

Radiology Corner: Case 1

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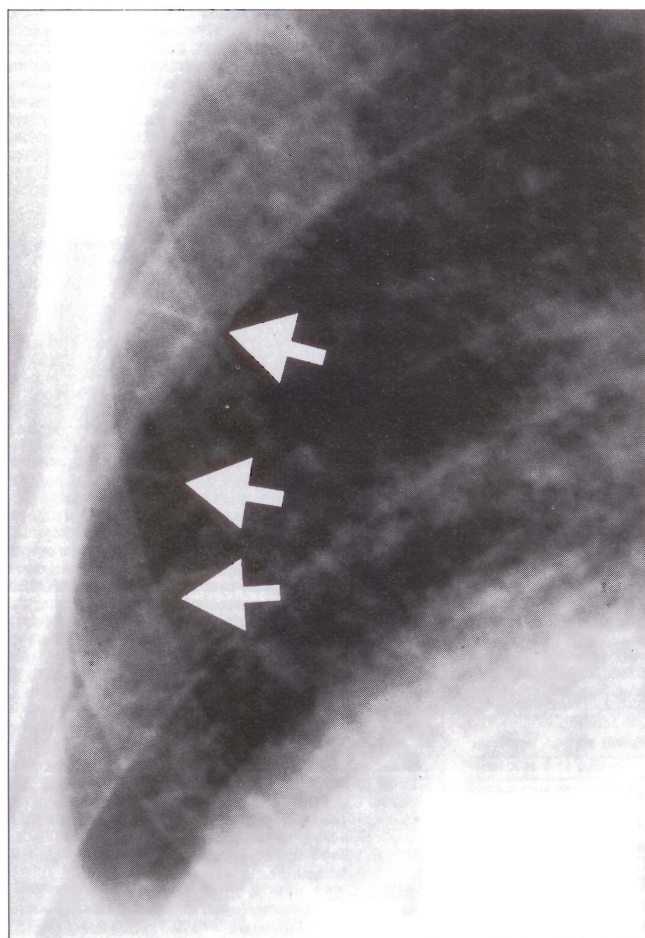
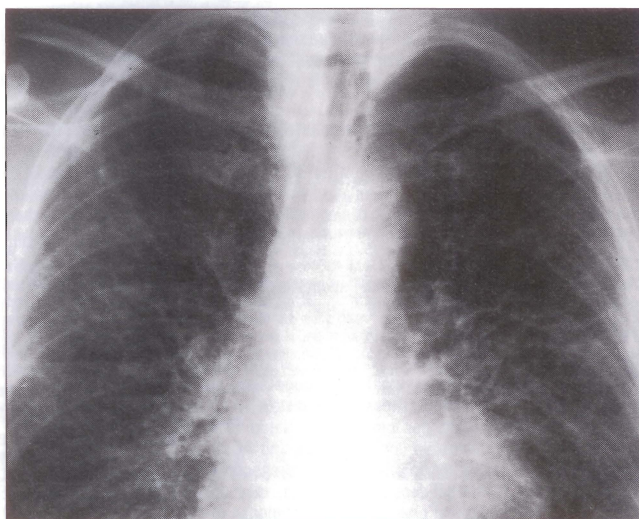


Figure 1) Posteroanterior and lateral chest radiographs. **Top right** Note diffuse predominantly reticular interstitial pattern; **Left** close-up of right costophrenic angle shows septal lines (arrows) are a prominent feature; **Right** blunting of posterior costophrenic angles suggests pleural fluid

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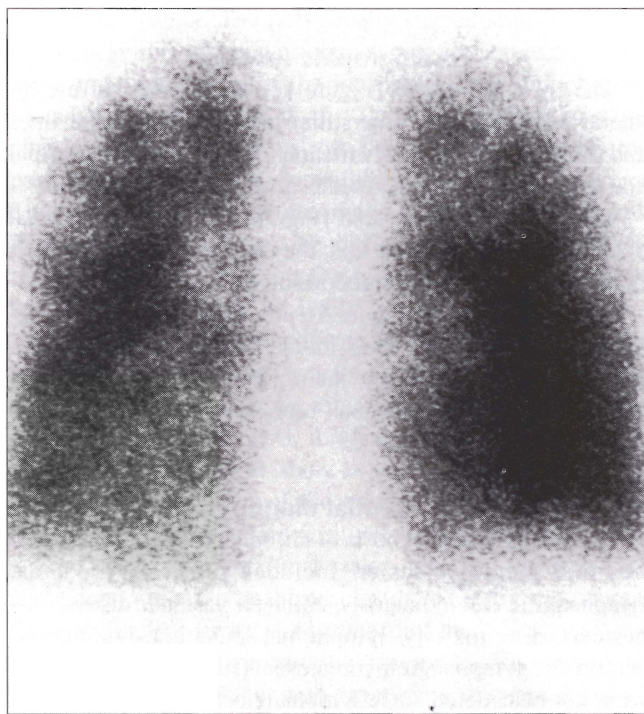
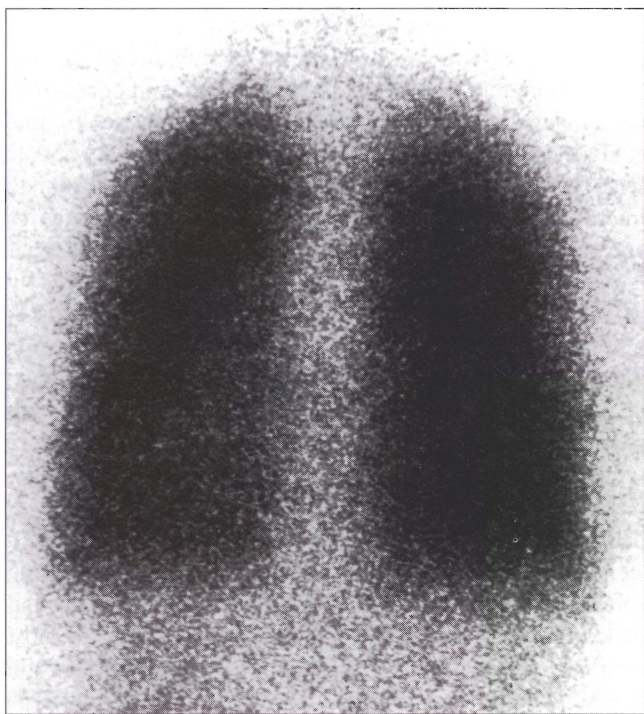


Figure 2) Ventilation/perfusion scan. (Left) The ventilation scan (^{133}Xe) shows normal ventilation while (right) perfusion scan ($^{99\text{m}}\text{Tc-MAA}$) demonstrates multiple subsegmental filling defects

HISTORY

A 56-year-old man was admitted with an eight-week history of worsening dyspnea and dry cough. In the last two weeks he experienced bilateral pleuritic chest pain. He had had surgery for gastric carcinoma two years ago. Otherwise, his past medical history was negative. He was on no recent medications.

Physical examination revealed a cachectic-appearing man with a heart rate of 110 beats/min and a respiratory rate of 30 breaths/min. Bibasilar fine crackles were heard. The rest of the examination was normal.

Laboratory tests revealed a normochromic, normocytic anemia.

A chest x-ray (Figure 1) and ventilation-perfusion lung scan (Figure 2) were performed. Based on these, the patient underwent high resolution computed tomography (HRCT) of the lungs (Figure 3). A chest radiograph eight months before admission was normal.

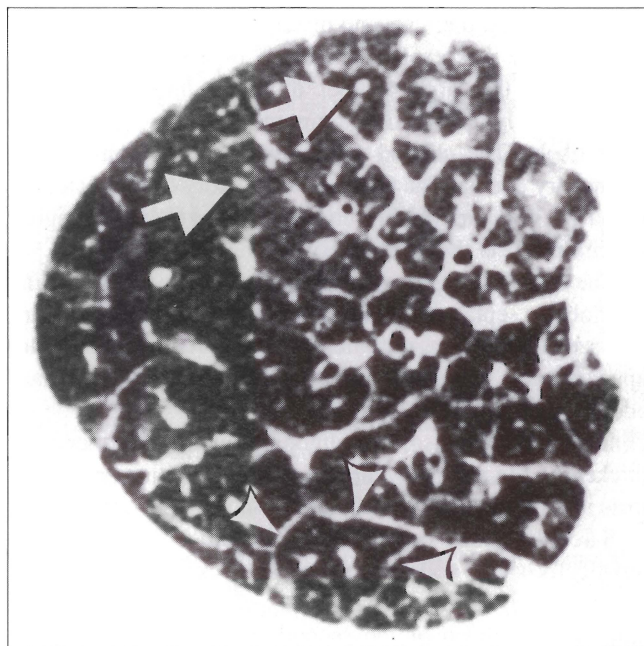


Figure 3) High resolution computed tomography scan of right lung; 1.5 mm cuts show prominent secondary lobule outlined by thickened interlobular septa (arrowheads) with thickened bronchovascular bundles (arrows). (L=-700 HU; W=1500 HU)

For radiographic findings and diagnosis, see over page.

ANSWERS

Radiographic findings

The chest radiograph (Figure 1) demonstrates diffuse bilateral reticulonodular interstitial lung disease, septal lines and small bilateral pleural effusions. The heart size is normal and there are no enlarged mediastinal or hilar lymph nodes.

The ventilation lung scan (Figure 2, left) shows normal distribution of ventilation and, the perfusion scan (Figure 2, right) shows multiple peripheral subsegmental defects.

In the HRCT scan (Figure 3), polygonal structures representing the secondary lung lobules are seen peripherally, indicating thickening of the interlobular septa. A central dot within some of the polygons represents thickening of the bronchiolar-vascular bundle.

Differential diagnosis

The differential diagnosis of chronic reticular lung opacities and pleural effusion includes pulmonary edema, lymphangitic carcinomatosis, collagen vascular disease, asbestosis, drug toxicity, lymphoma, leukemia, lymphangiectasia and lymphangiomyomatosis. The last two diagnoses were not considered since lymphangiectasia occurs in children and young adults, and lymphangiomyomatosis occurs in women of childbearing age. The diagnosis of lymphoma or leukemia is usually known by the time the interstitial lung pattern develops, and enlarged lymph nodes are a common feature of these diseases. Asbestosis was excluded because there were no calcifications or pleural plaques on the chest radiograph, and there was no history of exposure. Similarly, drug toxicity was excluded because of a negative history. Because of the large number of septal lines, pulmonary edema and lymphangitic carcinomatosis were considered most likely. The clinical picture was not in keeping with pulmonary edema. The past medical history of gastric carcinoma and nodularity on the chest x-ray makes **lymphangitic carcinomatosis the most likely diagnosis.**

The HRCT scan shows the characteristic thickening of the interlobular septa and the interlobular bronchovascular bundles seen in lymphangitic carcinomatosis. Although a similar appearance is sometimes seen in patients with pulmonary edema, differentiation of these conditions may be made on clinical grounds. Also, in pulmonary edema the interstitial thickening is smooth, whereas in lymphangitic carcinomatosis it may be nodular or beaded.

Sarcoidosis and coal-worker's pneumoconiosis characteristically show nodular interstitial thickening on HRCT; however, they are not usually associated with pleural effusion and distortion of the secondary pulmonary lobule is commonly seen.

Because of this patient's history of pleuritic chest pain a ventilation-perfusion (V-Q) lung scan was requested to rule out pulmonary embolism. It demonstrates V-Q mismatch with irregular peripheral perfusion defects outlining the bronchopulmonary segments. Several case reports have demonstrated this pattern of 'contour mapping' in patients with lymphangitic carcinomatosis (1-3).

DISCUSSION

Originally defined as tumour growth in the lymphatic system of the lungs, lymphangitic carcinomatosis is now more accurately described as interstitial carcinoma, since hematogenous spread of tumour is more common (4-6). In a study by Janower and Blennerhassett (7), 20 of 23 patients with lymphangitic carcinomatosis diagnosed at autopsy had microscopic tumour emboli in the small pulmonary arteries and arterioles; only 11 had metastatic tumour in the hilar lymph nodes. Thus, it was hypothesized that tumour usually reaches the lungs hematogenously and that the cells then multiply in the peripheral vessels. Tumour may then invade the adjacent lymphatics moving both peripherally and also centrally toward the hila. If tumour invades the adjacent lung parenchyma, pulmonary nodules are formed (7). The characteristic peripheral and subsegmental perfusion lung scan defects seen in lymphangitic carcinomatosis are consistent with pathology showing tumour emboli in the small vessels (2,3).

The most common primary tumours causing lymphangitic spread are adenocarcinomas of the stomach, breast, lung and pancreas. Often the primary site is unknown (8,9). The most common presentation is rapidly progressive dyspnea, and occasionally dry cough and hemoptysis. The prognosis is dismal and in one study of 62 patients, half died within three months (10).

To interpret the radiological findings of lymphangitic carcinomatosis, it is essential to understand the anatomy of the secondary lung lobule. The secondary lung lobule is the smallest discrete portion of lung surrounded by connective tissue septa (11). It is supplied by three to five terminal bronchioles and has the shape of an irregular polyhedron (12). Three components have been described: first, the interlobular septa marginate the lobule and contain pulmonary veins, lymphatics and connective tissue; second, the lobular core, which appears as a dot-like or linear branching opacity on HRCT, contains the pulmonary artery and bronchiolar branches and connective tissue; and third, the lobular parenchyma consists of alveoli, small airways, the pulmonary capillary bed and supporting connective tissue (9).

The interlobular septa are thickest peripherally or in a subpleural location where they normally measure approximately 0.1 mm. This is at the lower limit of HRCT resolution, and within the central lung they are thinner and more difficult to identify (9).

On a normal chest radiograph the secondary lung lobules are not visible. However, when fluid (eg, pulmonary edema), cells (eg, lymphangitic carcinomatosis), or fibrous tissue thicken the interlobular connective septa, septal lines become visible and the volume between two of these lines represents a secondary pulmonary lobule (13). Septal lines are often referred to as Kerley's B or Kerley's A lines (14). Kerley's B lines (Figure 4) are short, horizontally oriented lines extending to the pleural surface (15). Kerley's A lines, on the other hand, are linear opacities 2 to 6 cm long lying deep within the connective tissue of the lung parenchyma. Kerley's A lines usually extend to the hilum and never reach the visceral pleural surface.

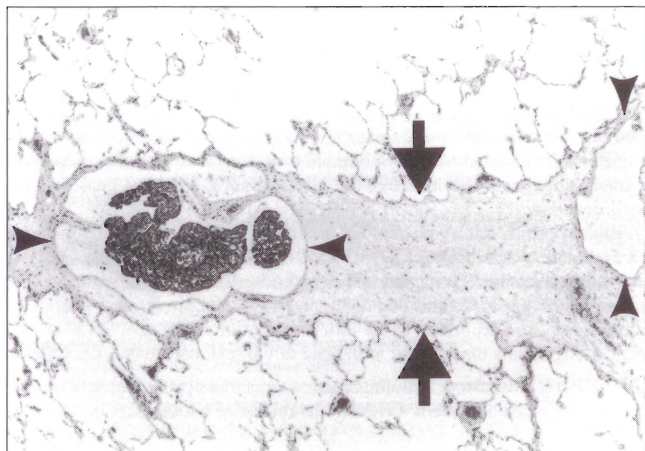


Figure 4) Open lung biopsy specimen showing lymphangitic carcinomatosis. The interlobular fibrous septum (arrows) is widened due to edema. It includes two dilated lymphatic channels (arrowheads). The one on the left includes large clumps of metastatic carcinoma cells. (Hematoxylin and eosin, x80)

Compared with plain chest radiographs and even conventional computed tomography, HRCT improves the sensitivity and specificity of diagnosis in lymphangitic carcinomatosis because of its higher spatial resolution. This is due primarily to the use of thin collimation or axial slices (1.0 to 1.5 mm) and to reconstruction of images of the lung parenchyma using a high spatial frequency algorithm (9). This increase in spatial resolution makes structures appear sharper.

The main HRCT findings in lymphangitic carcinomatosis

are well-defined polygonal bodies with a central dot or branching opacity. The polygonally shaped structure represents thickening of the interlobular septa while the central dot represents thickening of the intralobular peribronchiolar-vascular interstitium (9,12,16,17). Other findings may include similar thickening of the fissures and of the peribronchiolar-vascular interstitium in the perihilar areas. Thickening of these structures may be smooth and/or beaded. Disease distribution may be diffuse, patchy or unilateral. Preservation of normal lung architecture is a key feature of lymphangitic carcinomatosis (9).

An open biopsy obtained from the left lung in this patient shows lymphangitic carcinomatosis with thickened and edematous interlobular fibrous septa (Figure 4). The lymphatic channels in the subpleural, interlobular and perivascular fibrous tissue are dilated. Many contain clumps of metastatic carcinoma cells. Some of the tumour cells have a 'signet-ring' appearance and stain positively for mucin. The appearances are those of metastatic poorly differentiated adenocarcinoma. Given the past history of gastric carcinoma, the most likely primary site is the stomach.

In conclusion, lymphangitic carcinomatosis should be considered in any patient with progressive dyspnea, a known malignant tumour, and reticulonodular interstitial lung disease. Characteristic V-Q lung scan and HRCT results strengthen this diagnosis. Indeed, HRCT findings may obviate the need for pathologic diagnosis in patients with a known primary tumour and, in less typical cases, can direct the site of biopsy.

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