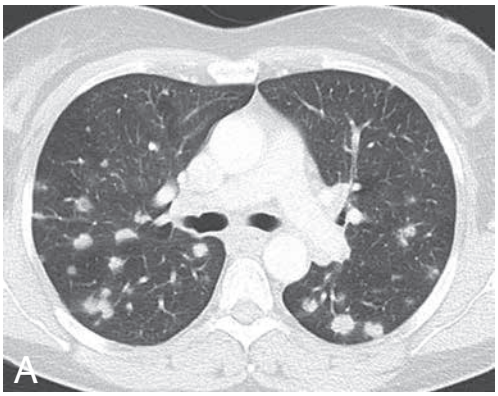
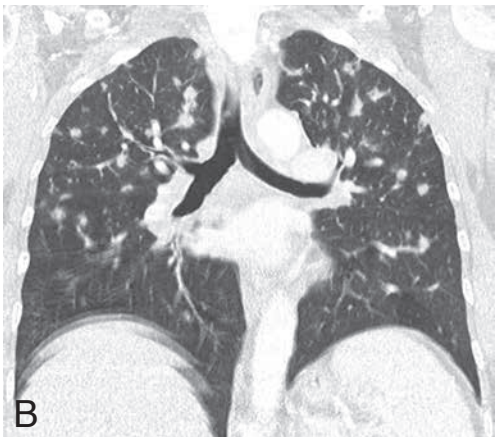
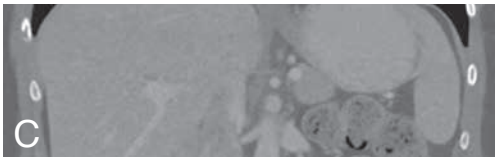
JBR–BTR, 2015, 98: 92.

IMAGES IN CLINICAL RADIOLOGY







Rare presentation of Langerhans cell histiocytosis

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A 51-year old woman consulted at our hospital with rightsided chest pain. A thoracic radiograph revealed multiple nodules in both lungs. These nodules were not present on a radiograph taken two months earlier during a routine check-up. A computed tomography of the thorax demonstrated multiple nodules varying from 3 to 15 mm (Fig. A). The nodules were spiculated and randomly distributed with predominance for the middle and upper lung zones (Fig. B). The mediastinal and hilar lymph nodes were slightly enlarged. Furthermore, a right sided osteolytic bone lesion was present anterolaterally in the eighth rib (Fig. C). Since the radiological differential diagnosis was very broad, a surgical lung biopsy and partial rib resection was performed. Histologically, the nodules showed infiltration of lymphocytes, eosinophils and Langerhans' cells. The osteolytic bone lesion showed typical findings of an eosinophilic granuloma. Both lesions were consequently a presentation of the same disease and the diagnosis of Langerhans' cell histiocystosis (LCH) with pulmonary involvement was made. Immunosuppressive therapy with glucocorticoids was initiated resulting in partial regression of the pulmonary nodules on a follow-up scan. Unfortunately, the patient refused to stop smoking and a recent scan showed disease progression.

Comment

Langerhans cell histiocytosis is a rare histiocytic disorder characterized by accumulations of large mononuclear cells forming granulomas in various organs. The disease can be divided into two groups based on single or multisystem involvement. Multisystemic disease is most common in young children whereas single organ involvement is more common in adults. When the lung is the primarily affected organ, the disease is called pulmonary Langerhans cell histiocytosis (PLCH), previously known as Histiocystosis X or eosinophilic granuloma of the lung. In the whole spectrum of LCH, PLCH is considered as somewhat atypical since it is associated with cigarette smoking in more than 90 percent of cases. Patients with PLCH usually present with respiratory or constitutional symptoms. Frequently the diagnosis is made incidentally on a routine chest radiograph. Our patient shows multisystemic disease but the pulmonary changes are most notably. Typically, in the first stage of pulmonary disease, multiple small nodules are present with middle and upper lung zone predominance. The nodules are normally around 5 mm in diameter. With disease progression, the nodules start to excavate and form cystic lesions. The advanced form is the most commonly seen by radiologists: multiple pulmonary cysts, usually less than 10 mm in diameter but often confluent, with variable wall thickness and middle and upper lung zone predominance, sometimes associated with some nodules. Multiple nodules lesions in the absence of cysts are only present in the early stage of LCH and are seldom seen in imaging. Multisystemic presentations like in our patient with a concomitant bone lesion are rare in older patients. Next to smoking cessation, glucocorticoid treatment is often effective in the treatment of this disease (1).

Reference

1. Mogulkoc N., Veral A., Bishop P.W., Bayindir U., Pickering C.A., Egan J.J.: Pulmonary Langerhans' cell histiocytosis: radiologic resolution following smoking cessation. *Chest*, 1999, 115: 1452.

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