Pulmonary Thromboendarterectomy in a Known Case of Antiphospholipid Syndrome

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A 23-year-old male was admitted to our hospital with complaints of shortness of breath, grade-III, chest pain, and palpitation for the last 3 months. He had one episode of hemoptysis 3 months back. He was a known case of primary antiphospholipid antibody syndrome (APS) with bilateral chronic pulmonary thromboembolism with chronic thromboembolic pulmonary hypertension (CTEPH) with the presence of anticardiolipin antibody. The patient had no history of joint pain, hematuria, and photosensitivity. He was on domiciliary oxygen therapy at 1 L/min to maintain an oxygen saturation of 93%, riociguat, ecosprin, hydroxychloroquine, and enoxaparin. The patient was referred to our hospital for pulmonary thromboendarterectomy (PTE).

On investigation, his two-dimensional (2D) echocardiogram showed global left ventricular hypokinesia with an ejection fraction of 25%, severe right ventricular systolic dysfunction, dilated right atrium and ventricle, dilated tricuspid valve annulus with severe tricuspid regurgitation, mild pulmonary regurgitation, high normal inferior vena cava with an absence of respiratory collapse, and pulmonary artery systolic pressure of 37 mm Hg. His computed tomography pulmonary angiogram (CTPA) revealed a thrombus in the right pulmonary artery. Bilateral lower limb venous Doppler showed no evidence of deep vein thrombosis. Doppler of the hepatic, portal, and inferior vena cava showed no evidence of thrombosis. The preoperative coagulation profile showed an activated partial thromboplastin time of 73.6 seconds, the International Normalized Ratio (INR) of 1.26, and a platelet count of 176,000/µL. The patient underwent PTE which was uneventful. The inferior vena cava filter was not inserted perioperatively in this case, as he had no evidence of deep vein thrombosis.[1] The patient was discharged on a postoperative day 11 on domiciliary oxygen 2 L/min. After 6 months of follow-up, the patient now has normal pulmonary artery systolic pressure and maintains an oxygen saturation of >94% on room air. Pulmonary artery systolic pressure can be expected to be high in patients with biventricular dysfunction. Pulmonary artery acceleration time can be a good indicator of pulmonary hemodynamics in cases of mild or insufficient tricuspid regurgitation.

APS is an autoimmune disorder characterized by the presence of recurrent arterial/venous thrombosis and fetal loss with the presence of antiphospholipid antibodies, namely, lupus anticoagulant or anti-Beta2 glycoprotein antibodies. APS is either primary or secondary which is in the absence or presence of other connective tissue disorders. The pulmonary manifestation of APS includes thromboembolism and pulmonary infarction, microvascular thrombosis, and alveolar hemorrhage. Recurrent pulmonary embolism may lead to CTEPH. Medical management of pulmonary hypertension is done but has little impact on survival. The prognosis of CTEPH is poor and the 5-year survival is 30% when the pulmonary artery systolic pressure exceeds 40 mm Hg.

Most of the patients with primary APS who underwent surgical PTE showed improved postoperative hemodynamics and quality of life.[2–4] PTE is a surgical procedure performed only in specialized medical centers. Lung transplantation is the only other hope of increasing survival in these patients. APS's unique features that complicate perioperative management include thrombocytopenia, specific anticoagulation

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needs, and increased risk of thrombosis and bleeding. The patients with APS who undergo PTE will need life-long anticoagulation with a target INR of 2 to 3. Neurological complications including stroke and delirium and severe thrombocytopenia were common in patients with APS after PTE.[5] Thrombotic events and bleeding complications were common in the postoperative period. Bleeding complications could be more because these procedures are performed under deep hypothermic circulatory arrest leading to coagulation abnormalities. Activated coagulation time (ACT) is an unsuitable indicator of adequate heparinization. An antiphospholipid antibody with lupus anticoagulant activity prolongs the phospholipid-dependent coagulation tests like activated partial thromboplastin time and ACT. Hence, it is difficult to monitor patients on heparin. Apart from PTE, other cardiac surgeries, including valve replacement, right atrial mass excision, and coronary artery bypass grafting in association with APS, have been described in the literature.[6]

Conflict of Interest
None declared.

References

1 Sasaki A, Suda R, Tanabe N. Efficacy and safety of inferior vena cava filters in patients with surgically treated chronic thromboembolic pulmonary hypertension. Eur Respir J 2019;54(suppl. 63):PA1463
2 Riedel M, Stanek V, Widimsky J, et al. Long term follow-up of patients with pulmonary thromboembolism. Chest 1982;81:151–157
3 Simonneau G, Azarian R, Brenot F, Dartevelle PG, Musset D, Duroux P. Surgical management of unresolved pulmonary embolism. A personal series of 72 patients. Chest 1995;107(1, suppl):52S–55S
4 Sandoval J, Amigo MC, Barragan R, et al. Primary antiphospholipid syndrome presenting as chronic thromboembolic pulmonary hypertension. Treatment with thromboendarterectomy. J Rheumatol 1996;23(04):772–775
5 Camous J, Decrombecque T, Louvain-Quintard V, Doubine S, Dartevelle P, Stéphan F. Outcomes of patients with antiphospholipid syndrome after pulmonary endarterectomy. Eur J Cardiothorac Surg 2014;46(01):116–120
6 Rawat RS, Mehta Y, Saxena P, Juneja R, Bhan A. Double valve replacement in a patient with antiphospholipid antibody syndrome. Ann Card Anaesth 2011;14(03):235–236

