CASE STUDIES

Presentation of atrial myxoma with unusual clinicopathological features

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Abstract

Atrial myxomas present with diverse clinical manifestations depending on its location, tendency to embolize, and systemic symptoms resulting from IL-6 production by tumor cells. We present here the case of right atrial myxoma, with persistent anemia and thrombocytopenia. Occurrence of thrombotic episodes with manifestations of antiphospholipid antibody syndrome (APAS) was noted and further investigations were carried out. Transthoracic echocardiogram revealed the presence of a right atrial mass with the possibility of an intracavitary thrombus or myxoma. Complete surgical excision and histopathological examination further confirmed the diagnosis of right atrial myxoma. The study emphasizes the importance of considering protean clinical manifestations including APAS and histopathological examination while diagnosing right atrial myxoma.

Introduction

Primary tumors of the heart are rare, in contrast to metastatic tumors. The prevalence of cardiac tumors at autopsy ranges from 0.001 to 0.3%. According to the literature evidence, myxomas are the most common primary tumors of the heart accounting for 20-50% of cases and are usually found in the left atrium.[1] Myxomas arising in other sites such as right atrium, ventricles, superior venacava or pulmonary veins are rare and are sometimes designated as 'atypical myxomas.' [2, 3, 4]

The clinical manifestations of atrial myxoma are diverse and the mean age of onset is between 30-60 years. The most common presentations of atrial myxoma can be broadly classified into three different ways: (1) Obstructive symptoms: dyspnea, cardiac failure, collapse and syncope due to obstruction of the mitral valve. (2) Constitutional symptoms: symptoms of autoimmune disease, vasculitis and various other non-specific symptoms. (3) Embolic symptoms: most common is cerebral emboli.[1]

We present here the case of a young female with a history of thromboembolism and persistent anemia with thrombocytopenia and positive antiphospholipid antibody (APA). Diagnosis of right atrial myxoma was suggested after considering differential diagnosis of atrial thrombus.

Case report

A 30-year-old female presented with sudden onset of generalized chest pain and nausea. She also had dyspnea of three months duration. There was a past history of amputation of left upper limb due to thrombosis of the left brachial artery. Peripheral blood examination revealed severe anemia and thrombocytopenia. Electrocardiogram and chest X-ray were normal. Coagulation workup carried out after admission revealed prolongation of prothrombin time (PT) to 24.2 sec. with international normalized ratio (INR) of 1.83 and activated partial thromboplastin time (APTT) to 76.8 sec. (control: 34.0 sec.). Prolongation of both parameters led to the suspicion of an immunological disorder. The immunological study disclosed the presence of positive lupus anticoagulant (LA) and high anticardiolipin antibody (ACA) titer. Transthoracic echocardiography (TTE) showed a pattern indicating right atrial myxoma (Fig.1).

Although the first diagnosis by TTE was atrial myxoma, the differential diagnosis of thrombus was also considered. The patient had undergone resection of right atrial mass and the specimen was subjected to histopathological evaluation. The tumor was friable and grossly gelatinous (Fig.2).

The biopsy report indicated the presence of stellate to elongated spindled cells. (Fig.3a). Extensive areas of organizing hemorrhage, myxoid changes, fibrosis, calcification, and ossification were noted. (Fig.3b). The patient survived for two weeks after surgery. Although the PT and APTT had normalized, persistent thrombocytopenia was noted. Despite the administration of packed cell and platelet infusions and she succumbed to cardiac arrest.

Fig.1: Transthoracic echocardiogram showing apical four chamber view with a right atrial myxoma

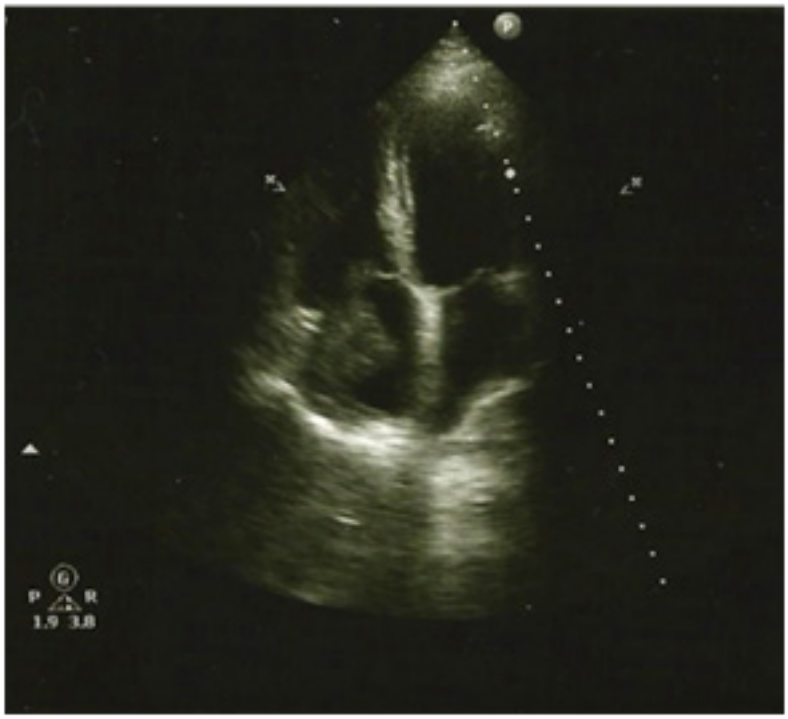


Fig.2: Friable and gelatinous fragmented mass

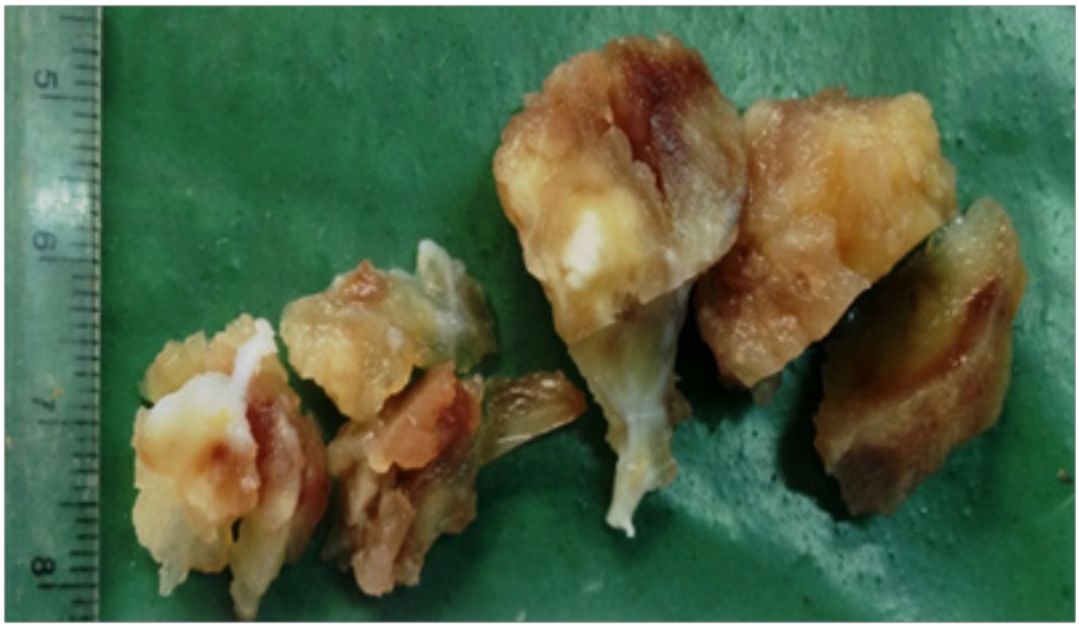
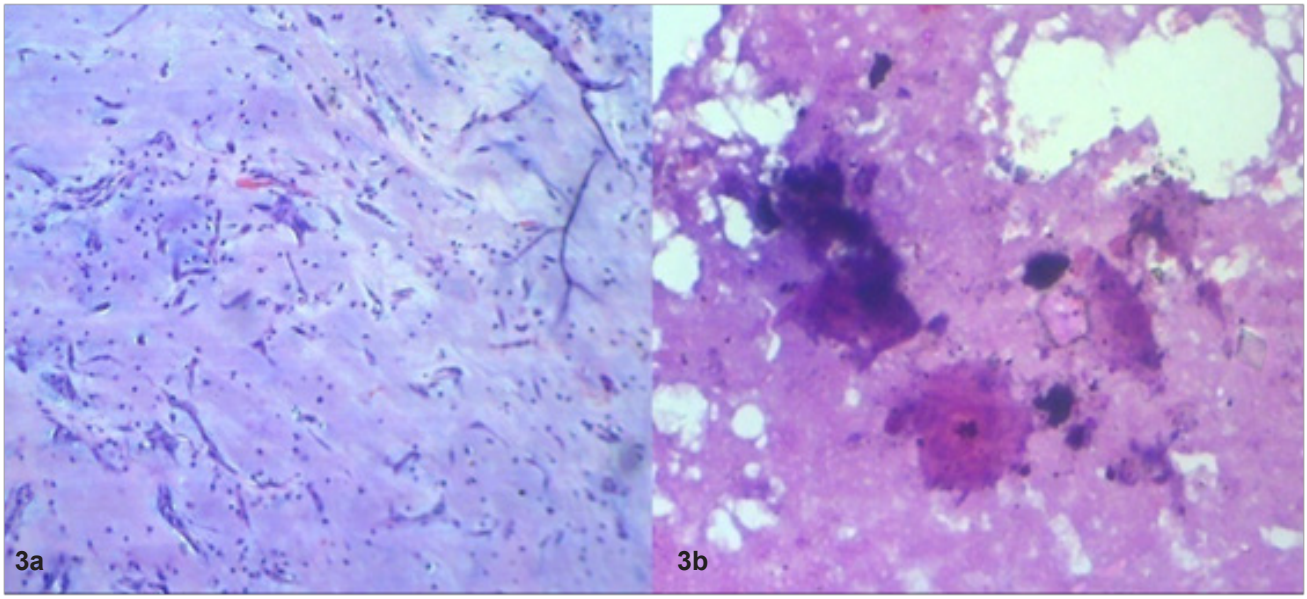


Fig.3a: Stellate and elongated myxoma cells in loose myxoid stroma (H&E, x100) Fig.3b: Foci of calcification (H&E, x400)



Discussion

Diagnosis of atrial myxomas is often challenging due to their asymptomatic nature or presentation of protean clinical manifestations. Constitutional symptoms and hematological abnormalities are seen in 90% of the patients with myxoma. Other symptoms of the disease, such as fever, weight loss, anemia, increased erythrocyte sedimentation rate, and hypergammaglobulinemia, can simulate connective tissue disease.[5]

Interleukin-6 (IL-6) secreted by right atrial myxomas stimulates the synthesis of acute phase reagents and promotes the differentiation of B lymphocytes, thereby inducing the synthesis of autoantibodies including APA.[5] In the current case, the normalization of coagulation abnormalities noted after the resection of the tumor suggests that the immunological reaction was caused by myxoma. The presence of APA has been related to the appearance of recurrent venous or atrial thrombosis, repeated miscarriages and/or thrombocytopenia constituting the antiphospholipid syndrome (APS).[5] Surgical excision followed by histopathological examination of the mass helped in concluding the diagnosis.

Atrial myxoma, usually found in the left atrium, is more prevalent among women.[1] Presence of cardiac symptoms and eosinophilia is found to be more common among male gender, while fever is more associated with female gender.[6]

Cardiac myxomas are fragile, gelatinous or myxoid tumors.[2] Myxoid matrix is composed of an acid mucopolysaccharide rich stroma in which polygonal cells (arranged singly or in small clusters) are scattered with fibrocytes, smooth muscle, lymphocytes, and plasma cells. Calcification, as noted in the current case, occurs in 10% of cardiac myxomas.[7] Metaplastic bone formation, as seen in the present case, is more common in right atrial myxomas.[8]

Conclusion

The present case study warrants the significance of considering atrial myxoma while investigating a patient presenting with thrombotic manifestations and abnormal hematological parameters. Early suspicion of diagnosis supported by TTE may assist in the diagnosis of myxoma.

Competing interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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