Pulmonary artery sarcoma or chronic pulmonary thromboembolism – A diagnostic dilemma and an anaesthetic challenge

Sir,

Chronic thromboembolic pulmonary hypertension (CTEPH) and pulmonary artery sarcoma (PAS) though different entities with diverse prognostic implications continue to post a diagnostic dilemma to the clinicians because of their similar clinical and radiological appearances. The common non-specific symptoms of PAS include dyspnoea, angina, cough, haemoptysis, anorexia, fatigue and dizziness, but they may have a slow onset. Clubbing, right heart failure (RHF) are rare late presentations.[1] Inflammatory markers namely erythrocyte sedimentation rate, lactate dehydrogenase and C-Reactive protein have been found elevated in some patients.[2]

We present a 44-year-old female with two-month history of exertional breathlessness and pedal edema. Transthoracic Echocardiography (TTE) revealed a mass suspected to be a thrombus producing a partial flow obstruction in the main and right pulmonary artery (PA) with a gradient of 31 mmHg and no flow in left PA. Computed tomography pulmonary angiogram (CTPA) showed hypodense filling defects in main and right PA and complete obstruction of left PA [Figure 1]. She was treated and discharged on anticoagulants. However, she returned in two weeks with signs of RHF. A repeat CTPA reconfirmed previous findings. A diagnosis of CTEPH was made and pulmonary endarterectomy (PEA) planned.

The anaesthetic preparation included inserting an internal jugular vein (IJV) triple lumen catheter, IJV HemoSphere®PA catheter via PA sheath (tip kept in right atrium) and a radial arterial line. Femoral arterial line, IJV sheath and femoral vein sheath were placed under local anaesthesia in anticipation of need of emergent cardiopulmonary bypass (CPB) or postoperative peripheral veno-arterial extracorporeal membrane oxygenator, so that they may be exchanged for cannulae over wire if required. Trans-oesophageal echocardiography (TEE) under general anaesthesia corroborated the TTE findings [Figure 2]. After establishing CPB and cardioplegic arrest, deep hypothermia was achieved by cooling down to 18ºC.

On incising the PA, the mass had a consistency of a tumour rather than that of an organised thrombus, arising from the posterior wall of main PA. Intra-operative frozen section biopsy confirmed it to be a primary mesenchymal tumour. All accessible mass was resected under a brief period of total circulatory arrest (TCA) and sent for histopathology. TCA prevented backflow of blood via the bronchial arteries and thus gave better bloodless surgical field.[3] The PA catheter was placed in the PA by the surgeon so as not to injure the bare intima. It was possible to wean the patient off CPB on moderate inotropic support.

HemoSphere® monitor was connected. Use of inhalational nitric oxide (iNO) is documented in PEA for CTEPH.[4] The pathophysiology of postoperative pulmonary hypertension is same in PAS and it exacerbates RHF, increasing perioperative mortality risk.[5,6] We conjectured that iNO would be useful as a preventive measure. As the recorded Pulmonary vascular resistance (PVR) was 143 dyn-s/cm^5 (on the high normal side), iNO was started at 20 ppm delivery dose via NOXtec® delivery system and was continued in postoperative period. It was stopped once the PVR came down to 85 dyn-s/cm^5. RV ejection fraction increased from 33% to 47%. Patient was extubated after 38 hours and was transferred to the ward on the third postoperative day. Histopathology and

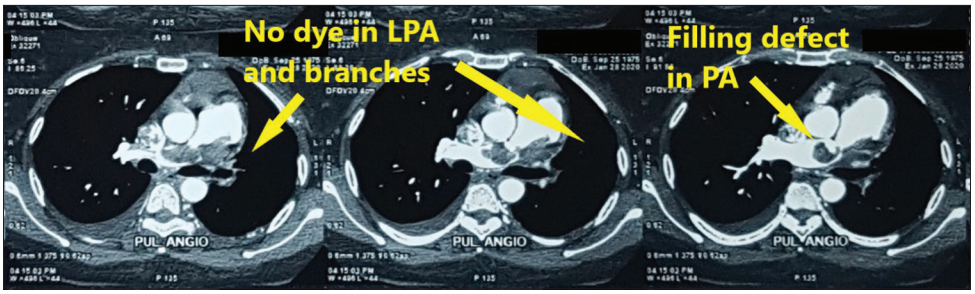

Figure 1: CTPA showing filling defect in pulmonary artery

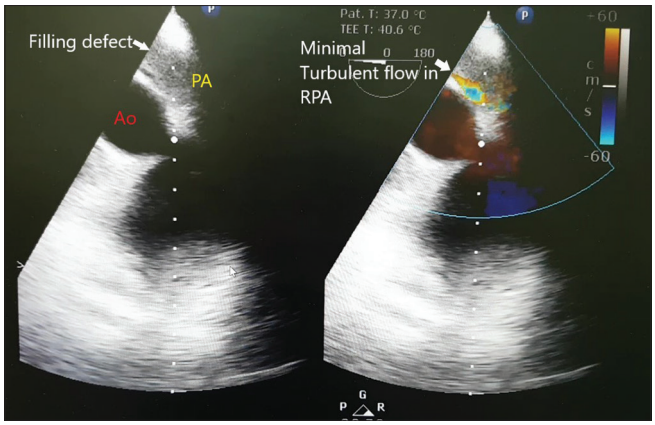

Figure 2: TEE mid-oesophageal ascending aorta short axis view showing filling defect in pulmonary artery with flow obstruction

positive immunohistochemical stains with Vimentin, Desmin and smooth muscle actin (SMA) confirmed the tumour as PAS. Further mainstay treatment with chemo-radiotherapy was started.[1]

The definitive management of CTEPH and PAS are different. While PEA has been shown to be the treatment of choice in CTEPH, adequate resection is the treatment in sarcoma that may even necessitate replacement of PA using a homograft. The anaesthetic management is similar in both cases with some modifications. Frequently PAS patients have cardiovascular instability due to either near total obstruction of PA by the mobile mass in the perioperative period or worsening RV failure. Postoperative bronchial bleed is also a dreaded complication. Unlike CTEPH, however, postoperative persistent pulmonary hypertension and RV dysfunction is rare and treatment should be titrated to the individual patient. High level of suspicion both on side of the surgeon and the anaesthesiologist and appropriate preparedness would help in proper diagnosis and treatment of this great mimicker.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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Submitted: 27-Sep-2020
Revised: 19-Oct-2020
Accepted: 31-Jan-2021
Published: 20-May-2021

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Access this article online	
Quick response code	Website: www.ijaweb.org
	DOI: 10.4103/ija.IJA_1235_20

How to cite this article: Jana S, Chowdhury SR, Narayan P, Das M. Pulmonary artery sarcoma or chronic pulmonary thromboembolism – A diagnostic dilemma and an anaesthetic challenge. Indian J Anaesth 2021;65:410-1.