RADIOLOGY OF THE MONTH

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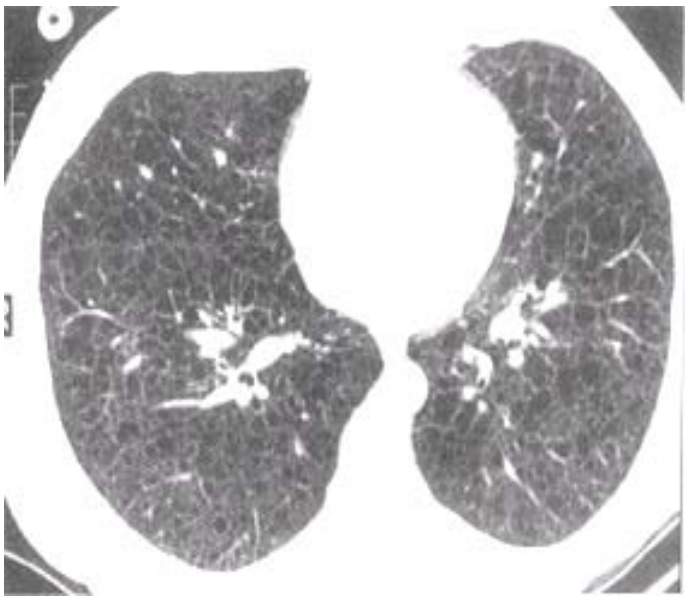
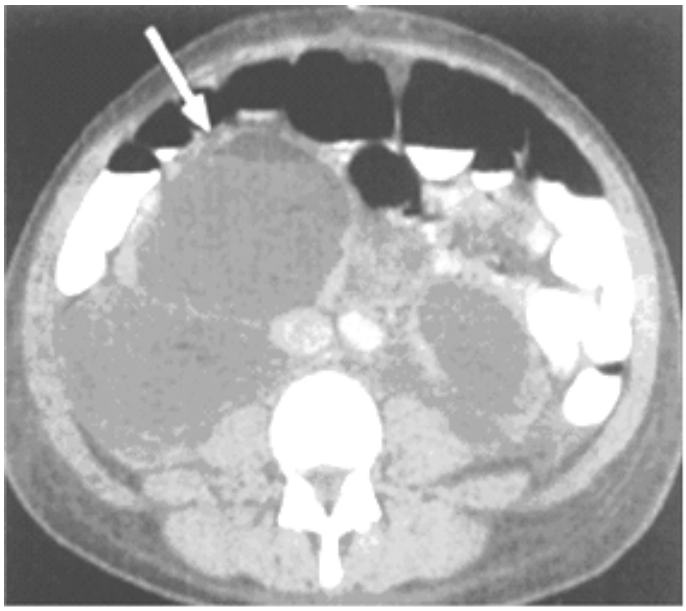
HISTORY

A 49-year-old premenopausal woman presented with acute onset of lower abdominal pain. Physical examination revealed that her abdomen was distended and non tender. Her white blood cell count and serum markers for ovarian cancer were normal, she had no history of malignancy or any chest symptoms; in particular, chest pain, cough, or dyspnea.

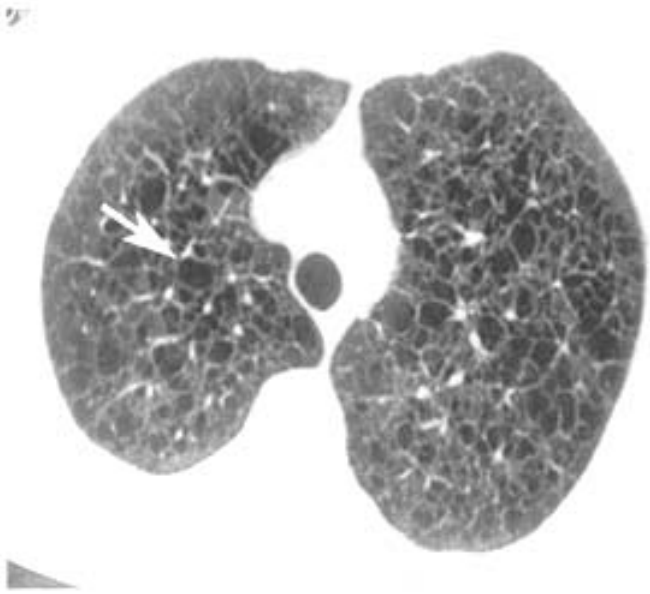
A computed tomographic (CT) examination of the abdomen and pelvis was performed. Helical CT was performed 150 mL of intravenous contrast material (iohexol, Omnipaque: Amersham Healthcare. Cork. Ireland) and 750 mL of oral contrast material (diatrizoate sodium, Hypaque; Amersham Health, Princeton, NJ). CT sections were 5 mm thick and were acquired from the top of the diaphragm through the ischial tuberosities using a LightSpeed 16 CT scanner (GE Medical Systems, Milwaukee, Wis).



Transverse CT image through the lung bases with lung window setting shows thin- walled lung parenchymal cysts (arrow).



Transverse CT image through the lower abdomen shows cystic retroperitoneal masses with fat- fluid level (arrow).



Transverse high resolution CT image demonstrates thin- walled lung parenchymal cysts (arrow indicates one of many) with intervening normal parenchyma.

IMAGING FINDINGS

Contrast - enhanced CT revealed large encapsulated low-attenuation retro peritoneal cystic masses in the abdomen and pelvis. Fat-fluid levels were present in some cysts. The kidneys, liver, spleen, and adrenal glands were normal. Images through the lower thorax revealed bilateral small thin-walled cystic lung parenchymal lesions.

High-spatial- resolution CT scanning enabled us to confirm the presence of diffuse, thin- walled cystic lung parenchymal lesions with intervening normal parenchyma. Ultrasonography-guided 18-gauge core biopsy and fine-needle aspiration biopsy (cloudy red fluid was aspirated) of the complex cystic right retroperitoneal mass was performed. Pathologic analysis confirm the diagnosis of lymphangiomyoma.

DIAGNOSIS

Lymphangioleiomyomatosis

DISCUSSION

The presence of low-attenuation retroperitoneal cystic masses (lymphangioleiomyomas) with fat-fluid levels, together with thin-walled lung parenchymal cysts, and small bilateral pleural effusions in a premenopausal woman—the most likely diagnosis is lymphangioleiomyomatosis (LAM). High-spatial- resolution CT scanning showed the presence of diffuse, thin- walled cystic lung parenchymal lesions with intervening normal parenchyma. Pathologic analysis confirmed the diagnosis of lymphangiomyoma.

LAM is a rare idiopathic disorder found almost exclusively in premenopausal women, and is characterized by the proliferation of abnormal smooth muscle cells (LAM cells) in the pulmonary interstitium and along the thoracic and abdominal lymphatics.(1)

Patients with LAM most commonly have dyspnea, pneumothorax, and cough at presentation. Chest pain, chylous pleural effusions, hemoptysis, and wheezing are less common at presentation but may develop during the course of the disease.(2) Extrapulmonary features include renal angiomyolipomas, lymph node masses, cystic soft-tissue masses (lymphangiomyomas), chybous ascites, and uterine fibroids leiomyomas.(3) In a large series of 80 patients with pulmonary LAM, 76% had positive abdominal findings at imaging: These findings included renal angiomyolipoma (54%), enlarged abdominal lymph nodes (39%), and lymphangiomyomas (16%). Less commonly, ascites (10%) and hepatic angiomyolipoma (4%) were present. Initial manifestation of LAM with abdominal pain is a common finding. Other manifestations of extrapulmonary LAM include palpable abdominal masses and ascites. If the diagnosis of extrapulmonary LAM precedes the diagnosis of pulmonary LAM, the patient usually develops thoracic symptoms within 1-2 years.(4)

Lymphangioleiomyomas result from the proliferation of smooth muscle cells in the lymphatic vessels, causing obstruction and dilation of the lymphatic vessels and resulting in cystic collections of chylous material.(5) At CT, dilated retroperitoneal lymph vessels may have thick or thin walls and may contain low-attenuation (3-25 HU) material.(6) Lymphangioleiomyomas may lie between and displace vascular structures in the retroperitoneum. Overdistention of lymph cysts may result in rupture and chylous ascites.

Radiologic diagnosis is difficult when LAM involves only the retroperitoneum or when it involves the retroperitoneum before it involves the lungs. When enlarged lymph nodes are seen in conjunction with lymphangioIeiomyomas, a neoplastic process— such as lymphoma, metastasis, or primary cystic retroperitoneal tumor— may be misdiagnosed. Old hematomas, abscesses, urinomas, and lymphoceles must also be considered in the differential diagnosis of an abdominal lymphangioleiomyoma. A fat-fluid level in n retroperitoneal cystic mass is a recognized feature of a mature teratoma.(7)

Pulmonary cysts have been present in nearly all cases of LAM reported to date. The mechanism of cyst formation in patients with LAM is controversial; both small airway obstruction and proteolytic destruction with amalgamation of alveoli have been proposed as possible mechanism.(8) High-spatial-resolution CT is both sensitive and specific in the diagnosis of pulmonary LAM, depicting characteristic round thin-walled cysts uniformly distributed

throughout the lungs.[9,10] The profusion of cysts and the quantitative CT scores correlate with impairment at pulmonary function testing and clinical condition. Langerhans cell histiocytosis can result in similar thin-walled cysts; however, the preferential distribution in the upper lungs and the presence of associated nodules in patients with Langerhans cell should help differentiate the entities.[11] While the clinical and radiologic features of LAM are characteristic, a definitive diagnosis is usually established with tissue biopsy, as the only medical therapy of benefit (hormonal therapy with medroxyprogesterone) will induce early menopause in these women, who are typically of child-bearing age. In most cases, a biopsy specimen is surgically removed from the lung. Reports suggest that transbronchial lung biopsy specimens and pleural fluid cytologic analysis may be sufficient for diagnosis in certain cases. Since LAM can involve extrapulmonnry sites, such as lymph nodes and abdominal or pelvic masses, a definitive diagnosis can occasionally be made with biopsy specimens obtained from such sites.[12]

All women with LAM should undergo a careful family history review and clinical examination for stigmata of tuberous sclerosis complex and should be seen by a clinical geneticist in cases of doubt.[2] Among the manifestations of LAM, pulmonary involvement is responsible for most of the morbidity and mortality. The disease is slowly progressive, leading to respiratory failure and death. Diverse treatments—including oophorectomy, tamoxifen, luteinizing hormone releasing hormone analogue, and progesterone—as well as combinations of these treatments have been tried. Among these, early administration of medroxyprogesterone is currently the most accepted medical treatment . When the patient's functional status declines, lung transplantation is the best therapeutic option.

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