Atrial Myxomas: A Case Report on Rare Benign Cardiac Tumour and its Histopathological Diagnosis

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Abstract

Cardiac tumours are a very rare entity of tumours which comprises of both malignant and benign group. The atrial myxomas are benign cardiac tumour which occurs most commonly. This case report illustrates a diagnosis of rare atrial myxoma through its characteristic signs, symptoms, most common site of occurrence and histopathological examination assisted by radiological examination.

The diagnosis is made by classical clinical presentation, histopathological examination along with 2D echocardiography to rule out the malignant activity. The clinical signs and symptoms vary with the location and size of tumour, which usually present as pulpy non-specific mass.

The histopathological study corresponded with classical pattern. Since the tumour may present as an emergency in the form of symptoms mimicking sudden cardiac death and other complications like embolization, prompt resection of tumour is the mainstay treatment associated with low mortality. However, the most common complication is recurrences hence the patient should be kept in regular follow up.

Keywords*: Atrial Myxoma, Histopathological examination, Benign tumour.*

Introduction

Primary cardiac tumours are rare. The incidence accounts for about 0.3% of patients with cardiac surgery. In cardiac tumours, the cardiac myxomas are most common accounting for about 50%.[1]Although the prevalence of cardiac tumours only with respect to autopsy is known which ranges from 0.001% to 0.3%, of more than 50% of benign cardiac tumours are myxomas. It has genetic origin in seven per cent of cases and rises as a component of a heritable disorder with some clinical manifestations. Over 72% of primary cardiac tumours are benign. In adults, the majority of benign lesions are myxomas.[2,3] Most common origin of cardiac myxomas is from the left atrium (about 75%) and less commonly the right atrium (18%), right ventricle accounts for (three per cent) and also very rarely from the left ventricle (three per cent). A small fraction of less than one per cent arises from the valves. Because of the non-specific symptoms, early diagnosis of cardiac myxomas can always be challenging.

Case History: A 42-year-old male patient was presented with complaints of shortness of breath associated with cough and pedal oedema for two months. His physical examination, on admission for cardiac surgery, revealed normal vital sign and neurological examination was normal. On gross examination, specimen was a reddish-brown jelly like pulp aggregating 16×2×1.5cm. A small whitish flap structure measured 1.5cm, as shown in Figure 1.

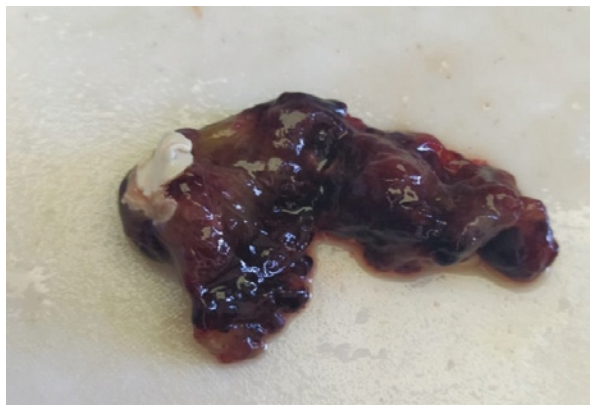

Figure 1: Reddish mass measuring 16×2×1.5cm

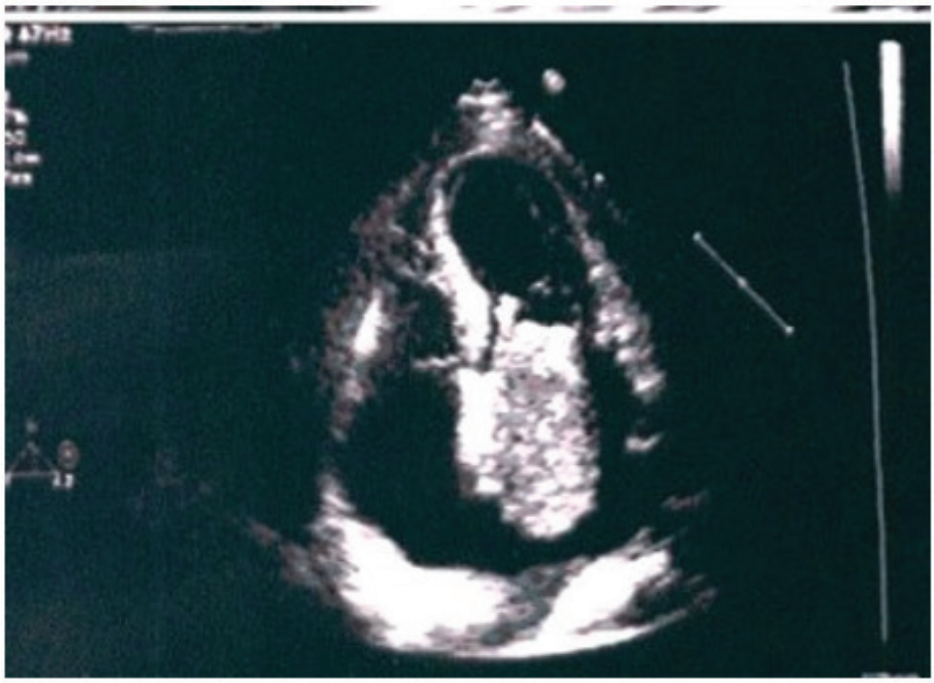

Figure 2: *2-D echocardiography revealed a left atrial tumour with no associated thrombus.*

[Figure3.jpg]

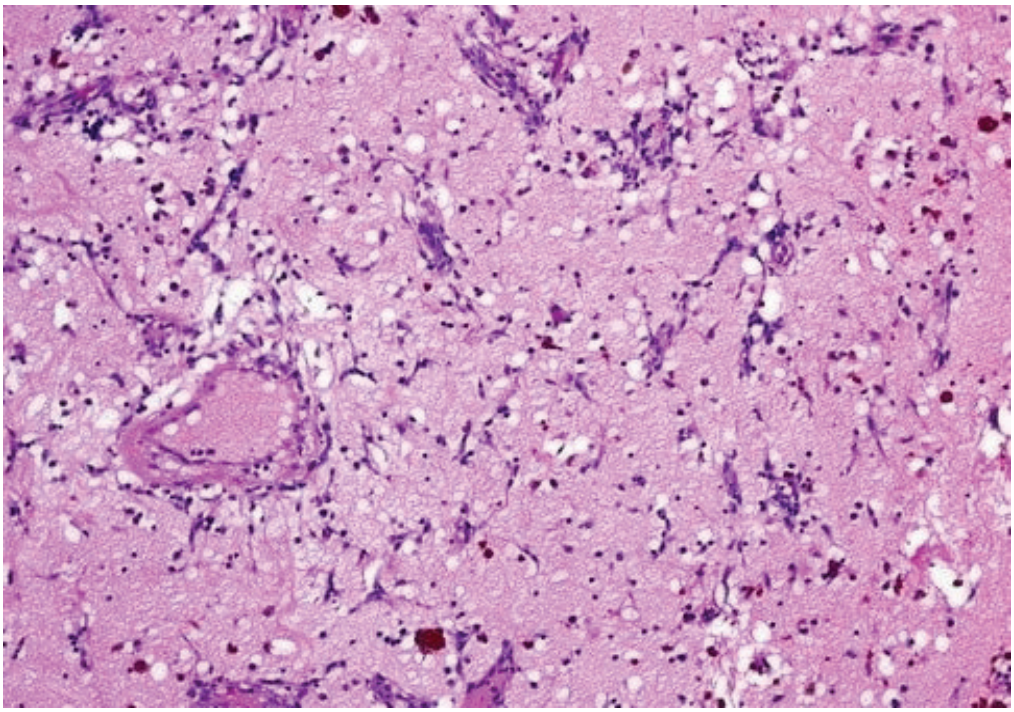

Figure 3: *The section shows stellate cells against abundant myxoidstroma. There is presence of hemosiderin laden macrophages along with delicate blood vessels.*

Discussion

With a rare incidence of 0.002% in general population, cardiac myxoma is the most common heart tumour, representing 50% of all cardiac tumours, and about 75% of them are in the left atrium.[5]

In our case as well, tumour arose from left atrium**.** Left atrial myxomas produce symptoms when they reach a weight of about 70 grams, right atrial myxomas to be symptomatic it can attain a weight of twice that of left atrial growth. Tumours size varies from 1 centimetre to 15 centimetres in diameter.[6]In our case, tumour size was 16 centimetres. The origin of myxoma is thought to originate from embryonic foregut, which is entrapped, therefore they arise from multipotentmesenchymal cells capable of both epithelial and neural differentiation.[7,8] Histologically it consists of mucopolysaccharidestroma containing tumour cells. Myxomas produce Vascular Endothelial Growth Factor (VEGF), which probably is responsible for angiogenesis and the early stages of tumour growth.[7,8]

The signs and symptoms depend upon the tumour size and involved heart chamber. Blood flow obstruction within the heart can produce heart murmurs and its associated clinical manifestations.[9]

Grossly, cardiac myxomas are polypoidal, round, or oval may be often pedunculated and soft gelatinous in consistency along with a smooth, villous, or friable surface.[10] Commonly observed symptoms and signs are dyspnoea, orthopnoea, paroxysmal nocturnal dyspnoea, pulmonary oedema, cough, haemoptysis, oedema, and fatigue.[11] On histopathological examination, tumour cells were shaped stellate, spindle, oval or round. The nucleus was shaped cylindrical or oval. There was no mitotic activity. The cytoplasm was stained pink. The pink or light blue staining myxoid material was filled with the tumour cells.[12]

Williams et al. (1970) demonstrated that myxoma cells produce a tumour stroma through its secretory function. They further concluded that stromal material is synthesized in the endoplasmic reticulum and Golgi zone which are known to be the site of acid mucopolysaccharide and glycoproteins production.[13,14,15]

In our case, classical histopathological features like background stroma of myxoid material and tumour cells in the form of stellate cells were observed.

For initial diagnostic evaluation, echocardiography is a simple, non-invasive, and largely available technique. With echocardiography, myocardium and cardiac chambers can be visualized which generally shows a presence of mass. In addition, echocardiography gives an indication of any other obstructive pathology which could be a source of emboli.[16]

Therefore, from the signs, symptoms, 2D echo, gross and histopathological examination, we concluded it to be a case of atrial myxoma.

Vaideeswa et al. (2012) analysed 20 patients of non-typical myxoma characterized by multi focal presentation, of whom four cases had recurrence, suggesting that young people with multiple lesions or family history are more likely to relapse. Such patients should always receive routine echocardiography examination throughout their life.[17]As risk factors for myxoma includes embolization including sudden cardiac death; prompt resection of tumour is advised once it has been initially diagnosed. The result of resection generally has a very good prognosis of under five per cent mortality rate.[18]

Conclusion

With the annual incidence of less than one per cent, atrial myxomas are one of the common benign cardiac tumours arising from most commonly the left atrium. The correct diagnoses of atrial myxomas by the aid of histopathological, radiological mean are necessary as it may be life threatening if misdiagnosed as malignancy or for that matter also benign and not followed up correctly.

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Conflict of Interest: Nil

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