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CASE REPORT

Pulmonary Thromboendarterectomy for Chronic Pulmonary Thromboembolism

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SUMMARY

Pulmonary embolism in terms of causes of mortality is the third most common cause, when talking about cardiovascular diseases, yet most undiagnosed or misdiagnosed one. Among causes of pulmonary thromboembolism protein C deficiency is a rare one and idiopathic most common entity. In general population the incidence of severe protein C deficiency is about 1 in 500,000-750,000 people. We present a case of chronic pulmonary thromboembolism in young female of 23 years. She presented with history of dyspnea, dry cough and bilateral DVT of legs. On pulmonary CT diagnosis of massive pulmonary thrombus was made and her pulmonary thromboendarterectomy was done in our center.

Conclusion: All patients with chronic pulmonary thromboembolic hypertension should be assessed for operability by proper referral to an experienced CTEPH (chronic thromboembolic pulmonary hypertension) team to determine if they are viable candidate for PEA (pulmonary endarterectomy). PEA is standard and recommended operative technique for treatment of CTEPH.

Keywords: pulmonary endarterectomy, pulmonary embolism, protein C deviciency

INTRODUCTION

To define pulmonary embolism we can say it is the obstruction of pulmonary vasculature by thrombus which has embolized from other site in the blood stream causing variety of symptoms depending upon dimensions and location of thrombus[1,8].

Taking account of the fact that incidence of pulmonary embolism is high even then it is difficult to diagnose it, primary reason being vagaries of signs and symptoms in its presentation. Due to difficulty of diagnosis, the true incidence of pulmonary embolism is still unknown, however it is estimated that almost 650,000 cases occur annually[2]. It may exhibit as vague a symptom as syncope or may present with 3 variants including pulmonary infarction, acute unexplained dyspnea, and acute corpulmonale. Pulmonary infarct occurs when a thrombus is embolized and completely hampers blood supply of a distal branch of the pulmonary circulation. Patients with this condition have symptoms of pleuritic chest pain, hemoptysis, rales, and abnormal findings on chest X-ray[3].

Fig 1: Thrombus after thromboendarterectomy

Unexplained sudden dyspnea may be the result of sub-massive pulmonary embolism without occurrence of pulmonary infarction. Obstruction of 60 to 75% of pulmonary circulation leads to acute corpulmonalesybdrome. These patients experience continuum of symptoms including shock, syncope, or sudden death. Here in our case patient presented with unexplained dyspnea and history of recurrent DVTs[4].

Common cause of VTE which leads to pulmonary embolism are, inactivity, blood vessel damage, medical and genetic conditions like cancer, protein C and S deficiency, pregnancy, contraceptive pills and HRT and last but not the least idiopathic[5,7].

Prompt diagnosis is a key to save a life. Once the diagnosis of pulmonary thromboembolus is made surgery is the answer for complete removal of thrombus.

Fig 2: Guidelines for the treatment of CTEPH

CASE REPORT

Twenty four years old female presented with history of dyspnea and bilateral leg DVT in March 2018 in our center. She was found to be in sinus tachycardia with stable blood pressure and maintaining oxygen saturation of 80% at room air. Her systemic examination was unremarkable. Patient had history of IVC filter implantation on 29 March 2018 due to recurrent DVTs. All routine blood tests were within normal range.ECG was also insignificant. Her echocardiography showed: Dilated right sided chambers. Pulmonary hypertension lying in category of moderate to severe. Her pulmonary CT scan showed clot in LPA (left pulmonary artery)

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and in its peripheral branches. After final diagnosis of pulmonary thrombosis, plan for pulmonary thrombectomy was made and done on 6th April 2018.

Fig 3: Pre-operative Echocardiography of the patient

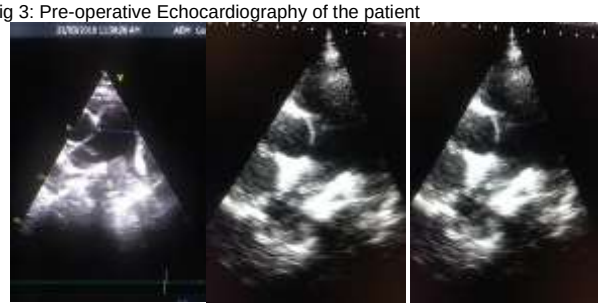

Fig 4: Pre-operative pulmonary CT showing clot in left pulmonary artery

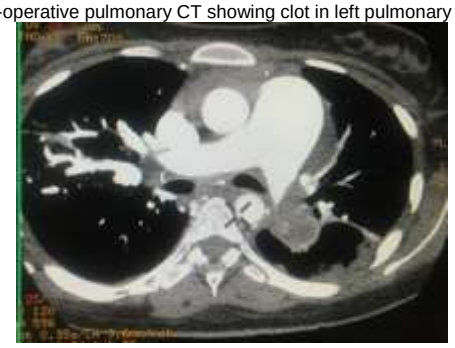

Fig 5: Pre-operative pulmonary CT showing clot in left pulmonary artery

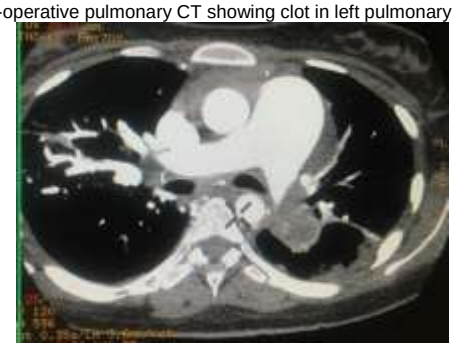

Fig 6: Post-operative picture of thrombus after complete thromboendarterectomy

Fig 7: Post-operative picture of thrombus after complete thromboendarterectomy

Surgical Technique: Median sternotomy was done as it is a prerequisite of surgery for adequate exposure.CPB was established between aortic and bicavalcanuulation. Myocardial protection was achieved with antegrade cold blood cardioplegia. After cross clamping, main pulmonary artery was opened and the incision was extended into left pulmonary artery.

Fig 8: Picture showing pulmonary arteriotomy

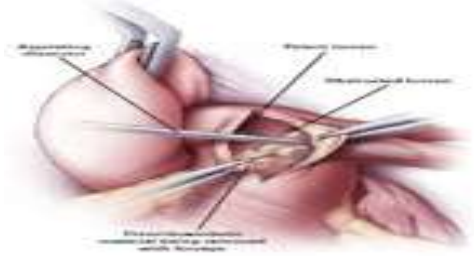

Fig 9: Per-operative view of pulmonary arteriotomy

Pulmonary thromboendarterectomy was performed in piece meal.After left sided thromboendarterectomy pulmonary artery was closed, cross clamp off, off CPB and routine closure was done.

Retrograde Pulmonary Perfusion: This is used to enable the complete extraction of residual thrombotic material from the distal branches of the pulmonary artery as it causes filling of pulmonary artery with blood in this way it also prevents pulmonary air embolism.In this technique left atrium or left superior pulmonary vein is filled with blood hence pulmonary artery is filled retrogradely (flushing of pulmonary circulation),during retrograde pulmonary perfusion lungs are inflated in order to facilitate the elimination of residual thrombus and air bubbles present in the distal branches of the pulmonary artery.

Postoperative outcome: Preop: CVP: 21mmHg postop CVP:9mmHg/ Patient was discharged on 25th POD with a good course of recovery and is on follow up with lifelong anticoagulation.

DISCUSSION

Pulmonary thromboembolism a squeal of VTE having multiple causes is one of the pronounced cause of mortality in hospital and outside as well. But due to its varied presentation and nonspecific nature of symptoms like dyspnea, cough, pleuritic pain, sub sternal chest pain, fever, syncope and hemoptysis, it remains undiagnosed[1].

To label it as event of massive pulmonary embolism signs includes tachycardia, raised JVP hypotension, right ventricular gallop rhythm, splitting of pulmonary component of 2nd heart sound, severe cyanosis and oliguria. Sub massive or medium pulmonary embolism has symptomatic variance of pleurisis, difficulty in breathing and coughing out blood. Chronic pulmonary thromboembolism illustrates signs of right sided heart failure whose symptoms comprise of light headedness, lethargy and early fatigability, fluid retention producing swelling in lower extremities or abdomen)[2].

In all cases one or more predisposing factors like history of recent surgery, traumatic injury, bed restricted patients, pregnancy, and pelvic trauma must be there which may aid in formulating the diagnosis of PTE.Protein C deficiency is among rare cause of PTE. Fundamentaldisplay of protein-C deficiency is venous thromboembolism but someprecipitating factor is usually present to cause PTE but in our case there were no precipitating factors to cause PTE[3-4].

Diagnosis of PTE includes:Clinical picture, routine blood investigations CXR (which may show different signs depending upon size, duration and site of PTE) echocardiography and pulmonary CT angiography[5]. CXR may show ranging from WESTERMARK sign (oligemic lung field distal to thrombus) to no finding on CXR.There may be appreciation of infarcted pulmonary area or signs of Right heart failure.ECG is not specific but helps to exclude other diagnosis.

Investigation of choice for definitive diagnosis of pulmonary embolism is Computed tomography pulmonary artery (CTPA) and it has made conventional angiography almost obsolete now. Once the diagnosis is confirmed treatment should be initiated immediately[6,9].

Treatment options include: Thrombolytic therapy, Anticoagulant therapy, Combination therapy or surgery. Depending upon severity and presentation of disease choice of treatment is made[10].

CONCLUSION

All patients with chronic pulmonary thromboembolic hypertension should be assessed for operability by proper referral to an experienced CTEPH (chronic thromboembolic pulmonary hypertension) team to determine if they are viable candidate for PEA (pulmonary endarterectomy). PEA is standard and recommended operative technique for treatment of CTEPH. The role of bridging with medical therapy is still not completely explored yet and it should be kept back for controlled investigation until further research.

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