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Left Atrial Myxoma: A Rare Tumor in a Child

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Abstract

Atrial myxomas are uncommon in the pediatric population but are very important due to pursuant atrioventricular valve obstruction and systemic embolic events. Concerning multiple symptoms ranging from congestive heart failure to neurological deficits, the diagnosis of this disease is complicated. This paper reports a case of left atrium myxoma in a child treated with a surgical excision of the tumor. Surgical excision appears to be curative, as shown in postoperative echocardiogram of this patient. To be precise, an immediate and timely diagnosis of myxoma by echocardiography, allowed the immediate surgical treatment of this rare, benign but potentially lethal cardiac tumor.

Key Words: Atrial myxomas, Child, Iran, Echocardiography.

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1- INTRODUCTION

Myxomas are the most common primary cardiac neoplasm; tumor usually grows in the left atrium (1-3). Myxomas mimic multiple cardiovascular diseases, however in about 20% of cases, myxomas are asymptomatic and are discovered as an incidental finding (4). Hence, cardiac myxomas are often misdiagnosed because of nonspecific symptoms (5). Patients with left atrial myxomas present symptoms when tumor reaches a weight of about 70g, tumors vary in size, ranging from 1–15 cm in diameter (4, 6). This tumor is more prevalent among women and the mean age of onset is between 30-60 years (7). Left myxoma is rarely seen in pediatrics but shows similar pathologic and clinical features (8). The most frequent cause in patients with left atrial myxomas is obstruction of mitral valve that may lead to syncope, so it is experienced by approximately 20% of patients (6). We reported a case of left atrium myxoma by unspecific presentation in a 5-year-old boy, finally treated by surgical removal.

2- CASE REPORTS

A 5-year-old boy presented with malaise, fatigue and weakness over a one-month period. A sudden syncope which lasted 20 minutes, led to emergent hospital admission. The patient was hospitalized after syncope in emergency room, where electrocardiogram showed sinus rhythm with depression of the ST segment and inversion of the T wave (**Figure.1**). Laboratory tests revealed the following results: WBC = 10.4 count/µl, Hb = 10.7 g/dl, PLT =30 000, ESR = 68 mm/h and urine test within normal limits. Biochemistry tests (BUN, Cr, Na, K, Ca, P) were normal but serum enzymes (AST=114 U/L, ALT =68 U/L) were elevated.

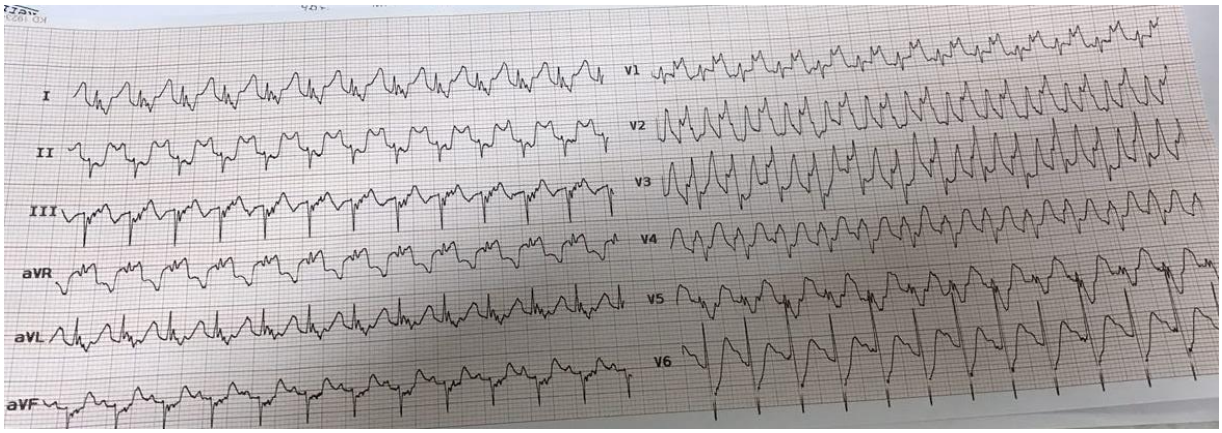

Fig.1: Electrocardiography showed depression of the ST segment and inversion of the T wave.

The patient had undergone resection of left atrial mass and septum interatrial through a median sternotomy approach, carefully. The intact atrial septal was closed and the patient was transferred to Intensive Care Unit (ICU). In macroscopic pathologic examination, the tumor was friable and jelly-like, its size was 6 × 5 × 2 cm^3 and weight was 25.3 g (**Figures 2 and 3**). Then the specimen was subjected to histopathological evaluation. Histological examination confirmed the macroscopic diagnosis of left myxoma and not thrombi. Follow-up echocardiogram after 12 months showed no recurrence of the tumor, and the patient had no symptoms after 12 months postoperatively (**Figure.4**).

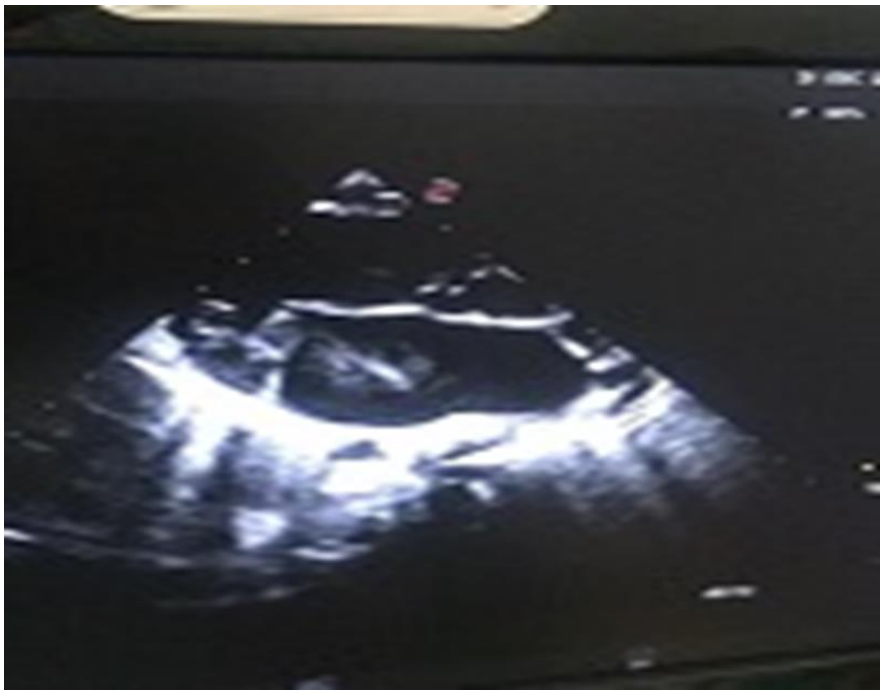

Fig.2: Echocardiography of a left atrial mass, attached to the atrial septum.

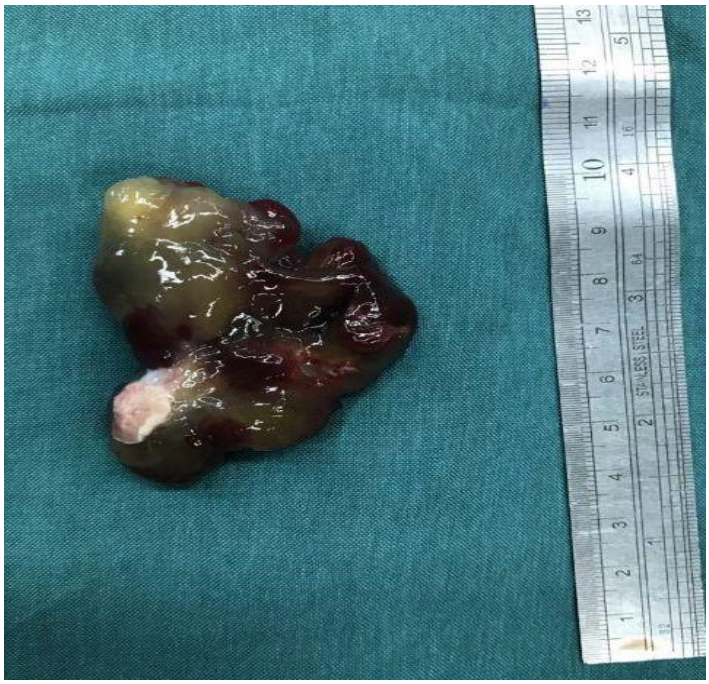

Fig.3: Size and appearance of atrial myxoma.

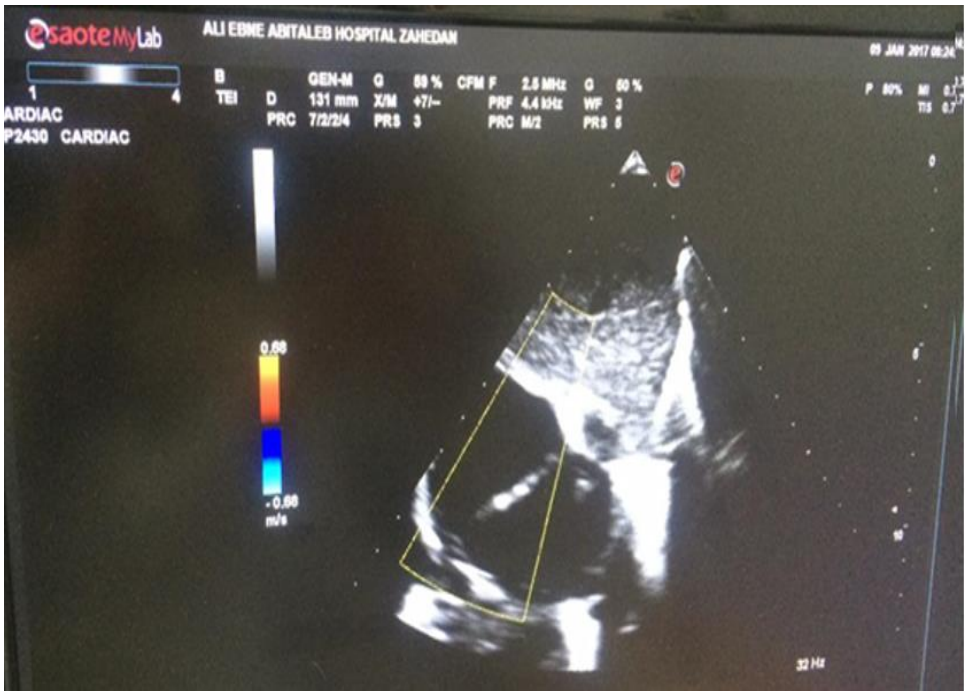

Fig.4: Postoperative Echocardiography.

3- DISCUSSION

According to the literatures, myxomas are non-cancerous primary tumors, frequently located in left atrium. Although left atrium myxoma is the most common tumor of the heart, it is rarely seen in the pediatric age group (9). Early diagnosis of myxoma in children and adults is very important, because it could be fatal if it is not surgically removed. Due to its many extra cardiac and confusing manifestations of disease, a careful investigation and suitable laboratory test (like anemia, thrombocytopenia, elevated erythrocyte sedimentation rate), and especially echocardiography are essential, both for differential diagnosis and for surgical planning (4). The diagnosis of myxoma in children is more complicated, because of the rarity of its occurrence in this setting. Therefore, in most cases the diagnosis occurs when children present with neurologic symptoms or with signs of embolization. Two-dimensional echocardiography is highly suggested for the diagnosis (10). Early diagnosis could mimic the chance of abnormalities related to embolization. Unlike medical treatment, surgical resection is the best choice for treatment of myxoma (11). The surgical specimen usually has an irregular frond-like gelatinous surface that should be completely and accurately removed (4). Bobo et al., presented recurrence of myxoma in a 15-year-old patient who underwent surgical procedure at 10 years of age (12). After a new resection, the patient did not experience disease recurrence over a 4-year follow-up period (12). Thus it is deduced from the evidence that children with known atrial myxomas must be followed closely, both for recurrence of the atrial tumor and for the delayed neurologic complications of aneurysm formation and hemorrhage (12). Careful and extensive surgical resection of the myxoma attached to atrial septum or atrial wall also can reduce the likelihood of recurrence in future (4). A case series by Al-Mateen et al., of children with cardiac myxomas causing cerebral emboli emphasizes the need for expedient diagnosis of cardiac myxomas; 6 out of 9 of these children had residual neurologic deficits and one died post-operatively (10).

This case emphasizes the importance of considering a cardiac myxoma in a patient with both systemic manifestations by unspecific presentations.

4- CONCLUSION

Atrium myxoma in pediatric ages is a rare complication; therefore, few studies have been performed in this field. Thus, differential diagnosis is essential in children, and echocardiography could help in this process. Once a differential diagnosis of myxoma has been done on imaging studies, prompt resection is necessary because of the risk of embolization or cardiovascular complications, including sudden death. Patients are at risk of recurrent myxoma; thus, long-term clinical and echocardiographic follow-up is mandatory. This tumor can be revealed by unspecific presentation especially in pediatrics group as we reported, and it is important to consider it as our differential diagnosis in a child with unusual signs and symptoms.

5- ABBREVIATIONS

ALT: Alanine transaminase;

AST: Aspartate transaminase;

CRP: C-reactive protein;

ESR: Erythrocyte sedimentation rate.

6- CONFLICT OF INTEREST: None.

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