JCRMHS

Case report

Journal of Clinical Case Reports, Medical Images and Health Sciences

Volume 9 Issue 4, 2025

Article Information

Received date: 15/02/2025

Published date: 07/03/2025

*Corresponding author

* Caio Cesar Cardoso, Federal University of Sao Paulo, Brazil.

Left Atrial Myxoma in a Patient with Pectus Excavatum: Case Report and Analysis of Surgical Approach

Caio Cesar Cardoso*, Dirceu Oscar Faelli Jr, Josiane de Souza Bezerra, Enzzo Vinícius Dias Queiroz

Federal University of Sao Paulo, Brazil.

Introduction

Myxoma is the most prevalent primary cardiac neoplasm, accounting for 50% of benign heart tumors in adults (1). Elderly women are more predisposed to the development of atrial myxomas, with a ratio of 3:1 between women and men, and left atrial myxomas are the most common (2,3). These left-sided tumors predominantly originate in the interatrial septum, at the edges of the fossa ovalis, and can mimic diseases of the mitral valve, presenting a physical examination similar to that observed particularly in valve stenosis (4,5).

Cardiac myxoma does not have a formally declared classification. However, the literature indirectly suggests it can be categorized based on tumor characteristics, including anatomy, size, location (right and/or left), morphological aspects, histology, mobility, and non-malignant neoplasms (6). Between 10% and 50% of myxomas are diagnosed incidentally during clinical exams (3). Myxomas larger than 4 cm have been associated with patients who present constitutional symptoms that mimic inflammatory disorders or connective tissue diseases, such as malaise, fever, and weight loss. These symptoms are more prevalent in women and in cases of large or multicentric myxomas (3,7). Tumors of 5 cm or more may suggest more severe cases.

Pectus excavatum is a common congenital deformity of the chest wall, with an incidence of 1 in every 300 to 400 births, showing a ratio of 2:1 between men and women (8). This condition, characterized by a concavity that brings the sternum and costal cartilages closer to the spine, is often asymptomatic, though it may present aesthetic deformities and/or breast asymmetry. In the presence of associated diseases, clinical manifestations may arise (9). Although there is no scientifically proven association between pectus excavatum and cardiac myxoma, it is observed that patients with pectus excavatum have a higher frequency of cardiac alterations.

Case Report

A 73-year-old female patient, Brazilian with parents born in Japan, with no known comorbidities, presenting pectus excavatum detected on physical examination. She reported a 2-year history of dyspnea on major exertion, which progressed over the last 3 months to dyspnea on minor exertion and orthopnea.

After clinical and complementary investigation, a left atrial myxoma was identified via transthoracic echocardiogram. Cardiac magnetic resonance imaging (MRI) was requested, with images shown below (Figure 1):

Given the clinical condition, surgical resection was indicated. Due to the presence of pectus excavatum, an anterolateral right thoracotomy approach was chosen, with the installation of extracorporeal circulation, cardioplegia, and access via left atriotomy.

The images below reveal the intraoperative appearance of the left atrial

DOI: 10.55920/JCRMHS.2025.09.001397

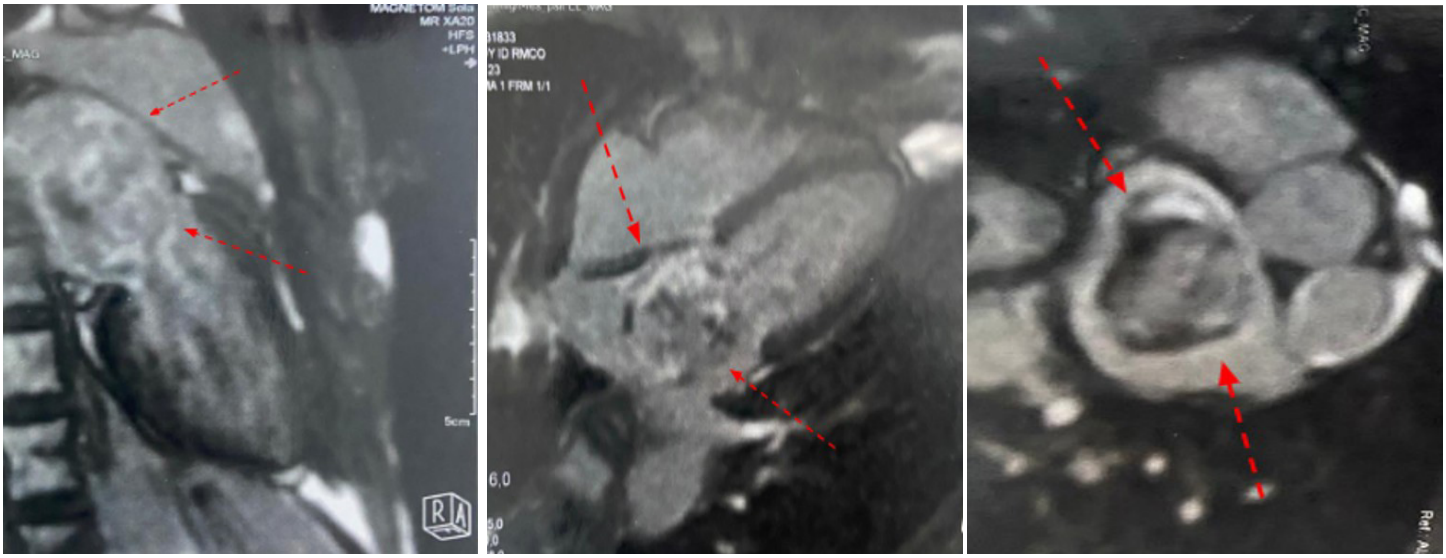

Figure 1: *Images from cardiac magnetic resonance imaging showing a tumor formation approximately 5 cm in diameter in the left atrium (red arrows), pedunculated in the interatrial septum, with a homogeneous structure and calcified borders, suggestive of a myxoma, occupying nearly the entire atrial cavity.*

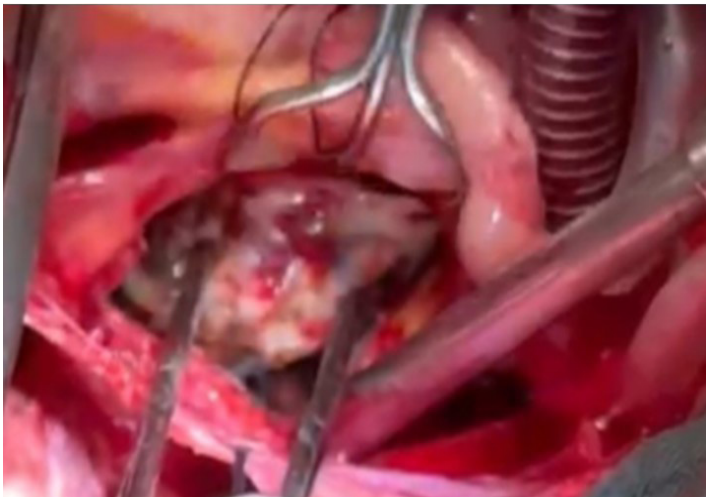

Figure 2: *Intraoperative view after left atriotomy, revealing, between the jaws of the anatomical clamp, a calcified tumor occupying almost the entire left atrium.*

tumor (Figure 2).

Resection of the tumor from the atrial walls was performed, along with resection of the interatrial septum, where the tumor was pedunculated (Figure 3).

After the resection of the tumor and the membranous portion of the interatrial septum, the correction of the resulting septal defect was performed using bovine pericardium, as shown in the image below (Figure 4).

The surgical procedure was completed with closure of the left atriotomy, placement of temporary epicardial pacemaker wires in the right atrium and right ventricle, release of aortic clamping, discontinuation of extracorporeal circulation, drainage of the right anterior and posterior pleura, and closure of the thoracotomy, with the image below showing the final appearance (Figure 5).

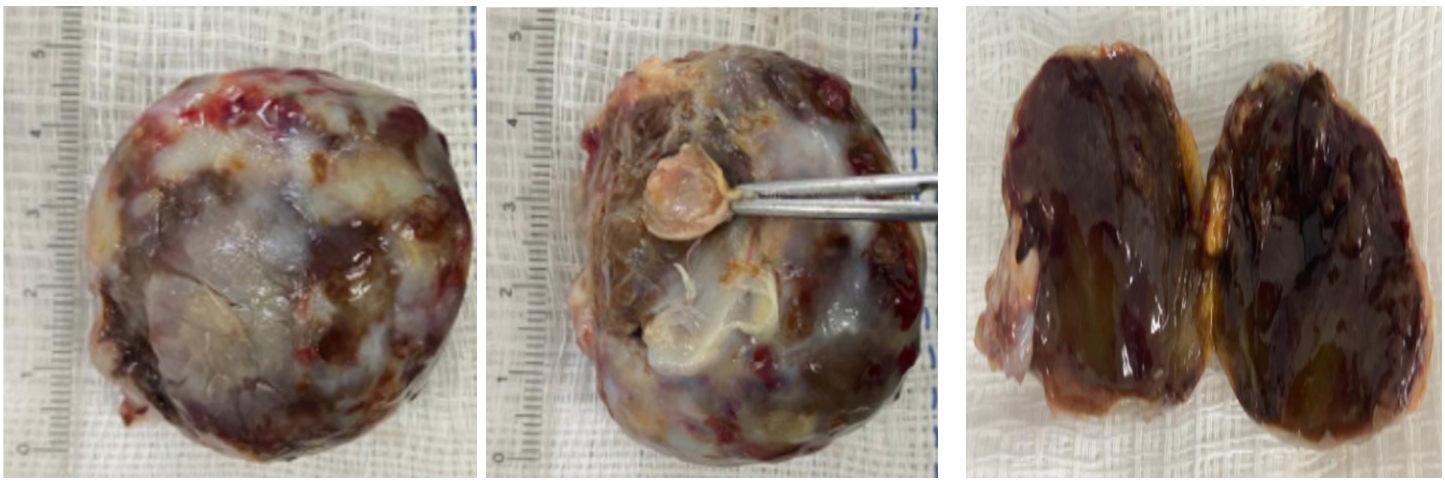

Figure 3: *Images related to the resected left atrial tumor. A. External appearance, showing pearly regions corresponding to calcification. B. Close-up, in the anatomical clamp, of the tumor's pedicle attached to the interatrial septum. C. Longitudinal opening of the tumor formation, revealing the calcified borders and the homogeneous appearance of the tumor.*

DOI: 10.55920/JCRMHS.2025.09.001397

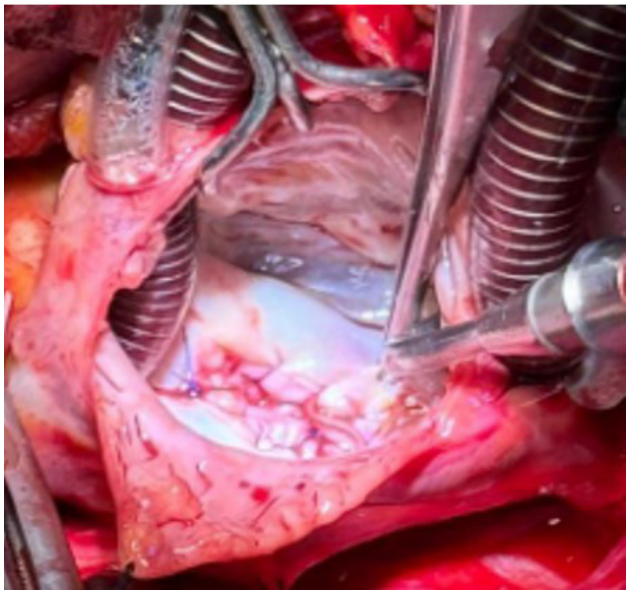

Figure 4: *Final aspect after correction of the septal defect resulting from the resection of the tumor and the fossa ovalis of the interatrial septum.*

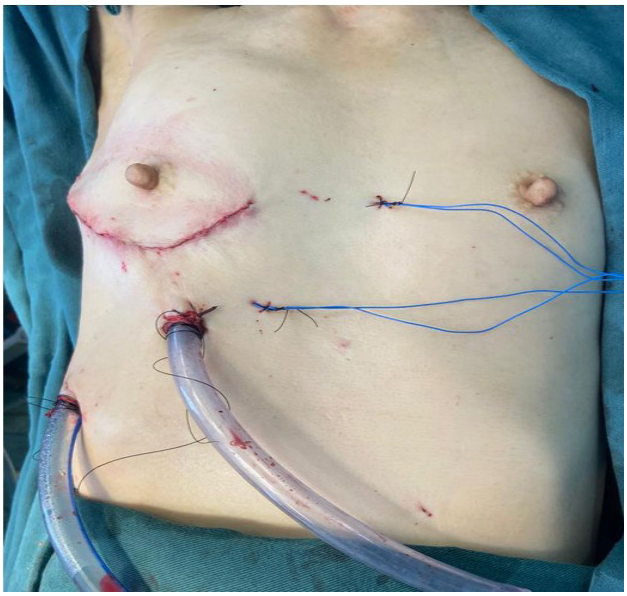

Figure 5: *Final appearance of the procedure, highlighting the patient's pectus excavatum and the incision in the right mammary sulcus for the surgical access via right anterolateral thoracotomy.*

The patient remained in the intensive care unit for 96 hours and was discharged from the hospital 7 days after the surgical procedure. At the 6-month follow-up, the patient was asymptomatic. The pathological examination of the surgical specimen confirmed the diagnosis of atrial myxoma.

Discussion

There is no correlation between myxoma and pectus excavatum, as described in the literature, and the lack of relevant evidence reinforces the idea that these conditions are independent. It is observed that the conditions are distinct.

Primary cardiac tumors, although rare, have myxoma as the most common, with an approximate annual incidence of 0.5 per million people. The origin of atrial myxoma is not completely established, but according to the current studies, myxomas arise from mesenchymal stem cells located in the subendocardial tissue near the fossa ovalis. Its etiology, in most cases, is sporadic, representing approximately 93% of all forms, and they frequently appear as isolated lesions in the left atrium in 75% to 85% of cases, with a low recurrence rate. Additionally, there is also a subgroup of familial myxomas associated with the Carney Complex, an autosomal dominant genetic syndrome (6,7).

On the other hand, pectus excavatum also has several theories regarding its etiology, the most recognized being excessive growth of the costal cartilages, which displaces the sternum posteriorly, causing the chest depression. This anomalous process is not fully understood, but it is known that the ossification centers of the sternum and the histological structure of the costal cartilages show normal patterns (9). "Funnel chest" is a congenital condition, often isolated, but it can also be associated with genetic syndromes of connective tissue disorders, such as Marfan syndrome or Ehlers-Danlos syndrome, which may present with this thoracic abnormality (10). Usually this condition is asymptomatic; however the reduction in intrathoracic volume can lead to cardiac displacement, causing cardiopulmonary symptoms such as breathing difficulties, palpitations, and decreased daily exercise capacity (11)

Based on this information, it is observed that cardiac myxoma is not a typical manifestation of pectus excavatum, and the genetic syndromes of both conditions are unrelated. Therefore, it is clear that there is no connection between the etiological, cellular, or genetic mechanisms within the known pathophysiological processes, as myxoma and pectus excavatum occur in distinct ways.

Cardiac myxoma is a rare condition, with a prevalence of about 0.03% in the general population. The annual incidence of cardiac myxoma ranges from 0.05 to 0.16 per 100,000 people. In Japan, the incidence rate of cardiac myxoma has remained unchanged over the past 20 years, showing no significant variation during this period (12).

According to the literature, although no specific racial predisposition has been identified, the deformity is more common in individuals of Caucasian descent, and less common in Asians, African Americans, or Hispanics. The incidence of "funnel chest" can reach 8 cases per 1,000 live births. However, the true incidence and prevalence of pectus excavatum are still not fully known and may be much higher than the currently recorded numbers (up to 5%, according to radiological reports), as large-scale population studies on the subject have not been conducted (10).

Therefore, since no studies have established a causal or associative relationship between myxoma and pectus excavatum, it is concluded that these conditions coexist rarely and without direct connection, reinforcing that their coincidence in an individual would be explained by chance, affecting them independently.

Cardiac Magnetic Resonance Imaging (CMRI) is essential for the accurate diagnosis of cardiac tumors, as it provides information on the heart's structure and hemodynamics. Additionally, it aids in surgical planning by determining the location, extent, and impact of lesions, allowing for a more effective therapeutic approach and a more positive prognosis. Moreover, it can generate detailed additional data on the heart's structure and hemodynamics, complementing the results of echocardiography. While echocardiography is commonly used as an initial test for identifying cardiac masses, it may be difficult to differentiate myxomas from other cardiac lesions, such as thrombi (13). CMRI functions as an essential non-invasive method for diagnosing primary cardiac tumors, offering images with better contrast resolution, a larger field of view, and the unique ability to distinguish lesions based on tissue characteristics. This exam also assists in determining the location, extent, and functional impact of the lesion. Currently, CMRI is the most reliable and accurate imaging technique, considered the "Gold Standard" for evaluating cardiac tumors, providing superior quality in terms of contrast resolution and tissue detail compared to other tests, such as transesophageal echocardiography. Thus, CMRI is essential for obtaining a conclusive diagnosis of cardiac masses, as well as determining therapeutic planning and patient prognosis. Large, benign tumors that cause symptoms are typically treated with complete surgical removal, and in most cases, individuals adapt satisfactorily to the surgical procedure. Patients with benign masses have a life expectancy comparable to the general population (14).

Currently, surgical removal of the atrial myxoma is the most recommended treatment, as there are no medications that can prevent tumor growth (6). Symptomatic myxomas need to be surgically removed as soon as possible after diagnosis, and the surgical approach should allow for minimal manipulation of the tumor, provide adequate access for complete lesion removal, including the pedicle area, allow assessment of all four heart chambers, and be both safe and effective. It is generally observed that the prognosis for cardiac myxoma surgery is very favorable, with mortality rates below 3% (5).

The patient's prolonged clinical course reflects the insidious progression of the atrial myxoma growth. Furthermore, the external calcification process and adhesions of the tumor to the left atrial wall support the extended evolution time of the condition before the patient sought medical assistance.

The choice of access to the thoracic cavity via right anterolateral thoracotomy was made due to both the pectus excavatum and the satisfactory access to the left atrium, the cardiac cavity of interest for tumor resection. The sternal reconstruction after a median sternotomy in a pectus excavatum patient might lead to a challenging and unique for the repair; the sternal adhesions and risk of cardiac injuries must be considered (15). In this case report, there was not the purpose of correction of pectus excavatum due to age, asymptomatic presentation and no patient's desire.

References

1. Rabiee Rad M, Ghasempour Dabaghi, G, Darouei, B (2024). Clinical and laboratory manifestations, ECG findings, and outcomes of right atrial myxoma: a systematic review of cases reported worldwide. Egypt Heart J 76, 125.
2. Bandaru V V, Talbot S L, Garcia R I, (2024). Year long Near-Syncope: Presentation of a Rare Right Atrial Myxoma. Cureus 16(4): e59070. ISLAM, A. K. M. Monwarul (2022). Cardiac myxomas: A narrative review. World J. Cardiol. , Dhaka, v. 14, n. 4, p. 206-219.
3. JATENE, Ieda B, FERREIRA, João Fernando M. ; DRAGER, et al (2022). SOCESP cardiology treatise. 5th edition. Barueri: Manole. E-book. p. Cover. ISBN 9786555765182.
4. Numaguchi, R. , Hashimoto, M. , Koshima, R. (2020). Surgical treatment of a giant right atrial myxoma. Surg Case Rep 6, 165.
5. Gao A, Yang J, Tian T, Wu Y, Sun X, et al (2023) Visual analysis based on CiteSpace software: a bibliometric study of atrial myxoma. Front. Cardiovasc. Med. 10: 1116771.
6. Cianciulli TF, Cozzarin A, Soumoulou JB, Saccheri MC, Méndez RJ, et al (2019). Twenty years of clinical experience with cardiac myxomas: diagnosis, treatment, and follow-up. J Cardiovasc Imaging. Jan; 27(1): 37-47.
7. Carroll MK, Powell AW, Hardie WD, Foster KE, Zhang B, et al (2024). Pectus excavatum: the effect of tricuspid valve compression on cardiac function. Pediatr Radiol. Aug; 54(9): 1462-1472.
8. MENDES, Paulo Roberto da Silva (2017). Pectus excavatum and breast asymmetry: correction with breast augmentation. Brazilian Journal of Plastic Surgery, v. 2, p. 225-230.
9. Janssen N, Coorens NA, Franssen AJPM, Daemen JHT, Michels IL, et al (2024). Pectus excavatum and carinatum: a narrative review of epidemiology, etiopathogenesis, clinical features, and classification. J Thorac Dis. Feb 29; 16(2): 1687-1701.
10. Norlander L, Anderzén-Carlsson A, Vidlund M, Sundqvist AS (2024) Experiences of living with funnel chest before corrective surgery: an interview study. PLoS ONE 19(7): e0304968.
11. Sega M, Yamashita M, Maruyama H, Taya Y, Ohgi K, et al (2024). Renal Embolism Associated with the Atrial Myxoma: A Case Report and Literature Review. Medicina (Kaunas). Apr 24; 60(5): 694.
12. Leivaditis V, Beltsios ET, Papatriantafyllou A (2024). Acute decompensated heart failure secondary to left atrial myxoma: a case report highlighting diagnostic challenges and multidisciplinary treatment. Cureus 16(7): e65177.

13. Haider I, Ullah H, Fatima M, Karim MS, Haq FU, et al (2023). Tissue characterization of benign cardiac tumors by cardiac magnetic resonance imaging, a review of core imaging protocol and benign cardiac tumors. Front Cardiovasc Med. Jun 27; 10: 1009411.

14. Jaroszewski DE, Gustin PJ, Haecker FM, Pilegaard H, Park HJ, et al (2017). Pectus excavatum repair after sternotomy: the Chest Wall International Group experience with substernal Nuss bars. Eur J Cardiothorac Surg. Oct 1; 52(4): 710-717.