Triple hit effect

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CLINICAL SUMMARY

A 65-year-old male presented with complaints of left-sided chest pain and increasing breathlessness over the last three months. He had cough and breathlessness for the last five years, for which he was taking oral theophylline. He was a tobacco smoker, with a smoking index of >300. There was no other significant medical, personal or family history.

On physical examination, he was sick-looking, with a body mass index of 17.74. Digital clubbing was present. Localized tenderness was present over the midaxillary region, on the left side. Pursed lip breathing was evident, with a respiratory rate of 22/minute. There was no cyanosis, palpable peripheral lymphadenopathy, engorged neck veins, pedal edema or the like. Oxygen saturation was 89% on room air by digital pulse oxymetry. Systemic examination of the respiratory system revealed bilateral distant breath sounds and bibasilar fine-end inspiratory crepitations.

INVESTIGATIONS

The chest radiograph (posteroanterior view) showed decreased lung markings at the upper zones with reticular shadows at the lower zones and a peripheral homogenous opacity having slightly irregular margins at the left mid zone [Figure 1]. Routine investigations of blood, including blood counts and biochemical tests were within normal range. The collagen profile was also normal. Spirometry revealed relatively preserved lung functions with forced expiratory volume in one second (FEV1) 68% of the predicted, forced vital capacity (FVC) 64% of the predicted, and an FEV1/FVC ratio of 82%. The six-minute walk distance test could not be completed by the patient owing to significant desaturation after three minutes. His diffusion lung capacity for carbon monoxide (DLco) was only 32% of the predicted. An electrocardiogram revealed sinus tachycardia and prominent 'p' waves. On echocardiography mild tricuspid regurgitation and moderate pulmonary arterial hypertension was detected.

A high-resolution computed tomographic (HRCT) scan showed centriacinar and paraseptal emphysematous changes with bullae in the bilateral upper lung fields. Subpleural reticular shadows were also seen at the lower zones, with patchy ground-glass opacities. A partially defined peripheral lung mass, 46 × 34 mm in size, with speculated margins having areas of necrosis, was also seen in the posterior segment of the left upper lobe, making an acute angle with the chest wall [Figures 2-4]. The lesion was also found to be eroding the fourth rib. CT-guided fine needle aspiration cytology of the lung mass revealed features of squamous cell carcinoma.

QUESTIONS

Q1. What is your diagnosis?
Q2. Briefly describe this syndrome.

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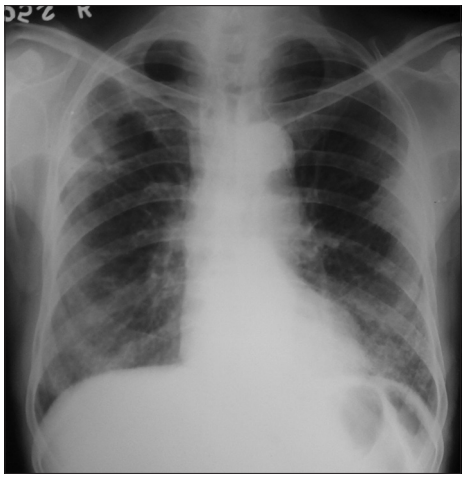

Figure 1: X-ray chest showing hypertranslucency at the upper zones, reticular shadows at the lower zones, and peripheral lung opacity at the left midzone

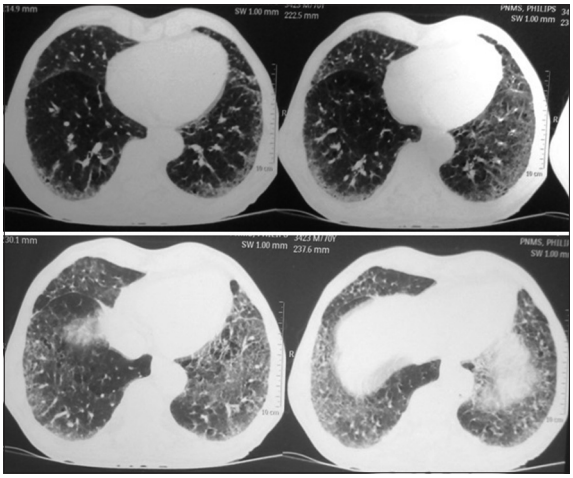

Figure 3: HRCT chest showing sub-pleural and basal reticular shadows with focal honeycombing

ANSWERS

A1. The diagnosis is, locally advanced non-small cell carcinoma lung (NSCLC) in a patient, a smoker, having the combined pulmonary fibrosis and emphysema (CPFE) syndrome.

A2. Combined pulmonary fibrosis and emphysema (CPFE) syndrome:

- *Definition*: Combined pulmonary fibrosis and emphysema (CPFE) is a clinical syndrome characterized by coexistent pulmonary fibrosis and emphysema in the same patient, with features of dyspnea, upper lobe emphysema, lower lobe fibrosis, and abnormalities of gas exchange. This is typically seen in a male who smokes and is frequently complicated by pulmonary hypertension[1]
- *Risk factors*: Environmental insult in the form of smoking is the most important risk factor seen in 98% of these subjects. Male sex is another important factor seen in 90% of the cases. Other associations observed are asbestos and other mineral dust exposure (i.e. coal dust, talc, silica, etc.), hypersensitivity pneumonitis, farmer's lung, connective tissue disorders (rheumatoid arthritis, systemic sclerosis, etc.), positive antinuclear antibodies, surfactant protein C mutation, exposure to rare earth element, recently a mutation in the telomerase component hTR, and so on[2]
- *Diagnosis:* The most important clinical symptom is resting/exertional dyspnea with a history of current/past smoking. Pulmonary function testing shows relatively preserved spirometric values, but unexpectedly low DLco. Hypoxemia on pulse oxymetry and pulmonary arterial hypertension are consistent findings. Chest x-ray and high-resolution computed tomography (HRCT) reveal upper lobe emphysema and lower lobe fibrosis. The incidental nodules or masses suggest associated malignancy. The pathological findings are, predominantly, the usual interstitial

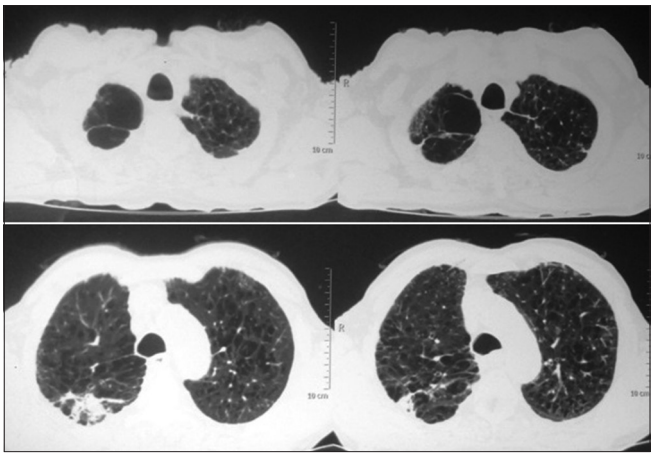

Figure 2: HRCT chest showing emphysematous changes at the upper lung fields

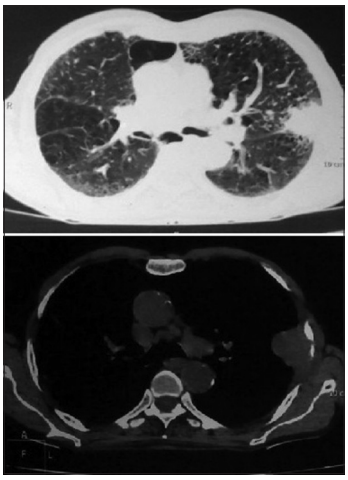

Figure 4: HRCT chest showing peripheral lung mass with rib erosion

pneumonia (UIP) pattern apart from a variety of other patterns

- *Complications*: Pulmonary hypertension is more frequent and severe in these patients compared to those having pulmonary fibrosis alone.[3] Lung cancer is significantly common in these patients owing to a 'triple hit' effect of smoking, emphysema and pulmonary fibrosis. Risk of acute lung injury is also more in such patients after lung resection surgery
- *Treatment*: There is no specific treatment for this condition. The therapeutic options includes smoking cessation, immune suppressive agents, oxygen to correct hypoxemia, lung transplantation, and so forth.[4]

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