

# Atrial myxoma in a primigravida presenting as Raynaud's phenomenon

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## Abstract

A 32-year-old primigravida at 8 weeks of gestation presented with gangrene of both great toe for 10 days. Two years back, she had an episode of Raynaud's phenomenon involving left ring finger diagnosed as primary Raynaud's phenomenon and was treated with nifedipine following which she improved. In the following months, she had 2-3 similar episodes of Raynaud's phenomenon in the upper extremity which were less severe. On diagnostic evaluation she was found to have an intra cardiac mass arising from mitral leaflet. We present a case of atrial myxoma initially presenting as an isolated Raynaud's phenomenon with a long asymptomatic period before the next clinical manifestation which to our knowledge is the first such report.

## Introduction

Cardiac masses range from non-neoplastic lesions to high grade malignancies and occur over a wide range of ages.<sup>1</sup> Cardiac myxoma is a rare disease with a variety of nonspecific clinical manifestations masquerading as other more common diseases. Atrial myxomas have been described as a proliferation of primitive mesenchymal cells that normally differentiate into endothelial and smooth muscle cells and fibroblasts. Myxomas are the most common type of primary cardiac tumor in all age groups, accounting for one-third to one-half of all cases at postmortem and about three-quarters of the tumors treated surgically. Myxomas can present with mechanical obstruction of blood flow, embolic phenomenon, and constitutional symptoms and signs. Pinede *et al.* in a study of patients with left atrial cardiac myxoma found 34% patients to have constitutional symptoms including fever, weight loss, or symptoms resembling connective tissue disease, due to cytokine (interleukin-6) secretion.<sup>1</sup> Atrial myxoma with Raynaud's phenomenon as the initial manifestation have been reported previously.<sup>2,3</sup> Thus presentation of atrial myxoma may mimic connective tissue disease.

## Case Report

A 32-year-old primigravida at 8 weeks of gestation presented with gangrene of both great toe for 10 days. Two years back, she had an episode of Raynaud's phenomenon involving left ring finger diagnosed as primary Raynaud's phenomenon from an outside clinic and was treated with nifedipine following which she improved. In the following months, she had 2-3 similar episodes of Raynaud's phenomenon involving the upper extremity which were less severe for which she did not take medical care. During this period she didn't experience any arthralgia, arthritis, fever, weight loss or any other symptoms suggestive of an connective tissue disease. She was found to be pale, all peripheral pulsations were palpable equally and blood pressure was 150/90 mmHg. Systemic examination was normal. Hemoglobin was 9.1 g/dL, total leucocyte count 24,800/ $\mu$ L with 85% neutrophils, platelet count 2.76 $\times$ 10<sup>9</sup>/L, ESR 25 mm in 1 h and CRP was high. The peripheral smear had normocytic normochromic anemia with neutrophilia. Urinalysis showed was normal. Biochemical parameters showed random blood sugar 131 mg, urea 55 mg/dL, creatinine 1.7 mg/dL, sodium 134 mmol/L, potassium 3.6 mmol/L and normal liver function test. Chest X-ray and electrocardiogram were normal. Ultrasonography of abdomen showed single live intrauterine gestation at 8 wks and bilateral hyperechoic kidneys. Arterial Doppler of lower limbs did not show significant stenosis. Her autoantibody profile including ANA profile, RA, cANCA, pANCA, VDRL were all negative. HIV, Hepatitis B and Hepatitis C serology were negative. IgG and IgM APLA, IgG and IgM beta2 glycoprotein and lupus anticoagulant tests were negative. Serum complement levels were normal. Trans thoracic echocardiogram demonstrated a 20 $\times$ 15 mm echogenic mass arising from mitral valve which prolapsed through the mitral valve causing a functional stenosis during diastole. The lobulated appearance of the mass and connection by a short stalk in trans thoracic echocardiogram was suggestive of an atrial myxoma (Figure 1). Her repeated blood cultures were negative. She was taken up for surgery but she developed a hospital acquired pneumonia went into sepsis and multi organ dysfunction, and despite all efforts she succumbed to her illness.

## Discussion

Primary cardiac tumors are rare, approximately three-quarters are histologically benign, and the majority being myxomas. Many tumors are now surgically curable; thus, early diagnosis is imperative. Cardiac tumors

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Key words: Atrial myxoma; Raynaud's phenomenon.

Contributions: the authors contributed equally.

Conflict of interest: the authors declare no potential conflict of interest.

Received for publication: 19 March 2015.

Revision received: 25 June 2015.

Accepted for publication: 30 June 2015.

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Licensee PAGEPress, Italy  
Rheumatology Reports 2015; 7:5915  
doi:10.4081/rr.2015.5915

may have a range of cardiac and non cardiac presentations; 50% of all benign cardiac tumors are atrial myxomas. In about a fifth of patients with the disorder, serious complications can arise through systemic arterial embolization in 30-40% of cases and obstruction to blood flow through the mitral valve leading to dyspnea and orthopnea in over 50% of patients.<sup>4</sup> The emboli have a predilection for the brain, but also can involve other organs, like the liver, spleen, kidney, retina, coronary arteries and the peripheral arteries. Embolic manifestations and constitutional signs and symptoms, including fever, weight loss, cachexia, malaise, arthralgias, rash, digital clubbing, Raynaud's phenomenon, hypergammaglobulinemia, anemia, polycythemia, leukocytosis, elevated erythrocyte sedimentation rate, thrombocytopenia, and thrombocytosis account for the frequent misdiagnosis of patients with myxomas as having endocarditis, connective tissue disease, or a paraneoplastic syndrome. Vasculitic manifestations of cardiac tumors causing diagnostic confusion and leading to wrong diagnosis was previously reported.<sup>4,8</sup> Cutaneous manifestations of cardiac myxoma can be classified into 3 subtypes: i) embolic cutaneous signs; ii) non-embolic cutaneous signs associated with auto-immune symptoms; and iii) cutaneous signs of a complex syndrome. Cutaneous features may be associated with autoimmune symptoms including Raynaud's phenomenon, malar erythema, and vasculitis, which can be explained by myxomatous secretion of interleukin-6.<sup>9</sup> The

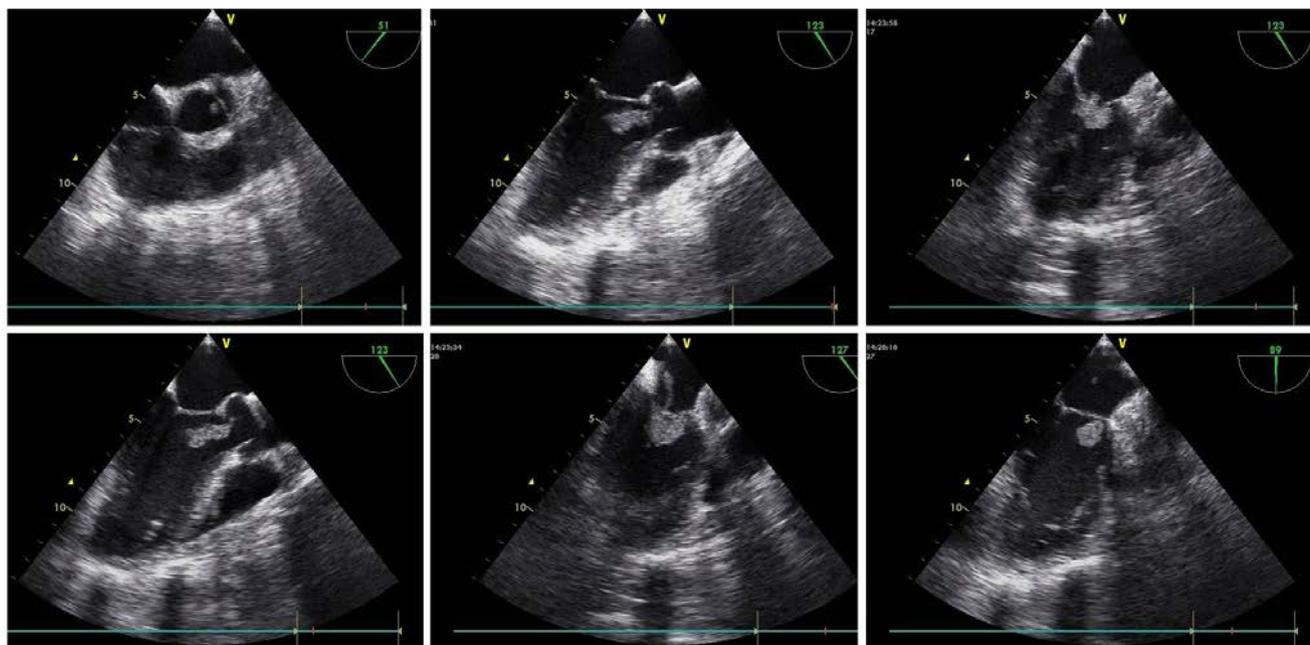


Figure 1. Trans thoracic echocardiogram demonstrated a 20×15 mm echogenic mass arising from mitral valve.

reported incidence of cardiac myxomas during pregnancy is extremely low with only 17 reported cases in the medical literature.<sup>10</sup> Standard therapy involves surgical resection given the potential for embolization.

The patient described here had a history of Raynaud's phenomenon involving left ring finger two years before the diagnosis of myxoma. She was evaluated for the same in an outside clinic and tests were done to rule out vasculitis, but an echocardiogram was not done. Since isolated Raynaud's phenomenon is a very rare presentation of cardiac tumours an echocardiogram should be considered in such patients before labelling as primary Raynaud's phenomenon. The fact that our patient was asymptomatic for a long period following her initial presentation makes this a silent killer. Because the tumor's histological benignancy contrasts with its malignant clinical behavior in terms of systemic embolization, early diagnosis and surgical intervention are essential.

## Conclusions

Atrial myxoma presenting initially with

symptoms resembling connective tissue disease including Raynaud's phenomenon have been reported previously. We present a case of atrial myxoma initially presenting as an isolated Raynaud's phenomenon with a long asymptomatic period before the next clinical manifestation which to our knowledge is the first such report. We recommend the readers to consider performing an echocardiogram in all patients presenting with isolated Raynaud's phenomenon.

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