Atypical presentation of left atrial myxoma: A case report

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Abstract

Atrial Myxoma is the most common primary benign tumour of the heart, commonly found in the left atrium. It typically presents in young females with characteristic features such as, constitutional symptoms, chest pain, and cardiac murmurs. However, atypical presentations can occur; causing a diagnostic challenge. This case report describes a 75-year-old male who visited the cardiology outpatient department of Dow Institute of Cardiology, Karachi on 18th April, 2023 with a left-sided atrial myxoma in late adulthood without typical features including constitutional symptoms, chest pain, syncope, dizziness, digital clubbing or neurologic findings. Further discussed are the diagnostic techniques used to find the tumour and the treatment strategy. This case report highlights the need for cardiologists to consider Atrial Myxoma as a potential diagnosis, even in the absence of typical symptoms, in elderly male population.

Keywords: Atrial Myxoma, Intracardiac tumour, Myxoma, Myxoma Resection.

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Introduction

Atrial Myxoma is the most frequently occurring primary benign cardiac tumour.[1] The most common site is the left atrium followed by the right atrium, particularly the atrial septum and the vena cava. Atrial Myxoma is three times more prevalent in middle-aged women than men.[2] Atrial Myxoma can be categorised into two epidemiological types: familial and sporadic. The sporadic type accounts for approximately 95% of all cases, whilst the familial type makes up the remaining percentage.[3] The familial type of atrial myxoma predominantly affects younger men and is characterised by the presence of multiple tumours. Unlike the sporadic type, familial myxomas can also be found in other cardiac chambers, not just limited to the atria.[4] The case of a 75-year-old man with a left-sided atrial myxoma presenting in his late adult life with atypical symptoms is documented. Atypical attachment of the myxoma and the associated new findings of myocardial bridging were found incidentally on angiography.

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Case Report

A 75-year-old patient presented to the cardiology outpatient department Dow Institute of Cardiology, Karachi on 18th April, 2023 with progressive shortness of breath, chest heaviness, palpitations and orthopnoea for 3 months. The patient displayed an atypical presentation with no typical symptoms like fever, syncope, dizziness, digital clubbing, or neurologic findings.

On physical examination, the patient vital signs were within normal limits. On auscultation, a mid-diastolic murmur and a third heart sound were audible at the apex. Certain investigations were conducted to establish the diagnosis. The patient was transferred to the Cardiac CCU for further evaluation. Initially, an ECG (Electrocardiography) was performed which yielded unremarkable results. A chest x-ray was obtained, showing no significant findings. Subsequently, an Echocardiography was conducted which revealed a large echogenic mobile mass (Left Atrial Myxoma) attached to the anterolateral commissure of the anterior Mitral Valve leaflet (size =30x24mm) as shown in (Figure 1). The left ventricle was of normal size with normal systolic function, and no regional wall abnormalities. However, the left atrium was mildly enlarged, measuring around 4.4cm. Additionally, the right ventricle was normal

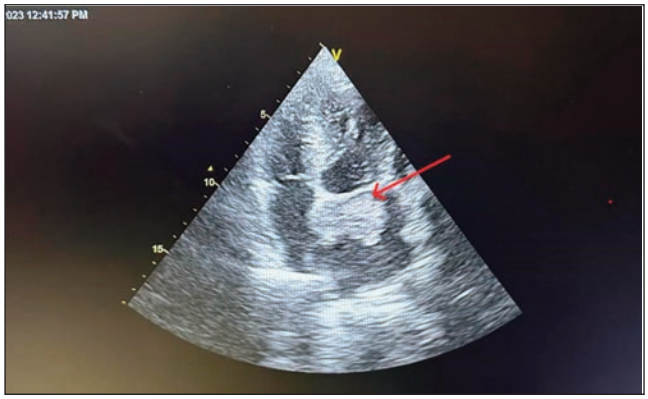

Figure-1: Pre - Op Four chamber Transthoracic Echocardiography showing clear demarcation between the Mitral valve and the Atrial Myxoma.

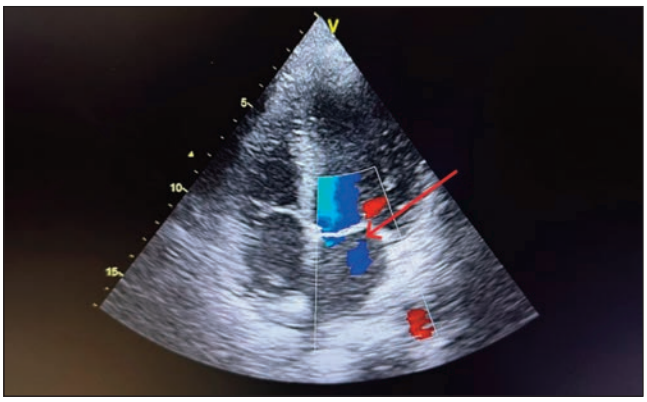

Figure-2: Post-Op Four chamber Echocardiography showing no myxoma with clear margins.

in size with normal systolic function. Upon examination for valvular abnormalities, the results were unremarkable. The myxoma was closely observed, noting its free movement in the left atrium during systole and protrusion into the left ventricle during diastole, occupying nearly the entire mitral orifice. Moreover, its stalk was not clearly visible due to its atypical attachment to the anterolateral commissure of the anterior mitral leaflet of the valve as depicted in (Figure 1). Angiography showed non-occlusive coronary arteries with mild to moderate bridging of the mid segment of Left Anterior Descending (LAD) Artery.

Following a multidisciplinary discussion, the surgical team decided on a Left Atrial approach to perform an atrial myxoma excision procedure on 20th April, 2023. The myxoma was attached to the anterolateral commissure of the mitral valve, near the anterior part of the inter-atrial septum, with an anatomically normal stalk. This atypical attachment presented a challenge for the surgeon, requiring careful excision of the tumour whilst preserving the mitral leaflet anatomy and preventing peri-operative mitral regurgitation, which could lead to mitral valve replacement. A biopsy of the tumour was performed, revealing a neoplastic lesion arranged in cord and nest. An individual neoplastic cell with abundant eosinophilic cytoplasm was also seen. The surrounding stroma was myxoid along with thick blood vessels, consistent with a myxoma diagnosis. Post-operative vital signs included a Blood pressure of 116/79 mmHg, a Glasgow Coma Scale of 15/15, with clear and audible bilateral chest sounds and an SpO2 reading of 98% without any additional heart sounds or murmurs audible. The Post-operative Echocardiography, as shown in (Figure 2) demonstrated clear margins with no iatrogenic Mitral Regurgitation.

Discussion

Atrial Myxoma is the most prevalent benign primary tumour of the heart. It should be suspected in patients presenting with symptoms of left-sided valvular obstruction or congestive heart failure[5] or even atypical symptoms, as seen in the presented patient. Unlike the usual presentation in young female, the reported patient was an elderly male. However, according to a case series spanning 35 years, it was found that Left Atrial Myxoma is very rare in the elderly. Bire F. et al. documented and shared their findings on 19 instances of left atrial myxoma in elderly patients over the age of 75. These cases were part of a larger group of 100 myxoma diagnoses, made between 1962 and 1997.[6] Symptomatic manifestations of left atrial myxomas arise when they obstruct the mitral valve, leading to peripheral embolization or inducing systemic effects.

Left atrial myxomas typically display a gross appearance characterised by different colours such as white, gray-white, yellowish, or brownish. They exhibit different shapes including polypoid, round or oval, and can be either pedunculated or sessile. The tumour consistency is gelatinous, and the surface may be smooth or villous in texture.[7]

Apart from their cardiovascular symptoms, individuals with myxomas often experience constitutional symptoms such as weight loss, fever and arthralgias.[8] However, this patient presented with none of these constitutional symptoms which is considerably rare. Myxoma without constitutional symptoms only occurs in about 34% of patients as seen in the case series conducted by Pinede et al.[7] Over 75% of myxomas arise within the left atrium, specifically near the mitral annulus or the border of the inter-atrial septum known as the fossa ovalis.[9] However in this patient, the myxoma was atypically attached to the anterolateral commissure of the anterior mitral leaflet of the valve, which also increased the chances of iatrogenic injury to the mitral valve leaflets.

Atrial Myxomas can be diagnosed with echocardiography alone or in conjunction with angiography, CT scan and MRI.[10] In this patient, during angiography, a mild to moderate bridging of the mid segment of LAD Artery was observed, which is also not a cardinal feature of atrial myxoma. Although, bridging was noticed during angiography, a non-surgical approach was chosen due to the potential for increased complexity. The risk of additional inadvertent damage to the heart tissue was also suspected during the surgical removal of the bridge, which would impair the outcome of surgery in an elderly man. Moreover, medical therapy for myocardial bridging is available, and symptoms can effectively be controlled. Later, a decision was made for surgical excision of the myxoma, as, an effective surgical excision of atrial myxoma can provide relief from immediate and ongoing symptoms with a low risk of complications and mortality. Upon confirmation of the diagnosis, it is recommended to

promptly perform surgical resection to prevent the potential occurrence of additional complications such as embolization or sudden cardiac death. Some cases of patients developing arrythmias following myxoma surgery have been reported[11] but the reported here did not develop any tachy or brady arrythmias.

Conclusion

This case emphasises on the importance of considering left atrial myxoma as a potential diagnosis, even when the patient presents with atypical symptoms and lacks the classical signs like chest pain, constitutional symptoms, syncope, dizziness, digital clubbing, or neurological findings. Early recognition, accurate diagnosis, and timely surgical intervention are crucial in preventing life-threatening complications such as cardiac obstruction, embolization, and sudden cardiac death associated with cardiac myxoma. In conclusion, this case underscores the significance of being vigilant for cardiac myxoma, particularly when presented with an atypical clinical presentation. Prompt recognition and management are vital in ensuring the best possible outcome for patients with this rare but potentially dangerous cardiac condition.

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Author Contribution:
MAS: Concept, design, data acquisition, analysis, interpretation, drafting and revision.
MII: Data acquisition, analysis, interpretation and revision.
MS, MK: Data acquisition, revision and final approval.
MTF: Supervision, revision and final approval.