Right ventricular thrombus in case of atrial septal defect with massive pulmonary embolism: A diagnostic dilemma

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A 55-year-old woman presented to the emergency services with a history of breathlessness and altered sensorium of 1 day duration. Other than a history of alcohol and tobacco abuse, her history was noncontributory. On examination, breathing was labored, respiratory rate was 24/min, heart rate was 110/min, and on pulse oximeter, saturation was 88%. She had no focal neurological deficits. The electrocardiography showed deep S-wave in lead I with inverted T-waves in lead III with right ventricular strain suggestive of probably pulmonary embolism, and chest X-ray showed cardiomegaly with right ventricular apex dilated right atrium (RA), ventricle, and pulmonary artery (PA). Arterial blood gas revealed a pH - 7.24, PO₂ - 58 mm of Hg with 6 L of oxygen, PCO₂ - 66.4 mm of Hg, and HCO₃ - 24.7 suggestive of type II respiratory failure. Biochemical investigations revealed elevated D-dimer, normal cardiac bio-markers, hypoalbuminemia, thrombocytopenia, and negative thrombophilia work-up. Liver enzymes were within normal limits. Lower limb venous Doppler did not reveal any thrombus. Ultrasound abdomen showed coarse echo texture of liver with no evidence of portal hypertension or cirrhosis. Transthoracic echocardiogram showed dilated RA and right ventricle (RV), RV thrombus, moderate RV dysfunction, pulmonary hypertension (right ventricular systolic pressure [RVSP]-58 mm of Hg), no regional wall motion abnormalities, and normal left ventricular function. Computed tomography (CT) pulmonary angiogram revealed thrombus in the main pulmonary artery (MPA) extending into the left pulmonary artery (LPA) with multiple pulmonary infarcts. CT brain was normal. The patient was started on anticoagulation and titrated to an international normalized ratio (INR) of 2–3 preoperatively. She was considered unsuitable for thrombolysis in view of altered mental status and alcoholic liver disease. She developed worsening respiratory failure and was intubated a week after admission. After a multidisciplinary review, the decision was taken to go ahead with surgical embolectomy.

Intraoperative transesophageal echocardiography (TEE) mid-esophageal five chamber view showed a 2 cm x 3 cm mobile ecchogenic mass in the RV cavity [Figure 1 and Video 1], mid-esophageal RV inflow-outflow view [Figure 2 and Video 2], and transgastric RV outflow view [Figure 3 and Video 3] revealed mobile thrombus 2 cm x 2 cm in the right ventricular outflow tract attached below the pulmonary valve. Transgastric right ventricular short axis view showed a 2 cm x 3 cm
echogenic mass in between the trabeculations of the RV [Figure 4 and Video 4], upper esophageal ascending aortic short axis view showed organized thrombus in right pulmonary artery (RPA) [Figure 5 and Video 5], mid-esophageal four chamber view showed dilated RA/RV, moderate RV dysfunction (tricuspid annular plane systolic excursion [TAPSE-11]), severe pulmonary hypertension (RVSP = 60 mm of Hg), and good left ventricular function. Mid-esophageal bi-caval view showed large (3 cm) ostium secundum atrial septal defect (ASD) which was shunting predominantly left to right [Video 6].

Surgical approach was via a midline sternotomy, cannulation was standard ascending aortic with bi-caval venous cannulation, and the minimum temperature was 28°C. Intraoperative findings were free thrombi in the RV, large organized clot load in MPA, LPA and RPA, no thrombi in RA/left atrium/left ventricle, and a large ostium secundum ASD. Pulmonary thrombectomy was done, and ASD was repaired with an autologous pericardial patch. The patient was weaned of cardiopulmonary bypass (CPB) with milrinone at 0.5 µg/kg/min, dobutamine at 5 µg/kg/min, adrenaline at 0.1 µg/kg/min, noradrenaline at 0.1 µg/kg/min, and vasopressin at 4 units/h. As nitric oxide is not available in our institute, we used milrinone nebulization 50 µg/kg. Post-CPB TEE showed no thrombus in MPA, RPA, LPA, and the RV. No residual ASD, mild tricuspid regurgitation (TR), RVSP-26, cardiac output of 4.3 l/min, and impaired RV function (TAPSE-8 mm). An inferior vena cava filter option was given to the relatives due to financial issues; it was not inserted.
Postoperative course

The patient was extubated 24 h later. She required noninvasive ventilation over the next 48 h, after which oxygen was gradually reduced to 2–4 L/min to maintain saturation in the 90s. As she had multiple pulmonary infarcts preoperatively, she required oxygen to maintain saturation. Inotropes were tapered over 72 h. On day 2, she developed atrial fibrillation which reverted with magnesium and amiodarone. Postoperative echo showed RA/RV/MPA dilated, no residual ASD, mild TR, RVSP-36, mild pericardial effusion, and impaired RV function. RV dysfunction was treated with digoxin, diuretics, and sildenafil. She was started on dalteparin 6 h after surgery, and oral anticoagulation was started to reach therapeutic INR. She was mobilized out of bed on the 4th postoperative day and was fully ambulated. Histopathological examination of the surgical specimen was reported as thrombus. By the 10th postoperative day, it was determined that the patient could be discharged on home oxygen, and plans were made for rehabilitation. A few hours before planned discharge, she suffered a sudden cardiac arrest and succumbed. The cause of death could not be determined, as the autopsy was denied by the family.

DISCUSSION

This patient had an unusual combination of massive pulmonary embolism, large ASD, absent deep vein thrombosis (DVT), and no paradoxical embolism. Acute pulmonary embolism occurs without documented DVT in about 6–7% of patients. These patients have an increased incidence of malignancy as compared to those with a documented DVT.[1] The other possibilities are complete dislodgement of peripheral venous thrombus, false-negative venous duplex, venous thromboembolism from pelvic veins, and in situ formation of thrombus in the RV and pulmonary arteries. Right ventricular thrombi are extremely rare, especially when not associated with thrombus in the left ventricle. Moderator band, papillary muscle, and coarse trabeculations in RV make the diagnosis of right ventricular thrombus very challenging. They have been described in the setting of autoimmune diseases (such as Behçet disease), hypercoagulability states (Factor V Leiden deficiency), in patients with right ventricular pacing leads, PA catheters, dilated or alcoholic cardiomyopathy, right ventricular infarction, and right ventricular arrhythmogenic cardiomyopathy.[2-4]

Interestingly, in this case, in spite of a large RA, large ASD, pulmonary hypertension, and moderate TR, she did not show any evidence of paradoxical embolism. In this case, whether to close the ASD or not and the right ventricular thrombus origin was a diagnostic dilemma, is it thrombus in transit or originating in situ in the RV due to the right ventricular dysfunction secondary to a large ASD and severe pulmonary arterial hypertension? In 1989, the European Working Group on Echocardiography identified three patterns of right heart thrombi. Type A thrombi are morphologically serpiginous, highly mobile masses moving within the RA, or ventricle, which often prolapse into the tricuspid or pulmonic valve. A point of attachment is often unseen or is visualized as a thin stalk. It is associated with DVT and pulmonary embolism. It is hypothesized that these clots embolize from large veins and are captured in-transit within the right heart in their path to pulmonary tree. Predisposing factors for this thromboembolism include prominent Eustachian valves, TR, low cardiac output, and pulmonary hypertension. Type B thrombi are nonmobile, mural, and believed to form in situ, in association with underlying cardiac abnormalities. Echocardiographically, these mural thrombi are less mobile, with a broad-based attachment to the heart wall, and occasional focal calcification. Type C thrombi are rare, share a similar appearance to a myxoma, and are highly mobile. Type B has a more favorable outcome and thrombolysis is not indicated. Thrombolytics are life-saving in type A.[5]

Therapeutic options for acute massive pulmonary embolism include anticoagulation, thrombolysis and catheter, or surgical embolectomy. The International Co-operative Pulmonary Embolism registry noted a mortality of 52% in patients with massive pulmonary

Figure 5: Upper esophageal ascending aortic short axis view, the arrow pointing at thrombus in the RPA. RPA: Right pulmonary artery, SVC: Superior vena cava
embolism and a strong correlation with RV thrombi, RV dysfunction, and congestive heart failure.\[6]\) In most institutions, the first line of treatment is usually thrombolysis (systemic or catheter directed). The contraindications for thrombolysis include recent stroke, surgery, or other possible sources of active bleeding. Thrombolysis can also cause thrombus fragmentation and distal embolization leading to chronic pulmonary hypertension.

Indications for surgical pulmonary embolectomy in this patient were the presence of a large free floating right ventricular thrombus, large clot burden, right ventricular dysfunction, the presence of an ASD, and worsening respiratory failure while on adequate doses of anticoagulation. The principal causes of postoperative mortality after pulmonary embolectomy are cardiac failure, CVA, and sepsis.\[7]\) Possible causes in this patient could be cardiac arrhythmia, hypoxic arrest, or recurrent pulmonary embolism.

**CONCLUSION**

Right ventricular thrombosis constitutes a rare, yet potentially fatal situation, whose optimal management remains to be evaluated. The differential diagnosis are tumors such as myxoma, carcinoid syndrome, and normal structures in the RV such as moderator band, trabeculation, and papillary muscles.

**Financial support and sponsorship**
Nil.

**Conflicts of interest**
There are no conflicts of interest.

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