

Case Reports

Large left atrial myxoma: Thorough clinical evaluation is the key

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Introduction

Cardiac tumour was first described by Columbus in 1562¹, but identifying a tumour in a living patient was documented in 1934². The first successful excision of cardiac myxoma was performed in 1955³. Paediatric cardiac tumour is a rare entity⁴. Most of them are benign in nature, but malignant and secondary tumours are also encountered very rarely⁴. Clinical presentation mainly depends on the location of the tumour but the nonspecific nature of the presentation makes it a diagnostic challenge⁴. This case report emphasises the importance of clinical evaluation when encountering a patient having nonspecific symptoms which cannot be explained by the clinical findings.

Case report

A 13-year old elder child of a healthy non consanguineous couple without a significant family history presented to medical attention following a syncopal attack while preparing for school. On further inquiry, she was having fainting episodes twice per month for the last 2 years, which

typically fits into a vasovagal syncope. She did not complain of palpitation or angina but was suffering from nonspecific headaches and muscle pains for last 8 months without any neurological sequel.

On examination, she was a healthy looking, adequately grown child without any dysmorphism. Her cardiovascular examination revealed a regular pulse rate and normal blood pressure without cardiomegaly. There was a mid-diastolic murmur best heard in the mitral area on auscultation suggesting an obstruction at the mitral level. Her chest x-ray (Figure 1) was normal. Her electrocardiogram (ECG) was normal without any rhythm abnormalities (Figure 2). Basic haematological and biochemical investigations, including haemoglobin and electrolytes, were normal.

The most striking finding on echocardiography was a large tumour attached to the interatrial septum prolapsing through the mitral valve (Figure 3).

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Figure 1: Chest x-ray of patient

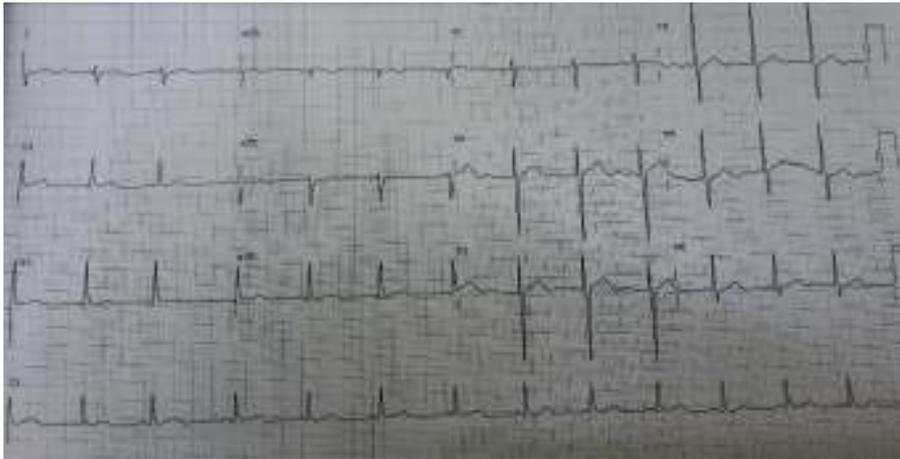


Figure 2: *Electrocardiogram of patient*

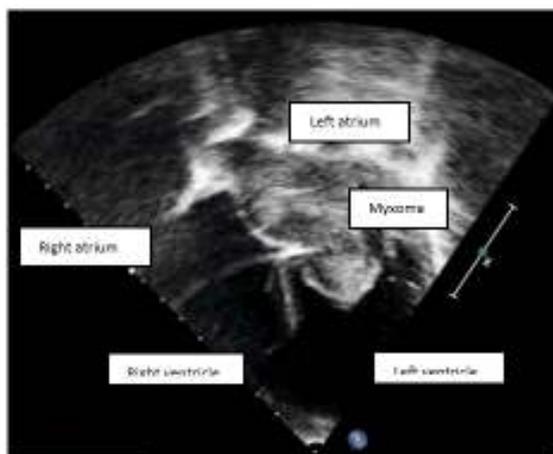


Figure 3: *Echocardiogram of patient*

Girl was taken for surgery and a very large, fragile myxoma, occupying the entire left atrium with a wide base from atrial septum was completely excised repairing the septum with pericardial patch closure. The sample was sent for histology, which confirmed atrial myxoma. Figure 4 is an echocardiogram subsequent to surgery.

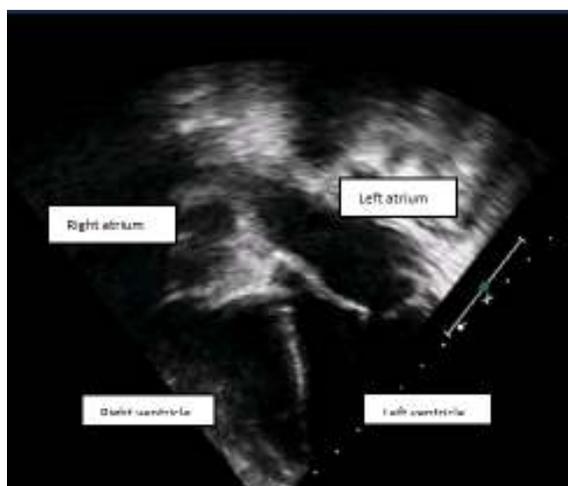


Figure 4: *Echocardiogram after surgery*

Discussion

General autopsy series revealed a cardiac tumour prevalence of 0.002 – 0.03%⁵ and autopsy series in children revealed an incidence of 0.027–0.08%⁶. However, echocardiographic databases including fetal studies, newborns, children and adolescence gave an incidence of 0.14 – 0.17%^{7,8}. According to previous autopsy reports, common primary cardiac tumours are rhabdomyoma (45%), fibroma (25%), myxoma (10%), intrapericardial teratoma (10%) and haemangioma (5%)⁴. Type of cardiac tumour incidence also varies with age, myxoma being the most prevalent tumour in adolescence⁴. Since the incidence of cardiac tumours is difficult to ascertain it is important to report every case encountered in clinical practice.

As clinical presentation varies markedly, investigations play a pivotal role in diagnosing cardiac tumours. Our patient had a normal ECG despite complaining of syncopal episodes. Echocardiography is the primary diagnostic modality which can also be used for follow up after the surgical excision⁹. It is noninvasive and accurate leading to the anatomic and haemodynamic diagnosis. Trans-oesophageal echocardiogram is useful in precise delineation of anatomy¹⁰. 3D echocardiography gives additional information such as tumour volume, precise relationship with adjacent structures and inflow/outflow tracts¹⁰.

Although magnetic resonance imaging (MRI) studies give superior details regarding the anatomy¹¹, we did not perform MRI in our child as we got enough information from the echocardiogram. Since the child was 13 years old she might not need general anaesthesia which is a major disadvantage in performing MRI. An important advantage of MRI is differentiation of the tumour type⁸, but in our scenario it was not an important concern since we decided to go ahead

with complete excision so that we can delineate the histology more precisely. Determining the tumour type is important in view of taking the management decision e.g. – conservative management in rhabdomyoma. On the other hand, detailed clinical evaluation will give insights to a correct diagnosis preventing an expensive study especially in developing countries. Tissue diagnosis is also possible using cardiac catheterisation, but with the significant risk of tumour embolisation¹². Therefore, histological assessment after the complete excision is justifiable.

Cardiac myxomas are commonly seen in the left atrium with a prevalence of 75% and they are friable, pedunculated and gelatinous tumours¹³. High calcification rates are seen in right sided tumours¹³, whereas our patient revealed no calcification. Malignant myxomas are rare, but can be identified by its histology, local invasion of primary site, regrowth of the tumour at the original site or different location and development of peripheral aneurysms¹⁴. Our patient did not reveal any of those features, but she needs regular follow up as there is a risk of recurrence. Clinical presentation of atrial myxoma can be due to cardiac obstruction, embolisation or systemic illness¹⁵. Unfortunately, most of them show nonspecific clinical features¹⁶ like in our patient. Therefore, late diagnosis is quite common. This patient's presentation could have been easily attributed to a vasovagal syncope, but thorough clinical examination paved the way to diagnosis emphasising the importance of clinical evaluation especially in a resource limited setting. Also, constitutional symptoms shown by the patient are described in the literature as a result of interleukin-6 secreted by the tumour¹⁶.

Critical obstruction and risk of systemic embolisation mandated early surgery in this large atrial myxoma. Entire myxoma was surgically excised and atrial septum was repaired successfully on the day after admission. Since familial occurrence is described in 7% of all myxomas¹⁶ echocardiographic examination was performed in parents and siblings and was found to be normal. Clinical and laboratory evaluation did not reveal any features suggestive of Carney complex¹⁷ in our patient. After successful surgery, patient is being followed up at our clinic paying attention to tumour recurrence and manifestations of a systemic illness. This case emphasises the importance of thorough clinical evaluation and a high index of suspicion in children with non-specific cardiac symptoms in resource limited setting.

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