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Left Atrial Myxoma in an adolescent male: A case report

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ABSTRACT: **Introduction**
Myxomas are the most common benign primary cardiac tumour in adult population. They have been found to be more frequent among females. They often present with cardiovascular, constitutional and embolic manifestations.
Here we present a case report in which left atrial myxoma was found in an adolescent male, a rare population group to be affected. Males more frequently were found to present with embolic manifestation but here this boy presented with constitutional symptoms.
Conclusion : An early echocardiographic examination played an important role in diagnosis and management of this patient. Thus emphasizing the importance of a proper clinical examination in every patient and an early echocardiographic examination of any patient presenting with fever, other constitutional symptoms & cardiac murmur
KEYWORDS: Benign primary cardiac tumour, myxoma, left atrial myxoma, adolescent male, constitutional symptoms.

I. INTRODUCTION

Primary cardiac tumours are extremely rare (1) and 75% of primary cardiac tumours are benign(2-7).The most common benign primary cardiac tumour in adults is Myxoma and other common benign lesions include papillary fibroelastomas and lipomas.

Myxomas Histologically, are composed of scattered cells within a mucopolysaccharide stroma. The cells originate from a multipotent mesenchyme that is capable of neural and endothelial differentiation(8).Myxomas produce vascular endothelial growth factor(VEGF), which probably contributes to the induction of angiogenesis and the early stages of tumour growth(9,10)

In children, rhabdomyoma and fibromas are the most common tumour(11) and myxomas are rarely seen, accounting for only 9-15% of all cardiac tumours from birth to adolescence.

Myxomas have an autosomal dominant transmission and more commonly affect females.

Cardiac tumours may cause symptoms through a variety of mechanism like – Embolization, obstruction of circulation through the valves, causing regurgitation because of interference with valve structure & function, direct invasion of myocardium, resulting in impaired left ventricular function, arrhythmias, heart block, or pericardial effusion with or without Tamponade; invasion of lung causingpulmonic symptoms, mimicking bronchogenic carcinoma(12); constitutional symptoms

The cardiovascular manifestations like symptoms of mitral valve obstruction, depends upon the anatomic location of the tumour. Approximately 80% of myxomas originate in the left atrium and the most of the rest is found in right atrium.(13-17)

Constitutional symptoms like fever, weight loss and laboratory abnormalities (anemia, raised—ESR , CRP or globulin level) suggest presence of a connective tissue disease and is frequently found in patients with myxoma usually attributed to production of various cytokines and growth factors by the tumour.
Here we present a case of Left Atrial Myxoma in a 14 year old boy who presented with fever and shortness of breath.

II. CASE REPORT

A 14 year old malefrom Patna, Eastern part of India came to Medicine OPD, AIIMS Patna with complaints of fever for 3 months with progressive dyspnoea on exertion.
He was apparently well 3 months back when he developed sudden onset high grade fever associated with chills & body ache. Fever was continuous relieved with medicine only for few hours but no documentation of temperature was done. There was an associated dyspnoea on exertion (NYHA CLASS III)
Past medical history of tubercular lymphadenitis at 6 years of age and has taken full course of anti-tubercular drugs. No other significant history of illness in past.
No history of any significant illness in past.

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On examination he was febrile , had pallor , no cyanosis/ clubbing /icterus/ pedal edema.Temperature.-102℉, Pulse-112/min, Respiratory rate-24/min, BP-130/80 mm hg, JVP- raised, No organomegaly.

In cardiovascular system examination apex was not displaced, S1, S2 were normal, but a mid- diastolic murmur was heard. No opening snap/presystolic accentuation were appreciated.

Patient was admitted with a provisional diagnosis of RHD with MS, ?IE and Echocardiography(figure 1a & 1b) was planned along with other investigations.(Table 1)

ECG(figure 3) was suggestive of sinus tachycardia with P-pulmonale, while Chest X-Ray(figure 4) was within normal limits.

On echocardiography(figure 1a & 1b) there was a pedunculated mass of about 9.2 sq cm attached to inter-atrial septum & distal part of AML(Anterior mitral leaflet), in left atrium, bulging into left ventricle Suggestive of left atrial myxoma.

Presence of left atrial tumour was further confirmed on a cardiac MRI.(figure 2a-2d)

After about 3 months, surgery was done withgovernment grant as patient belonged to an economically weaker section.

He underwent left atrial myxoma removal and IAS reconstruction with Dacron patch.

The left atrial tissue was sent for histopathological examination where the tumour was confirmed to be myxoma.

As Myxoma needs prompt resection to prevent the risk of embolization or cardiovascular complications, including sudden death (16,21,23), patient was immediately referred to a higher centre with such facility.

After about one month of surgery patient came to us for follow up. He was asymptomatic and was doing well.

Table 1(Investigations)	
HEMOGRAM Hb(g/dl) MCV(fl)/MCH(pg)/MCHC(g/dl) TLC N/L/E/M (%) ESR	Suggestive of Microcytic anemia 7.9 81/23.7/29.3 8200/microL 62/26/4/8 70mm/hr
PBS	MICROCYTIC HYPOCHROMIC ANEMIA, malarial parasite not seen
ASO titre	210 IU/ml
RBS	120 mg/dl.
LFT Bilirubin (mg/dl) Total Direct Indirect SGPT/SGOT/ALP (U/L) TP/albumin/globulin (g/dl) A/G	 0.36 0.14 0.22 9/14/329 8.41/3.1/5.29 0.589
Blood urea (mg/dl) Creatinine (mg/dl)	28.8 1.05
Malarial antigen	NOT DETECTED
RK39	NEGATIVE
ECG	SINUS TACHYCARDIA,RAE (figure 3)
Blood C&S	NO GROWTH
USG abdomen	Suggestive of CYSTITIS
CHEST X-RAY	Within normal limits (figure 4)
Echocardiography	There was a pedunculated mass

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	of about 9.2 sq cm attached to inter-atrial septum & distal part of AML, in left atrium, bulging into left ventricle Findings suggestive of left atrial myxoma.(figure-1a & 1b)
Cardiac MRI	Signal suggestive of tumor. Left atrial tumour (figure 2a-2d)
Histopathology of exised left atrial tissue	Myxoma with cystic changes.

Figure 1a

Figure 1b

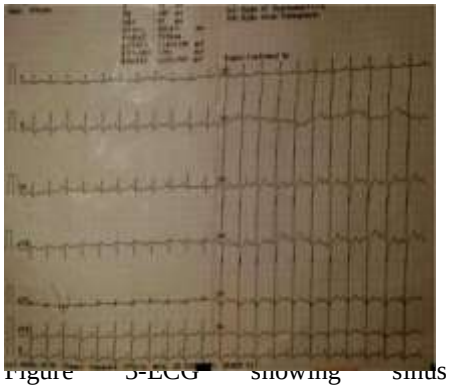

Figure 3-ECG showing sinus tachycardia, p-pulmonale

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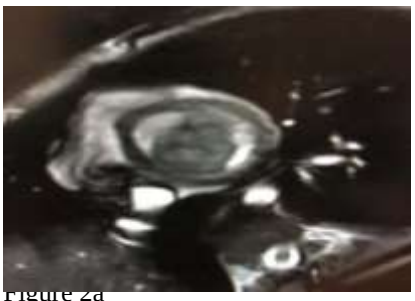

Figure 2a

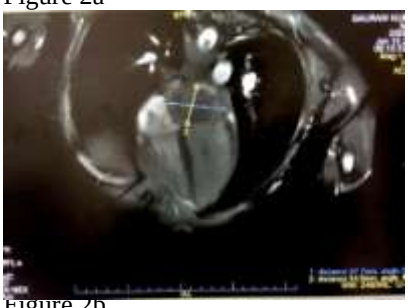

Figure 2b

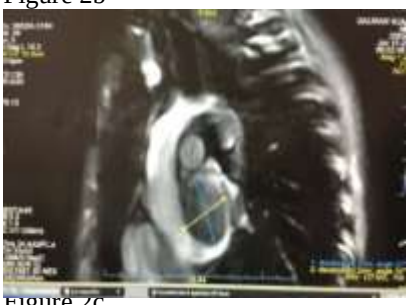

Figure 2c

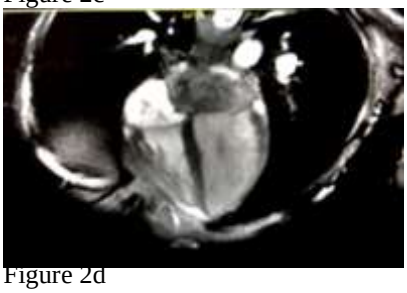

Figure 2d

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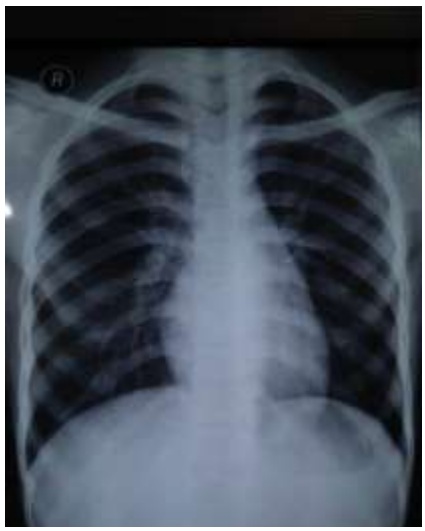

Figure 4- Chest X Ray PA view

III. DISCUSSION

Primary cardiac tumours being extremely rare(1) makes this a rare case. In one series of about 12,000 autopsies only seven were identified with an incidence of < 0.1% (18)

Myxoma is the most common benign primary cardiac tumour in adults, while in children, rhabdomyoma and fibromas are the most common tumour(11). In children myxomas are rarely seen, accounting for only 9-15% of all cardiac tumours from birth to adolescence.

In this case patient belongs to the adolescent age group where myxoma is rarely seen. Pinede L et al.(19) in a series of 112 patients found 72 women suggesting a female preponderance. Other large series of patients with myxomas , also found a predominance of women (60 to 70 percent) (14,16-17,20-21).

Myxoma being more frequent in female population and adult age group in most of the studies makes this case—a rare case presenting in a rare population group

Pinede L et al.(19) also illustrated different sign & symptoms(19) in his study— Cardiovascular symptoms were present in 67 percent. Most commonly, these resembled symptoms of mitral valve obstruction. Although auscultatory abnormalities were found in 64 percent, the classic tumour "plop" was identified in only 15 percent.

Constitutional symptoms (e.g., fever, weight loss) were seen in 34 percent of patients. Keeling IM et al.(16) also reported similar incidences of cardiovascular, embolic, and constitutional symptoms.

Cardiovascular symptoms of mitral valve obstruction were more frequent in the above studies. Similarly, symptoms and signs like exertional dyspnoea and a mid- diastolic murmur in our patient were most likely due to the obstruction of the mitral valve orifice by the tumour mass.

Embolization, usually systemic but can be pulmonic. Aortic valve and left atrial tumors were associated with greatest risk of embolization(22).

Pinede L et al. in the above series(19)found that evidence of systemic embolization was present in 29 percent of patients, and 20 percent had neurologic deficits. He also found that though myxomas were more common amongst women, men were more likely to have evidence of embolization.

Our patient being male with left atrial tumor was more likely to present with embolic manifestation but luckily there was no evidence of embolization.

Presence of dyspnoea on exertion in the background of predominant constitutional symptoms, like prolonged fever and laboratory anomalies like anemia, raised—ESR, presence of cardiac tumour could have been easily missed or its diagnosis delayed,but a proper clinical examination, revealing a mid-diastolic murmur and an early echocardiographic study saved the day.

The early echocardiographic examination played a very important role in diagnosis and management of this patient, emphasizing the need for proper clinical examination in every patient and an early echocardiographic examination of any patient presenting with fever, other constitutional symptoms & cardiac murmur.

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