A Case Report of Pulmonary Endarterectomy in a Tertiary Care Hospital in Eastern India

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Abstract: Pulmonary endarterectomy (PEA) is the preferred surgical treatment for patients with chronic thromboembolic pulmonary hypertension (CTEPH), aimed at removing obstructive thromboembolic material from pulmonary arteries, thereby reducing pulmonary vascular resistance, alleviating pulmonary hypertension, and improving right ventricular function. This case report discusses a 23-year-old female with antiphospholipid syndrome, presenting with symptoms such as shortness of breath, pedal edema, and jaundice. Diagnostic evaluations revealed severe pulmonary hypertension and chronic thrombosis. The patient underwent successful bilateral PEA, resulting in significant clinical improvement. The case underscores the importance of careful patient selection, meticulous surgical planning, and postoperative management in achieving favorable outcomes in CTEPH treatment.

Keywords: CTEPH, pulmonary endarterectomy, antiphospholipid syndrome, pulmonary hypertension, thromboembolic disease

1. Background

Pulmonary endarterectomy (PEA) is the treatment of choice to relieve pulmonary artery obstruction in patients with chronic thromboembolic pulmonary hypertension (CTEPH). It is a complex surgical procedure with the principle of removal of obstructive thromboembolic material from the pulmonary arteries to reduce pulmonary vascular resistance, relieve pulmonary hypertension (PH) and correct right ventricular dysfunction. In the majority of patients there is symptomatic and prognostic benefit.

However, not all patients with CTEPH are suitable for treatment with PEA. Operability assessment is not always easy, being largely subjective and based on experience. Such a case was referred to our Centre. After meticulous evaluation and planning we went for surgery. The patient is currently doing well and undergoing regular follow up in our opd. Patients who are unsuitable for surgery maybe candidates for medical therapy.

2. Case Summary

A 23 year unmarried female patient, a diagnosed case of Anti - phospholipid syndrome on medication presented with B/L pedal edema and leg pain along with shortness of breath for 2months and yellowish discoloration of skin for 1month. On examination she had raised JVP, jaundice, B/L pedal edema, hepatomegaly, ascites, loud P2, ejection systolic murmur over left sternal border.

Her ECG (A) shows Rt. atrial enlargement and Rt. axis deviation. Chest X - ray (B) shows globular enlarged heart with gross enlargement of Rt. atrial shadow, central pulmonary arterial dilatation, with 'pruning' (loss) of the peripheral blood vessels.2D echocardiography (C) shows RA, RV dilatation, severe pulmonary hypertension, severe TR. CT Angiography of pulmonary arteries (D) shows sub - acute pulmonary thrombo - embolism in postero - basal segmental branches of right lower lobe pulmonary artery and chronic thrombosis of left pulmonary artery (complete occlusion)

with pulmonary arterial hypertension. Pulmonary function test shows early small airway obstruction.

Blood tests show raised Anticardiolipin antibodies IgG and IgM (ELISA), raised Anti - beta - 2 - glycoprotein - I antibodies IgG or IgM (ELISA), Raised liver enzymes, raised conjugated bilirubin.

USG color Doppler of B/L Lower limbs shows irregular wall thickening with filling defect noted in B/L Femoral veins and Left popliteal vein.

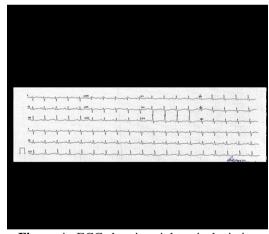


Figure A: ECG showing right axis deviation

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Figure B: CXR shows globular enlarged heart with gross enlargement of Rt. Atrial shadow, central pulmonary arterial dilatation, with 'pruning' (loss) of the peripheral blood vessels

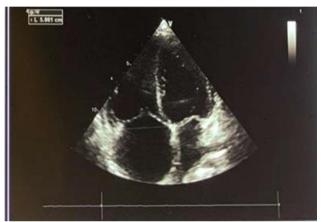


Figure C: 2D Echo shows RA, RV dilatation, severe pulmonary hypertension, severe TR



Figure D: CT Pulmonary Angiography shows sub - acute pulmonary thromboembolism in postero - basal segmental branches of right lower lobe pulmonary artery and chronic thrombosis of left pulmonary artery (complete occlusion)

Operative procedure:

The operation is performed through a median sternotomy incision to approach both lungs, with cardiopulmonary bypass (CPB) (established by ascending aortic and caval cannulation) enabling hypothermia to 20°C and circulatory arrest. Cooling to 20°C was carried out gradually over 90 min. Under DHCA right pulmonary arteriotomy was performed first, and the endarterectomy plane identified, followed by progressive dissection distally to remove the endarterectomy (specimen). The procedure was bilateral, and on completion

of the right PEA, bypass was resumed and the patient reperfused while the arteriotomy was closed so that the procedure can be repeated on the left side, with circulatory arrest being initiated as necessary. Total cross clamp time was 110mins.

Patient was rewarmed and weaned off CPB without any untoward event.

The patient was shifted to the ICU and kept on ventilator support. She was gradually weaned off and extubated after 48 hours. She was discharged on 7th post - operative day.



3. Discussion

The primary site of initiation of a thrombo - embolic event is debatable. Incomplete resolution and organization of the thrombus. infection. inflammation with genetic predisposition and in - situ thrombosis can lead to vascular occlusion and stenosis. Simultaneously, the sheer stress in non - obstructed vessels and vascular remodeling causes increased pulmonary vascular resistance and pulmonary artery pressures causing CTEPH. This progression to pulmonary hypertension after the initiating event may be attributed to pulmonary vascular remodeling in the entire pulmonary vasculature including both major and small vessels.

Risk factors for the development of CTEPH include malignancies, chronic inflammatory disorders, combined coagulation defects and antiphospholipid antibody syndrome. Idiopathic PE, young age, large perfusion defect and recurrent embolism are associated with increased propensity to CTEPH according to the literature. There are also instances where CTEPH has occurred even in the absence of any prior PTE.

4. Conclusion

CTEPH is a progressive and potentially lethal disease. Medical therapy is palliative and only surgery is definitive. Pulmonary endarterectomy (PEA) is the treatment of choice and lung transplantation should be considered only when PEA is contraindicated. Multidisciplinary collaboration allow appropriate patient selection, rigorous surgical technique, and adequate postoperative management. All these aspects represent the key to the success in the treatment of CTEPH. After PEA, quality and expected length of life are similar to the age - matched general population and the only therapy required is oral anticoagulation.

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List of abbreviations

- PEA Pulmonary endarterectomy
- CTEPH Chronic thromboembolic pulmonary hypertension
- JVP Jugular venous pressure
- B/L Bilateral
- RA Right Atrium
- RV Right Ventricle
- · DHCA Deep hypothermic circulatory arrest
- PTE Pulmonary thromboembolism

Declarations

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