Combined pulmonary fibrosis and emphysema with lung cancer: Triple calamity

Sir,

Combined pulmonary fibrosis and emphysema (CPFE) has been increasingly understood to be a distinct entity characterized by the presence of histological or radiological evidence of lower lobe fibrosis and upper lobe emphysema, commonly seen in males and smokers, with reasonably preserved lung volumes and disproportional impairment in gas exchange.[1] The development of lung cancer in this group of patients has been reported, and various studies have looked at how this group of patients differ from patients with lung cancer without CPFE.

A 67-year-old male presented with increasing dyspnea of 2 weeks' duration associated with nonproductive cough. He had been having exertional dyspnea for over a year and has been on inhaled salbutamol therapy as needed. He had stopped smoking cigarettes 5 years ago and had smoked for over 25 years. Clinical examination revealed pandigital clubbing, tachypnea with a respiratory rate of 20/min, room air saturation of 92%, and bibasilar end-inspiratory crepitations along with diminished intensity of breath sounds over bilateral apices. Blood investigations revealed a total serum protein and albumin of 8.3 and 3.7 g per deciliter (g/dL), respectively (normal range of 6.6–8.3 and 3.5–5.2 g/dL, respectively), with normal complete blood counts, coagulation profile, and renal and liver function tests. Arterial blood gases revealed a pH of 7.38, pCO_2 of 44.4 mmHg, pO_2 of 75.6 mmHg, and HCO_3 of 25.8 mmol/L. He could not perform a spirometry in view of dyspnea.

Chest X-ray posteroanterior view showed bilateral lower zone reticular shadows with a left upper zone lung mass and paucity of lung markings in the right upper zone [Figure 1]. High-resolution computed tomography (CT) of the thorax showed bilateral lower lobe subpleural microcystic changes, upper lobe centrilobular and paraseptal emphysema, and a left upper lobe spiculated heterogeneous lung mass measuring 2.5 cm × 4.8 cm × 4.2 cm with mediastinal lymphadenopathy ($T_3N_3M_0$) [Figures 2 and 3]. CT-guided biopsy of the left lung mass revealed squamous cell carcinoma. He was subjected to four cycles of chemotherapy (intravenous docetaxel and carboplatin) at 3 weekly intervals, following which whole-body positron emission tomography scan was done and revealed stable disease. He was then subjected to 35 cycles of radiotherapy. Repeat CT showed stable disease, and after a multidepartmental team review, it was decided to stop any further therapy in view of poor performance status. At this point of time, he required regular home supplemental oxygen to maintain a room air saturation of above 90%.

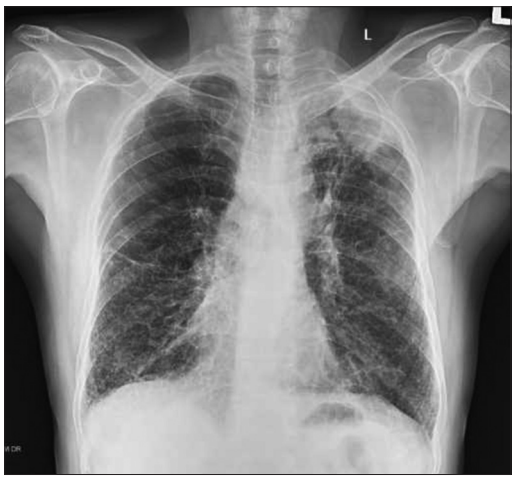

Figure 1: Chest X-ray posteroanterior view shows bilateral lower zone reticular shadows with a left upper zone lung mass and paucity of lung markings in the right upper zone

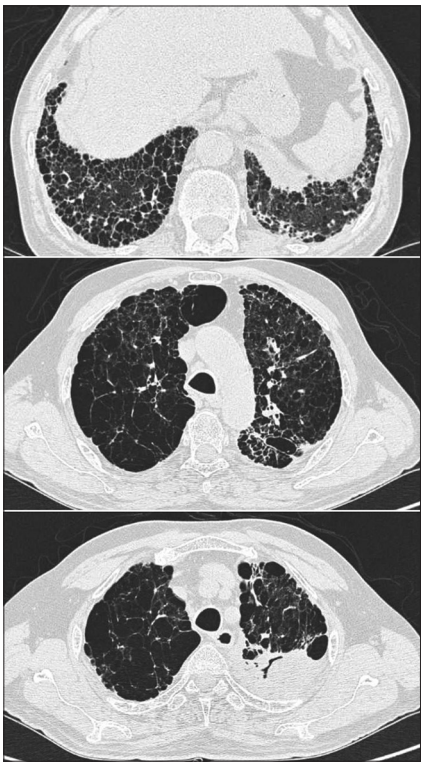

Figure 2: High-resolution computed tomography of the thorax showing: (top) bilateral lower lobe microcystic changes with architectural distortion at the level of cardiophrenic recess, (middle) paraseptal and centrilobular emphysema bilaterally, in addition to subpleural microcystic changes in the right upper lobe at the level of arch of aorta, and (bottom) similar emphysematous changes bilaterally with a left upper lobe lung mass with spiculated margins, at the level of left brachiocephalic vein crossing over the midline

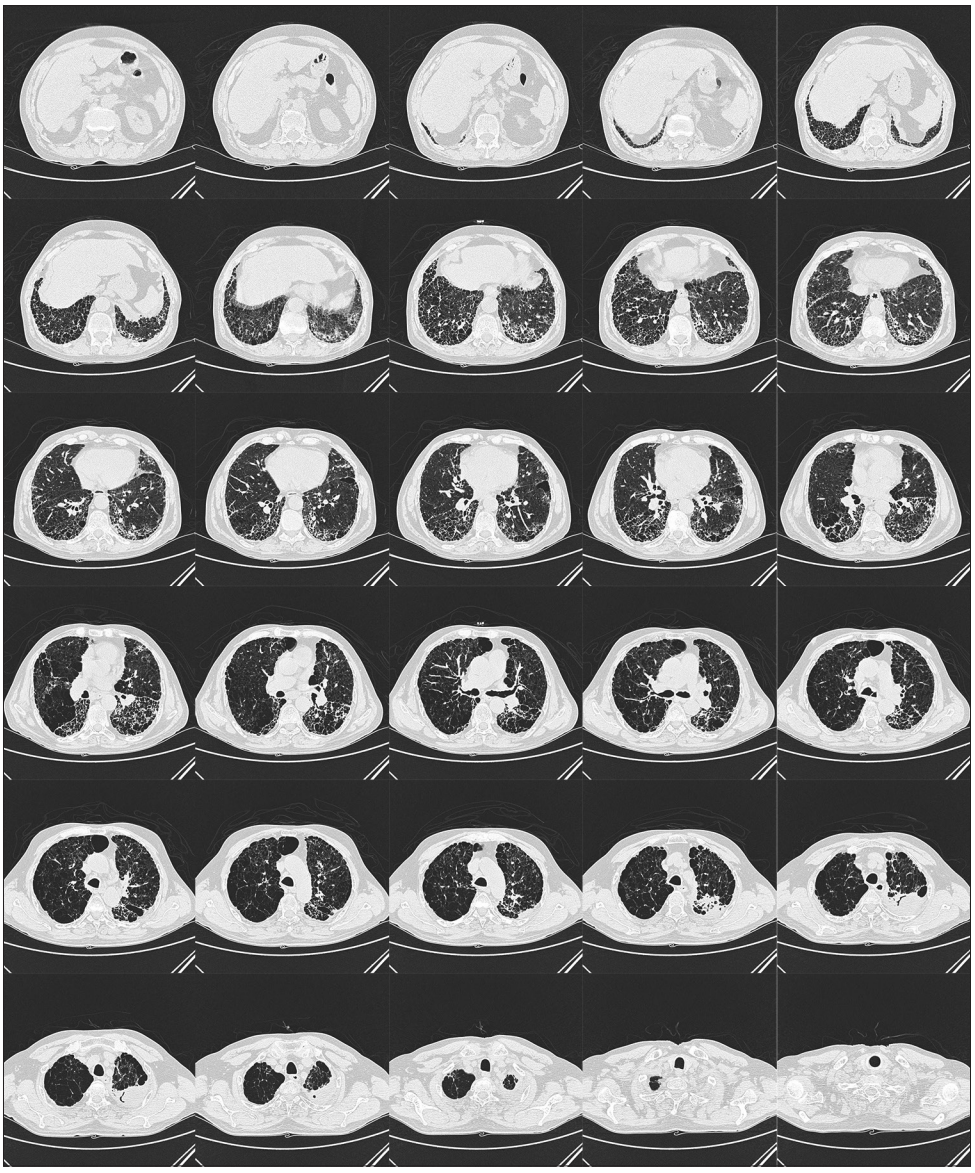

Figure 3: GIF image shows high-resolution computed tomography images of the thorax from caudal to cranial showing bilateral lower lobe fibrosis and bilateral upper lobe emphysema with a left upper lobe lung mass with spiculated margins

Pulmonary hypertension and lung cancer are known to complicate CPFE in its natural course.[1] Compared to patients with lung cancer without CPFE, lung cancer patients with CPFE are likely to be older, predominantly male, and ex- or current heavy smokers. Histologically, squamous cell carcinoma is more common than adenocarcinoma, with lower lobe preference and advanced pathological stage common in these patients. Higher overall mortality and incidence of postoperative complications in patients post tumor resection in operable candidates, along with poorer overall survival has also been reported in these patients.[1-3]

Squamous cell histology of lung cancer is more significantly associated with tobacco use and may explain why it is more common in patients with CPFE, which in turn is characterized by a combination of fibrosis and emphysema, both again, being strongly related to tobacco use.[1,4] Lower lobe preference is attributed to the causal role of fibrosis to carcinogenesis and explains why a similar radiological presentation is seen in patients with lung cancer and lung fibrosis alone without emphysema.[2,5] Our patient, on the contrary, had upper lobe malignancy, which is not unexpected, considering that dysplastic epithelium can also develop in emphysematous lung parenchyma. Our patient was also not a candidate for surgery in view of advanced stage and received chemotherapy instead.

We believe this case to be one of the few publications from India describing lung cancer in a patient with CPFE. Literature has been published on this combined entity from other countries, but information gathered from such earlier literature, and replicated above, may not be representative of, or be extrapolated to, patients diagnosed

with CPFE and lung cancer in our country. This case serves to ignite an interest in physicians in India, treating such patients, to record and generate information on this relatively rare entity so as to understand, predict, and probably propose appropriate interventions, including chemotherapy and surgery, that may improve clinical outcomes in them.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Irfan Ismail Ayub, Natraj Manimaran

Department of Pulmonary Medicine, Sri Ramachandra Medical College and Research Institute, Chennai, Tamil Nadu, India.
E-mail: iia@rediffmail.com

REFERENCES

1. Jankowich MD, Rounds SI. Combined pulmonary fibrosis and emphysema syndrome: A review. Chest 2012;141:222-31.
2. Li C, Wu W, Chen N, Song H, Lu T, Yang Z, *et al*. Clinical characteristics and outcomes of lung cancer patients with combined pulmonary fibrosis and emphysema: A systematic review and meta-analysis of 13 studies. J Thorac Dis 2017;9:5322-34.
3. Koo HJ, Do KH, Lee JB, Alblushi S, Lee SM. Lung cancer in combined pulmonary fibrosis and emphysema: A systematic review and meta-analysis. PLoS One 2016;11:e0161437.
4. Khuder SA. Effect of cigarette smoking on major histological types of lung cancer: A meta-analysis. Lung Cancer 2001;31:139-48.
5. Zhang M, Yoshizawa A, Kawakami S, Asaka S, Yamamoto H, Yasuo M, *et al*. The histological characteristics and clinical outcomes of lung cancer in patients with combined pulmonary fibrosis and emphysema. Cancer Med 2016;5:2721-30.

Access this article online

Quick Response Code:

Website: www.lungindia.com

DOI: 10.4103/lungindia.lungindia_225_19

How to cite this article: Ayub II, Manimaran N. Combined pulmonary fibrosis and emphysema with lung cancer: Triple calamity. Lung India 2019;36:468-70.