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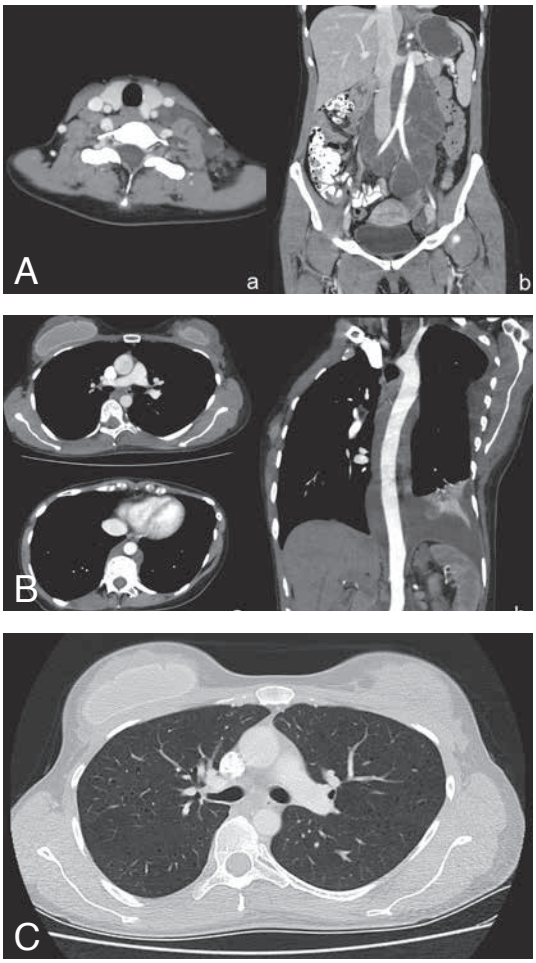
IMAGES IN CLINICAL RADIOLOGY



Extrapulmonary manifestation of lymphangioleiomyomatosis

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A 31-year-old female patient presented at the department of hematology with painful nodal swellings in the neck region for 2 months. Four weeks before presentation, she had an airway infection during which the nodes were swollen. Since 10 days she suffered from dyspnea, a dry cough and a burning ache in the chest. There was no fever, neither hemoptoe.

At clinical examination painful supraclavicular nodes on the left side and along the sternocleidomastoid muscle on the right side were found and interpreted as lymph nodes. A blood investigation showed known ferriprive anemia but no reactive lymphocytosis. Chest x-ray was normal (not shown) despite a clear scoliosis.

Some weeks later the patient mentioned that the nodes appeared to be fluctuating in volume, especially to be enlarged in a supine position or when bending over. There was increasing discomfort when wearing her seatbelt. An ultrasound of the painful neck region was performed and showed multiple fluid collections (not shown), probably mutual communicating. The collections enlarged in a supine position compared to a prone position. There was no flow in the collections.

Subsequently, a CT of the chest and abdomen was done and showed multiple thin-walled cystic masses along the sternocleidomastoid and scalene muscles (Fig. Aa), descending in the mediastinum (Fig. Ba). These cystic masses continued along the thoracic aorta to the abdominal retroperitoneum and further down along the iliac arteries, to stop at the inguinal canal (Fig. Ab). The luminal density of these masses measured 40HU in the thorax and 10HU in the abdomen. In the abdomen late enhancement of the thin cystic wall could be noted.

Along these findings the patient had multiple thin walled cysts in the lung with a perihilar predominance (Fig. C), which were the key to make the diagnosis of lymphangioleiomyomatosis or LAM.

No renal angiomyolipoma was found, nor was there any chylous ascites. No symptoms of tuberous sclerous complex were found. This was confirmed by MRI. The patient had no history of pneumothorax.

Two months after the initial diagnosis, the patient presented at the emergency department with dyspnea, coughing, hemoptoe and burning chest ache. CT showed a chylothorax which was treated with thoracentesis (Fig. Bb). Pleural fluid was positive for chylomicrons.

Comment

LAM is a disease that presents typical in women of reproductive age. It is a proliferation of disorderly smooth muscle growth in the lungs, kidneys and the lymphatics. The pathogenesis is unknown but the latest data suggest that there is loss of tumour suppression function of some proteins, or that there are abnormalities in the enzymes for the synthesis of catecholamines. It is also clear that estrogen plays a role in the disease as it is almost only observed between menarche and menopause and it is known that there is disease progression during pregnancy and disease cessation after ovarectomy.

The pulmonary manifestations are the typical formation of small lung cysts. The atypical findings are pulmonary hypertension, pneumothorax and chylothorax. The extrapulmonary manifestations include chyloperitoneum, lymphangioleiomyomas, renal angiomyolipomas and meningiomas. Our patient showed lymphangioleiomyomas in the lower neck extending to the mediastinal and retroperitoneal lymphatic system, and lung cysts.

Patients with LAM can present with progressive dyspnea, spontaneous pneumothorax, hemoptysis or other more common pulmonary complaints as wheezing, coughing or chest pain.

The 10-year survival rate ranges from 49-79%. It is a progressive disease and leads to respiratory failure. LAM can occur with increased frequency in patients with tuberous sclerosis complex (TSC), an autosomal dominant disorder due to mutations in the TSC1 or TSC2gene.

LAM can be diagnosed using CT, which is perfect for detecting the typical lung cysts, lymphangioleiomyomas and renal angiomyolipomas. Other techniques as MRI and ultrasound can also aid to the diagnosis.

References

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