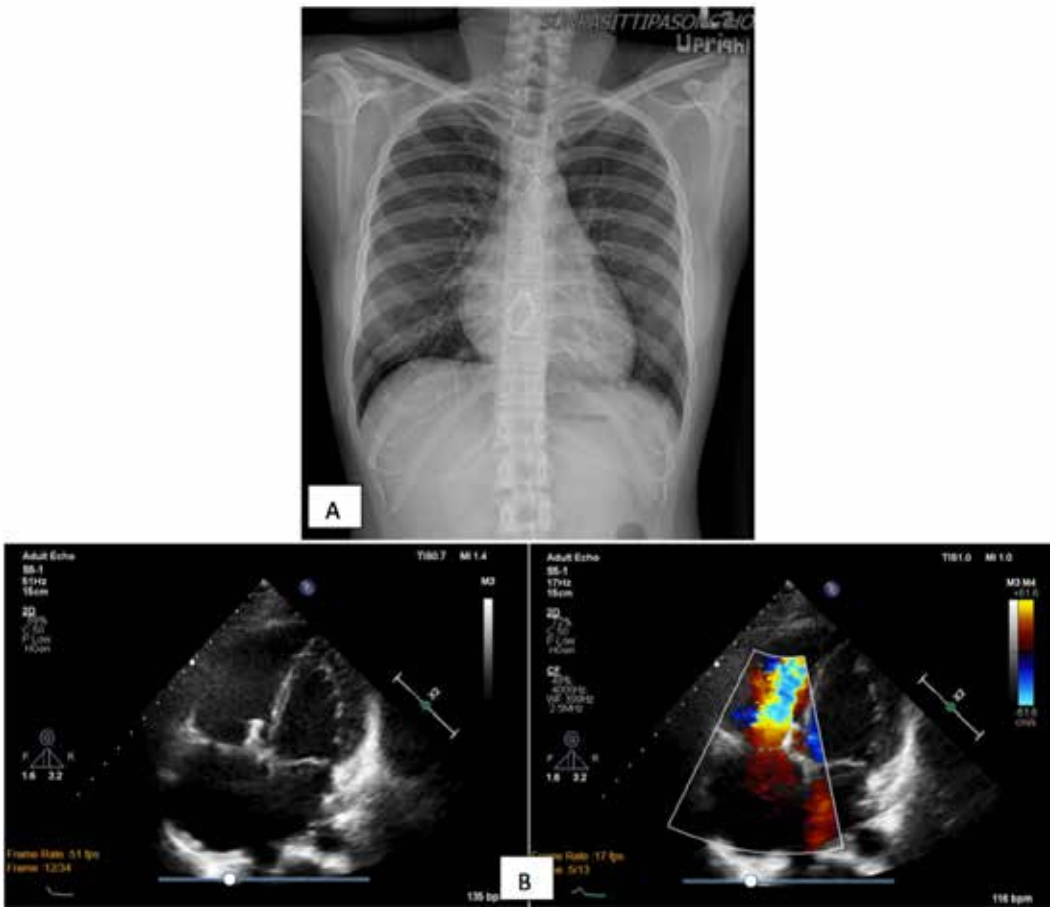

Figure 5: A: Chest X-ray (Post-Operative), B: Echocardiogram

Discussion

Primary tumors of the heart are exceedingly rare, occurring in only 0.0017% to 0.19% of unselected patients at autopsy,[1, 2, 3] with myxomas being the most common primary cardiac tumors. Although myxomas have been reported in all age groups and both sexes, they predominantly affect women in their third to sixth decade of life and are usually benign. These tumors most frequently occur in the atria, with around 75% arising in the left atrium (LA) and 15% to 20% in the RA. While most LA myxomas are located on the border of the fossa ovalis, they can originate from any location on the atrial wall. In comparison, myxomas arising from cardiac valves are rare.[4, 5]

The first successful excision of a LA myxoma was reported in 1955 by Carfoord C. It is crucial to perform surgical excision without delay once a cardiac myxoma is diagnosed due to the constant risk of thromboembolic events.[6, 7, 8] The surgical treatment for atrial myxoma is typically considered definitive, providing good outcomes, with a low risk of morbidity and mortality (from 0% to 3%). While recurrence of atrial myxomas is infrequent, sporadic myxomas can recur in approximately 1% to 3% of patients, usually occurring around 2.5 years after the initial surgery. However, the risk of recurrence is higher in the case of familial myxomas, ranging from 12% to 20%. Consequently, regular echocardiographic follow-up is strongly recommended for patients with familial myxomas. If a recurrence does occur, the myxoma should be surgically resected.[4]

In this case, the patient underwent TV replacement with a tissue valve due to severe tricuspid regurgitation, and severe annulus dilatation. The choice of a tissue valve was based on the current studies showing no significant hemodynamic differences between mechanical and tissue valves.[9] Additionally, using a tissue valve allows for a shorter duration of warfarin treatment, typically only three months. During the six-month post-operative follow-up, there was no evidence of tumor recurrence or the presence of any mass or clot in the RA. The

tricuspid bioprosthetic valve functioned well, and in line with standard practice for tissue valve follow-up, regular echocardiographic assessments are recommended at 12 to 24 months.

Conclusions

This case report highlights the rarity and potential severity of right atrial myxomas when they become large enough to protrude the tricuspid valve and cause tricuspid valve regurgitation. Early diagnosis and treatment are crucial to improving patient outcomes and preventing further complications.

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