Case report

Role of imaging in the evaluation of a large left atrial myxoma: A case report and review of literature

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Abstract

Cardiac myxomas of giant size are rare. Their clinical presentation depends on the rapidity of growth of the mass and pressure effects which it produces. Echocardiography alone is sufficient in most cases for initial diagnosis and to prepare for surgical removal. Modern imaging is warranted in cases with atypical features like huge size or non-visibility of stalk or signs of infiltration. We present one such case of a giant left atrial myxoma which was evaluated with MRI in this report.

Key words: Cardiac tumors, myxoma, imaging of cardiac tumors

Mass lesions occupying the entire left atrium is a rare occurrence. Only a few cases of cardiac myxomas of giant size are reported. Their clinical presentation depends on the rapidity of growth of the mass and pressure effects it produces. Echocardiography alone is sufficient in most cases for initial diagnosis and to prepare for surgical removal. Modern imaging is occasionally performed in cases with atypical features like huge size or non-visibility of stalk or signs of infiltration. We present one such case of a giant left atrial myxoma which was evaluated with MRI as the stalk could not be identified in echocardiography.

Case report

A 45-year-old Indian woman admitted for an episode of pre-syncope. She also endorsed exertional breathlessness (NYHA class 3) for the last 3 months; but denied chest pain, palpitations, orthopnoea or paroxysmal nocturnal dyspnea. She had low grade subjective fever and myalgia 4 months prior to presentation. On examination, vitals were stable (blood pressure 110/70mm Hg, heart rate 88 beats per minute, respiratory rate varied between 18 and 20 /minute) and she was afebrile. There was mild pallor. Jugular venous pressure was normal. Precordial examination revealed normal apex. Cardiac auscultation revealed loud P2 and a short mid-diastolic murmur. Lungs were clear to auscultation. Abdomen was benign and there was no hepatosplenomegaly.

The electrocardiogram was within normal limits (Fig 1). Chest x-ray showed normal cardio-thoracic ratio with no suggestion of any chamber enlargement. There was mild dilatation of main pulmonary trunk and cephalisation of the pulmonary venous pattern (Fig 2). The transthoracic echocardiography showed a huge filling defect indicative of a possible mass occupying almost entire left atrium, measuring 6 x 8 cm. It was hyper echoic and no stalk could be identified. The gradient across mitral valve was 20mm Hg (Fig 3A, B, C). Magnetic Resonance Imaging (MRI) study confirmed the mass which had heterogenous density predominantly with intermediate signal in T1W study and mildly

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hyperintense on T2W images and it protruded with each diastole through the mitral valve suggesting that it could be a myxoma (Fig 4). Coronary arteriography showed normal coronaries and there was minimal tumour blush (Fig 5).

Fig 1. Electrocardiogram shows sinus tachycardia and signs of left atrial enlargement.

She underwent successful surgical removal of the mass. The resected specimen was large, globular, gelatinous but firm and had a wide stalk. It measured 7 x 7 x8 cm. Resection margins were clean. There were no adhesions. Cut surface showed areas of gelatinous white and yellowish substance with a little calcification. Histopathological examination showed ovoid to spindled tumor cells arranged in cords, micro trabeculae, nests and perivascular bands within a myxoid background confirming the diagnosis of myxoma. Post-operative echocardiography showed no residual tumor tissue with normal left and right ventricular function.

Fig 2. Chest x-ray shows normal cardio-thoracic ratio with no suggestion of any chamber enlargement. Note that there is mild dilatation of main pulmonary trunk and cephalisation of the pulmonary venous pattern

Fig 3 A, B. The transthoracic echocardiography shows a huge filling defect indicative of a mass occupying almost entire left atrium, measuring 6 x 8 cm. It is hyperechoic and no stalk is identified.

Discussion

Primary cardiac tumors are uncommon with a reported incidence of 0.0017 to 0.19% in autopsy series\(^7\). Approximately 75% of them are benign. About half of all cardiac tumors are myxoma, which are almost always benign\(^8\). As many as 80% of them occur in left atrium\(^9\). Most of them are moderate sized (2 to 6 cms) and a show a stalk attached to the interatrial septum close to fossa ovalis\(^10\). Cardiac myxomas are either typical or atypical. Typical cardiac myxomas are almost always single with a striking predilection to arise within the left atrium. Atypical myxomas can be multiple, may arise from the right side of heart or left ventricle. Lesions can be sessile or pedunculated with a stalk of variable sizes (1-10cms)\(^11\). They commonly present with nonspecific symptoms like low grade fever, malaise and anemia or maybe detected during echocardiography in patients with suspected mitral stenosis\(^12\). Common Symptoms include dyspnea, orthopnoea and paroxysmal nocturnal dyspnea or pulmonary edema (mimicking mitral stenosis) or may present with complications like distal embolization (resulting in stroke)\(^13\). Slow growing large myxomas may be asymptomatic\(^14\). Only a few cases of huge left atrial myxomas have been reported so far\(^6,15,16\). Sometimes, the clinical picture can be dramatic with drop attacks due to prolapse of the tumor through the mitral valve (MV) needing emergency surgery\(^6,15,16\).

Chest radiograph may show features suggestive of mitral stenosis (MS). Calcification may be seen sometimes. In the evaluation of left atrial masses, transthoracic echocardiogram (TTE) and 2 dimensional trans-esophageal echocardiogram (TEE) are the initial investigations of choice. Real time 3 dimensional TEE is increasingly being used in clinical practice as well. TTE diagnoses the presence of a mass, can determine its size and most of the
anatomical details. On echocardiography lesions are hyper echoic and a stalk may be visible. The mass can usually be differentiated from large thrombus and other tumor–like lesions and this determination is important prior to surgical intervention. On TTE, myxomas are seen as mobile masses attached to endocardial surface by a stalk. If a narrow stalk is not visible, the diagnosis by echocardiography alone can be challenging as in our case. Further imaging with real time 3D TEE or cross sectional imaging (computerized tomography or MRI) is essential in such situations. TEE more accurately delineates the tumor structure and site of attachment as well as potential satellite lesions. MRI and Computerized tomography (CT) are very important imaging tools to precisely show the extent of the mass and its morphology. The resectability can be pre-assessed with confidence based on these images. In our patient the attachment of the mass to the interatrial septum could be made out on MRI which was not clear on echocardiography.

Contrast enhanced CT scan (CECT) usually shows heterogenous enhancing mass. On CT scan, the average attenuation of LA myxoma mass is 43Hounsfeld Units, closely mimicking a thrombus. MRI reveals heterogeneously hypointensities in T1W and hyperintensities in T2W images. On contrast MRI variable enhancement will be seen. Heterogeneity is due to necrosis, repeated hemorrhage, and dystrophic calcification. There may be variable enhancement, differentiating the mass from thrombus, which is non enhancing mass on both CT and MRI. Thrombus is usually located in the atrial appendage and may sometimes be found in body of left atrium. Thrombi are associated with atrial fibrillation, enlarged LA, prosthetic mitral valve or low cardiac output state. Imaging is performed to determine if there is invasion of tissue plane. The growth rate of myxoma is variable-sometimes may not show any growth at all; but growth rates can vary between 1.3 to 6.9mm/month in diameter. Neovascularity may be detected on angiography suggesting a cardiac tumor (unlike organized thrombus). Some myxomas can undergo regressive changes, hemorrhage and rarely osseous metaplasia and such masses have to be differentiated from organized thrombi. 3D real time TEE can be a very useful tool preoperatively which can give surgeons a better understanding of the anatomical characteristics of any tumor.

Conclusion

Left atrial myxomas when present with atypical features like huge size or non-visibility of stalk should be evaluated by modern imaging techniques like MRI.

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References