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Case Report

Internal Medicine Section

Pulmonary Artery Aneurysm Thrombosis with Combined Pulmonary Fibrosis and Emphysema: A Case Report

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ABSTRACT

We report a rare case of Pulmonary Artery Aneurysm (PAA) thrombosis with Combined Pulmonary Fibrosis and Emphysema (CPFE) with pulmonary hypertension. A 75-year-old male presented with haemoptysis, dyspnoea, clubbing and bilateral fine end inspiratory rales on examination. He was diagnosed to have PAA thrombosis with CPFE on the basis of computed tomographical angiography and high resolution computed tomography. He was then managed conservatively with pirfenidone for the interstitial lung disease.

Keywords: Dyspnoea, Haemoptysis, Pulmonary hypertension

CASE REPORT

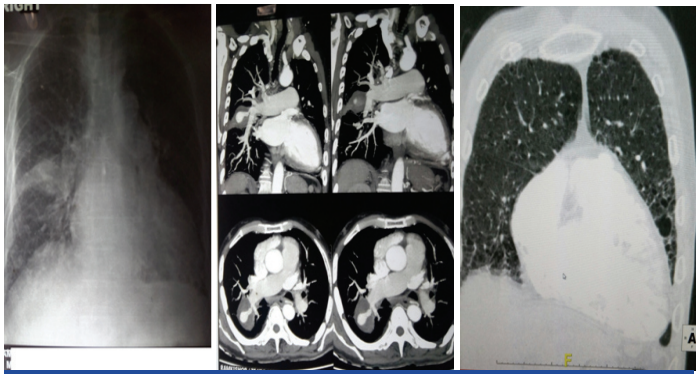
A 75-year-old male presented in the outpatient services in the Department of Pulmonary Medicine at Lokmanya Tilak Municipal Medical College and General Hospital. He presented with four episodes of haemoptysis (less than 10ml) over 1 month duration. Patient also complained of progressively increasing exertional dyspnoea of 1 year duration. The patient denied history of other respiratory complaints as well as other systemic complaints. Clinical criteria of Behçet's disease (e.g., oral-genital ulcers and ocular affection) were absent. The patient gave history of bidi smoking at the rate of 1bundle per day for 50 years (50 pack years). The patient denied history of similar illness in family.

General examination revealed pulse 84 beats per minute, respiratory rate 18 per minute and blood pressure 110/70 mmHg. Clubbing was present while pallor, cyanosis, icterus and lymphadenopathy were absent.

Respiratory examination revealed bilateral fine end inspiratory rales heard in infrascapular area. Other systems examined were within normal limits.

Routine Laboratory investigations like complete blood count, erythrocyte sedimentation rate in first hour, blood sugar, renal and hepatic functions were within normal limits. Serum immunological profile e.g. cANCA, pANCA, rheumatoid factor, antinuclear antibody and antibody to HIV were negative.

Spirometry could not be performed because of active haemoptysis and risk of aneurysmal rupture. The arterial blood gas was within normal limits with pH of 7.409, pCO_2 of 38, pO_2 of 64, HCO_3 of 24 and saturation of 92%.



[Table/Fig-1]: Chest X-ray showing Right para hilar opacity with convergence sign positive. **[Table/Fig-2]:** CECT Thorax showing right descending pulmonary artery aneurysm thrombosis. **[Table/Fig-3]:** HRCT Thorax showing bilateral upper zone emphysematous changes with interstitial thickening in bilateral lower lobes.

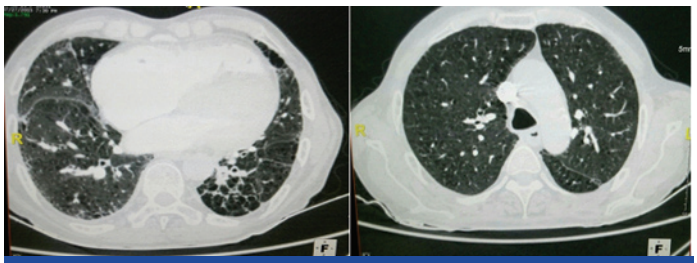
A 2-D echo revealed pulmonary artery hypertension with pulmonary artery systolic pressure of 65mm Hg , dilated right atrium and right ventricle and degenerative affection of aortic and mitral valve with mild aortic regurgitation. No evidence of thrombosis was seen.

The chest radiography [Table/Fig-1] revealed right midzone opacity with hilum convergence sign positive and reticular shadows bilaterally in all the zones.

Contrast-Enhanced Computed Tomography Thorax [Table/Fig-2]: A well defined heterogenous predominantly hypodense lesion (HU +40 to +45) measuring approximately 4.3 x 3.9 x 3.3cm (anteroposterior x mediolateral x superoinferior) in the superior segment of right lower lobe which showed a 1.6 x 1.5 x 1.7 cm (anteroposterior x mediolateral x superoinferior) sized central area of contrast opacification {(avg HU +206) on arterial phase followed by gradual decrease in enhancement (avg HU +128) on venous phase corresponding the enhancement of pulmonary vasculature} with peripheral non enhancing component of average thickness 1.4 cm. It was supplied by one of the segmental branch immediately arising from descending right pulmonary artery. This lesion was suggestive of right pulmonary artery aneurysm with partial thrombosis. Cardiomegaly was noted. Pulmonary vasculature appears dilated with main pulmonary artery measuring 31mm, right pulmonary artery measuring 28mm and left pulmonary artery measuring 25mm suggesting pulmonary hypertension.

Extensive areas of centriacinar and paraseptal emphysematous changes in apical segments of bilateral upper lobes with diffuse inter and intra lobular septal thickening and early changes of honeycombing in bilateral lower lobes suggest of CPFE [Table/Fig-3,4].

The patient refused for surgery as well as embolisation. So he was managed conservatively with haemostatics as he was having haemoptysis. He was treated with pirfenidone for CPFE.



[Table/Fig-4]: HRCT Thorax showing bilateral upper zone emphysematous changes with interstitial thickening in bilateral lower lobes along with honeycombing.

DISCUSSION

Pulmonary artery aneurysm (PAA) is a very rare condition found in approximately 1 of every 14 000 autopsies [1-3]. Its natural history is yet not well known and there are no clear guidelines about its optimal management [1]. Besides combined pulmonary fibrosis with emphysema (CPFE) is also rarely recognized clinically and radiologically. This syndrome classically occurs in male smokers and is often complicated by pulmonary hypertension, acute lung injury, and lung cancer [4]. However occurrence of pulmonary artery aneurysm thrombosis in CPFE has not been reported. So we report a case of patient with CPFE with pulmonary hypertension with pulmonary artery aneurysm thrombosis.

In 1947, Deterling and Clagett published a review of proximal PAAs over a period of 100 years and the incidence was reported to be 0.0073% [5]. It can be idiopathic or due to several pathologies, such as congenital cardiac anomalies (in more than 50%) associated with pulmonary hypertension, pulmonary valve stenosis, Behcet's disease and generalized vasculitis, infections and trauma [1]. PAA associated with significant primary or secondary pulmonary hypertension poses a high risk of dissection and rupture [6,7]. This patient also had pulmonary hypertension mostly secondary to underlying CPFE. Besides thrombosis of pulmonary artery aneurysm is also very rare. The prevalence of CPFE has been suggested to be between 5% and 10% of cases of diffuse interstitial lung diseases [8].

CONCLUSION

So here we report a rare case of CPFE with pulmonary artery aneurysm thrombosis.

Furthermore there has been no case report on co-existence of CPFE with pulmonary artery aneurysm thrombosis with pulmonary hypertension. So this case report may enhance the understanding and may add to the literature on pulmonary artery aneurysm and CPFE.

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