Right atrial (RA) myxoma is a rare entity, and RA thrombus is still rarer. Tricuspid valve vegetation can mimic an RA mass and is seen more frequently than either a tumor or a thrombus. RA thrombus is a rare manifestation of severe thromboembolic disease. In situ RA thrombi are usually immobile whereas secondary RA thrombus is mobile. Mobile RA thrombus usually coexists with massive pulmonary embolism (PE).1 Protein C deficiency is a hematologic abnormality with propensity to thromboembolic manifestations.

Case Report
A 32-year-old woman—nonhypertensive, nondiabetic, nonasthmatic—presented with shortness of breath class III–IV and swelling of both the lower limbs for the previous 10 days. The patient had history of acute left upper limb ischemia 2 years back for which she was admitted. Electrocardiogram (ECG) and echocardiogram were normal at that time. Diagnostic angiogram showed normal coronaries and left axillary artery thrombus with total cutoff for which she received catheter-guided thrombolysis with streptokinase. The patient was discharged in stable condition with pulses restored in the left upper limb with advice for workup for prothrombotic state. She was admitted 1 month back at an outside hospital with shortness of breath class II and fever of 15 days. On evaluation, she was found to have an RA mass that was presumed to be RA myxoma after a transthoracic and transesophageal echocardiography, and she underwent surgical excision of the mass. The discharge summary from the cardiothoracic surgeon mentioned the RA mass as a mobile pedunculated mass with the stalk attached to the inferior vena cava–right atrium (IVC-RA) junction, occluding the tricuspid valve without much infiltration and was removed successfully. Histopathology showed a well-circumscribed mass, very focally covered by flat/cuboidal cells on the surface. Large areas of coagulative necrosis were present. There was tiny volume of viable tumor consisting of few stellate cells arranged in the cord, intimately admixed with slit-like blood vessels with areas of fresh hemorrhages and inflammatory cells. It was reported as cardiac myxoma with extensive infarction and necrosis.

Postoperatively, the patient continued to have breathlessness of class II and presented to the authors’ institute with the present complaints. At presentation, she was hemodynamically stable, and clinical examination showed clear lungs with raised jugular venous pressure (JVP). Cardiovascular
However, this description holds not a common site for myxoma), histopathology—not classic thrombus, RA mass attached to IVC-RA junction (which is abnormal collagen profile, the authors thought of RA mass as thrombus rather than myxoma. Histopathology slides were reviewed at the institute, and the pathologist unanimously opined them as thrombus. The patient was discharged on oral anticoagulants with advice of further workup for thrombotic state. She lost regular follow-up and was noncompliant with medication.

One year later, the patient again presented with shortness of breath of class III and bilateral pedal edema of 3 months duration. Clinically, there were features of right-sided heart failure. On evaluation, ECG was suggestive of RV strain (Fig. 4). Two-dimensional (2D) echo showed dilated RA, RV with RV dysfunction, severe TR with severe PAH (RVSP = 96 mm Hg). Chest X-ray showed the same features as before (Fig. 5). Repeated CT pulmonary angiogram was done as there was exacerbation of symptoms, which showed chronic pulmonary thromboembolism with recanalization in right lower lobe pulmonary arteries and marked attenuation of left lower lobe pulmonary artery likely sequelae of chronic thrombosis (Fig. 6). Complete blood picture and renal parameters were normal. ANA and anticardiolipin (aCL) antibodies were negative, homocysteine levels were normal, and protein S was normal. However, protein C was very low 29% (normal 70–130%). Anti-PAH drug therapy was started, and oral anticoagulation tapered to ideal international normalized ratio (INR) was continued.

Discussion

Myxomas are the most common intracardiac tumors. Atrial myxoma accounts for 35 to 50% of primary cardiac tumors of which only 20% arise from the RA. A typical RA myxoma is solitary pedunculated arising from the fossa ovalis or base of the interatrial septum. Occasionally they arise from the atrial wall, appendage, and very rarely tricuspid and eustachian valve. The diagnosis is usually made by 2D echocardiography; however, transesophageal echocardiography is superior for delineation of size, exact location, morphology, and the point of attachment.

The classic echocardiographic features to differentiate myxoma from atrial thrombus are that the thrombi are irregular, laminated, and immobile with a broad base attached to the posterior atrial wall. However, this description holds good only for in situ atrial thrombi and not for secondary atrial thrombi. Secondary atrial thrombi are usually from the peripheral veins and are hence mobile. These are also referred to as “emboli in transit” as they on their way to the pulmonary arteries. They are described as spherical, coiled, grapelike, ovoid, worm-like, or serpiginous masses moving within the RA. When large, they may prolapse through the tricuspid valve into the RV. Often, these masses appear free floating with no attachment site. Such mobile thrombi are very often mistaken for RA myxoma.

In this patient, features favoring RA thrombi are history of repeated thromboembolic episodes, initial normal echocardiogram (less likely in case of myxoma), and abnormal
attachment of the mass (to the IVC-RA junction). Large stel-
late cells with vacuolated cytoplasm in the myxoid back-
ground on histopathologic examination is pathognomonic
of myxoma. However, the classic description was missing
in the initial histologic report commenting as probable
myxoma.

Intracardiac thrombus could be the manifestation of pri-
mary cardiac abnormality or hematologic disorder or rheu-
matologic disease. It is common in those with indwelling
catheter in the RA such as those on hemodialysis. Echocar-
diographic features suggestive of primary cardiac pathology
such as hypokinetic chambers, valvular abnormality, and
spontaneous echo contrast were absent in this case, exclud-
ing cardiac source of thromboembolism.

Cardiac thrombus in the absence of primary cardia-
c disease is less likely, but it can occur in the setting of
hypercoagulable state. The risk of intracardiac thrombosis
is increased in patients who have systemic lupus erythe-
matosus (SLE) and who either show positive test results for
lupus anticoagulant activity or have medium or high lev-
els of anticardiolipin antibodies. A retrospective study of
637 patients found that venous thrombosis with PE was
more frequent in persons who had lupus anticoagulant ac-
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Collage profile showed some abnormality, but the rheuma-
tologist opined that it was not specific for any rheumato-
logic disorder.

The most likely cause for thrombus formation in this
patient was protein C deficiency that was 29% against a
normal of 70 to 130%. Protein C has both anticoagulant and
profibrinolytic properties. It inactivates factors Va and VIIIa,
and protein S is a cofactor for these actions. Hence proteins
C and S deficiency predisposes to the formation of arterial
and venous thrombi. These patients usually develop ve-
nous thrombotic complications between the ages of 15 and
40 years with a high incidence of deep venous thrombosis
(DVT) and PE. Thrombosis can also occur in the cerebral, ret-
inal, mesenteric, and renal veins and the inferior vena cava.
Presentation as right-side cardiac thrombus is the result of
fragmentation of the thrombus and subsequent pulmonary
embolization.

Many other rare mimics of RA myxoma have been de-
cribed in literature. Khanna et al reported mimicker of
RA mass due to sinus of Valsalva’s aneurysm. An asym-
tomatic 55-year-old man of unruptured sinus of Valsalva’s
aneurysm of noncoronary cusp was on medical follow-up.
At 2-year follow-up, there was thrombus formation in the aneurysm, mimicking RA tumor on 2D transthoracic echocardiography. Cardiac CT showed filling defect in the aneurysm suggestive of thrombus. Garg et al report the case of a 53-year-old woman with RA mass mimicking myxoma, which the histopathologic evaluation revealed to be Rosai-Dorfman disease (sinus histiocytosis with massive lymphadenopathy) of the RA. Garg et al and Li et al presented a case of RA mass due to paraganglioma that is pedunculated like in this case. Li et al demonstrated pedunculated mass mimicking RA myxoma, turned out to be a thrombus. Salani et al presented the case report of a 19-year-old patient with chronic kidney disease due to chronic glomerulonephritis, in hemodialysis (HD) by central catheter, with the incidental finding of a mass of 28 × 16 mm in the RA. The diagnosis of thrombus, infective endocarditis, or myxoma was considered. After 180 days of anticoagulation, there was significant reduction in mass.

RA thrombus is seen in 4% of patients diagnosed as PE and is associated with increased mortality. Treatment options are controversial as there are limited data for comparison. Initial treatment with heparin and follow-up with oral anticoagulation form the primary cornerstone of treatment. Large thrombi require surgical excision as thrombectomy may not suffice. The mortality rate associated with no therapy, anticoagulation therapy, embolectomy, and thrombolysis were 100%, 29%, 24%, and 11%, respectively. Surgery may be insufficient in case of multiple thrombi in which thrombolysis is favored despite the risk of migration of thrombi. Low-dose and prolonged fibrinolysis is recommended to reduce the risk of thrombus migration.

**Conclusion**

The diagnostic dilemma of an RA mass could persist despite all investigations and histopathology unless clinical scenario is taken into consideration. RA thrombus should be considered in the close differential diagnosis of RA mass, especially in the clinical context of multiple thromboembolic events. Treatment should be individualized owing to lack of evidence. Patients should be evaluated for rheumatologic disorders and hypercoagulable state. All patients should be evaluated for proteins C and S deficiency in the presence of unprovoked DVT and PE.
References


