Left Atrial Myxoma in a Seven Year Old Boy - A Case Report

ARUP DEB AJITBHUSANDEV
Department of Cardiology,
CHRISTIAN MEDICAL COLLEGE

Abstract:
Primary cardiac tumors of the heart are rare across all age groups. These tumors present with non specific symptoms and are great mimickers of various cardiac and extra cardiac diseases. Thereby, they are among most challenging disease entity to diagnose and the clinician should maintain a high index of suspicion to diagnose these tumors. Among primary cardiac tumors myxoma is the most common variety which commonly occur in Left Atrium. They commonly occur in middle aged Female patients with common age range is 30 to 50 years of age. We hereby, present the case of a seven year old male child who presented to us with fever of unknown origin along with easy fatigability. Clinical examination was positive for mitral area diastolic murmur and a diastolic heart sound. He was found to be anaemic with elevated acute phase reactants. His echocardiography revealed a large pedunculated mass arising from mid part of inter atrial septum protruding into left ventricle during diastole. He underwent complete surgical excision of the left atrial mass under cardiopulmonary bypass. Histopathology of the mass revealed it to be myxoma. Follow up echocardiography revealed no residua of the left atrial mass and symptomatic improvement clinically. The present case report demonstrates the presentation of Left Atrial myxoma with systemic non specific symptoms at an unusual early age. Hence, cardiac myxoma should always be kept in mind when dealing with fever of unknown origin even in pediatric population. Keyword: Left Atrial Myxoma, Male Child, Fever

Primary cardiac tumours are among the most challenging disease entities to diagnose due to their rarity and highly variable and non specific clinical presentation. Clinicians need to maintain a high index of suspicion for rare entities like primary cardiac tumors, especially when atypical features are present. A thorough history, clinical examination along with appropriate laboratory tests especially transthoracic or trans oesophageal echocardiography is of paramount importance. Primary cardiac tumors are great masqueraders of many commonly encountered cardiac and systemic
diseases. Depending on their size, location, mobility and friability, they produce myriad signs and symptoms. However, their overall presentations can be broadly divided into these following categories: systemic manifestations, embolic manifestations, cardiac manifestations. We hereby discuss a case of a seven-year-old boy who presented to our out patient department with fever for more than three weeks and shortness of breath.

CASE DESCRIPTION:
A seven-year-old boy presented with three weeks of fever and history of easy fatigability. He also complained of shortness of breath for the last 2 months for which he was not able to compete with his peers in sporting activities. He had an uneventful childhood prior to the present ailment. He was born by spontaneous vaginal delivery without any perinatal complications. He was second of two siblings, the elder brother was 12 years old and enjoying good health. On examination his height and weight were appropriate for his age. There was pallour without cyanosis, clubbing, icterus and lymphadenopathy. There were no skin pigmentation nor there were any evidence of neurocutaneous markers. His pulse rate was 96/minute, regular, normal in volume and character, all peripheral pulses were palpable with no radiofemoral delay. Blood pressure was 96/60 mmHg in both upper limbs and 106/60 mmHg in lower limbs. Jugular venous pulse was normal in pressure and waveforms. Cardiovascular system examination revealed apex beat localized at fifth intercostal space at mid clavicular line with no palpable precordial impulse or thrill, normal first and second heart sounds. There was an early diastolic sound followed by a low frequency diastolic murmur best heard at the apex in upright posture. His respiratory system examination was normal. Abdominal system examination did not reveal any hepatosplenomegaly or free fluid in the abdomen.

Baseline laboratory analysis revealed anaemia and elevated acute phase reactants. Hemoglobin 9.4 gm%, P.C.V 30.3%, WBC Count 18,500 cells/mm³, Platelet Count 1,53,000 cells/mm³, ESR 34 mm/hour, CRP 23.2 mg/litre, Creatinine 0.3 mg%, Fasting Glucose 80 mg/dl, TSH 2.773 iIU/ml, SGOT 37 U/L, Sodium 136 m mol/litre, Potassium 3.9 m mol/Litre.

ECG: showing sinus tachycardia.
CXR : Normal Cardiothoracic ratio, clear lung fields

ECHO PLAX view (mass in left atrium protruding into left ventricle)

ECHO (Apical Four Chamber view): Left Atrial mass attached to interatrial septum

ECHO Apical Four Chamber View (Post Surgery): No Residual Left Atrial mass

Histopathology of the Left Atrial Mass: spindle and stellate cells in abundant myxoid stroma

DISCUSSIONS:
Primary tumors of the heart are rare across all age groups with reported prevalence of 0.001% to 0.03%. Secondary involvement of the heart by extracardiac tumors are much more common than primary cardiac tumors. Among primary cardiac tumors, myxoma is the most commonest variety. (1) The diagnosis of primary cardiac tumor is frequently very challenging. The symptoms associated with primary cardiac tumors are non-specific and commonly mimic other more common systemic and cardiac conditions. The clinical manifestations that are produced by primary cardiac tumors overall can be divided into four general categories: systemic manifestations, embolic manifestations, cardiac manifestations and phenomenon secondary to metastatic diseases. Patients with primary cardiac tumors may experience constitutional symptoms of fever, chills, fatigue, malaise and weight loss. These symptoms mimic those of several connective tissue diseases and vasculitides. (2) Routine laboratory tests may reveal evidence of leucocytosis, polycythemia, anaemia, thrombocytosis, thrombocytopenia and increased erythrocyte sedimentation rate. Among the benign primary cardiac tumors, cardiac myxomas are the most notorious for causing the systemic manifestations. Elevated serum interleukin-6 (IL-6) frequently found in these patients is believed to mediate such symptoms. (3)
Myxoma, the most common type of primary cardiac tumor, accounts for 30% to 50% of all primary tumors of the heart. It most commonly presents in adults 30 to 50 years of age, although it can occur across all age groups. Sixty five percent of cardiac myxoma occur in women and 10% of them are familial, thus routine screening of first degree relatives of myxoma patients is recommended. They are histopathologically benign with tumor cells arising from multipotent mesenchymal cells. Clinically, patients with symptomatic cardiac myxoma can have a variety of non-specific findings, however, majority of the patients will present with at least one of the classic triad of obstructive cardiac, embolic and constitutional symptoms. Our patient presented with constitutional symptoms at a relatively early age than classic myxoma. Carney’s complex is an autosomal dominant syndrome characterized by myxoma formation in cardiac and several extra cardiac locations and associated with spotty skin pigmentation, endocrine hyperactivity. Cardiac myxoma associated with Carney syndrome show no age or gender predilection, can be single or multiple, can occur in any intra cardiac location and tend to recur with a rate of 20%. In contrast, sporadic cases of cardiac myxoma tend to occur in women of middle age and as isolated lesions in the left atrial aspect of inter atrial septum. Although our patient was a male child of seven years age, he did not have any syndromic associations. His echocardiography revealed a large mass attached to inter atrial septum and protruding to left ventricle through mitral valve during diastole. He underwent total excision of the left atrial mass under cardiopulmonary bypass. Post operative echocardiogram revealed no residual mass in the left atrium. Histopathological examination of the excised mass revealed tumors arranged in cords, nests and singly dispersed spindle and stellate shaped cells set in an abundant myxoid stroma along with binucleate and multinucleated forms with areas of haemorrhage, fibrous exudates and inflammatory infiltrate with the excised mass being reported as left atrial myxoma.

In conclusion, the present case demonstrates the unusual presentation of left atrial myxoma at an early age with fever of unknown origin. Thus, this diagnosis should always be borne in mind when dealing with fever of unknown origin even in pediatric population.


7 Braunwald’s Heart Disease - A Text book of Cardiovascular Medicine, 9th edition, Pages 1638-1640.