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CARDIAC MYXOMA: THE DECODED CARDIAC MYSTERY IN HISTOPATHOLOGY

Pathology

Dr. Swati Nayek*	Junior Resident, Department of Pathology, Burdwan Medical Collegeand Hospital, Purba Bardhaman. *Corresponding Author
Dr. Soumyadip Das	Junior Resident, Department of Pathology, Burdwan Medical College and Hospital, Purba Bardhaman.
Dr. Shreosee Roy	Assistant Professor, Department of Pathology, Burdwan Medical Collegeand Hospital, Purba Bardhaman.
Prof. (Dr.) Anup Kumar Boler	Professor, Department of Pathology, Burdwan Medical College and Hospital, Purba Bardhaman.
Dr. Saptarshi Roy	MS, MCh(CTVS).

ABSTRACT

Cardiac myxoma is a rare neoplasm but, the most common among all cardiac tumors. It occurs most commonly in left atrium. Here we report a case of left atrial myxoma presenting with symptoms of mitral valvular obstruction. A 40 years old woman presented to the Cardiology outpatient clinic with complaints of frequentepisodes of syncope. On examination, the patient had irregular feeble pulse, bradycardia and some added heart sounds. Patient was advised certain oral medications but she did not show much improvement. ECG and Echocardiography were advised as further investigations. Echocardiography revealed a pedunculated mass arising from the roof of the left atrium. A provisional diagnosis of myxoma was made. Patient was referred to the cardiovascular surgery department where the mass got excised by an open thoracotomy . Histopathological examination of that excised specimen confirmed the diagnosis of a soft tissue tumor of heart, cardiac myxoma.

KEYWORDS

Cardiac myxoma, Histopathological examination.

INTRODUCTION:

Primary cardiac tumours are rare. Heart and mediastinum are usually the site of metastases from a distant primary malignant neoplasm. Most primary cardiac tumours are benign. Cardiac myxoma is the most common true intracardiac neoplasm but its presentation could be diverse. It is usually benign in nature. It is considered to be arising from subendocardial vasoformative reserve cell remnants or multipotent primitive mesenchymal cells in the fossa ovalis. Here we present a sporadic case of cardiac myxoma.

Case Report:

A 40 year old female, non-smoker, non-diabetic, previously asymptomatic presented with frequent episodes of syncope and exertional dyspnea (NYHA Grade1) for the past one year. Examination of the patient revealed irregular pulse , bradycardia, and fluctuating blood pressure. The patient had tachypnea (Respiratory Rate 20/min) and had bilateral normal vesicular breath sounds. Auscultation of precordium revealed loud first heart sound and added heart sounds which were clinically attributed to mitral valve obstruction. No other significant abnormality could be detected upon clinical examination of the cardiovascular and respiratory systems respectively. Examination of the gastrointestinal system and nervous System were within normal limits. All the hematological parameters were within normal limits, except for a raised ESR. Urgent 2D echocardiography unveiled a pedunculated mass hanging from the roof of the left atrium blocking the outlet of left atrium (Fig 1). Patient was then referred to cardiothoracic surgery unit. The mass got excised by an open thoracotomy and subsequently sent for histopathological examination. Post operative period was uneventful and the patient was discharged upon stabilization after 10days.

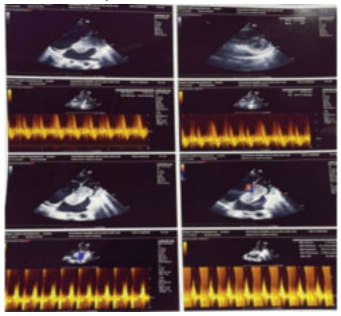

Fig.1: Echocardiographic findings

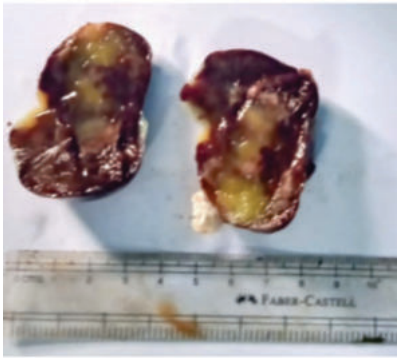

Fig2: Cut-section of specimen on grossing

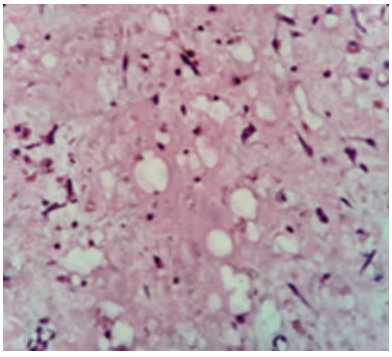

Fig.3: H & E section(40X)

On gross examination, the mass was well circumscribed, smooth, glistening brown, lobulated and measured 6cmX3.5cmX2cm. Cut surface was gelatinous (Fig.2). Microscopy (Fig. 3) showed a hypocellular lesion composed of bland cells in cords and small nests in a myxoid background. Few stellate lepidic cells with abundant eosinophilic cytoplasm and indistinct cell borders were also noted along with occasional inflammatory cells. All of these findings in Hematoxylin &Eosin stained sections confirmed the mass to be a cardiac myxoma.

DISCUSSION:

Often symptoms can misguide the clinician, likely when a cardiac neoplasm imitates mitral valve stenosis. Since the therapeutic avenues are absolutely different, it offers no less than a challenge in diagnosis. The word “myxoma” comes from 'muxa' and 'oma' , that means tumor consisted of mucus. Though there is a controversial role of HSV infection in pathogenesis of cardiac myxoma, it can be sporadic in 80% cases followed by familial. About 75% of cardiac myxomas are located in the left atrium.[1]

As few myxomas frequently recur and display aggressive characteristics, some refer to it as 'Malignant Myxoma' but it is a misnomer.

Cardiac myxoma is the most common primary tumor of heart. In the sporadic form, the tumor typically occurs in middle aged women. In familial cases there is mutation in PRKAR1A gene presenting as Carney complex and its subsets ;NAME syndrome [Nevi, Atrial myxoma, Myxoid neurofibroma and Ephelides] and LAMB syndrome [Lentigines, Atrial myxomas, Mucocutaneous myxomas and Blue nevi].

Clinical presentation typically includes the triad of intracardiac obstruction, embolic events and few constitutional symptoms.[2]In this case, our patient presented with the typical symptoms of mitral valve obstruction. Occasionally left atrial myxomas exhibit moderate pulmonary hypertension and their right counterparts, venous congestion. Cardiac Myxoma can increase concentration of interleukin-6 in patient's serum which may possibly potentiate metastasis of cardiac myxoma.

Histology manifests polygonal, stellate lepidic cells arranged in cords and nests in abundant mucopolysaccharide along with inflammatory cells, hemorrhage, hemosiderin and some perivascular tufts. Occasional fibrosis, calcification, extramedullary hematopoiesis and glandular structures can also be noted.

Cardiac myxoma shows strong diffuse positivity for calretinin. Variable positivity for S100 can be shown by myxoma cells scattered within myxoid stroma and positivity for CD31 and CD34 due to vessels. A minority of myxomas stained positive for antismooth muscle actin(staining the cells that immediately surround the myxoma cells) and *Ulexeuropaeus*agglutinin I (staining the endothelial cells of vascular channels).This tumor shows always negativity for KP-1 and Cytokeratin .[3]

Although cardiac myxoma is a benign neoplasm and curable by surgical removal only, reports on its malignancy are well-known, which include local relapse, local invasiveness, distant metastasis. The potential for malignant transformation is controversial, despite the publication of some reports of sarcomas transforming from recurrences.[4]

Although cardiac myxoma is the most common cardiac neoplasm, its confusing clinical symptoms may pose a diagnostic challenge as in our case. This patient presented with symptoms of mitral valve obstruction which could be due to several other etiology such as mitral valve prolapse, Rheumatic heart disease, infective endocarditis, cardiomyopathy etc. s The visibility of a mass on echocardiography followed by its surgical removal and subsequent hitopathological examination of the excised specimen could only furnish the ultimate diagnosis.

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