A Rare Case of a Diffuse Parenchymal Lung Disease – A Case Report

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ABSTRACT
Diffuse parenchymal diseases (DPLD) have varying presentations. We report a case of rare presentation of DPLD in a 68 year old male. The patient had clinical and radiological pictures similar to sarcoidosis. A systematic approach in clinico-radiological evaluation combined with an interventional approach and efficient histopathology examination lead us to diagnose lymphangitis carcinomatosis and treat primary malignancy as well. Lymphangitic carcinomatosis is morphologically defined by the presence of tumor within pulmonary lymphatics. Definitive therapy for lymphangitic carcinomatosis is directed at treating the primary tumor. We conclude that a high level of suspicion and systematic work up will help to diagnose DPLD precisely.

Keywords: DPLD, Sarcoidosis, lymphangitis Carcinomatosis.

INTRODUCTION
Diffuse parenchymal lung diseases (DPLD) encompass over 140 different diseases of the lung parenchyma that can basically affect all three compartments of the lung (endothelium – interstitium – epithelium) and eventually lead to increased cellularity or to an increased amount of connective tissue in the terminal portion of the lung.

Although the prevalence of DPLD is rather rare (67.5/100,000 for females and 80.9/100,000 for males), many more patients die each year as a result of DPLD as compared to asthma, which is approximately a hundred times more common.

DPLD have many features in common: these include, firstly, the principal clinical symptom of exertional dyspnoea and later, in addition, dyspnoea at rest, the restrictive ventilatory impairment, the diffusion disorder and arterial hypoxaemia on exertion and later at rest, and finally demonstration of the typical changes on high-resolution computed tomography (HRCT). Correct classification of the actual underlying disease requires considerable effort on the part of the patient and doctor alike and represents one of the most challenging differential diagnoses in internal medicine. Here we describe an unusual and rare manifestation of a DPLD.

Case report
An 68 year old Male from a rural area presented with complaints of progressive breathlessness (grade 3 MMRC at presentation), dry cough—which was present for past 6 months and worsened for past 2 weeks. Patient also had fever for past 2 weeks. Patient had cerebrovascular event four year back and is on corresponding medications. Patient is a reformed smoker and no other history was of significance. General physical examination revealed Pallor, grade 3 Clubbing and bilateral pitting Pedal
edema. Fine crepitations and Rhonchi were noted on respiratory system examination. Rest of the systemic examination was normal. Complete haemogram showed anaemia and peripheral blood smear showed dimorphic anaemia. Mantoux test was non-reactive and dilated right atrium and ventricle with severe tricuspid regurgitation with moderate PAH and left ventricular ejection fraction fraction of 60% was observed in echocardiography. Chest radiograph showed an hyperinflated chest with reticulo - nodular shadows in the mid & lower zones (Figure 1). Sputum for AFB was negative & Culture didnot show any growth. High resolution CT chest (Figure 2) showed diffuse nodularity distributed in peribronchovascular and perilymphatic pattern with thickend interlobular septa. Patchy consolidation in lingular segment with minimal pleural thickening, bilateral apical fibrosis and level 3(A) and 4 L medistinal lymphnodes were also noted.

Bronchoscopy showed diffuse nodularity of the mucosa and rest of the segmental openings were patent. Transbronchial lung biopsy (TBLB) was done from right lower lobe and biopsy was taken from endobronchial nodules too. Histopathology showed malignant cell clusters in the lymphatic channels in subepithelium. Lymphatic channels of varying sizes were seen which are plugged with carcinoma cells.(Figure 3).

Immunohistochemistry (IHC) was done to correlate the primary tumour. Ck7, TTF1, Napsin A were Positive (Figure 4&5). This was more in favour of Primary Adenocarcinoma Lung. Hence a final diagnosis of Lymphangitis carcinomatosis with Primary Adenocarcinoma lung was made. The patient was started on Gefitinib 250 mg OD in view of current clinical condition. Patient had reported for follow up, with chest xray ( showing decrease in radiological shadows). (Figure 6)

Discussion

Lymphangitis carcinomatosis was first described by pathologist Gabriel Andral in 1829 in a patient with uterine cancer1. Lymphangitic carcinomatosis is morphologically defined by the presence of tumor within pulmonary lymphatics2. Tumor cells can invade the interstitium resulting in thickened bronchovascular bundles and interstitial septae. The most common primary sites are the breasts, lungs, colon, and stomach3,4,5. Radiological features include thin lines several centimeters in length radiating from the hila, transverse lines about 3 cm in length at the lung bases and extending into the pleura and a fine meshwork throughout the lung giving a reticular pattern. All three types of Kerley lines should be sought for, in the diagnosis of lymphangitis carcinomatosis4.

Pulmonary lymphangitic carcinomatosis can mimic sarcoidosis radiologically. Nodular thickening and ground-glass attenuation are seen in 30%–60% of patients with sarcoidosis. The nodules in sarcoidosis mainly involve central regions of the middle and upper lobes of the lungs. In contrast, changes usually occur in the lower lobes in pulmonary lymphangitic carcinomatosis. Although imaging studies may suggest sarcoidosis, the diagnosis should be confirmed by biopsy.

Definitive therapy for lymphangitic carcinomatosis is directed at treating the primary tumor. This may involve surgical resection, systemic chemotherapy, radiation therapy or any combination. Supportive therapy frequently involves the administration of oxygen, mechanical ventilation, and inotropic support, when needed. The prognosis for patients with Lymphangitis carcinomatosis is extremely poor with less than half surviving past 3 months6.

We report this case, owing to fact that lymphangitis carcinomatosis is rare and diagnosis needs high levels of suspicion and contiguous work up. A systematic clinico – radiologic examination coupled with appropriate tissue biopsy and pathological examination is needed to identify such diseases.

Conclusion:

Lymphangitis carcinomatosis is a rare presentation of DPLD. A proper clinico – radiological examination , and meticulous tissue biopsy and histopathological examination will aid in the diagnosis.
References:

- Janower ML and Blennerhassett JB. Lymphangitic spread of metastatic cancer to the lung. Radiol 1971; 101: 267
Figure 1 - Chest Xray (on presentation)

Figure 2 – HRCT Thorax

Figure 3 - Malignant cells in lymphatic spaces (Arrow)

(400x, H&E stain)

Figure 4 - IHC – TTF-1
Figure 5 - IHC – Napsin A

Figure 6 – Follow up Chest xray