A Case of Syphilitic Interstitial Pulmonary Fibrosis

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Diagnosis of late pulmonary syphilis was made in a 72-year-old woman on the basis of her medical history of untreated genital syphilis, serological and radiological findings, and the response to therapy. The diagnosis was confirmed two years later by autopsy findings. There was a good correlation between chest x-ray radiography, computed tomography scans, and autopsy findings. In the diagnosis of sarcoidosis and other diseases with similar radiological interstitial lung pattern, syphilitic interstitial pulmonary fibrosis should be carefully excluded.

Key words: chest radiography; interstitial lung disease; pulmonary fibrosis; syphilis, latent

Usually painless and often unnoticed chancre (ulcer) of the primary syphilis appears at the site of infection approximately 10-90 days after contact. Secondary syphilis develops approximately 7-10 weeks after the initial infection, and its flu-like symptoms and a general rash may again remain unnoticed. The infection then becomes latent, and may progress to tertiary syphilis after 5-35 years. Although syphilitic involvement of the lungs was first described more than a hundred years ago, only a few cases of tertiary pulmonary syphilis have been reported so far (1). Tertiary syphilis is a result of damage to internal organs which may include the brain, spinal cord, heart, and rarely the lungs. The patient’s history, clinical findings, and detection of spirochetes in the tissue were the basis for the diagnosis. There are simple nontreponemal tests for screening, and complex treponemal tests for the confirmation of the diagnosis (2). Four types of pulmonary lesions in an acquired tertiary syphilis have been described (3): a) gummas, ranging from small multiple lesions to a single large formations; b) syphilitic pulmonary fibrosis of non-specific character; c) syphilitic chronic interstitial fibrosis and; d) gummatous ulceration of the trachea and bronchi. The first and third variety may have a bronchiectatic form. We present here the case of tertiary lues of the lungs and the aorta in an elderly woman.

Case Report

A 72 year-old woman complaining of breathlessness, dry cough, and slightly elevated body temperature (37.4 °C) was presented to the Department of Pulmonary Diseases. She was conscious, mobile, and dyspneic on admission. Physical examination revealed dullness over the lungs and diffused bilateral crepitation. Erythrocyte sedimentation rate was 110 mm; RBC 3.2x108/L; Hb 106 g/L, and WBC and hepatogram were within the normal range. Chest x-rays and high resolution computed tomography scans showed a diffuse reticulonodular lung pattern, mostly in the middle and lower lung zones, and bilateral hilar enlargement (Fig. 1). Ultrasonography of the heart revealed a 5-cm enlargement of the ascending aorta. Bronchoscopy showed dilated bronchial three at the level of the 3rd and 4th branch. Sputum and fluid lavage were sterile. The test for the Mycobacterium tuberculosis was negative. Pulmonary function tests showed a serious restrictive pattern. Transbronchial biopsy of pulmonary parenchyma showed normal bronchial wall with a mild lymphocytic infiltrate of fibrotic interstitial tissue. Extensively thickened fibrotic alveolar septum was also observed in the biopsy specimen. At that point, a more detailed anamnestic data were obtained. The patient reported a small vulvar lesion 30 years before, which was treated with a short penicillin treatment. According to these information, the serological tests for syphilis were performed. The findings were positive: venereal disease laboratory test (VDRL)=512 U, and Treponemal hemaglutination antibody test (TPHA)= 1280 U. Intravenous penicillin therapy was initiated, but was discontinued because of the onset of an allergic reaction to penicillin. The patient was then treated with Erythromycin (250 mg capsules 4x2/50 days). Six months later, TPHA level was decreased, whereas VDRL remained unchanged. After two years the patient was admitted again because of a serious cardiorespiratory insufficiency. Two hours later she died in cardiorespiratory arrest. Pathological findings at autopsy revealed dilatation of the ascending aorta and a 10-mm aneurysmatic sac on the posterior wall. A fibrotic scar
on the posterior wall of the left ventricle (measuring 5 cm) was also found. The lungs were free, with nodular surface in the form of a small oval and round elevated areas about 10 mm in diameter. On the cut surface, the lungs were firm, red and brown in color, with solid fibrotic areas peripherally. A microscopic examination showed pulmonary edema, intraalveolar exudation, hyaline membranes, and infiltration of the alveolar septa with mononuclear cells. Pneumocyte type II hyperplasia was also observed, appearing as cuboidal or columnar cells lining the alveolar spaces. There was an organization of the intraalveolar exudate by fibrous tissue in the central areas of both lungs. The intraalveolar septa were thick, with mononuclear cells infiltrate of the interstitium, suggesting a co-existing pneumonitis (Fig. 2).

**Figure 1:** Chest radiography of a 72-year old women with tertiary syphilis revealing a fine reticulonodular bilateral basilar lung pattern and bilateral hilar enlargement. [view this figure]

**Figure 2:** Interstitial fibrosis and epithelization of the damaged alveoli, and generalized interstitial chronic inflammatory cell infiltration. [view this figure]

**Discussion**
The syphilitic pulmonary pneumonitis is a complication of the secondary syphilis (4-6). In our case, the patient showed the combination of two different pulmonary lesions: diffused pulmonary fibrosis and bronchiectasis with pneumonitis. Decrease in the TPHA value after therapy and positive serologic tests confirmed that specific treponema antigen tests, such as TPHA, more frequently give positive results and therefore are more reliable as a supportive evidence of late syphilis (7). Symetrical hilar enlargement with reticulo- nodular appearance of the lungs on a chest radiograph can be misinterpreted because this type of changes may occur in sarcoidosis with advanced pulmonary fibrosis. Lung involvement has a similar radiologic pattern to sarcoidosis and other diseases (coal worker’s pneumoconiosis, silicosis, idiopathic diffuse interstitial pulmonary fibrosis, malignant lymphoma, and secondary syphilis in an HIV-infected patient) and must be included in the differential diagnosis (8). High-resolution tomography scans of the lung, in addition to showing morphological changes that were found at autopsy, revealed the existence of numerous small fibrous nodules on the visceral pleural surface. Syphilitic lung involvement should therefore be included in the differential diagnosis of radiological interstitial lung pattern.

**References**

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