Unilateral Chest Wall Edema with Associated Pleural Effusion: Unusual Sign of Primary Retroperitoneal Lymphoma

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Isolated retroperitoneal malignant lymphoma may be asymptomatic until late in its clinical course, or, alternatively, may be manifest by early systemic symptoms such as fever, pruritus or weight loss. In some instances, these rapidly enlarging tumors present with local signs, attributable to compression of nearby structures by growing lymph node masses (1).

We wish to call attention to an unusual clinical presentation of retroperitoneal lymphoma — namely unilateral chest wall edema and ipsilateral pleural effusion.

Case Report:

A 67 year old woman was admitted to the hospital because of fatigue and exertional dyspnea. She was well until two months earlier, when she developed progressive weakness, loss of appetite and shortness of breath. She had lost 8 kg during the previous few months. On examination the patient appeared pale and thin. Except for a slight tachypnea the vital signs were normal. A slight infiltration of the skin and subcutaneous tissue was visible in the lower third of the left hemithorax, particularly between the posterior axillary and the scapular lines. The skin fold, measured by a dermal caliper, was 1.8 cm in this area, and 0.6 cm at the corresponding contralateral site. Dullness and impaired transmission of fremitus were detectable at the base of the left hemithorax. Pleural puncture vielded 800 cc of serous fluid. The liver, spleen and external lymph nodes were not enlarged. Ascites was absent. Examination of the breasts was unremarkable.

The hematocrit was 30%, the white blood cell count was 5300 per mm³, with 47% neutrophils,

48% lymphocytes and 5% monocytes. The erythrocyte sedimentation rate was 75 mm per one hour. The plasma calcium, alkaline phosphatase, albumin and globulin were unremarkable. X-ray films of the chest revealed an opacity at the base of the left hemithorax consistent with pleural effusion (Fig. 1). The pleural fluid contained 4.1 g/dl protein, 2400 erythrocytes and 530 lymphocytes per mm3. No malignant cells were found in repeated cytological examinations of the fluid and in a pleural biopsy specimen. Bacteriological cultures were negative. Gastroscopy, sigmoidoscopy, barium enema, intravenous urography, isotopic scintiscans of the liver, spleen and bones were normal. Liver needle biopsy and bone biopsy showed no microscopic abnormality. Computerized tomography (CT) of the abdomen revealed a large multilocular retroperitoneal mass (Fig. 2). Cytological study of a fine needle aspirate from the mass was consistent with malignant lymphoma. Frequent pleural punctures were necessary to relieve dyspnea from rapidly recurring effusion. It was improved only after chemotherapy with concomitant regression of the retroperitoneal mass. Three months later unilateral chest wall edema was gone.

Discussion

Unilateral chest wall edema (UCE) is a variant of local edema formation. It is observed in a multitude of conditions such as boils, cellulitis, erysipelas, venous obstruction, trauma, arteriovenous fistula, and lymphatic obstruction caused by malignant growth (2,3). In the patient described herein, the association of UCE with ipsilateral pleural effusion suggest a common etiology for both phenomena. Known

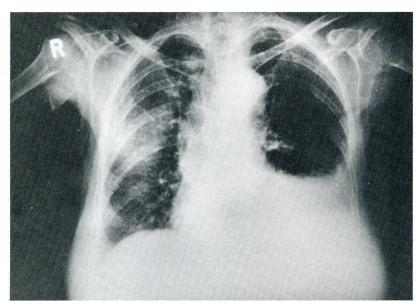
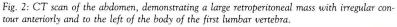
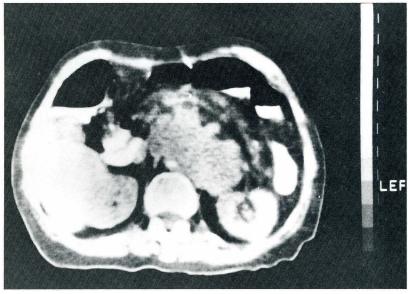


Fig. 1: Radiograph of the chest, showing left pleural effusion.





causative factors include purulent pleurisy (4), iatrogenic extravasation of pleural fluid following paracentesis with inflammatory reaction in the subcutaneous tissue (5), or neoplastic seeding of the pleura with secondary impairment of lymph flow. These factors were excluded by microscopic and bacteriologic study of the fluid, absence of paracentesis prior to appearance of UCE, and the lack of tumor cells in the pleura. However, after detection of retroperitoneal lymphoma and appropriate chemotherapy, the retroperitoneal mass regressed and both UCE and pleural effusion disappeared. This evolution suggests that these latter manifestations were directly related to the retroperitoneal mass and/or indirectly to regional lymphatic infiltration by tumor.

Lymphography and phlebography are useful techniques to identify the mechanism of peripheral edema in patients with retroperitoneal lymphoma. Obstruction of the inferior vena cava by mass compression (6) or neoplastic plugging of lymph vessels in the leg (7) are recognized causes of peripheral edema, but these diagnostic maneuvers are not applicable to the truncal area.

A reasonable explanation for the pathogenetic relationship between retroperitoneal lymphoma and UCE with pleural effusion derives from the anatomy of the lymphatic vessels of the pleura and thoracic wall. Lymphatics from the visceral and parietal pleura, and chest wall drain in three main directions: to retrosternal lymph nodes, para-aortic intrathoracic nodes, and transdiaphragmatically to para-aortic intraabdominal lymph nodes at the level of the kidneys (8). Plugging of lymph trunks subdiaphragmatically, or lymphatic compression by tumor mass potentially interferes with downward flow of lymph from the chest wall and pleura. The end result favors accumulation of lymph in the pleural cavity and subcutaneous tissue overlying the chest wall.

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