Case Report

Pulmonary embolism due to the right atrial thrombus mimicking atrial myxoma in protein C deficiency patient

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Abstract

Introduction: Atrial masses are uncommon, and mainly consist of tumours, vegetations and thrombi. Despite the availability of several imaging modalities, it may still be difficult to distinguish between them. Right atrial thrombi are rare but associated with high mortality. We describe a case of a large right atrial thrombus with pulmonary embolism, in a protein C deficient patient. Right atrial thrombus mimicked atrial myxoma on TEE examination. CT thorax with contrast demonstrated thrombosis in right pulmonary artery and its branches. Right atrial clot was successfully removed surgically and pulmonary embolism treated conservatively with full recovery of symptoms.
Keywords: protein C, atrial myxoma.

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INTRODUCTION

Mobile right heart thrombus is a severe but rare presentation of thromboembolic disease and usually coexists with an already massive pulmonary embolism. Mortality rates for mobile right heart thrombus are high at over 40%; death usually occurs when thrombotic masses move towards the pulmonary trunk[1]. Protein C deficiency has been reported to be a risk factor for thrombosis in multiple organs as a result of inactivation of factor of Va.

CASE REPORT

47 YRS old male patient admitted with complaints of left sided chest pain since 3 days. It is constricting in nature, radiating to left arm and present at rest. Dyspnea on minimal exertion since 3 days. No past history of hypertension and diabetes mellitus. No similar complaints in past. No history of prolonged immobilisation or major surgery in recent past. No history of any addiction. Patient was admitted in ICU. On admission pulse was 104/min, blood pressure was 140/90 mmHg and oxygen saturation was 96%. Jugular venous pressure was normal. Cardiovascular examination revealed normal apex beat, no thrill and parasternal heave, S1/S2 normal; no murmur was audible. Systemic examinations were unremarkable.

Lab investigation

Hemoglobin was 18.1, total leukocytes count was 13900. ESR was 8.0. Renal function test was normal. Troponin I was negative. Cardiac markers were negative. PT INR was 1.3. Total cholesterol was 146, total triglyceride was 46, HDL was 14, LDL was 122 and VLDL was 92. Serum homocystein (20 umol/l) was slightly raised. Protein C was decreased 26% (normal limit 75-160%). Protein S was 59%. Antithrombin was normal. Lupus anticoagulant was negative. APLA IgG and igM was negative. ECG showed sinus rhythm with sinus tachycardia with T wave inversion in V1 to V3 with RV strain pattern (Fig.1). Chest X-ray appeared normal. 2D-echo revealed 2.3 X 3.0 cm right atrial mass which was

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round in shape and mobile (Fig.2). It was protruding into right ventricle through tricuspid valve in diastole. RA/RV mildly dilated. No RWMA. Normal LV function. On Doppler examination revealed Moderate TR with PASP 68 mmHg. For better evaluation of right atrial mass transesophageal echocardiography (TEE) was planned. TEE revealed 2.3 X 3.0 cm pedunculated mass in right atrium which was attached to right a trial free wall (Fig.3). Smooth surface and pedunculated nature of mass in favour of right atrial myxoma. Doppler of both lower limbs venous system was normal. CT thorax with contrast showed well defined non-enhancing intraluminal hypodense filling defect in right main pulmonary artery measuring 3.6 x 2.8x 3.8 cm extending in the anterior descending branch of right pulmonary artery and proximal ascending branch of pulmonary artery with few calcification and peripheral passage of contrast s/o acute to subacute thrombus (Fig.4). Patchy wedge shaped pleural based areas of consolidation with central air lucencies in the anterior basal, posterior basal segments of right lower lobe and anterior segment of right upper lobe s/o pulmonary infarct. Diagnosis of pulmonary embolism due to right a trial mass with protein C deficiency was made. Patient was treated with low molecular weight heparin. He responded well to treatment, symptoms free after 5 days of treatment. Patient with Pulmonary embolism with a trial mass has high mortality rate, so surgically removal of right a trial mass is planned. Before surgery, coronary angiography was performed to rule out clinically significant coronary artery disease. Coronary angiography was normal. The patient subsequently underwent surgery. After the institution of cardiopulmonary bypass, the right atrium was opened, and the large mass was removed from the free wall (Fig.5). Gross examination of the operative specimen revealed multiple masses with irregular surfaces (Fig.6); histologic examination showed an organized thrombus with a fibrin network and focal calcifications (Fig.7). The patient experienced no postoperative complications. Patient was discharged on oral anticoagulant warfarin.

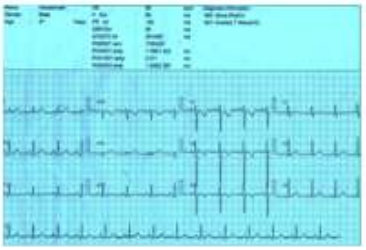

Figure 1: ECG showing Right ventricular strain pattern

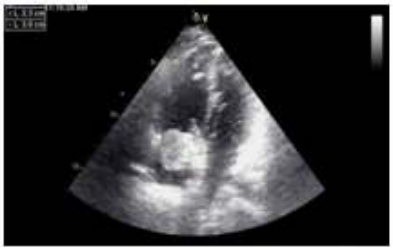

Figure 2: Transthoracic echocardiography showed 2.3 X 3.0 cm right atrial mass in apical four chamber view

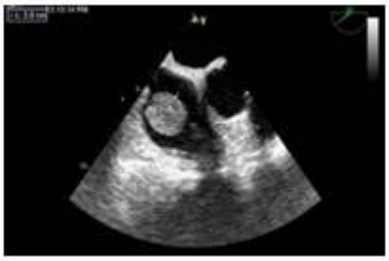

Figure 3: Transesophageal echocardiography showed 2.3 X 3.0 cm right atrial pedunculated mass attached to right atrial free wall in mid esophageal parasternal short axis view.

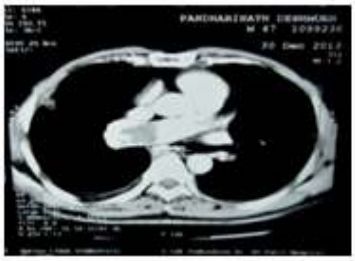

Figure 4: Computed tomography of thorax with contrast showed well defined non-enhancing intraluminal hypodense filling defect in right main pulmonary artery suggestive of acute thrombus.

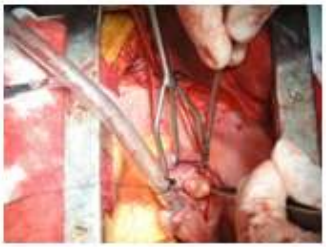

Figure 5: Intraoperative picture of removal of mass from right atrium

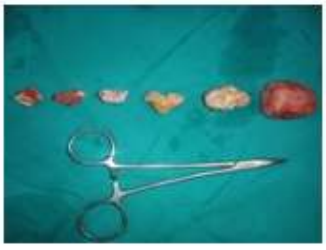

Figure 6: Gross specimen showed multiple masses with irregular surfaces

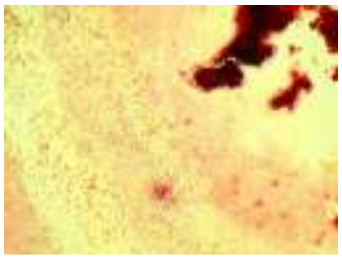

Figure 7: Histopatholgical examination showed an organized thrombus with a fibrin network and leukocytes infiltration

DISCUSSION

The differential diagnosis of right atrial mass includes vegetation, tumor, and thrombus. Echocardiography can be used for characterization of the mass by morphologic shape, appearance, site of attachment, type of margin, and presence or absence in the left atrial appendage. Transesophageal echocardiography is superior to transthoracic echocardiography. The most common primary cardiac tumor is the myxoma. 15% of atrial myxomas arise in the right atrium, and they are usually attached to the inter atrial septum[2]. Although in our patient the attachment site was the free wall of the right atrium, atrial myxoma was a possible diagnosis because of smooth surface and pedunculated nature of mass. Right atrial thrombi have been described in patients with atrial fibrillation/flutter, central venous catheters, or pacemaker leads. In-situ right atrial thrombi are usually immobile, attached to the atrial wall with occasional calcification. Secondary right atrial thrombi are often mobile as they have propagated from the peripheral veins, and are in transit to embolize into the pulmonary arteries. Thus, these mobile right heart thrombi have often been referred to as "emboli in transit"[3]. Mobile right atrial thrombi have been described as spherical, coiled, grapelike, ovoid, worm-like or serpiginous masses moving within the right atrium, and if large, they may prolapse through the tricuspid valve and into the right ventricle. Often, these masses appear free-floating with no attachment site. While nearly all of the detected cases of mobile right heart thrombi are diagnosed when echocardiography is performed in patients with suspected PE or proven PE, the true incidence of mobile right heart thrombi may be difficult to ascertain. In patients with PE, it is not common to detect thrombi in the right atria during echocardiography, and in unselected patients with PE, about 4% of patients have right atrial thrombi patients with PE is associated with increased mortality, little is known about optimal management of this difficult clinical situation[4]. The treatment of choice remains controversial with limited data to compare the various options. In a meta-analysis that included 177 cases, the overall mortality rate was 27%. The mortality rate associated with no therapy, anticoagulation therapy, embolectomy, and thrombolysis were 100%, 29%, 24%, and 11%, respectively[5]. Cardiac thrombus may be a complication of primary cardiac, hematological and rheumatological disease. Cardiac-sourced thromboembolism can be predicted on the basis of echocardiographic, clinical, electrocardiographic and laboratory assessments, such as the presence of spontaneous echocontrast, large hypokinetic cardiac chambers, mitral stenosis, history of thromboemboli, atrial fibrillation and increased coagulation markers. In our case, no cardiac pathology tending to thromboembolism was found. The most likely cause for thrombus formation was the decreased protein C. The treatment of atrial masses typically includes surgical resection. In cases of atrial thrombi, anticoagulation therapy along with a hypercoagulable workup is recommended. Thrombi in the absence of underlying cardiac disease are distinctly less common, but may occur in the setting of hypercoagulable states such as autoimmune disease, pregnancy, and certain malignancies Protein C is the central component of a major antithrombotic regulatory system with both anticoagulant and profibrinolytic properties. It inactivates factors Va and VIIIa. Protein S is as a cofactor for these actions of Protein C. Protein C and Protein S deficiencies are genetic traits predisposing to the formation of venous clots. Protein C deficiency is one of several hereditary abnormalities of haemostatic proteins that have been described in patients with propensities to thromboembolic complications. Major morbidity is often seen in these patients. The diagnosis of various aspects of hereditary protein C deficiency in terms of clinical presentation and genetics (both homozygous and heterozygous states) is important. In heterozygotes, plasma levels of protein C are usually 35– 65% normal, whereas most normal individuals have levels between 70% and 130%[6]. Protein C-deficient patients usually develop venous thrombotic complications between the ages of 15 and 40 years with a high incidence of deep venous thrombosis and pulmonary embolism. In addition to the deep veins of the lower extremities, thrombosis can also occur in the cerebral, retinal, mesenteric and renal veins and the inferior vena cava. Acquired Protein C and S deficiencies have been reported in patients with deep venous thrombosis, pulmonary embolism, acute disseminated intravascular

coagulopathy, post-operative state, severe liver disease, malignancy, infection, hemolytic-uremic syndrome, adult respiratory distress syndrome and vitamin K deficiency. Treatment of symptomatic patients is initial heparin therapy followed by coumarin. After multiple thrombotic events, lifelong oral anticoagulant therapy is necessary. Symptomatic presentation in right side cardiac thrombus is fragmentation of the thrombus and subsequent pulmonary embolization. The spontaneous prognosis of pulmonary embolism associated with mobile intra-cardiac thrombus is poor, and choice of a therapeutic strategy is often difficult. The literature on treatment and therapeutic management of mobile right heart thrombus with pulmonary embolism gives no clear consensus. Treatment should be individualized according to the extent of intracardiac thrombus (number and sizes of clots and their mobility and morphology), likelihood of pre-existing pulmonary embolism, the patient's cardiopulmonary reserve, co-morbid conditions, and local expertise with treatment modalities. Thrombectomy might have been incomplete in view of the TEE findings. The outcome of untreated mobile thrombus is poor. Surgical removal should be considered for large thrombi[7]. In the event of mobile multiple intra-cardiac thrombi, surgery might not be a good choice for their complete removal. In these conditions, thrombolytic therapy might be preferable. Fibrinolysis is generally efficient but exposes the patient to the risk of migration of the thrombus. Intravenous fibrinolytic treatment for multiple intracardiac mobile thrombi should be given in low doses and for long times to avoid subsequent migration of the intra-cardiac thrombus[8].

CONCLUSION

The detection of right heart thromboemboli during echocardiography may have diagnostic and therapeutic implications. The presence of mobile right atrial thrombi in patients with PE portends poor prognosis with cardiopulmonary collapse due to PE. Therefore, treatment should be started immediately as any delay in administering therapy may be lethal. The optimal therapy remains controversial given absence of randomized trials. Protein C and S deficiencies should be investigated in all patients without history of risk factors for PE. Analysis of protein C and S activities are important to avoid underdiagnosis of this pathology.

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