ISSN- O: 2458 - 868X, ISSN–P: 2458 – 8687
National Library of Medicine - ID: 101731606

International Journal of Medical Science and Innovative Research (IJMSIR)
IJMSIR : A Medical Publication Hub
Available Online at: www.ijmsir.com
Volume – 9, Issue – 6, December – 2024 , Page No. : 121 – 124

Cardiac Myxoma- A Medical Enigma

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Citation this Article: Veerpal Kaur, Sukhjinder Kaur, "Cardiac Myxoma- A Medical Enigma", IJMSIR - December - 2024, Vol – 9, Issue - 6, P. No. 121 – 124.

Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

Cardiac myxomas are rare primary cardiac tumours presenting with diverse range of symptoms. Generally, the patients recover well if these tumours are diagnosed early and timely surgical intervention is provided. We present a case of an 85-year-old female who presented with complaints of chest pain and palpitations since 2 months. A Transthoracic Echocardiogram revealed a large mass attached to the interatrial septum protruding into the left ventricle. The possibility of intracardiac thrombus was given. The mass was excised and sent for histopathology which confirmed it as cardiac myxoma. However, the patient couldn't survive post surgical resection.

Keywords: Atrial myxomas, cardiac tumours, benign, primary.

Introduction

Cardiac myxoma (CM) is a benign tumour, accounting for 30-50% of all the primary heart tumours, with an yearly incidence of 0.5 per million population. Mostly, these tumours show female predominance and occur in third to sixth decades but age is no bar. These tumors frequently develop in the left atrium followed by right atrium, few arise in both atria and rarely in ventricles. CM can be smooth, round, polypoid, papillary or irregular in appearance. They can be sessile or pedunculated with diameter ranging from 1-15 cm. These tumours arise from multipotent mesenchymal cells in the subendocardial space.

Atrial myxoma can present with diverse range of symptoms such as fever, malaise, anorexia, arthralgia, weight loss, obstructive symptoms leading to dyspnoea, chest pain, palpitations or can embolise resulting in syncope and stroke. [1]

Rarely, atrial myxomas can occur as component of the carney complex exhibiting unusual skin pigmentation and tumours in endocrine tissues such as adrenal glands, thyroid, testes, and ovaries. Surgical resection is the primary treatment for CM. [2] Echocardiography is the first-line imaging technique for diagnosis. CT scans or cardiac MRI may be needed for detailed evaluation. Histopathological assessment confirms the diagnosis. [3]

An interatrial myxoma was diagnosed in an 85 year old lady in our hospital. When she presented to the emergency department, she was suspected of carrying an atrial thrombus being responsible for her chest pain and palpitations but when the mass was finally excised following laboratory and radiological investigations, it

was diagnosed as cardiac myxoma histopathologically. Being a private tertiary care hospital, autopsies are not conducted in our hospital, so the number of cardiac specimens are limited to those patients who are operated for serious heart ailments. As such the cardiac myxomas are rare entity on their own, hence it was an incidental finding in cardiac mass received post-operatively in our department.

Case Presentation

An 85-year-old female presented to the Emergency department of Adesh Medical College and Hospital, Shahbad (Ambala) with chief complaints of chest pain and palpitations since 2 months. The chest pain was gradual in onset, progressive and aggravated since 15 days. She was a known case of type 2 diabetes and hypertension with no family history of heart disorders. On arrival, the patient was irritated but conscious to time, place and person. The vitals were stable. On auscultation, bilateral normal bronchovesicular breath sounds heard. Abdomen was soft and no organomegaly was noted on palpation. However, Cardiovascular system examination revealed loud S1, soft S3 with a plopping sound in the mitral area and a mid-diastolic murmur. A Transthoracic Echocardiogram revealed a large mass attached to the interatrial septum which protruded into the left ventricle during diastole, interatrial septum thickness=1.2 cm, Ejection fraction=45%, mitral valve moved posteriorly during diastole, concentric left ventricular hypertrophy and akinesia of IVS. A coronary angiogram showed no coronary artery disease.

She was referred to the CTVS/plastic surgery department for emergency atrial mass excision and the excised mass was sent to the pathology department. The histopathology department received brown black globular soft and friable soft tissue mass measuring 4*3*2 cm. (figure 1). On cut section, gelatinous and hemorrhagic areas were identified. Microscopically, sections examined revealed a benign tumour comprising of stellate shaped myxoma cells lying singly and also forming cords and nests at places. (figure 2). Intervening stroma showed myxoid changes, extensive areas of hemorrhage and hemosiderin laden macrophages. Presence of Gamna gandy bodies and multiple foci of lymphoplasmacytic infiltrate were also noted. No mitosis/ necrosis/atypia was noted. Gamna gandy bodies stained blue with Perl's stain.(figure 3).

Post-operatively, the patient developed wound dehiscence 2 days later for which bilateral pectoralis major muscle flap was done. She was shifted to CTVS ward for further management and regular dressings were done. The patient had wound dehiscence 9 days later for which rectus abdominis flap was done under general anaesthesia. Regular dressings were done but she had discharge from the wounds for which culture sensitivity was sent and antibiotics were given accordingly. Laboratory investigations were suggestive of sepsis and Acute Kidney Injury. Hence, the patient was taken for wound debridement under general anaesthesia. She was shifted to Intensive Care Unit and relatives were informed about the poor prognosis. One day later, her condition deteriorated as she was hypotensive and developed oliguria. She was put on ionotropic support. She had cardiopulmonary arrest, three cycles of Cardiopulmonary resuscitation were given. Unfortunately, despite all efforts, she couldn't be revived and she succumbed to death.

Discussion

The cardiac myxoma is the commonest benign cardiac tumor, followed by fibroelastoma, fibromas, rhabdomyoma, hemangioma, and lipomas. Metastatic cardiac tumors are more frequent than primary tumors. Primary malignant cardiac tumors are also rare and can

be broadly divided into sarcomas, malignant lymphomas, and pericardial mesothelioma.[4]

Sporadic and familial cases have been identified. Mid-aged women are frequently affected with tumor in left atrium in the sporadic cases whereas in the familial variety, young men are commonly affected and the tumours are less commonly located in the left atrium. [5]

These tumours present with non-specific symptoms or with obstructive or embolic complications. However, the clinical presentation of primary cardiac tumors depends on their site, size, mobility and infiltration of adjacent structures. The obstructive features include left and right sided heart failure, with dyspnea, orthopnea, paroxysmal nocturnal dyspnea, ascites, hepatomegaly, peripheral edema, intermittent syncope, dizziness, or sudden death. These tumors have the tendency to fragment and embolize to various organs, most commonly to cerebral and retinal arteries. Up to 12% of these tumors are diagnosed incidentally or at postmortem examination.[7]

Trans-Thoracic Echocardiography (TTE) or Trans-Esophageal Echocardiography (TEE) are useful modalities to detect cardiac tumors. However, both TTE and TEE cannot provide differentiation between thrombus and tumors. Multi-Detector Computed Tomography (MDCT) scanners provide excellent anatomic details about cardiac muscle with coronary artery configuration. Due to high radiation exposure with MDCT, Cardiac Magnetic Resonance (CMR) imaging has evolved as new tool for assessment of the tumour.[4]

Histologically, these tumors can undergo secondary changes such as fibrosis, thrombosis, and calcification or can demonstrate atypical histologic and cytologic features like nests of epithelioid cells, foci of glandular cells, and atypical lymphoid cells associated with Epstein-Barr virus (EBV), ectopic presence of thymic nests which can undergo malignant transformation very rarely. [6]

Cardiac myxomas which show the presence of gamna gandy bodies and fibrosis most commonly present with non-embolic clinical features as seen in our case. [8]

Immunohistochemically, about 80%–90% of CM express vimentin, Notch1, α smooth muscle actin (α-SMA), calretinin, caldesmon and tenascin C. Prognosis of primary benign cardiac tumors is excellent following surgical excision. [7]

Recurrence of myxomas is very rare and the risk is correlated with young age, family history of myxoma, inadequate resection, intraoperative implantation, or multicentre growth. [9

Conclusion

Early diagnosis and timely surgical resection of cardiac myxomas can save the life of patients. Post operatively these patients perform well with rare instances of tumor reccurence. In our case, the patient didn't experience heart issues till old age but her health worsened over a short span of time. Although the prognosis of these tumours is generally good after resection but old age, pre-existing comorbidities leading to immunocompromised state might have contributed to complications post operatively leading to her death.

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Legend Figures

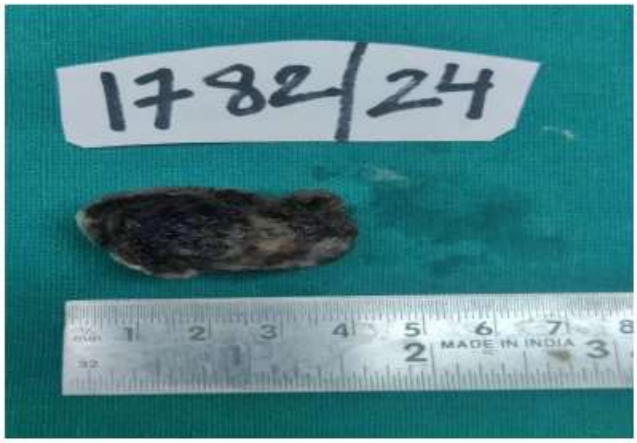

Figure 1: Gross specimen of cardiac mass

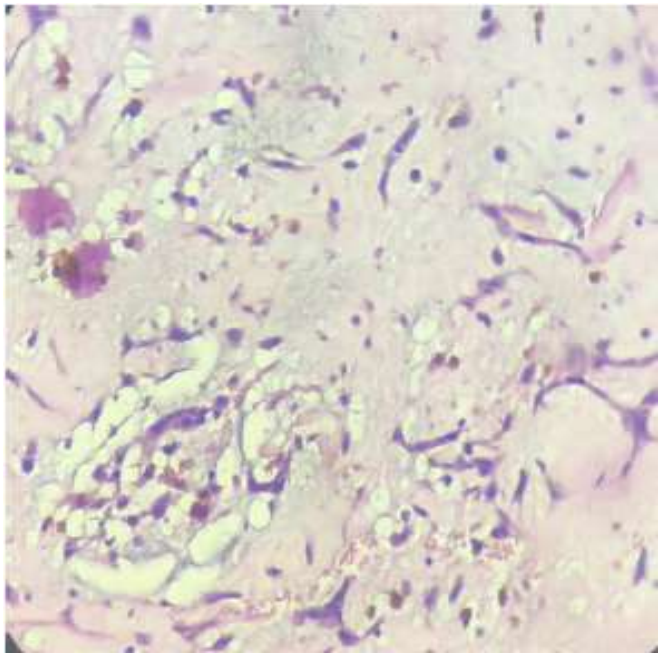

Figure 2: stellate shaped myxoma cells

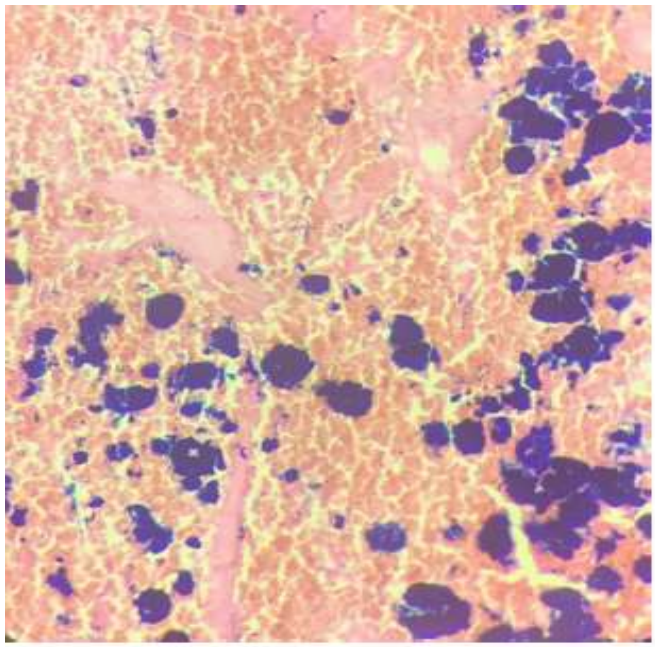

Figure 3: Gamna gandy bodies (blue): Perl's stain