DOI:10.31557/APJCN.3251.20260826

CASE REPORT

Clinical Characteristics and Histopathological Features of Cardiac Myxoma: A Case Report and Review of the Literature

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Abstract

Background: Primary cardiac tumors are extremely rare, with an estimated prevalence of 0.001%–0.03% in the general population. Approximately 75% of these tumors are benign, and cardiac myxoma (CM) represents the most common type. These tumors may originate from any cardiac layer or valve and can present with a wide range of clinical manifestations depending on their location. In some cases, they remain asymptomatic and are detected incidentally. **Case Presentation:** A 43-year-old female presented with a two-week history of progressive dyspnea and persistent dry cough. Echocardiography revealed a large, mobile, pedunculated mass in the left atrium attached to the interatrial septum. Following surgical excision, histopathological examination confirmed a benign left atrial myxoma measuring 4.8 × 3.2 × 3.6 cm. Microscopic examination demonstrated spindle-shaped, ovoid, and stellate cells embedded within a myxoid stroma, accompanied by extravasated erythrocytes, hemosiderin-laden macrophages, and dystrophic calcification. No features of malignancy were identified. **Conclusion:** Cardiac myxoma is a rare but important benign cardiac tumor, most commonly occurring in the left atrium. Clinical manifestations vary considerably according to tumor location and size, ranging from asymptomatic cases to obstructive, constitutional, or embolic presentations. Accurate diagnosis requires correlation between clinical findings, imaging studies, and histopathological examination.

Keywords: Cardiac myxoma, Primary cardiac tumor, Cardiac neoplasm, Histopathology, Carney complex

Asian Pac J Cancer Nursing, 21-23

Submission Date: 05/29/2026 Acceptance Date: 07/01/2026

Introduction

Primary cardiac tumors are extremely rare, with an estimated prevalence of approximately 0.001%–0.03% in the general population. Nearly 75% of these tumors are benign [1].

Cardiac tumors may arise from any layer of the heart, including the endocardium, myocardium, and pericardium, as well as from valvular structures. Their clinical presentations are highly variable, ranging from incidental findings in asymptomatic individuals to symptoms related to tumor location and size. Common manifestations include dyspnea, chest pain, palpitations, and embolic events such as stroke [2].

Echocardiography is considered the primary diagnostic modality for the initial evaluation of cardiac tumors, whereas magnetic resonance imaging (MRI) and computed tomography (CT) provide additional anatomical characterization and assist in surgical planning [3].

Cardiac myxoma (CM) is the most common primary cardiac neoplasm and accounts for approximately 75% of benign cardiac tumors. Although its exact cellular origin remains uncertain, current evidence suggests that CM develops from pluripotent subendocardial mesenchymal cells [4].

CM occurs more frequently in females and is commonly diagnosed between the fifth and seventh decades of life. Most cases are sporadic; however, approximately 10% of cases are associated with familial predisposition. Familial cardiac myxomas are frequently linked to genetic syndromes such as Carney complex [1].

The left atrium is the most common site of occurrence, accounting for approximately 75% of reported cases. Clinical manifestations depend on tumor size, mobility, and anatomical location. Patients may remain asymptomatic, whereas others may develop obstructive symptoms,

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including dyspnea caused by impaired intracardiac blood flow and pulmonary congestion. Constitutional symptoms, such as fever, fatigue, and anemia, occur in approximately 20% of cases and may mimic infectious endocarditis [2].

Case Presentation

An archived pathology specimen from a 43-year-old female patient admitted to Sohag University Hospital was reviewed. The patient had a two-week history of progressive dyspnea and persistent dry cough.

Clinical examination and transthoracic echocardiography revealed a large, heterogeneous, mobile, and pedunculated mass attached to the interatrial septum and protruding into the left atrium. The mass was surgically excised and submitted to the Pathology Laboratory of the same hospital for histopathological evaluation.

Gross examination revealed a solitary, polypoid mass measuring approximately 4.8 × 3.2 × 3.6 cm. The lesion showed a smooth, glistening external surface with focal areas of thrombus formation.

Microscopic examination demonstrated stellate, ovoid, and spindle-shaped cells with central round nuclei, bland cytological features, and indistinct cell borders. The cells were arranged in cords, tubules, and small aggregates within a myxoid stroma (Figure. 1). The stroma contained prominent extravasated erythrocytes and hemosiderin-laden macrophages. Degenerative changes, including dystrophic calcification, were also observed (Figure. 2). No cellular atypia, necrosis, or mitotic activity was identified. Based on the clinical, radiological, and histopathological findings, a diagnosis of left atrial myxoma was confirmed.

Discussion

Primary cardiac tumors are extremely rare, with the majority being benign. Cardiac myxoma (CM) is widely recognized as the most common primary benign cardiac tumor in clinical practice. Although papillary fibroelastoma has been reported as one of the most frequent benign cardiac tumors in recent classifications, several clinical series and retrospective studies have demonstrated that CM represents the predominant type of surgically treated primary cardiac tumor. In a multicenter

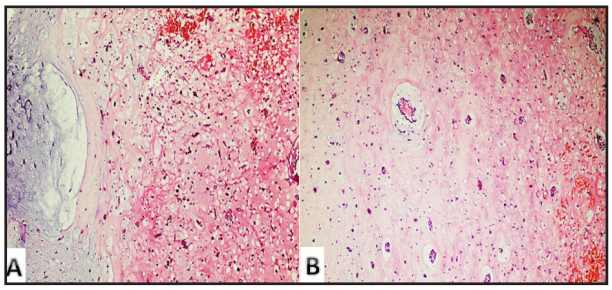

Figure 1. H&E-stained Sections of Left Atrial Myxoma. The tumor cells are arranged in cords and small aggregates (A: ×40) and tubules (B: ×100) within a myxoid stroma.

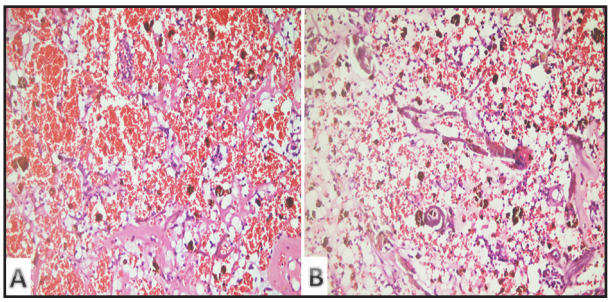

Figure 2. H&E-stained Sections of Left Atrial Myxoma Showing Abundant Extravasated Erythrocytes and Hemosiderin-laden Macrophages (A), and Degenerative Changes in the Form of Dystrophic Calcification (B) (×200).

retrospective study of 274 patients with cardiac tumors, CM accounted for approximately 70% of cases, followed by papillary fibroelastoma (10%) [2]. Other studies have similarly identified CM as the most common primary cardiac neoplasm [4, 5].

In the present study, an archived pathology specimen and histological sections from a case of left atrial myxoma attached to the interatrial septum were re-evaluated in a 43-year-old female patient. This case illustrates the typical clinical, radiological, and histopathological characteristics of CM.

CM occurs more frequently in females and is commonly diagnosed between the fifth and seventh decades of life. The true incidence of CM is difficult to determine because some cases remain asymptomatic and are discovered incidentally. However, available literature estimates a prevalence of approximately 0.03% in the general population. Most CMs occur sporadically, whereas approximately 10% are associated with familial syndromes, particularly Carney complex. Carney complex-associated CMs usually occur at younger ages and may demonstrate different clinical characteristics compared with sporadic cases [5].

The pathogenesis of CM remains incompletely understood. Earlier hypotheses suggested that CMs represented organized thrombi, largely because thrombotic material is frequently observed on the tumor surface [6, 7]. However, subsequent genetic studies have demonstrated chromosomal alterations and molecular abnormalities in both sporadic and familial CMs, supporting their neoplastic origin and association with pathways involved in cardiac development [1].

The left atrium is the most frequent site of occurrence, accounting for approximately 75% of reported cases, while more than 90% of CMs arise from either the left or right atrium [1, 5]. In contrast, valvular myxomas are extremely uncommon, representing less than 1% of all cases.

The patient in this report presented with progressive dyspnea and persistent dry cough. Clinical manifestations of CM depend mainly on tumor size, mobility, and anatomical location. Left atrial myxomas may obstruct mitral valve inflow, resulting in pulmonary congestion and symptoms of left-sided heart failure, including dyspnea. Although CMs are histologically benign, their clinical behavior may be significant because tumor mobility

and surface thrombus formation can result in systemic embolization. Embolic events most commonly involve the cerebral circulation and may lead to ischemic neurological complications [5].

Constitutional symptoms, including fever, fatigue, anemia, and arthralgia, occur in approximately 20% of patients and are thought to be associated with cytokine production, particularly interleukin-6 (IL-6), by tumor cells. The combination of constitutional symptoms and thromboembolic manifestations may mimic infective endocarditis and delay diagnosis [1].

Grossly, CMs can be categorized into polypoid and papillary types. Polypoid lesions are more commonly associated with obstructive symptoms, whereas papillary lesions are considered more fragile and have a greater tendency toward fragmentation and embolization [5].

Histologically, CM is characterized by scattered stellate, spindle-shaped, or ovoid cells with bland nuclei and indistinct cytoplasmic borders embedded within a myxoid mesenchymal stroma. Tumor cells may form cords, tubules, small aggregates, or poorly cohesive arrangements. Common associated findings include extravasated erythrocytes and hemosiderin-laden macrophages. Secondary degenerative alterations, including dystrophic calcification and, rarely, ossification, may also occur [2]. Rarely, CMs may exhibit unusual histological features, including glandular differentiation. In a 10-year study of 38 excised CMs, three cases demonstrated glandular differentiation characterized by columnar cells, cytoplasmic vacuolization, and gland-like architectural patterns [4].

Surgical excision remains the treatment of choice for CM and is associated with excellent outcomes in most patients [8]. Reported postoperative complications include infection, pericardial effusion, and arrhythmias [4]. Recurrence following complete surgical removal is uncommon, occurring in approximately 2% of sporadic and familial cases, although recurrence is more frequently observed in patients with familial disease or incomplete excision [1].

In conclusion, cardiac myxoma is a rare benign primary cardiac tumor that most commonly arises in the left atrium but can occur in other cardiac locations. Clinical manifestations vary according to tumor size, mobility, and anatomical site, ranging from incidental findings to obstructive symptoms or embolic complications. Despite its benign histological characteristics, CM may cause significant morbidity due to its potential for obstruction and systemic embolization. Accurate diagnosis requires integration of clinical presentation, imaging findings, and histopathological evaluation.

Acknowledgments

Not applicable.

Authors' Contributions

Conception and design: Maisa Hashem Mohammed. Acquisition, analysis, and interpretation of data: Maisa Hashem Mohammed. Drafting of the manuscript: Maisa Hashem Mohammed. Approval of the final manuscript version for publication: Maisa Hashem Mohammed.

Conflict of Interest

The author declares that there are no conflicts of interest.

Data Availability

All relevant data are included in the manuscript.

Ethical Considerations

As this study was based entirely on pre-existing retrospective data, a waiver of informed consent was requested and granted. Patient confidentiality was maintained by anonymizing all identifiable information prior to analysis.

Funding

Not applicable.

Study Registration

Not applicable.

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