Diagnosis of the case presented in the previous edition

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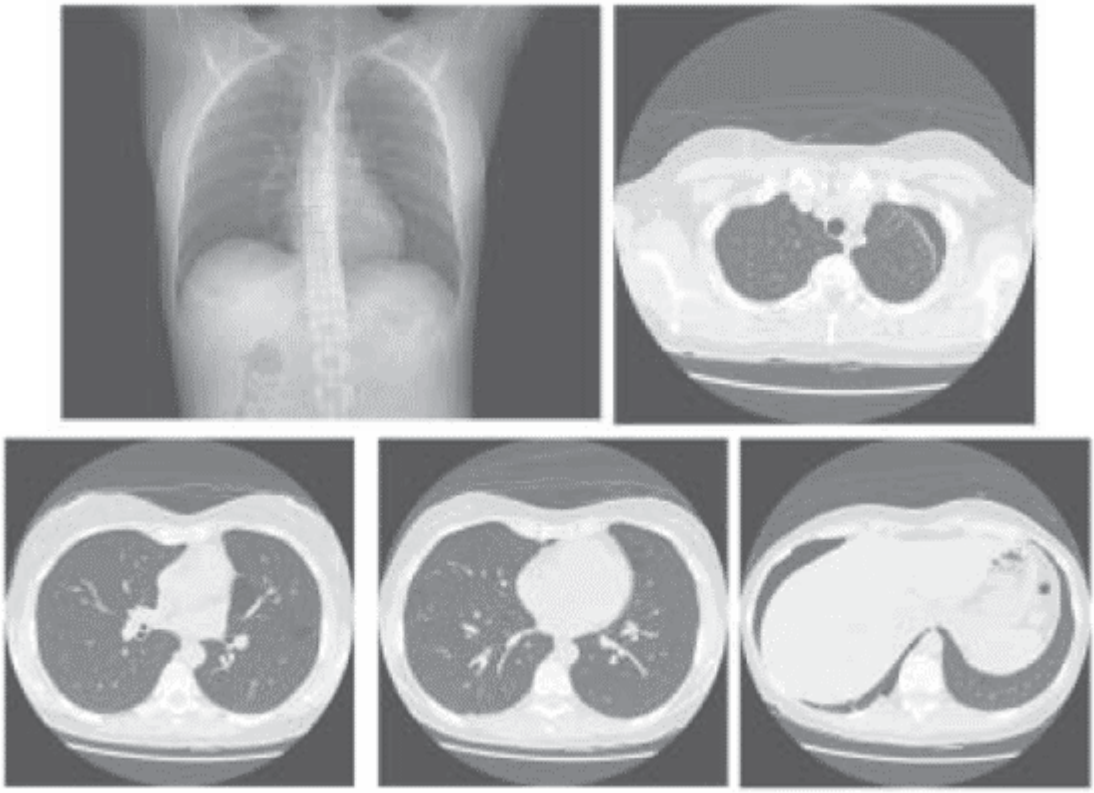
LYMPHANGIOLEIOMYOMATOSIS



COMMENTS

Lymphangioleiomyomatosis (LAM) is an idiopathic disease that mainly affects women of childbearing age. It is characterized by non-neoplastic and atypical proliferation of smooth muscle cells, especially in the pulmonary parenchyma, resulting in vascular and airway obstruction, pulmonary cysts and progressively greater impairment of respiratory function. In approximately half of all cases, LAM may involve concomitant renal angiomyolipomas, appearing sporadically or as part of the tuberous sclerosis complex (TSC), which also includes mental retardation, convulsions and skin lesions.

Symptoms of LAM commonly include dyspnea, cough and thoracic pain. In many cases, the initial manifestation is pneumothorax. According to some authors, the prevalence of pneumothorax may be as high as 68%, compared with 29% for chylothorax.

In virtually all cases, simple radiographs present alteration in the form of a diffuse reticular pattern, and larger cysts may also be seen. Pneumothorax and pleural effusion may occasionally be observed. Lung volume is maintained or increased.

The imaging method of choice is computed tomography, preferably high-resolution computed tomography, which will show air-filled thin-walled cysts measuring 0.2 to 5.0 cm in diameter with diffuse distribution. Lymph nodes may be enlarged, and pleural effusion, pneumothorax and pulmonary opacities with ground-glass patterns may be

present. The abdominal CT may reveal renal angiomyolipomas and enlargement of additional lymph nodes.

The main differential diagnoses are those that lead to pulmonary hypertransparencies. In most cases, pulmonary emphysema can easily be distinguished from LAM since emphysema does not present the walled cysts seen in LAM and is predominantly found in the upper lobes. Histiocytosis of Langerhans cells is the main differential diagnosis, although also found mainly in the upper lobes and rarely involves the costophrenic sulci, producing cysts that are usually irregular and nodular. In addition, histiocytosis almost exclusively affects smokers. Lymphoid interstitial pneumonia with follicular bronchiolitis represents another differential diagnosis, although it presents fewer cysts and it is usually related to collagenosis or AIDS.

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