Case 3359

Eurorad

Mac Leod syndrome and bronchogenic cyst

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Section: Chest imaging
Imaging Technique: CT
Imaging Technique: MR
Imaging Technique: Nuclear medicine conventional
Case Type: Clinical Cases
Authors: Oliva M, Paolicchi A
Patient: 8 years, female

Clinical History:

An 8-year-old female patient presented to the Pediatric Department with a history of consecutive bronchial pneumonia from the age of 1 year. She had cough and a defective thorax development. The spirometry showed a restrictive syndrome.

Imaging Findings:

A conventional chest radiograph was taken, which demonstrated a hyperlucent left lung with bronchiectasis on the inferior left lobe, and a mass in the anterior mediastinum (Fig. 1a, b). A contrast enhanced CT scan showed hypoplastic left pulmonary artery, cystic bronchiectasis and reduced density of the left lung parenchyma (Fig. 2a, c, d). Besides, the CT scan showed the presence of a paratracheal cystic mass compressing the mediastinal structures (Fig. 2b). The Magnetic Resonance Imaging confirmed the presence of a paratracheal cystic mass compressing the left main bronchus, the left pulmonary artery, and the left atrium (Fig. 3a, b). The ventilation-perfusion scintigraphy showed a marked hypoperfusion of the left lung (Fig. 4a, b). Pulmonary angiography showed a hypoplastic left pulmonary artery compared to the right side.

Discussion:

Mac-Leod syndrome (syn.: Swywer-James-syndrome) is a manifestation of postinfectious obliterative bronchiolitis. The involved lung or portion of the lung does not grow normally and is slightly smaller than the opposite lung (2). Radiographically, pulmonary hyperlucency caused by overdistended alveoli combined with diminished arterial flow characterizes its imaging appearance. The lung is expected to grow by progressive alveolarization in a child between the age of 2 and 8 years. Thereafter, lung growth is related to hyperexpansion of the existing alveoli. Mac-Leod syndrome is characterized by diminished vascularity, arrest of progressive growth and alveolarization of the lung and resultant hypoplasia . Multifocal areas of air trapping may be seen. Patients with Mac-Leod syndrome have a small lung, compensatory overexpansion of the contralateral lung, peripheral bronchi and bronchioles "pruned" secondary to obliterative bronchiolitis, a mosaic pattern of hyperlucency on CT, and small vessels and vascular occlusions in the abnormal areas (3). Adults with Mac Leod syndrome may be asymptomatic or may have cough, recurrent respiratory infection and haemoptysis. Bronchogenic cysts are lesions of congenital origin derived from the primitive foregut and are the most common primary cysts of the mediastinum. They are often unilocular, and contain

clear fluid or, less commonly, hemorrhagic secretions or air. They are lined by columnar ciliated epithelium, and their walls often contain cartilage and bronchial mucous glands. It is unusual for them to have a patent connection with the airway, but when present, such a communication may promote infection of the cyst by allowing bacterial entry. Some bronchogenic cysts are asymptomatic. The most frequent symptoms are cough, fever, pain, and dyspnea. Tracheobronchial compression and pulmonary infections can occur in children because of the relatively soft tracheobronchial tree (1). Complications of bronchogenic cysts are frequent . Most of the complications result from compression of adjacent structures. Infection is a common complication, especially in cysts with bronchial communications. The cyst can rupture into the trachea, the pericardial cavity, or the pleural cavity. Pneumothorax is not a rare complication and is usually accompanied by pleuritis. Severe haemoptysis is rarely reported. This is a rare case of Mac-Leod syndrome associated with bronchogenic cyst. It is not clear if the Mac leod syndrome is secondary to compression on the left main bronchus and pulmonary artery.

Differential Diagnosis List: Mac Leod syndrome and bronchogenic cyst.

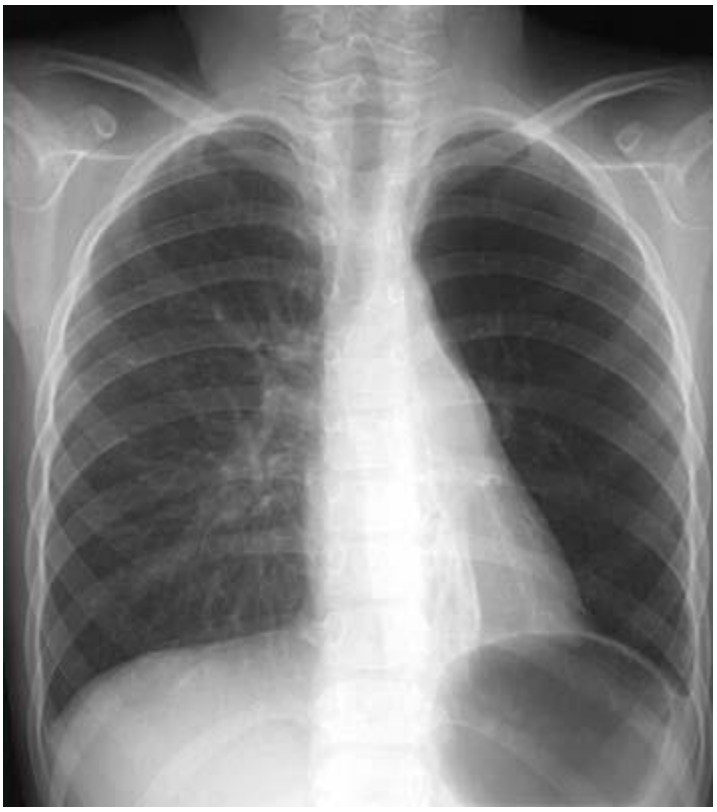
Final Diagnosis: Mac Leod syndrome and bronchogenic cyst.

References:

Vasilios D.Kollias,Jeorge H.Kantidakis et All
Large tension bronchogenic Cyst in adult mimicking the Swjer-James syndrome.
Respiration 1998; 65:411-413. (PMID: 9782227)
Lucaya J,Gartner S,Garcia-Pena P et All.
Spectrum of manifestations of Swjer-James-Mac Leod syndrome.
J Comput Assist Tomogr 1998; 22:592-597. (PMID: 9676450)
Norton KI,Mendelson DS,Hodes D et All.
Computed Tomography findings in the Swjer-James syndrome.
Clin Imaging 1989; 13(1):48-50. (PMID: 2743192)

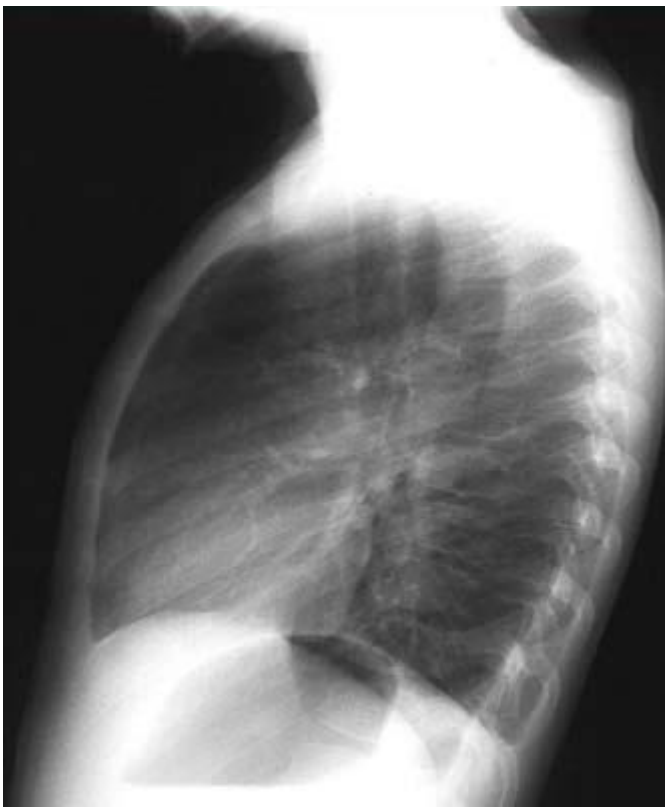
Figure 1

a



Description: Conventional p.a. chest radiograph showing a hyperlucent left lung with bronchiectasis on the inferior left lobe. **Origin:**

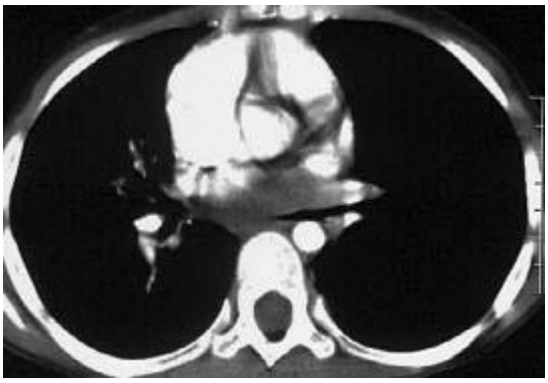
b



Description: Conventional lateral chest radiograph showing the presence of a mass in the anterior mediastinum. **Origin:**

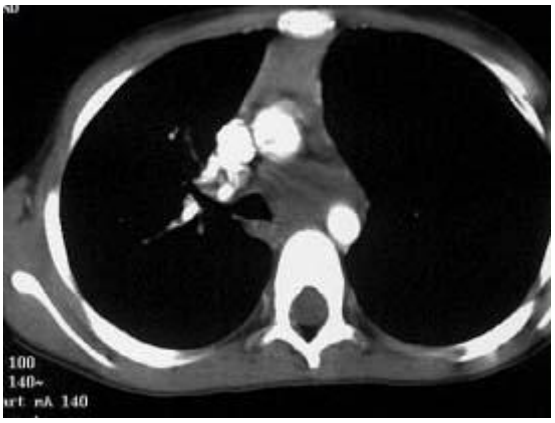
Figure 2

a



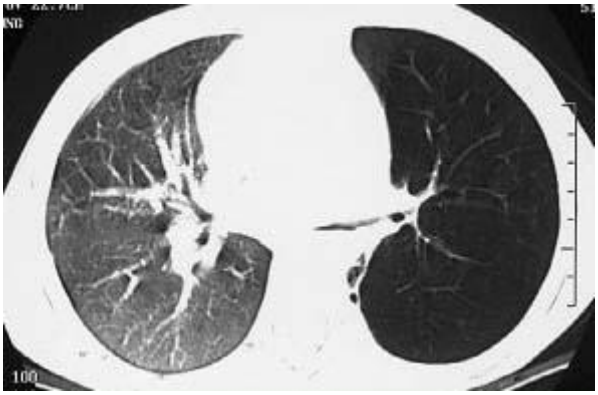
Description: A CT image demonstrating a hypoplastic left pulmonary artery. **Origin:**

b



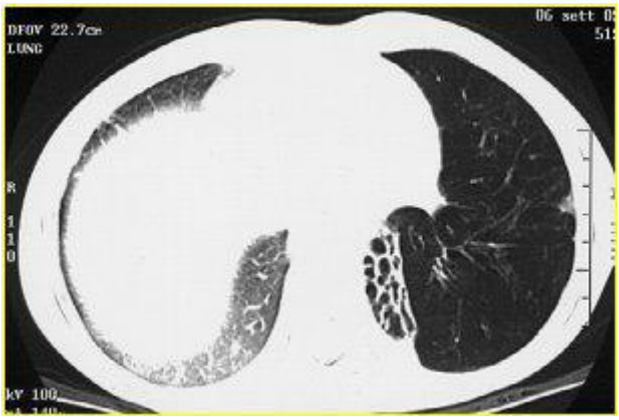
Description: A CT scan showing the presence of a paratracheal cystic mass compressing the mediastinal structures. **Origin:**

c



Description: A CT image demonstrating reduced density of the left lung parenchyma. **Origin:**

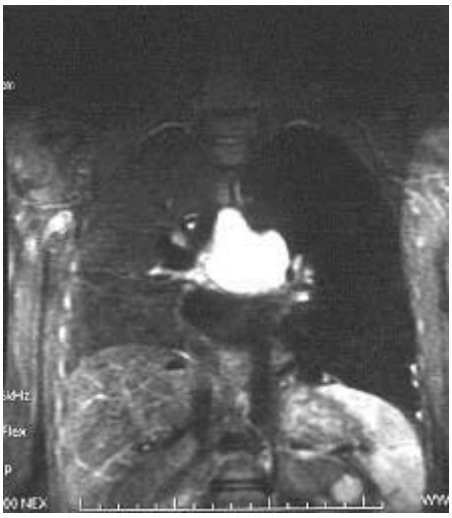
d



Description: A CT image demonstrating cystic bronchiectasis on the inferior left lobe. **Origin:**

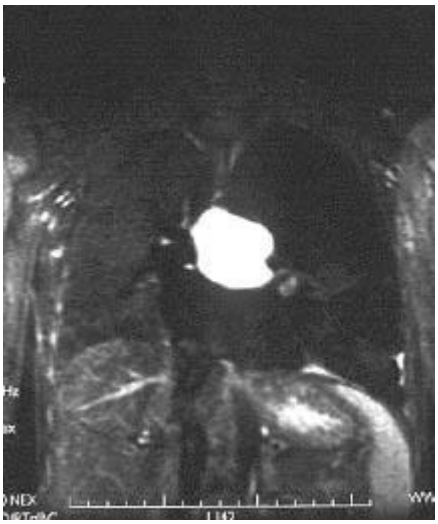
Figure 3

a



Description: A coronal T2-weighted MR image demonstrating the presence of a high signal mass in the left paratracheal region. **Origin:**

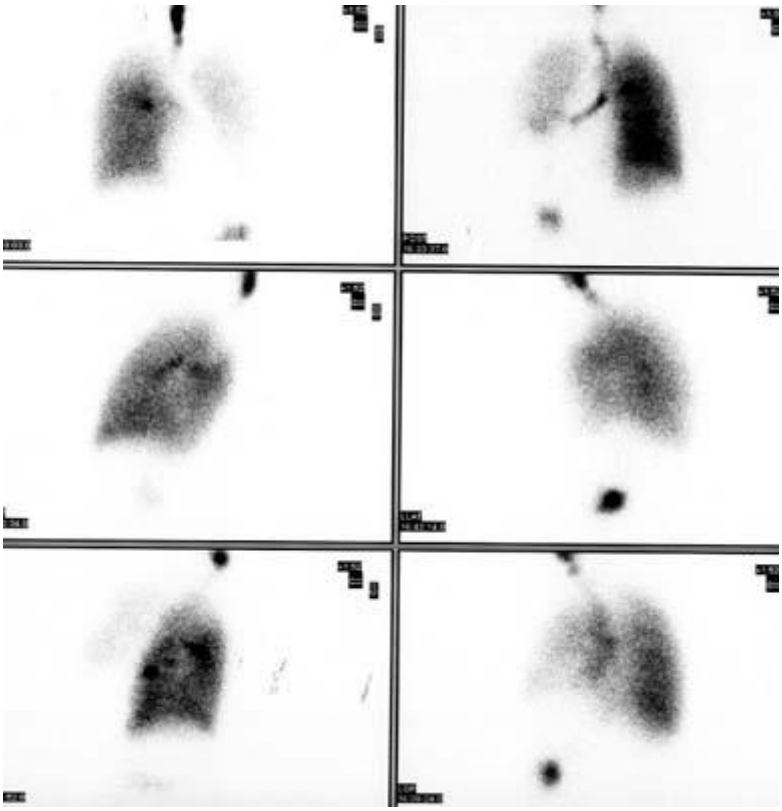
b



Description: A coronal T2-weighted MR image demonstrating the presence of a paratracheal cystic mass compressing the left bronchial tube, the left pulmonary artery and the left atrium. **Origin:**

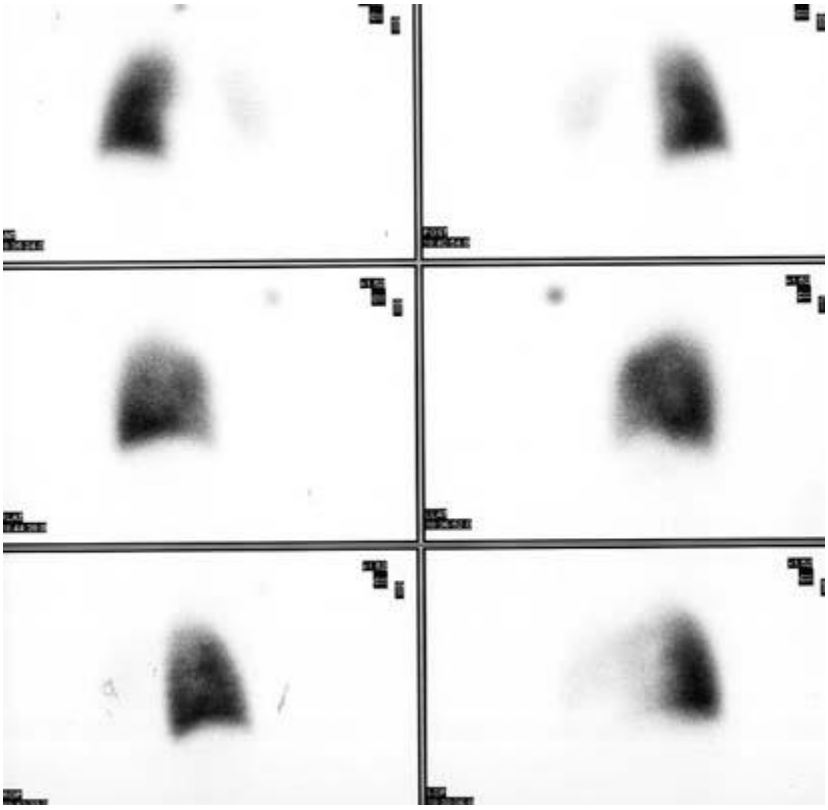
Figure 4

a



Description: The ventilation-perfusion scintigraph showing a reduced ventilation on the left lung (ventilation scintigraph). **Origin:**

b



Description: The ventilation-perfusion scintigraph demonstrating a marked reduction of perfusion on the left lung (perfusion scintigraph). **Origin:**