FOCAL FIBROSING MEDIASTINITIS

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Key-word: Mediastinitis

Background: A 38-year-old Cameroonian refugee was admitted to the Emergency Department for dyspnea, cough, chest pain and hemoptysis.

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Work-up

Chest radiograph (Fig. 1) (PA-view) shows right hilum enlargement and para-hilar parenchymal opacities.

CT scan of the thorax (Fig. 2) shows on unenhanced CT scan at the level of the right hilum (A) calcifications in hilar soft tissues. Contrast-enhanced CT scan at the level of the right hilum (B) demonstrates a hilar soft tissue infiltration compressing the interlobar artery. On maximum intensity projection (MIP) reformatted image in the coronal plane (C) and maximum intensity projection (MIP) reformatted image in the axial plane (D) the “pseudo-tumoral” pattern of the hilar mass that compresses arterial structures, including the right interlobar pulmonary artery and the internal segmental pulmonary artery of the middle lobe is clearly visible.

Note absence of enhancement at the level of the pulmonary vein of the middle lobe.

The infiltrate in the middle lobe corresponds to an infarction.

Radiological diagnosis

Based on morphology and localization of the lesion on CT scan of the thorax, the diagnosis of focal fibrosing mediastinitis was made.

Discussion

Fibrosing mediastinitis (FM) is a rare benign disorder caused by proliferation of acellular collagen and fibrous tissue within the mediastinum. The consequence is compression or obstruction of mediastinal structures such as the pulmonary veins, pulmonary arteries, central airways, superior vena cava and esophagus.

The precise cause and pathogenesis of FM is unknown in most cases, and links to infectious or noninfectious causes remain speculative.

A focal and diffuse type of FM are distinguished. The focal type (82% of cases) usually manifests on CT or MRI as a localized, partially calcified (63%) mass in the paratracheal or subcarinal regions of the mediastinum or in the pulmonary hila. The right side of the mediastinum is more commonly involved than the left. FM is considered to result from an abnormal immunologic reaction to organisms such as Histoplasma capsulatum or Mycobacterium tuberculosis and in rare cases to other infectious diseases including aspergillosis, mucormycosis, blastomycosis and cryptococcosis. The diffuse type (18%) manifests as a diffusely infiltrating, often noncalcified mass that affects multiple mediastinal compartments. It is mostly due to an auto-immune disease associated with other idiopathic fibroinflammatory disorders such as retroperitoneal fibrosis, sclerosing cholangitis, Riedel thyroiditis, and pseudotumor of the orbit.

Conventional chest radiographs underestimate the extent of disease, showing distortion and obliteration of normally recognizable mediastinal interfaces or lines.

On CT scan, FM manifests as an infiltrative mass of soft-tissue attenuation that obliterates normal mediastinal fat planes and encases adjacent structures. On contrast-enhanced scans the degree of enhancement of the mass is variable. Contrast-enhanced CT is also useful for depicting encasement or obstruction of pulmonary arteries and veins.

Fibrosing mediastinitis manifests on T1-weighted MR-images as a heterogeneous, infiltrative mass of intermediate signal intensity. The appearance on T2-weighted MR-images is more variable: regions of both increased and decreased signal intensity are frequently seen in the same lesion.

The most common symptoms include cough, dyspnea, recurrent pulmonary infections, hemoptysis, and pleuritic chest pain. Treatment consists in systemic antifungal, antituberculosis, and/or corticosteroid treatment. Endovascular or endobronchial stent can be inserted to restore the bronchial or vessel lumen. Fibrosing mediastinitis often has an unpredictable course, spontaneous remission or exacerbation of symptoms being reported. The mortality rate is higher than 30%.

Bibliography