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Case Report

Unusual Presentation of a Rare Disease

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Abstract: Intra-cardiac tumors are rare with estimated prevalence in autopsy series of 0.02%. Atrial myxoma is the most common primary cardiac tumor. The diagnosis can be challenging and high index of suspicion is required for prompt diagnosis and recognition, as symptoms and signs are nonspecific. Atrial myxoma is one of the few emergency diagnoses in cardiac surgery that require urgent consultation and resection to avoid serious embolic complications. We present a case of atrial myxoma with recurrent embolization. We reviewed the literature for prevalence, clinical presentation, diagnostic challenges and treatment of atrial myxoma.
Keywords: Atrial myxoma, Left atrial tumor

INTRODUCTION

Atrial myxoma is a rare disease. It is the most common benign primary cardiac tumor. It accounts for 50-70% of all primary cardiac tumors [1]. We present a case of 50 years old, female with recurrent abdominal pain for 6 months and multiple emergency visits with no clear diagnosis who was found to have left atrial myxoma with recurrent liver, spleen and kidney embolizations. This case highlights the challenges with making a diagnosis of left atrial myxoma, as diagnosis can be elusive. We outline the approach to clinical presentation, diagnosis and management.

CASE PRESENTATION

A 50 years old female presented with recurrent abdominal pain for 6 months. She had recurrent visits to the local emergency department without clear diagnosis for the etiology of her abdominal symptoms. Serum biochemistry, complete blood count and abdominal x-rays were all normal and the patient was treated conservatively with pain medications. Past medical history was significant for atrial septal defect that was surgically repaired in 2001. At her sixth visit to a local emergency department, abdominal ultrasound was obtained and revealed hypoechoic lesions in the liver and kidney. An abdominal CT scan revealed multiple lesions in the liver, kidney and spleen consistent with arterial emboli. (Figure 1A, 1B). A transthoracic echocardiogram (TTE) reveals a left atrial mass attached to fossa ovalis, measured 52x18 millimeters, protruding from the left atrium to the left ventricle through the mitral valve in diastole (Figure 2) consistent with left atrial myxoma. Cardiac surgery was consulted for consideration of an urgent open heart surgery and atrial myxoma resection on the same day of diagnosis. The patient was taken to the operating room in the next 24 hours and the tumor that was resected was sent to pathology and the diagnosis of atrial myxoma was confirmed. The patient recovered from the surgery without any postoperative complications.

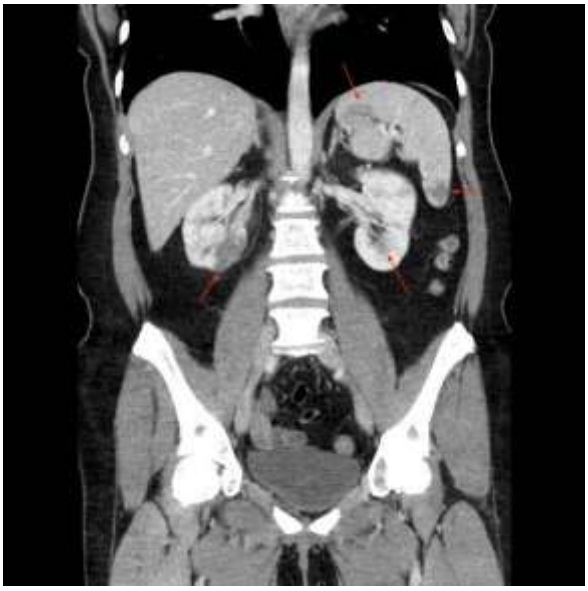

Fig-1A: CT abdomen shows multiple spleen and kidney embolization (Arrows)

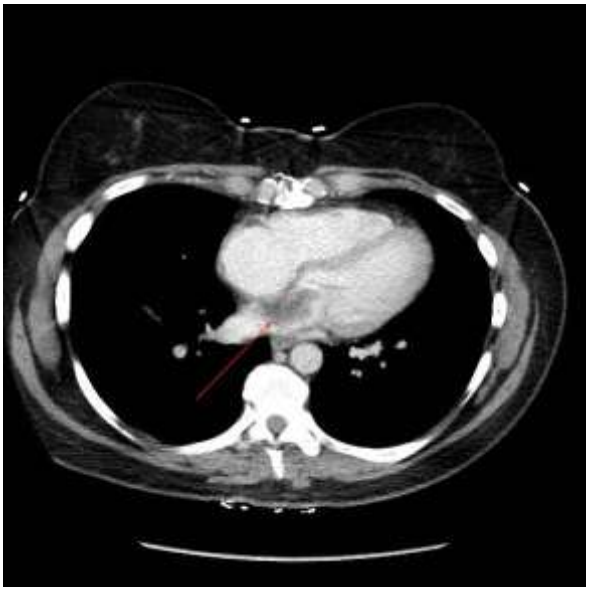

Fig-1B: CT scan shows left atrial myxoma

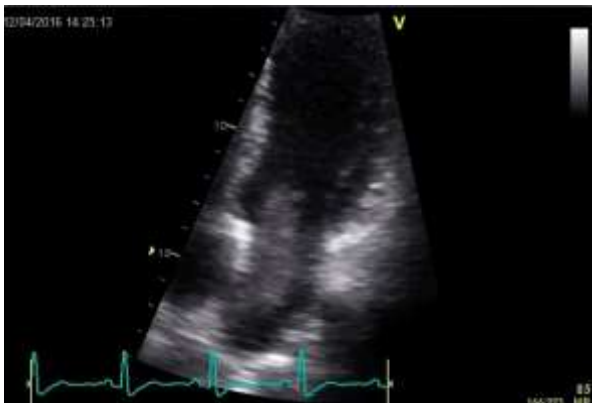

Fig-2: Apical 4 chamber view shows left atrial myxoma

DISCUSSION

Atrial myxoma is the most common primary benign cardiac tumor. It accounts for 50%-70% of all primary cardiac tumors. It occurs in third to sixth decade of life. Myxomas are more common in women, with a 3:1 female to male ratio. It is typically located in the left atrium in about 85% of cases, followed by the right atrium, the right ventricle and rarely the left ventricle. Most cases are sporadic, however, 7% of atrial myxoma are part of Carney complex, which is autosomal dominant disease manifested with recurrent, multiple atrial myxoma, endocrine, breast, cutaneous, and neural tumors and skin pigmentations. The recurrence rate of atrial myxoma after excision is about 3% in sporadic cases and 22% in carney complex [2].

Clinical presentations vary, with most patients being asymptomatic and diagnosed incidentally based on echocardiogram, cardiac MRI or CT scan performed for other reasons. The second most common group of symptoms are caused by the myxoma obstructing the mitral valve and producing functional mitral stenosis, which includes positional syncope, shortness of breath when lying on left side and pulmonary edema. Constitutional symptoms such as low-grade fever, fatigue, weight loss, arthralgia and skin rash are common.

Stroke is the most dreadful complication and 25.9% of patients with atrial myxoma will develop stroke [3]. Peripheral embolization to liver, spleen, kidney and lung is rare and reported in 3.7% of patients [3].

Physical examination includes diastolic rumbling sound at the mitral valve consistent with functional mitral stenosis, tumor plop sound that is a low pitched diastolic sound after the second heart sound produced by myxoma prolapsing through the mitral valve.

Differential diagnosis for atrial mass include primary benign or malignant tumors, tumor metastasis, atrial thrombus, vegetation, lipomatous hypertrophy of the atrial septum and artifact. Normal structure such as Eustachian tube and prominent crista terminals can be mistaken for atrial tumors.

Transthoracic echocardiogram (TTE) is the primary modality for diagnosis. Features of myxoma on TTE include gelatinous, pedunculated mass attached with a stalk to fossa ovalis with a smooth, villous, or friable surface, atrial myxoma protruding into the left ventricle during diastole, which produces functional mitral stenosis. Echocardiographic contrast helps to confirm the diagnosis and enhances the sensitivity and specificity of TTE. Complete lack of enhancement of the atrial mass on contrast echo suggests thrombus, while partial enhancement suggests myxoma and complete enhancement suggests malignant tumors.

Cardiac MRI or cardiac CT scan can be useful to establish or confirm the diagnosis if it remains uncertain after echocardiogram. Cardiac MRI is helpful to delineate the anatomy, the extent of the tumor and the surgical planning for resection.

THERAPY

Surgical resection is the definitive treatment and is curative in most patients. The diagnosis of Atrial myxoma constitutes an emergency diagnosis and requires emergent cardiac surgery consult. Delay in diagnosis or resection may result in new embolization or death. Recurrence rate of the tumor is 3% in sporadic cases; however, it is higher in patients with Carney complex and approaches 22%. TTE surveillance is indicated to monitor for reocurrence.

Recurrence may be due in part to unrecognized incomplete tumor resection or metastasis of myxoma to other parts of heart or blood stream during the resection.

CONCLUSIONS

Atrial myxoma is a rare tumor and is a cardiac surgical emergency. High index of suspicion is required for prompt diagnosis. Early diagnosis has important therapeutic and prognostic implications. Delayed diagnosis may result in catastrophic complications such as stroke or death. Clues to atrial myxoma include syncope upon sitting from lying position, dyspnea while lying on the left side, embolic stroke or systemic embolization and the presence of tumor plop on physical exam. Surgical resection is the definitive treatment and lifelong surveillance is required to monitor for recurrence.

AUTHOR CONTRIBUTIONS

All authors contributed to case design, data collections, literature review, writing up the case and approval of the final draft

ABBREVIATIONS

Transthoracic echocardiogram (TTE), Magnetic resonance imaging (MRI), Computerized tomography (CT scan)

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