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Left atrial myxoma – A case report

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Abstract

Primary cardiac tumors are rare, with an estimated incidence of 0.0017–0.3% in autopsy series. The most common primary cardiac tumor in adults is the left atrial myxoma, typically presenting as a benign neoplasm. Approximately 75% of myxomas arise in the left atrium, originating most often from the interatrial septum near the fossa ovalis. Clinical manifestations are varied and often non-specific, ranging from constitutional symptoms (fever, malaise, and weight loss) to obstructive symptoms (dyspnea and syncope) or embolic events (stroke). Surgical excision is the curative treatment.

Keywords: Left atrium, Myxoma, Cardiac tumors

1. Introduction

Atrial myxoma is the most common primary cardiac neoplasm. Over 72% of primary cardiac tumors are benign. In a 7% of cases, the tumor has a genetic origin and arises as a component of a heritable disorder with some clinical manifestations. The prevalence of cardiac tumors at autopsy is 0.001–0.3%, of which more than 50% are myxoma.[1-3]

These are benign neoplasms arising from primitive multipotent mesenchymal cells. Two-thirds of Carney complex-associated cardiac myxomas exhibit mutations in PRKAR1A. PRKAR1A mutations occur in both familial and sporadic forms of computer numerical control but have not been described in isolated (non-syndromic) cardiac myxomas. Although sporadic myxomas do not show consistent genetic alterations, familial syndromes associated with myxomas have activating mutations in the *GNAS1* gene or null mutations in PRKAR1A.[4]

2. Case representation

A 55-year-old female patient came with complaints of multiple episodes of headache and dyspnea for the past 1 week. She had a history of weight loss. There was no history of cough, syncope, or fever. Her vitals and other laboratory investigation parameters were within normal limits. The X-ray showed mild cardiomegaly. Echocardiography showed a left ventricular ejection fraction of approximately 60% with atrial myxoma, likely with pulmonary valve regurgitation and pulmonary arterial hypertension (PAH). It is highly suggestive of a mass obstructing flow in the left atrium, commonly seen with a large myxoma, which can lead to increased left atrial (LA) pressure, causing dyspnea and eventually pulmonary hypertension.

3. Pathological findings

The patient underwent surgical excision of the mass. The pathology findings are as follows:

3.1. Gross examination

The images [Figures 1 and 2] show fragments of resected tissue, consistent with the gross appearance of a myxoma. Excision of the LA mass was done and received as a tissue fragment altogether measuring 5 × 3 × 2 cm. The surface appeared firm, grayish to brownish. The cut surface showed hemorrhagic, friable, with a variegated area with a gelatinous appearance. The fragments were relatively large, with one measuring approximately 4 cm along the ruler. The surface appears irregular, a characteristic of the papillary/villous type of myxoma, which is often associated with a higher risk of embolism.

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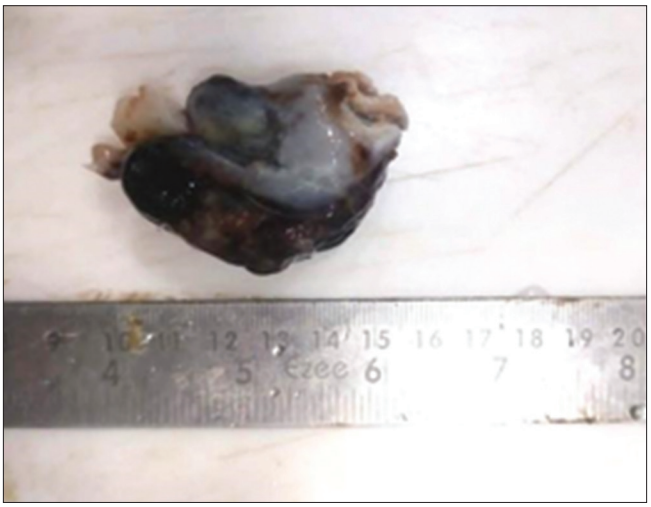

Figure 1: Gray-brown to blackish soft-tissue pieces 3 × 5 cm soft tissue

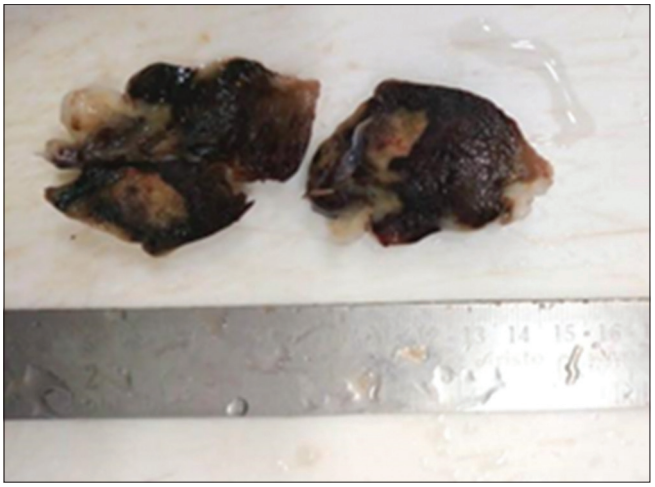

Figure 2: Gelatinous, myxoid areas. Patchy hemorrhage, soft friable consistency

3.2. Microscopic examination

The hematoxylin and eosin-stained slides [Figures 3 and 4] reveal the classic histological features of an atrial myxoma:

(i). Cells: Sparse, bland-looking myxoma cells (often called lepidic cells) scattered within the myxoid background. These cells have a characteristic stellate (star-shaped), polygonal, or fusiform (spindle-shaped) morphology with pale cytoplasm and bland nuclei. They are often seen in small nests or cords

(ii). Background: Abundant, loose, pale-staining, myxoid (gelatinous) stroma, which is the defining feature of the tumor

(iii). Vascularity: Numerous, often thin-walled blood vessels, which are a common finding

(iv). Other features: Areas of hemorrhage and hemosiderin-laden macrophages (seen as brown/yellow pigment) are present, which are also typical of this tumor type. No evidence of significant nuclear atypia or mitotic activity is noted, confirming the benign nature

(v). Special stains: Alcian blue [Figures 5 and 6] showed blue staining of background acid mucopolysaccharide.

4. Discussion

The most common primary tumor of the heart is the cardiac myxoma, which frequently occurs in the left atrium. The tumor may cause symptoms of LA obstruction and systemic embolization.[5] Myxomas occur most often in patients aged 30–70, with female predominance, and in families with a tendency to develop myxomas.[6] About 85% of myxomas occur in the left atrium, 10% in the right atrium, and 5%in the ventricles.[7] Atrial myxomas produce three types of clinical presentation: obstructive, constitutional, and embolic. Obstruction mimics mitral and tricuspid valve disease and occurs due to blockage of the atrioventricular valves. The tumor can mimic mitral stenosis, and occasionally an early diastolic rumbling sound called "tumor plop" may be heard during auscultation. Diastolic murmurs are the most common murmur heard in up to 89% of

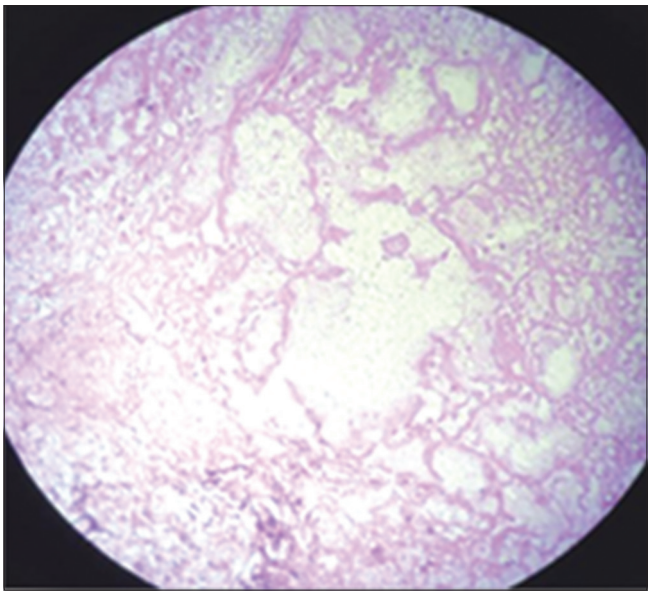

Figure 3: Abundant myxoid stroma

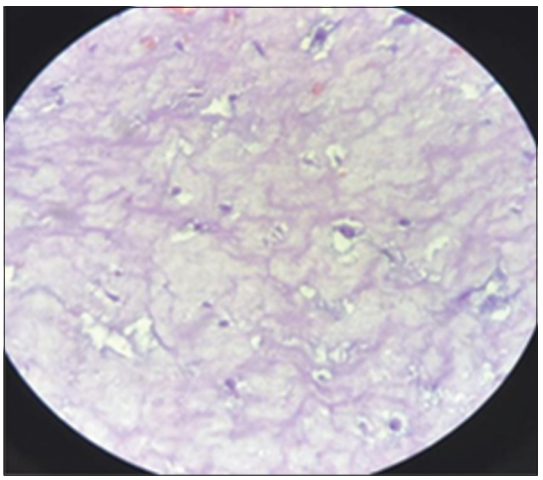

Figure 4: Scattered stellate/spindle cells

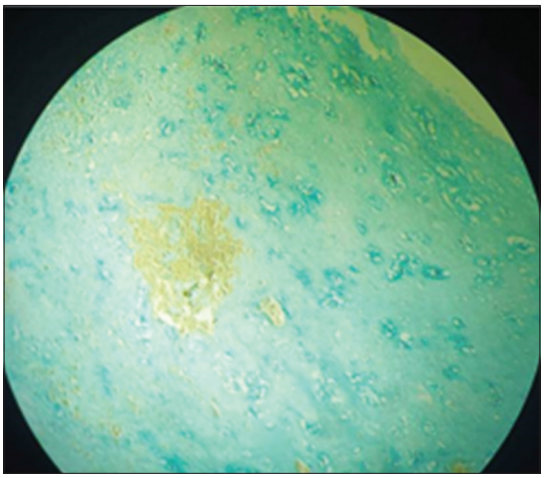

Figure 5: Alcian blue positive

patients with cardiac myxoma. The patient presented with classic symptoms (dyspnea and headache) and echocardiographic findings consistent with a LA myxoma, a diagnosis confirmed by both the gross and microscopic pathology. The myxoma's

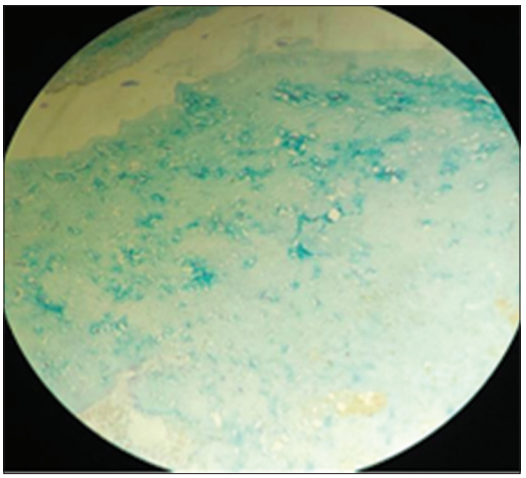

Figure 6: Alcian blue positive

obstructive effect on the mitral valve inflow tract is indicated by the finding of mitral stenosis function (MSF) and resultant PAH on the echocardiogram.

The gross appearance of the resected tissue (irregular and friable) suggests a papillary morphology, which makes it prone to fragmentation and subsequent systemic embolization (explaining the patient's headache, a potential sign of cerebral embolism).

Treatment for myxoma is prompt surgical excision to relieve obstruction and prevent systemic emboli. The low rate of recurrence and generally excellent prognosis post-surgery make this a highly treatable condition. Follow-up echocardiography is essential to monitor for potential recurrence.

5. Conclusion

This case details the presentation, diagnosis, and surgical pathology of a large LA myxoma in a 55-year-old female. The case highlights the importance of echocardiography in the initial diagnosis and the classical features of the tumor on gross and histopathological examination. Surgical resection remains the gold standard treatment.

6. Source of funding

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7. Conflicts of interest

There are no conflicts of interest.

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