Indian J Hematol Blood Transfus (June 2016) 32 (Suppl 1):S242–S245
DOI 10.1007/s12288-015-0614-2

CASE REPORT

Pulmonary Hypertension and Pulmonary Aspergilloma-Coexistence of Two Rare Sequelae of Pulmonary Embolism

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Received: 20 July 2015 / Accepted: 19 October 2015 / Published online: 26 October 2015

Abstract We report a 42 year old non-smoker male who presented with progressive exertional dyspnoea, productive cough with streaky hemoptysis and progressive pedal edema. His physical examination, ECG, chest X-ray and 2D-ECHO revealed features suggestive of right heart failure and pulmonary hypertension. On further evaluation for the cause of pulmonary hypertension, his CT pulmonary angiography revealed features of chronic pulmonary thromboembolism with calcified thrombus in the main pulmonary artery along with pulmonary hypertension. Incidentally the CT also revealed a cavity in the right lung with soft tissue within it. A, trans-thoracic needle aspiration of this tissue was suggestive of an aspergilloma. This is a rare case report of co-existence of two uncommon complications of pulmonary embolism-chronic thromboembolic pulmonary hypertension and pulmonary Aspergilloma in the same patient.

Keywords Chronic thromboembolic pulmonary hypertension · Aspergilloma · Deep venous thrombosis · Pulmonary infarction

Introduction

The natural history of pulmonary embolism (PE) dictates that most thrombi undergo spontaneous resolution (with lysis and organization) with a near complete restoration of perfusion within 1 year. [1, 2] Chronic thromboembolic pulmonary hypertension (CTEPH) occurs in about 1.5 % of cases. Aspergilloma is an extremely rare complication of pulmonary embolism listed with a frequency of around 1 % [3, 4]. We present a case in which PE is complicated by both CTEPH leading to corpulmonale and aspergilloma as a sequel of pulmonary infarction. The co-existence of these two rare sequelae of PE has not been reported earlier, to the best of our knowledge.

Case Report

A 42 year male patient was admitted to our hospital with a 5 months history of exertional dyspnoea, productive cough with streaky hemoptysis and progressive bilateral lower limb swelling. He was a non-smoker and he gave history of amputation of the left great toe for gangrene in the past. On admission his physical examination revealed tachycardia (heart rate 120 per minute), tachypnea (respiratory rate 26 per minute), hypotension (BP of 90/60 mmHg), raised JVP with prominent "a" wave, bilateral pedal edema and oxygen saturation of 90 % at room air. Systemic examination revealed crackles in bilateral basal areas and non-tender hepatomegaly palpable 4 cm below right costal margin with span of 16 cm. Cardiovascular examination revealed loud pulmonary component of the second heart sound, and right ventricular third heart sound. ECG showed sinus tachycardia, right axis deviation, P pulmonale and right ventricular hypertyrophy. Chest radiograph revealed cardiomegaly, right atrial and ventricular enlargement with fullness of pulmonary bay. Patient was initially managed as right heart failure with diuretics.

The haematological (Hb 11.3 gm%, TLC 7700/mm^3, P77, L18, E03, M02, Platelets 1.23 lac/mm^3) and biochemical investigations did not reveal any significant

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abnormality. His base line coagulogram revealed PT test 15, control 13, PTTK test 27 control 30 and INR 1.23. 2D-ECHO showed dilated right atrium and right ventricle with pulmonary hypertension. The pulmonary artery systolic pressure was 54 mmHg. There was evidence of mild tricuspid regurgitation. Left ventricular function was however normal.

CT pulmonary angiography (Fig. 1) showed a well-defined non- enhancing filling defect (2.2 × 1.7 × 1.8 cm) at bifurcation of main pulmonary artery (MPA) adherent to its posterior wall with calcification and interventricular septum (IVS) convexity towards left ventricle (MPA = 33.3 mm ascending aorta = 24.4 mm) suggestive of pulmonary hypertension. Incidentally the same CT showed thick walled cavity in superior segment of right lower lobe with soft tissue content within and mild pericardial effusion. Rest of the lung was normal. Doppler study of lower limbs showed calcified plaque in common femoral vein at right common femoral bifurcation suggestive of chronic deep venous thrombosis (DVT). Prothrombotic which included work-up included screen for ANA, APLA, ACLA, serum homocysteine levels, Factor V Leiden mutations and Anti thrombin levels, Protein C & S levels done after 3 months did not reveal any abnormality. He underwent CT guided transthoracic needle aspiration from the lung mass the culture of which grew yellowish green colonies after 48 h incubation at room temp in Sabouraud Dextrose agar (Fig. 2). Mount from colonies on Lacto-phenol cotton blue (LCB) (Fig. 2) showed *Aspergillus flavus*. Screening for tuberculosis which included analysis of sputum and transthoracic lung aspirate for acid fast bacilli, Mycobacterial cultures and PCR were negative. In addition, cytological evaluation of trans-thoracic lung aspirate did not show any evidence of Tuberculosis.

Patient was diagnosed as chronic DVT right lower limb with pulmonary embolism and CTEPH. Since there was no other evidence of chronic lung disease and considering the location of the aspergilloma, it was presumed to be a sequel of pulmonary infarction. Patient was managed with anticoagulation, pulmonary vasodilators and diuretics. Patient improved symptomatically with the treatment. Pulmonary endarterectomy was considered but not resorted to in view of general condition of the patient.

Discussion

Proximal DVT is the commonest cause of PE [1]. After the acute episode, the pulmonary thrombus undergoes spontaneous resolution in survivors with part lysis of the clot and part organization and recanalization. Though minor perfusion defects remain, the restoration of perfusion is near complete in more than 90 % patients. Around 1 % patients may develop CTEPH after pulmonary embolism. The risk factors for development of CPTEH include recurrent, unprovoked or idiopathic PTE, underlying thrombophilia, raised Factor VIII levels and significant perfusion defect at presentation. Others include underlying malignancies, infected cardiac shunts and connective tissue disorders such as antiphospholipid antibody syndrome [5]. It has been found that nearly half of the patients of CTEPH do not give history of having received anticoagulation for PE. However, a comparative study of treated and untreated patients of PE did not show any significant differences in

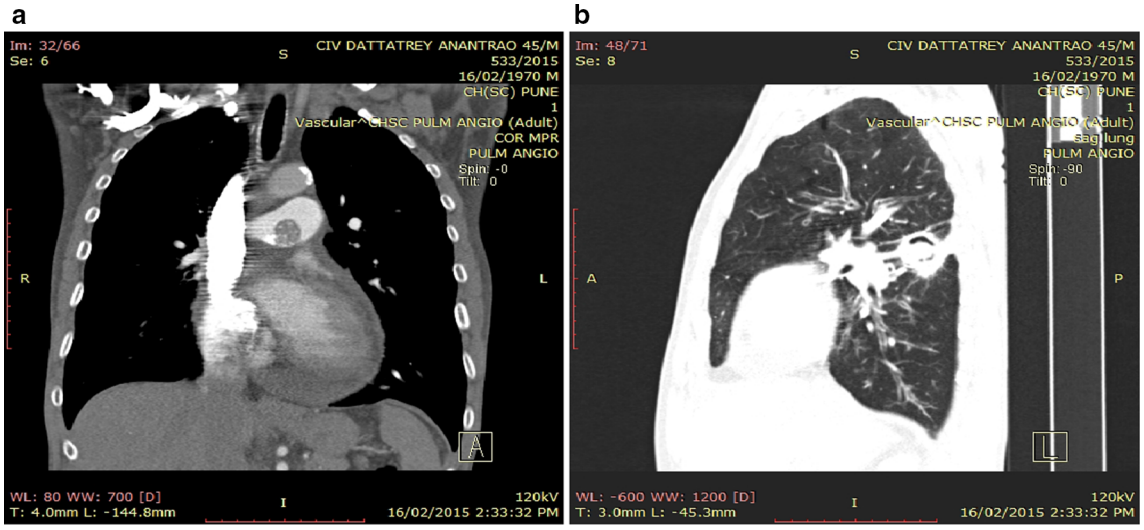

Fig. 1 **a** CT pulmonary angiography showing well defined non-enhancing filling defect (2.2 × 1.7 × 1.8 cm) at bifurcation of main pulmonary artery (MPA) adherent to its posterior wall with calcification. **b** Thick walled cavity in superior segment of right lower lobe with soft tissue content within and mild pericardial effusion

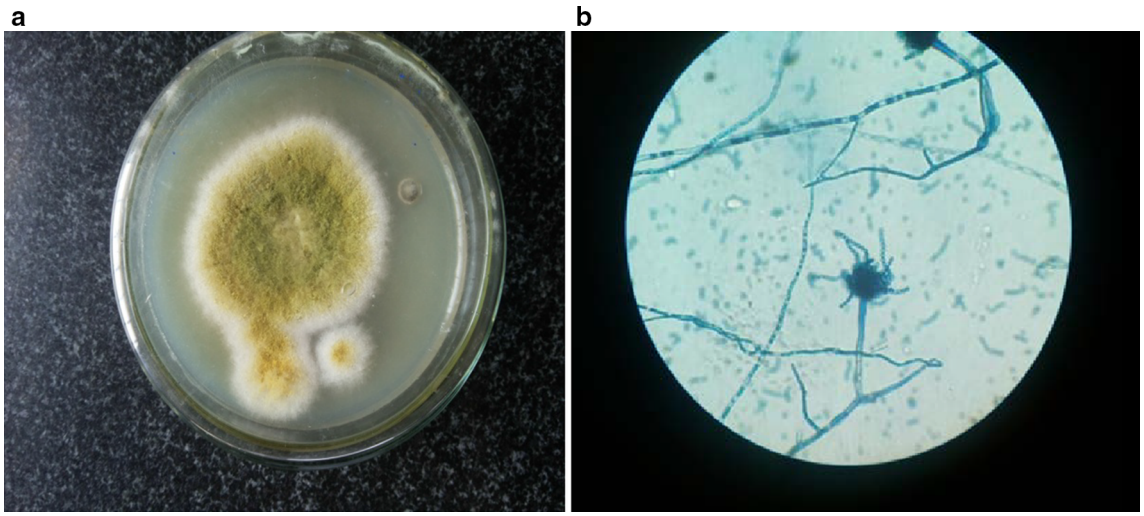

Fig. 2 **a** *Yellowish green* colonies after 48 h incubation at room temp in Sabouraud Dextrose agar. **b** Mount from colonies on Lactophenol *cotton blue* (LCB) showing septate hyphae branching suggestive of *Aspergillus flavus* (color figure online)

their mean pulmonary artery pressures after 3 months of PE [6].

Exact pathogenesis of CTEPH is unknown with endothelial dysfunction and abnormal fibrinolytic mechanisms often implicated as causes [5]. CTEPH presents as right sided heart failure with complaints of dyspnoea, pedal edema, chest pain and near-syncope or syncope. Physical examination may reveal subtle findings like narrowing of S2 with a loud pulmonary component. Later signs of overt right ventricular dysfunction may develop. Few cases may have presence of flow murmurs over the lung fields, and this is a unique finding of CTEPH. Increased pulmonary pressures can be demonstrated by standard tools. History of past PE is often not there and diagnosis is made while a hunt is being made for secondary causes of pulmonary hypertension. Ventilation/perfusion scan and CT pulmonary angiography are preferred modalities to establish the diagnosis of CTEPH while right heart catheterization and pulmonary angiography remain the gold standard. Pulmonary artery endartrectomy is the treatment of choice if surgically resectable. Medical management is indicated in cases which are inoperable or as bridge to surgery. Effective drugs include endothelin receptor antagonists, phosphodiesterse inhibitors or prostanoids; all having equal efficacy in symptomatic improvement [5].

Pulmonary infarction in pulmonary thromboembolism has incidence of 10–32 % in various studies [1, 5]. Infarction is uncommon since lung parenchyma has alternative sources of oxygen in the form of bronchial arteries and airway oxygen. It accounts for less than 10 % of all cavitary lung diseases. Mean duration of development is 2–63 days. There is no specific location. Initially cavity is aseptic but there is 50 % chance of superinfection. Common organisms being gram negative bacteria. Aspergilloma in an infarct cavity is rare [7].

This was a rare happenstance of two uncommon complications of pulmonary embolism occurring together. The fact that the original episode of pulmonary embolism was missed and that he did not receive anticoagulation might have been a possible culprit. However, as mentioned earlier, it has not been universally shown that treated and untreated patients, among survivors of PE have different outcomes [6]. There are many lacunae in the knowledge of natural history and outcomes of PE and one of the reasons is that a large number of patients remain undiagnosed. Retrospective analysis of data from large registries and hospital records would probably be of assistance in bridging these lacunae.

Conclusion

CTEPH and aspergilloma are both rare sequelae of pulmonary embolism and co-existence of the two is being reported for the first time. Both of these sequelae may have life threatening consequences but often have only subtle clinical and radiological findings. Awareness and high index of suspicion is therefore required, during follow-up of survivors of pulmonary embolism, to ensure that these rare complications are diagnosed timely and appropriate treatment is instituted.

Compliance with Ethical Standards

Conflict of interest None.

References

1. Konstantinides SV, Torbicki A, Agnelli G et al (2014) ESC guidelines on the diagnosis and management of acute pulmonary embolism. Eur Heart J 2014(35):3033–3073
2. Miniati et al (2006) Survival and restoration of pulmonary perfusion in a long-term follow-up of patients after acute pulmonary embolism. Medicine 85:253–262
3. Regnard et al (2000) Aspergilloma: a series of 89 surgical cases. Ann Thorac Surg 69:898–903
4. Passera et al (2012) Pulmonary aspergilloma clinical aspects and surgical treatment outcome. Thorac Surg Clin 22:345–361
5. Morris TA, Fedullo PF (2010) Pulmonary thromboembolism. In: Murray JF, Nadel JA, Mason RJ et al (eds) Textbook of respiratory medicine, vol 51, 5th edn., SaundersElsevier Inc, Philadelphia, pp 1186–1216
6. Stein et al (1995) Untreated patients with pulmonary embolism—outcome, clinical, and laboratory assessment. Chest 107:931–935
7. Gadkowski LB, Stout JE (2008) Cavitary pulmonary disease. Clin Microbiol Rev 21(2):305–333