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Giant Left Atrial Myxoma Revealed by Neurological Manifestation in a Young Female Patient

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Abstract:- Cardiac myxoma is a prevalent benign neoplasm that arises inside the cardiac tissue and exhibits a gradual growth pattern. The annual incidence rate of cardiac myxoma is approximately 0.5–1 cases per 1,000,000 persons. The fragmentation of a cardiac tumor may be influenced by cerebrovascular events. Ischemic stroke is a relatively infrequent occurrence in pediatric populations and can occasionally be attributed to cardiac myxoma. In instances of ischemic stroke, neurological impairments are observed as a result of emboli or thrombi originating from the myxoma. Echocardiography is a valuable tool for promptly diagnosing myxoma and facilitating immediate surgical removal of the myxoma. We report a 17 years old female with neurological deficit weakness in the upper and lower right extremities since 2 months ago as the chief complaint with intermittent shortness of breath. Haemodynamic parameters was stable. The patient underwent brain imaging with result a lesion of the left lentiform nucleus, transthoracic echocardiography and trans esophageal echocardiography with oscillating septated mass on left atrial, attached to the anterior mitral leaflet (AML). Primary cardiac tumors are a very uncommon cause of cardiac embolic events. An atrial myxoma, a benign growth, can form on either the left or right side of the upper part of the heart. Untreated atrial myxoma can lead to adverse effects such as arrhythmia, pulmonary edema, and emboli formation that obstruct blood flow in the heart's veins. Ischemic stroke causes a specific functional impairment. Neurological symptoms in stroke can be an extra cardiac manifestation of atrial myxoma. The use of transthoracic and tran seso phageal is important in diagnosing atrial myxoma.

Keywords:- *Cardiac Tumors; Myxoma; Stroke; Hemiparesis; Echocardiography.*

I. INTRODUCTION

Left atrial myxomas are rare cardiac tumors that can present with a variety of symptoms, including neurological manifestations. These tumors, although uncommon, can lead to significant morbidity and mortality if not promptly diagnosed and managed. The case report titled "Giant Left Atrial Myxoma Revealed By Neurological Manifestation in a Young Female Patient" highlights a scenario where a young female patient's left atrial myxoma was initially detected due to neurological symptoms rather than typical cardiac signs [1]. Neurological manifestations of left atrial myxomas can vary widely and may include symptoms such as visual disturbances, speech impairments, strokes, memory loss, and even hemiparesis. These presentations can sometimes be misleading, as seen in cases where patients initially present with embolic strokes or other neurological deficits without typical cardiovascular symptoms [2]. Imaging studies play a crucial role in the diagnosis of left atrial myxomas with neurological manifestations. Neuroimaging techniques have been instrumental in identifying cerebral embolisms, aneurysms, and intra parenchymal metastases associated with these tumors [3]. Additionally, echocardiography, both transthoracic and trans esophageal, is essential in visualizing the cardiac mass and its effects on surrounding structures, aiding in treatment planning [3]. The management of left atrial myxomas with neurological implications often involves a multidisciplinary approach. In some cases, emergency surgery may be necessary to prevent further cerebral embolization and mitigate the risk of stroke [4]. Thrombolysis and embolectomy have been employed in acute stroke settings to address embolic events caused by these tumors. Surgical resection remains the cornerstone of treatment for left atrial myxomas, especially in cases where the tumor poses a risk of embolization or hemodynamic compromise.

The most prevalent type of primary cardiac neoplasm is cardiac myxoma, accounting for approximately 30% to 50% of all primary heart tumors. It has an annual incidence rate of 0.5 per million individuals. The left atrium (LA) is responsible for almost 70% of cardiac myxomas, whereas the right atrium (RA) accounts for 18%. The prevalence of biliary myxomas in the context of cardiac myxomas is less than 2.5%. The majority of myxomas exhibit one or more of the elements, manifesting as either embolic or obstructive. Cardiac myxoma serves as a reservoir for emboli that can be directed towards the central nervous system and other regions within the vascular tree. Nevertheless, in the absence of any prior cardiac issues, nonspecific systemic symptoms and small embolic events can go unnoticed. Atrial myxoma was identified in all subjects using echocardiography. There exists a correlation between atrial myxomas exhibiting an uneven surface and an increased susceptibility to embolic events. In the absence of cardiovascular risk factors, atrial myxomas frequently manifest as cerebral infarction. The primary focus of these tumors is on the middle cerebral artery. The irregular texture of myxomas appears to be associated with embolic events. The utilization of echocardiography has the potential to improve the detection and timely treatment of atrial myxomas. In summary, it is crucial to take into account left atrial myxomas when evaluating patients who exhibit

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unexplained neurological symptoms, as these tumors can emerge with a range of neurological symptoms. Prompt identification, suitable imaging techniques, and timely action are essential in effectively managing these situations and enhancing patient outcomes.

II. LITERATURE REVIEW

A. Incidences of Cardiac Myxoma

Cardiac myxoma is a rare primary cardiac tumor with varying reported incidence rates. Studies have shown that the incidence of cardiac myxoma ranges from 0.06% to 1.74% [5]–[7]. The rarity of cardiac myxoma underscores the importance of further research to better understand the epidemiology of this condition.

B. Ischemic Stroke in Children and Cardiac Myxoma

Cardiac myxoma can lead to ischemic stroke, even in children. A study reported a case of left atrial myxoma with ischemic stroke complications, highlighting the potential for cardiac myxomas to cause serious neurological manifestations, including strokes [8].

C. Embolic Complications of Cardiac Myxoma

Cardiac myxomas are known to be a source of embolic complications, with reported incidences of embolic events ranging from 0.06% to 77.14 [6], [7]. These embolic events can lead to various complications, including strokes and limb ischemia.

D. Echocardiography for Diagnosing Cardiac Myxoma

Echocardiography plays a crucial role in diagnosing cardiac myxomas. It is a valuable imaging modality for detecting these tumors and assessing their characteristics, aiding in the timely diagnosis and management of patients with cardiac myxomas [9].

E. Atrial Myxoma and Neurological Manifestations

Atrial myxomas can present with neurological manifestations, with studies reporting that 9-22% of patients with atrial myxomas suffer from embolic strokes [10]. These neurological manifestations underscore the importance of considering cardiac myxomas in the differential diagnosis of patients presenting with neurological symptoms.

F. Surgical Resection of Cardiac Myxoma

Surgical resection is the mainstay of treatment for cardiac myxomas. Studies have shown that patients with cardiac myxomas may experience embolic events before surgery, emphasizing the importance of timely resection to prevent further complications [11], [12].

III. CASE STUDIES AND PRATICAL APPLICATIONS

A 17 years old female was admitted to the hospital due to neurological deficit with sudden diminished strength of the upper and lower right extremities since 2 months ago with intermittent headache, dizziness, fatique and nausea. No seizure had been witnessed. There was intermittent shortness of breath. There was no palpitation, chest pain and blueness. There was no history of cardiac tumor in family. On initial examination, the patient was well oriented, had stable vitals with blood pressure 110/60 mmHg, regular pulse 88 beats per minute, repiratory rate 20 times per minute with oxygen saturation 98-99%. Hemodynamic parameters within normal limits. During cardiac auscultation, an early diastolic murmur grade 2/6 was detected, and the motoric strength was measured at 3 locations for both the upper and lower right extremities. The stroke score obtained from the National Institutes of Health Stroke Scale (NIHSS) was 5. Electrocardiography findings revealed sinus rhythm, heart rate 75 beats per minute, regularly, normoaxis, P wave 0.06 second, PR interval 0.20 second, QRS complex duration 0.06 second, Benign Early Repolarization. Chest X-Ray findings indicated bronchovascular features of both lungs within normal, there were no visible infiltrate or consolidation spots in both lungs. The shape and size of the heart within normal limits. There were no abnormalities in laboratory examination.

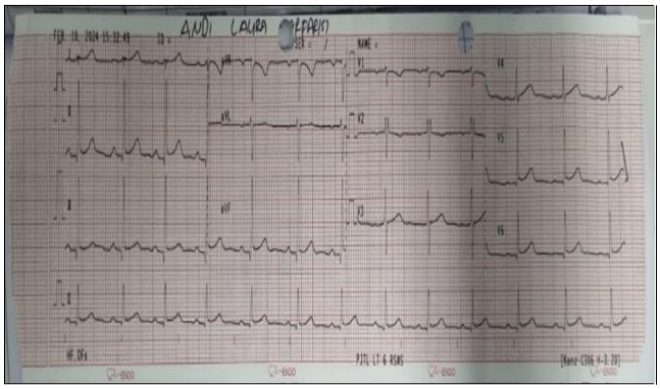

Fig 1 Electrocardiography

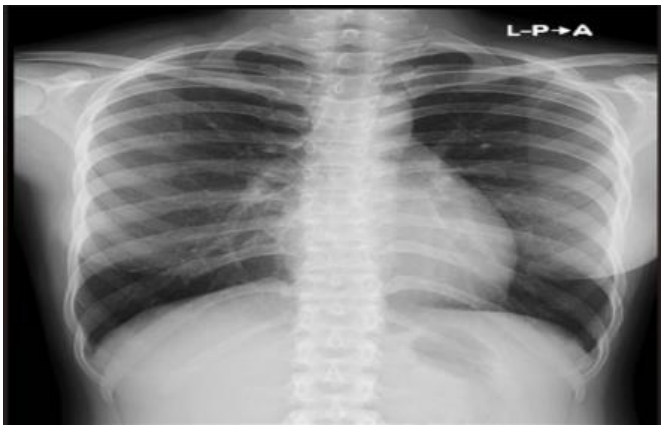

Fig 2 Chest X-Ray

Transthoracic echocardiography revealed a solid oscillating mass with size 54 mm x 30 mm in the left atrium leading to mitral valve occlusion. This mass was prolapsing into the left ventricle during diastole. Other echocardiography examination results were found situs solitus, all pulmonary vein retrieved to left atrial, AV-VA concordance, normal cardiac dimension, interatrial septum intact, interventricular septum intact, normal function of tricuspid valve, pulmonary and aortic valves, Confluence pulmonary arteries, and normal left and right ventricular function.

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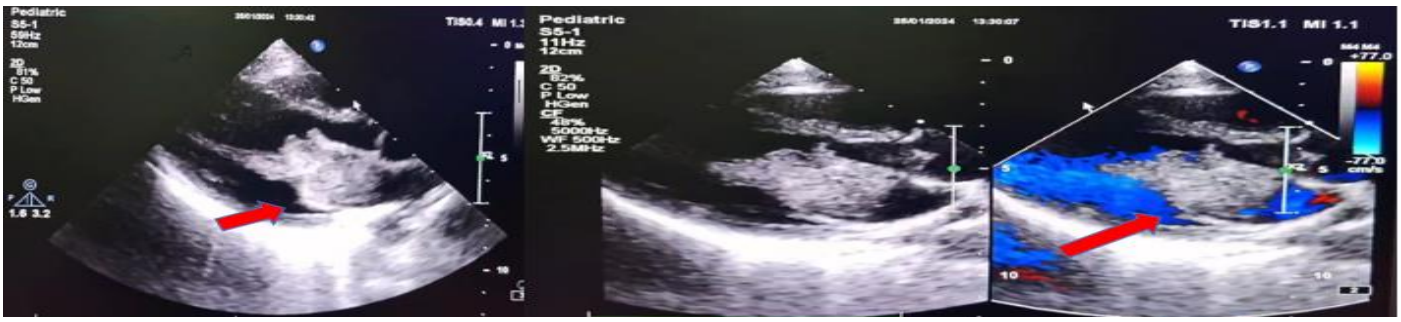

Fig 3 Transthoracic Echocardiography Parasternal Long Axis View (Red Arrow)

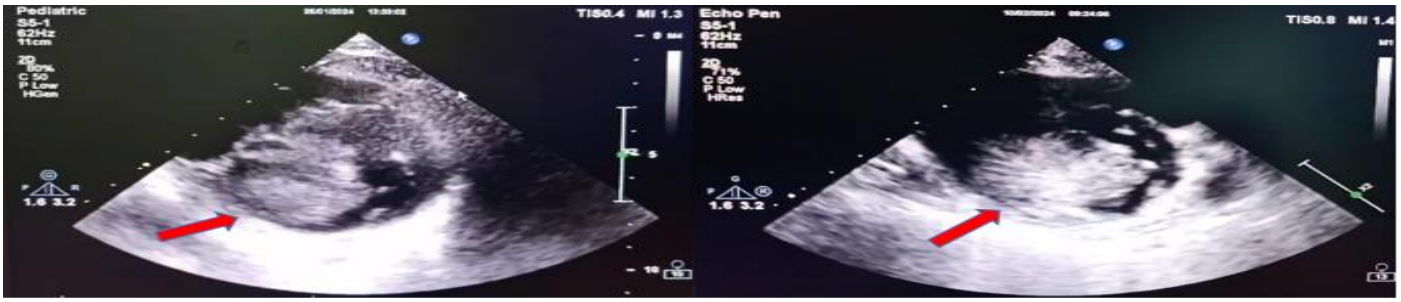

Fig 4 Transthoracic Echocardiography Parasternal Short Axis View (Red Arrow)

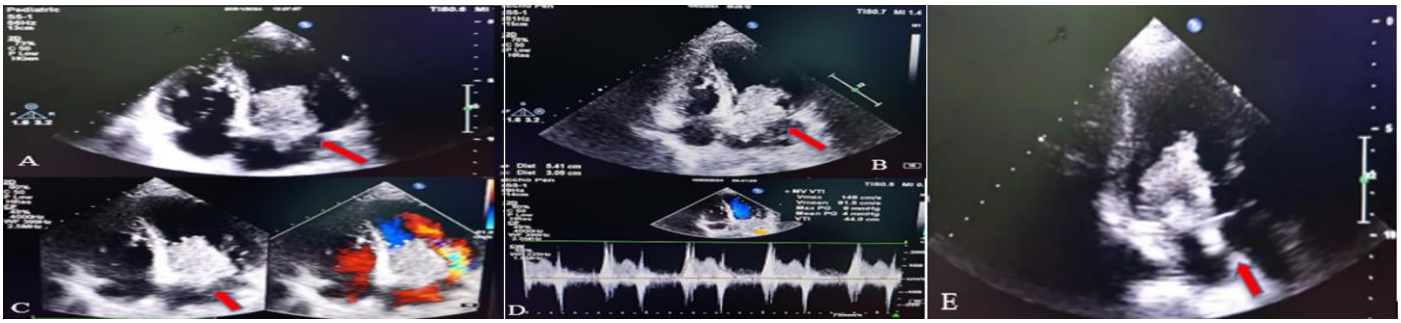

Fig 5 Transthoracic Echocardiography A-C. Apical Four Chamber View, D. Mild Mitral Stenosis, E. Apical Three Chamber

It confirmed with Trans esophageal echocardiography with conclusion multiple septated mass attached to the anterior mitral leaflet (AML) suggestive for cardiac myxoma of the mitral valve with differential diagnosis multiple fibroelastoma of mitral valve.

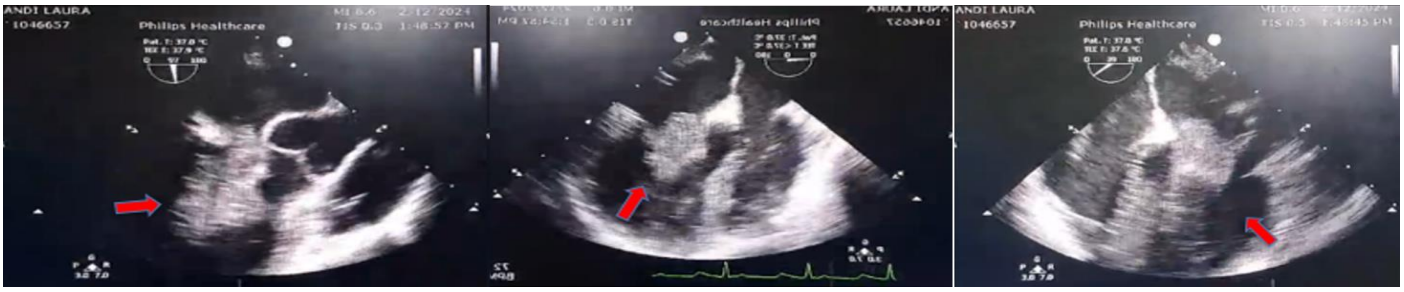

Fig 6 Trans esophageal Echocardiography

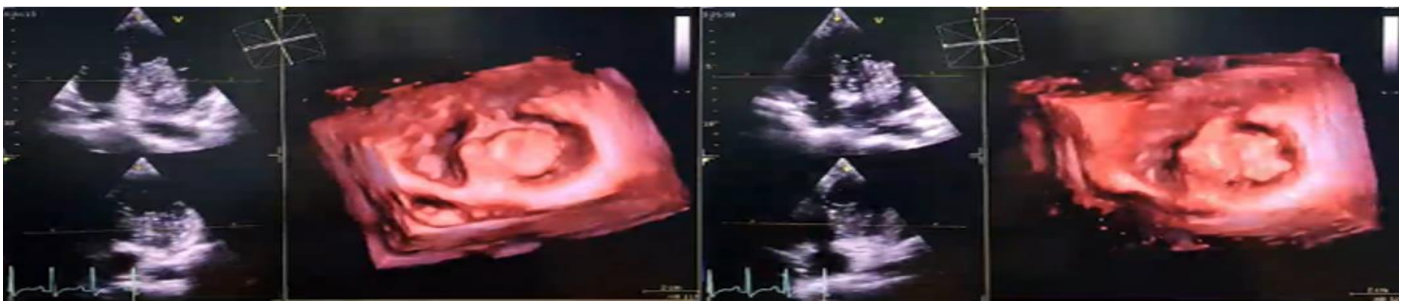
Fig 7 Transthoracic Echocardiography 3D

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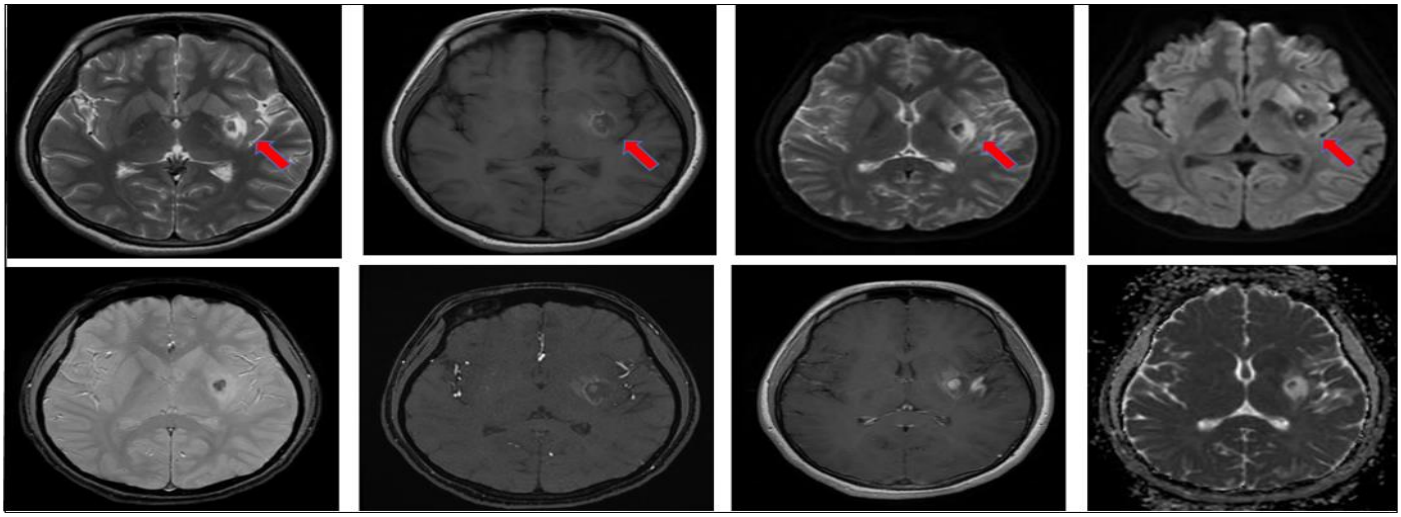

Fig 8 Brain Magnetic Resonance Imaging

Magnetic Resonance Imaging (MRI) of her brain demonstrated an early subacute process of central region and chronic at the edges with conclusion a left lent form nucleus infarction, sulci and gyri within normal limits, position of the interhemispheric fissure appears normal in the midline, ventricular system, subarachnoid space, cerebellum and pons within normal limits.

IV. DISCUSSION

Cardiac tumors in pediatric patients are exceedingly uncommon. At large children's referral facilities, the incidence of primary cardiac tumors was found to range from 0.001% to 0.003% among admissions. The distribution of males and females is equal. Primary cardiac tumors are infrequently observed in individuals of all age cohorts, with an incidence rate of less than 0.1% in a comprehensive analysis of over 12,000 postmortem examinations. Around 75% of primary cardiac tumors are classified as benign, with a predominant mesenchymal origin. Among these tumors, cardiac myxoma constitutes over 50% of the cases [13]–[15].

Cardiomyxomas represent the prevailing form of primary cardiac neoplasm, predominantly observed in the adult population. Endocardial (mesenchymal) cells are thought to be the source of their origin. The occurrence of the typical myxoma is predominantly observed in the left atrium, accounting for 75% of instances. However, it is worth noting that 20% of cases emerge in the right atrium, while the other 5% manifest in the ventricles. The myxo ma is typically connected to the interatrial septum around the fossa ovalis through a stalk or pedicle. However, there have also been reports of attachments to the mitral valve. Tumors that originate in the left atrium (LA) have a tendency to infiltrate the atrial lumen, resulting in the manifestation of mitral stenosis or mitral regurgitation. This condition, sometimes referred to as the 'wrecking ball' phenomenon, leads to the destruction of the valve due to tumor protrusion. The presence of left atrial tumors might potentially mimic the symptoms of mitral valve disease, leading to the development of heart failure and/or secondary pulmonary hypertension. [16].

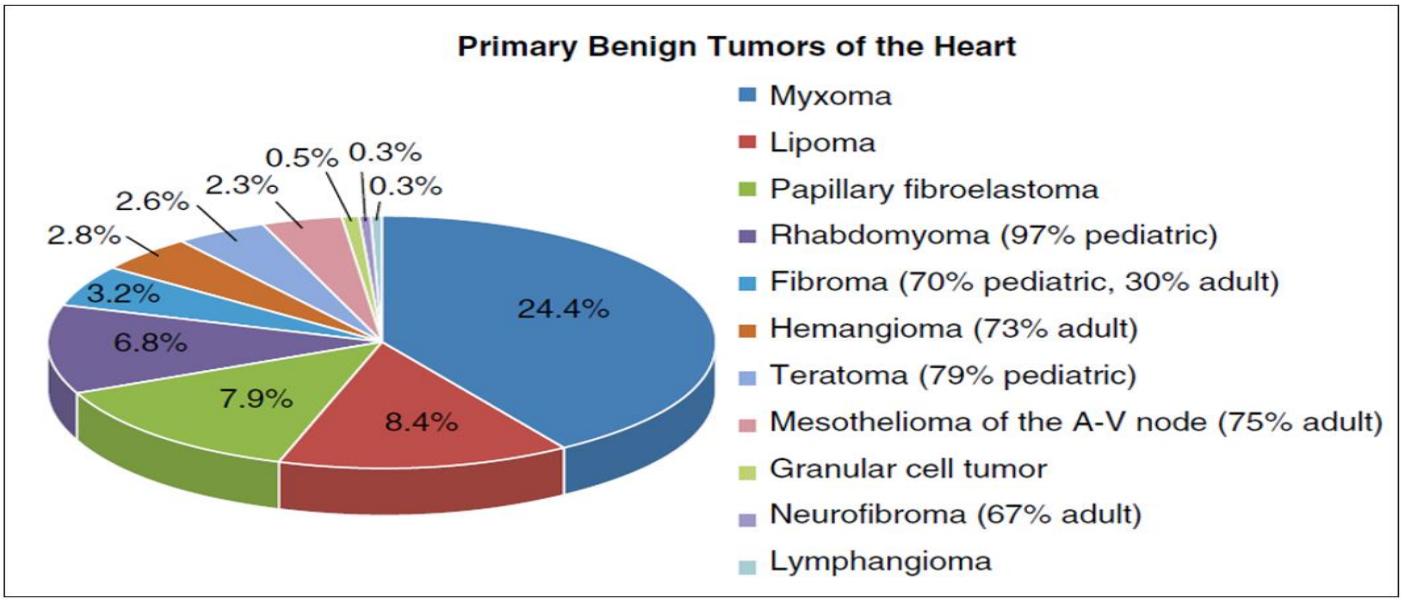

Fig 9 Primary Benign Tumors of the Heart

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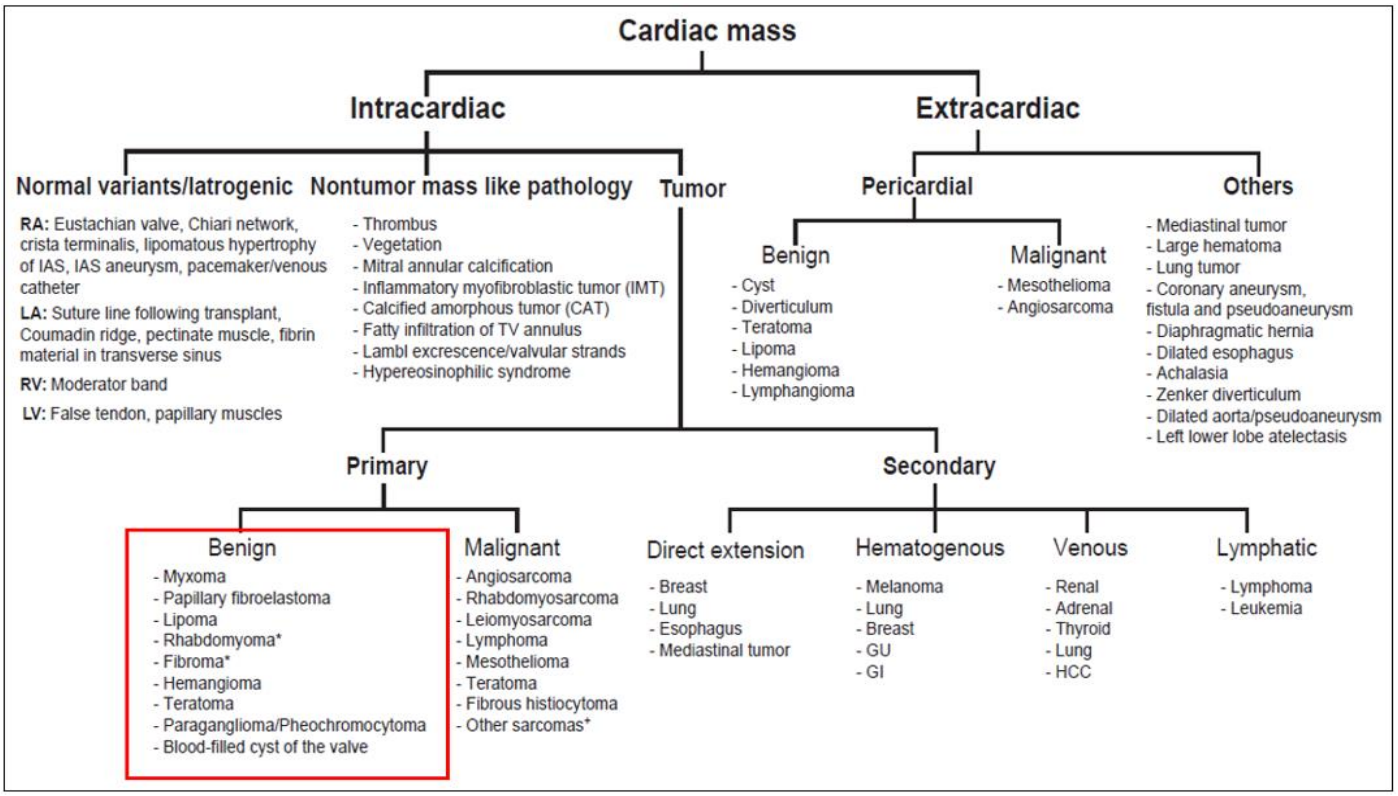

Fig 10 Overview the Classification of Cardiac Mass and Normal Variants

There are two fundamental classifications for cardiac masses: secondary and primary. Cardiac tumors generally do not exhibit clear signs and symptoms unless they interfere with the regular operation of the heart. Therefore, it is approximated that only 5 to 10 percent of cases may be classified through clinical assessment, while the remainder cases may be inadvertently recognized during the evaluation of seemingly unrelated illnesses or through physical observations. Patients who have been diagnosed with cardiac tumors may display symptoms that are associated with the cardiovascular system. These symptoms include obstruction, disruption of valvular structures resulting in regurgitation, direct invasion of the myocardium leading to impaired contractility, arrhythmias and conduction disorders, pericardial effusion, or embolization. The aforementioned symptoms may also possess constitutional characteristics [17], [18].

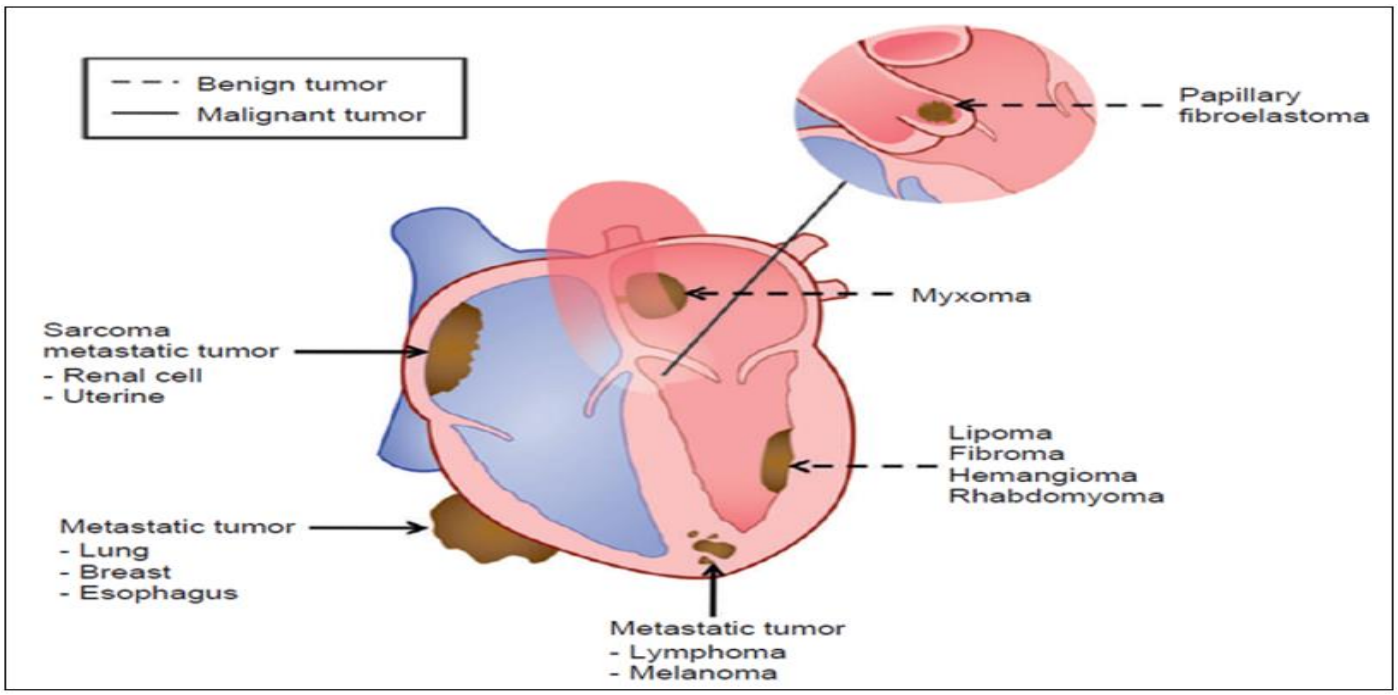

Fig 11 Schematic of the Typical Locations for Common Tumors in the Heart

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The patient presented at the hospital with neurological impairments in the upper and lower right limbs, as well as occasional dyspnea. These findings indicate that the neurological impairment may be attributed to the cardiac condition, specifically atrial myxoma. Left atrial pedunculated myxomas have the ability to move freely in response to blood flow entering and exiting the mitral valve, particularly due to the force of gravity. This movement can potentially interfere with the valve's functionality. When the myxoma obstructs the valve's functionality, there is an instantaneous cessation of blood flow, potentially leading to pulmonary congestion. The mitral valve may sustain injury, leading to the occurrence of backflow and subsequently causing a heart murmur. Nevertheless, it is possible for either blood clots originating from the surface of the myxoma or fragments of the myxomas to detach and subsequently result in obstruction within the blood artery [19]. The obstruction has the potential to result in cerebrovascular stroke, pulmonary embolism in the pulmonary vessels, and acute limb ischemia in the vessels of the lower extremities. In the present study, the patient exhibited symptoms of sporadic headache, dizziness, fatigue, and nausea. The observed symptoms may potentially indicate constitutional manifestations of myxoma, as well as the presence of emboli or thrombi resulting in small cerebral strokes. The physical examination revealed the presence of an early diastolic murmur grade 2/6 during cardiac auscultation. Additionally, the motoric strength of the upper and lower right limbs was measured at 3 locations. The stroke score obtained from the National Institutes of Health Stroke Scale (NIHSS) was 5. The occurrence of neurological manifestations in individuals diagnosed with atrial myxoma ranges from 25% to 45%. These manifestations can appear as secondary cerebral infarction, cerebral hemorrhage, and, less commonly, subarachnoid hemorrhage. Syncopes (28%) and epileptic fits (12%) are additional neurological signs that have been documented [18], [19]. The primary cause of neurological symptoms associated with myxomas is cardioembolic events, which can be initiated by the movement of a tumor fragment or the detachment of a clot from the tumor. The myxoma is connected to the interatrial septum (IAS) by a stalk and can be sufficiently large to result in obstruction of the atrioventricular (AV) valve (rarely regurgitation) due to the tumor protruding over the annulus during ventricular filling diastole [17].

Table 1 Clinical Features in Intracavity Tumors

Location	*Clinical features*	*Symptoms*	*Examples*
Left atrial	Left ventricular inflow obstruction	Dyspnea, paroxysmal nocturnal dyspnea, orthopnea, syncope, sudden cardiac death (SCD) (may have postural variation) especially myxomas	Myxoma Fibroma Undifferentiated sarcoma Osteosarcoma
	Embolism—central nervous system, coronary, peripheral, retinal	Stroke, myocardial infarction (MI)	
Left ventricle	Left ventricular outflow obstruction	Angina, syncope, SCD, murmur on examination	
	Systemic embolism	Stroke, SCD, MI	Papillary fibroelastoma
	Intramural—arrhythmias	Ventricular tachycardia, ventricular fibrillation, atrioventricular blocks	Fibroma
Right atrium	Benign superior vena cava syndrome (obstruction)	Right heart failure, i.e. peripheral edema, ascites	Myxoma
	Tricuspid stenosis/regurgitation	Murmur increasing with respiration	Angiosarcoma
		Cyanosis, R-L shunt through patent foramen ovale	
Right ventricle	Right ventricular inflow and outflow obstruction	Dyspnea, appropriate murmurs, syncope	Fibroma
	Pulmonary embolism	Right heart failure	Rhabdomyoma
		Pulmonary hypertension	Hamartoma

Table 2 Diagnostic Triad in Presentation of Atrial Myxoma [20]

Feature	Manifestations	Frequency, % of patients
Obstructive symptoms[6,7]	Heart failure, dyspnea, syncope, sudden death (rare)	54–95
Constitutional symptoms[3,6]	May mimic autoimmune disease or vasculitis: myalgia, arthralgia, weight loss, fatigue, fever, Raynaud's phenomenon, finger clubbing	34–90
Embolic phenomena[6-8]	Emboli may travel to any organ, but 73% reach central nervous system, including spinal cord	10–45

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The incidence of ischemic stroke in individuals under the age of 18 is exceedingly uncommon. Myxomas with an irregular surface had an increased propensity to induce embolisms. It is considered that fracturing small fragments of Myxoma can lead to embolic events. The irregular surface facilitates the fragmentation of Myxomas, resulting in an increased number of contact sites and subsequent embolism. The neurological manifestations associated with Myxomas primarily stem from cardioembolic events, which can be attributed to either the migration of a tumor fragment or the detachment of an attached clot from the tumor. Ischemic conditions are characterized by diminished blood flow and oxygen delivery, resulting from an embolism inside the central nervous system. This reduction in blood flow ultimately culminates in the occurrence of a stroke. The secretion of vascular endothelial growth factor (VEGF) by Myxoma plays a crucial role in promoting angiogenesis in the early stages of tumor progression. At a macroscopic level, Myxomas manifest as masses that exhibit yellowish, white, or brown hues. Upon examination, it is common to observe the presence of thrombi covering these masses. The surface of the Myxoma is adorned with a multitude of intricate or exceedingly intricate, gelatinous villi characterized by fragile extensions that exhibit a proclivity for spontaneous rupture and are associated with embolic occurrences [8], [21]. In this particular instance, the utilization of transthoracic and trans esophageal echocardiography unveiled the presence of a pedunculated mass measuring 54 mm × 30 mm within the left atrium. This mass exhibited an uneven shape and possessed a friable surface, extending towards the mitral valve. Echocardiography data reveal two distinct anatomical manifestations of atrial Myxomas. The initial manifestation is characterized by a solid and circular shape with a stationary surface, whereas the subsequent manifestation exhibits a poly poid, soft, uneven form and a surface that is prone to fracturing. The latter phenomenon is correlated with an increased likelihood of embolization, reaching a maximum of 58%. Had echocardiography been conducted during that period, it is plausible that cardiac Myxoma could have been identified at an earlier stage and at a reduced size. Upon admission, echocardiography was promptly conducted following the manifestation of neurological dysfunction. Histopathological investigation is necessary to distinguish cardiac Myxoma from other potential diagnoses, including intra-cardiac thrombus, papillary fibroelastoma, lipoma, and rhabdomyoma, in addition to echocardiography [8], [21].

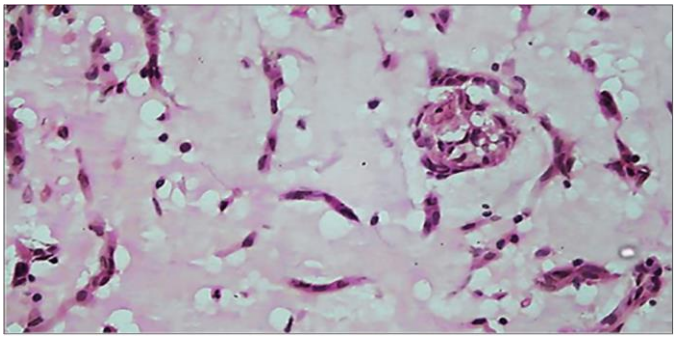

Fig 12 Photomicrograph of the Surgical Specimen Showing the Microscopic Features of Atrial Myxoma. Stellate Shaped Myxoma cells Embedded in a Myxoid Stroma

Surgical surgery is the exclusive therapeutic approach for cardiac cancers that need surgical intervention. Surgical intervention is advised for patients who exhibit any form of inlet or outlet obstruction, as well as those who manifest symptoms indicative of heart failure or ventricular arrhythmias that fail to improve with traditional therapeutic approaches. Once a diagnosis of Myxoma is confirmed through imaging studies, it is imperative to promptly perform resection in order to fully eliminate the Myxoma. This is crucial due to the potential hazards of embolization or cardiovascular complications, such as sudden death. Prompt surgical intervention is crucial to minimize the risk of future tumor embolism and valve obstruction, which are highly consequential complications. Prompt removal of cardiac Myxoma is crucial in order to mitigate the risk of life-threatening systemic embolism. Nevertheless, the existing guidelines lack clarification regarding the precise timing of cardiac surgery or the administration of preoperative anticoagulation and antithrombotic medication. This omission can be attributed to the scarcity of reported instances of Acute Ischemic Stroke associated with atrial Myxoma. There exists a substantial association between a prolonged time period between stroke and tumor removal surgery and the occurrence of stroke recurrence [14], [17]. In this instance, the patient received antiplatelet medication and citicolin from the neurology department, without undergoing further surgical intervention. The occurrence of cardiac Myxoma in pediatric patients is infrequent; however, it can occasionally lead to ischemic stroke and other problems arising from thrombi or emboli. Prompt and accurate diagnosis is vital for children, as the most efficacious treatment currently involves the immediate excision of the tumor. Based on our data, it is recommended to perform echocardiography in cases where children display abnormal neurological deficits.

V. CONCLUSION

Atrial Myxomas are the most prevalent primary cardiac tumors. Patients with atrial Myxoma may exhibit a range of signs and symptoms. Stroke is a non-cardiac symptom of atrial Myxoma. Therefore, it is imperative to assess the existence of atrial Myxoma in individuals who have experienced a stroke. TTE and TTE have a crucial role in the diagnosis of atrial Myxoma. Ischemia is a medical disorder characterized by a decrease in blood flow and oxygen delivery. A stroke occurs in the central nervous system due to diminished blood supply following an embolism. An ischemic stroke, a prevalent sign of atrial Myxoma, can occur in any location. The occurrence of an ischemic stroke results in a significant impairment of functional abilities.

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