Langerhans Cell Histiocytosis: Bizarre Pulmonary Cysts

Histiocitose de Células de Langerhans: Quistos Pulmonares Bizarros

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Abstract

Langerhans cell histiocytosis (LCH) or histiocytosis X, is a rare disorder characterized by an abnormal increase in histiocyte cells, components of the immune system. Signs of LCH depend on the extent and location of the disease. A male predominance is observed.

Keywords

Cysts; Lung; Histiocytosis.

Images of Interest / Imagens de Interesse

19-year-old male patient, smoking 5 packs/year, working in an aviary, with a history of daily dry cough for 2 years and sudden episode of right pleuritic thoracalgia. Chest X-ray showed bilateral reinforcement of the pulmonary reticulum. The high resolution computed tomography of the thorax revealed exuberant bilaterally distributed pulmonary cysts with predominance by the upper lobes, with irregular and bizarre morphology, the largest reaching 3-4cm of greater diameter (Fig. A and B). The functional ventilatory study demonstrated obstruction of the small airways with increased residual volume and a slight decrease in the diffusion capacity of carbon monoxide. He underwent bronchofibroscopy with bronchoalveolar lavage, whose immunohistochemical examination was positive for CD1a cells, allowing to establish the diagnosis. In adults with histiocytosis, the respiratory tract is most frequently involved and pulmonary lesions may be the only manifestation of the disease. Imaging studies may reveal pulmonary (sometimes cavitated) nodules, irregularly shaped cysts, fibrosis, pneumothorax, and pleural effusion.
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Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Protecção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial.

References
