

Case Report:

Pulmonary Lymphangitis Carcinomatosis In Primary Oesophagogastric Carcinoma

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Abstract:

Pulmonary Lymphangitis carcinomatosis is an unusual metastatic manifestation of Oesophagogastric carcinoma and it occurs due to diffuse spread of the tumour to the pulmonary lymphatic system. We described a case of a 28 year old woman, presenting with gradually progressive dyspnoea and cough where results of Chest X-ray and HRCT thorax were consistent with features of Lymphangitis carcinomatosis. Upper GI endoscopic evaluation showed a tumour originating from oesophagogastric junction and extending to cardia of stomach. Biopsy from tumour revealed adenocarcinoma. As there is no definitive therapy to this condition, patient was managed conservatively only to succumb few days after hospitalisation.

Key Words: Pulmonary Lymphangitis carcinomatosis; Oesophagogastric neoplasm; Adenocarcinoma

Case Presentation:

In April 2012, a 28 year old housewife with no comorbid illness admitted to our Pulmonary Medicine ward with gradually progressive breathlessness over 5 months. There was dyspnoea even at rest prior to admission. Dyspnoea was associated with cough and scanty mucoid expectoration. There was no history of fever, haemoptysis or chest pain. Patient recalled an incident of exposure to dust in a paddy field before onset of the symptoms. In review of systems, she mentioned difficulty in swallowing solid food since 10 days and loss of appetite since 2 months. She also complained of occasional mild abdominal pain over last 6 months though she denied any history of vomiting, jaundice or altered bowel habit. She was a non-smoker, and denied any recreational drug use. There had been no history of recent travelling or infectious contact. She was having two children with history of last child birth 2 years back. No recent abnormality in menstrual cycle was noted. There was no similar history at past or in the family. On examination, she was of thin built, afebrile and found to have tachycardia, tachypnoea and hypoxia. Blood pressure was 130/80 mm of Hg. There was no lymphadenopathy or clubbing. Examination of breast was normal. There was bilateral wheezes and

inspiratory fine crepitations on auscultation.

Initial Chest X-ray showed coarse reticulonodular opacities in both lungs (fig.I) and a diagnosis of presumed hypersensitivity pneumonitis was made.



Fig. I: Chest X-ray : Bilateral coarse reticulonodular infiltrates in lungs

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Patient was put on supplemental oxygen with bronchodilator and systemic corticosteroid. Her routine blood tests were unremarkable except for a Hb of 10.4 gm/dl. Sputum was found to be negative for AFB or any other organism. Routine urine test was within normal range and patient was HIV seronegative. Spirometry could not be performed appropriately as patient was dyspnoeic and her effort was poor.

Subsequently HRCT of thorax was performed. HRCT scan demonstrated coarse nodular interlobular septal thickening and peribronchovascular thickening with ground glass opacity and mild right pleural effusion which is consistent with the features of Lymphangitis carcinomatosa (fig. II).

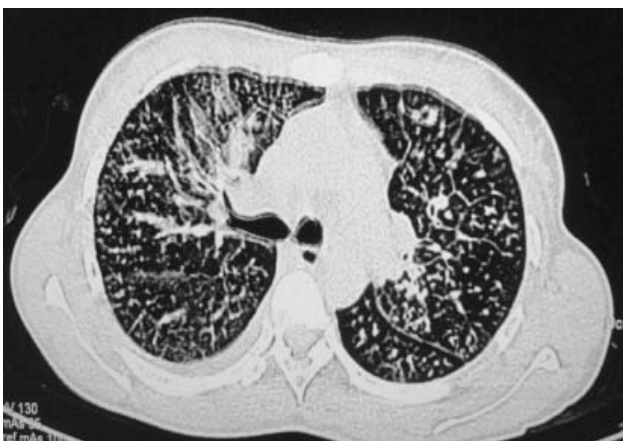


Fig. II: HRCT scan showing coarse nodular thickened interlobular septa and peribronchovascular interstitium with ground glass opacities and right pleural effusion

In search for a primary tumour, an Ultrasonography of abdomen and pelvis was done which revealed presence of bilateral solid ovarian masses along with mild ascites and mild right pleural effusion (fig. III).

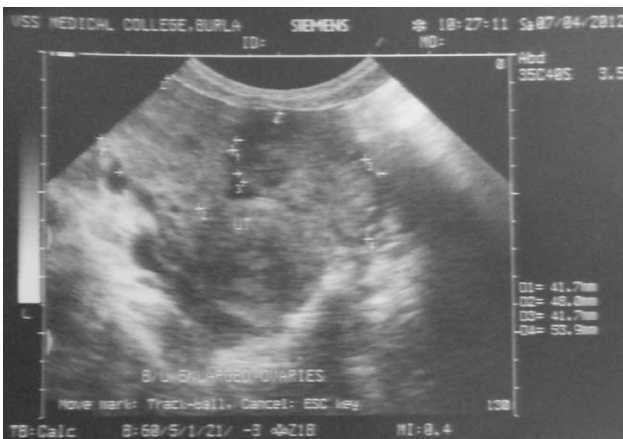


Fig.III:Ultrasound of abdomen & pelvis showing bilateral enlarged ovaries

Based on ultrasound findings, possibility of a Krukenberg tumour was suspected and an Upper GI endoscopy was done which showed presence of an Oesophago-gastric neoplasm - starting at Oesophago-gastric junction and extending distally to involve 3/4th of cardia and adjoining fundus and corpus of the stomach (fig. IV).



Fig IV a

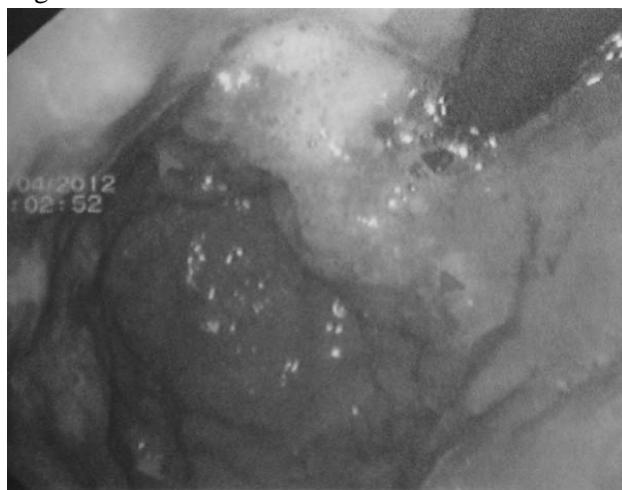


Fig IV b

Fig.IV: Upper GI Endoscopy showing

- a) Nodular ulcerated lesion at juxta EGJ
- b) Lesion extending to involve cardia, fundus and corpus of stomach

Biopsy was taken from the above mentioned nodular ulcerated lesion which revealed invasive adenocarcinoma on histological examination (fig. V).

Poor prognosis and incurable nature of the disease was explained to patient party and they refused to go for lung biopsy due to financial constraint and poor general condition of the patient. She did not respond to supportive therapy and succumbed within 7 days of hospitalization.

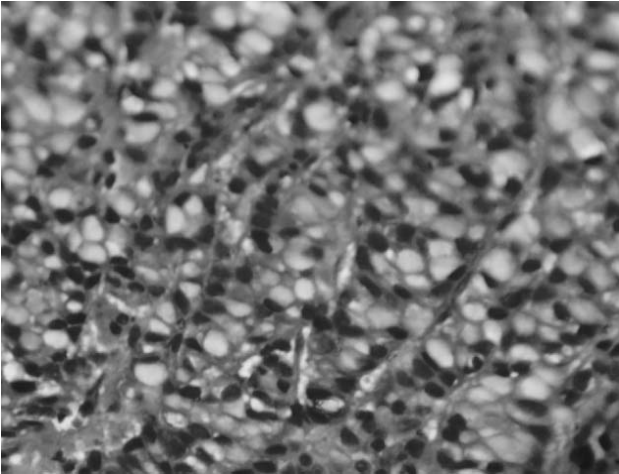


Fig V: Histological section of biopsy taken from oesophagogastric mass showing features of adenocarcinoma

Discussion:

Pulmonary lymphangitis carcinomatosa (PLC) is a metastatic lung disease characterised by the diffuse infiltration and obstruction of the pulmonary parenchymal lymphatic system by tumour cells¹. Infiltration of the pleural, peribronchial, and perivascular lymphatics by neoplastic cells is a condition first noted by Andral in 1829. PLC occurs in 6-8% of metastatic lung cancers². Highest incidence was found in the age range of 40-49¹ and it is uncommon in younger patients as noted in our case. The primary tumours most commonly associated with PLC are: breast (33%), stomach (29%), lungs (17%), pancreas (4%) and prostate (3%)^{1,3}, though Yang et al reported that stomach cancer is by far the most frequent primary lesion followed by the bronchus². Other described sites with PLC are cancers from colon, pancreas, kidney, cervix, thyroid, larynx and hypopharynx³. Most of the case studies revealed adenocarcinoma as the commonest type of malignancy causing PLC which corroborates with the findings in our patient.

Pathophysiology of LC occurs as a result of the initial haematogenous spread of tumour to the lungs, with subsequent malignant invasion through the vessel wall into the pulmonary interstitium and lymphatics. Tumour then proliferates and easily spreads through these low-resistance channels⁴. In case of gastric carcinoma, the tumour probably spreads along the paraesophageal lymphatics, hence through the hilar anastomosis into lung parenchyma as well as through haematogenous tumour emboli⁵.

Patients with pulmonary lymphangitis carcinomatosa often present with breathlessness and a non-productive cough with crepitations and without features of consolidation. The onset of pulmonary symptoms may precede diagnosis of the primary tumour; however, the frequency of this presentation is unknown. Dyspnea secondary to pulmonary involvement may be the only symptom of an occult nonpulmonary malignancy⁶ and such cases may put up a challenge in diagnosis as happened in above mentioned case where initially a diagnosis of hypersensitivity pneumonitis was suspected.

Typical radiographic finding in lymphangitis carcinomatosa is an interstitial infiltrate that is usually coarse and patchy but may be diffuse and symmetric². Bilateral coarse bronchovascular markings with irregular outline are seen predominantly in the lower lobes of the lungs; however, changes may be unilateral or asymmetric. Other characteristics of PLC on the radiograph are septal Kerley's lines, hilar lymphadenopathy and pleural effusion^{2,7}. But Chest radiographs appear normal in 30-50% of patients with histologically proven disease⁵.

Computed tomography scan of thorax reveals :

- Smooth (early stage) and nodular (late stage) thickening of interlobular septa and peribronchovascular interstitium with polygonal arcades with thickened limbs from thickened septa of adjacent lobules
- pleural effusion (30-50%) and lymphadenopathy (30%)
- Normal lung architecture is maintained
- Ground-glass appearance from interstitial oedema or extension of the tumour into the parenchyma

Recently PET/CT has gained a lot of importance due to its high specificity in detection of pulmonary lymphangitis carcinomatosa.

The differential diagnosis is interstitial lung disease, primary malignancy in the lung, pulmonary sarcoidosis and hypersensitivity pneumonitis. Other differential diagnostic considerations include pulmonary oedema, opportunistic infection, radiation fibrosis, drug-induced lung disease etc. In most instances, however, the differential diagnosis is not difficult if occupational histories, sign-symptoms and other clinical information are available⁸.

Pulmonary function tests in patients with PLC usually show a restrictive ventilatory defect, a reduction

in diffusing capacity and reduced compliance ⁷. Transbronchial biopsy is the procedure of choice for a definitive diagnosis ⁹.

Optimal treatment of lymphangitis carcinomatosis is neither well defined nor encouraging, but a trial of chemotherapy and/or intravenous steroids may be warranted. Desigan et al reported a case of a young adult with PLC from occult gastric cancer, who was treated with 5-fluorouracil, doxorubicin and mitomycin, with remarkable improvement ¹⁰.

As described in above case, lymphangitis carcinomatosis traditionally has carried a poor prognosis. Of the 60 patients reported by Yang and Lin ², half

died within 3 months and only one seventh survived beyond 6 months. In a study conducted by Dennstedt et al, six patients with PLC from occult stomach carcinoma were described and the average duration of survival after the first hospital admission was 22 days ⁶. Liam et al reported three cases of PLC with gastric carcinoma as primary site, where all three patients died within 20 days of hospital admission ⁷. In conclusion, it can be said that though Lymphangitis carcinomatosis is an unusual primary presentation of an occult carcinoma in young patients, its diagnosis should be considered when a patient presents with clinical and radiological features of Interstitial lung disease.

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