Upper Lobe Fibrocavitary Disease in a Patient With Back Pain and Stiffness*

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(A 37-year-old white man presented with a productive cough, anorexia, weight loss, and night sweats of 4 months' duration. He also complained of back pain with stiffness. The back pain was worse at night. He was treated for pneumonia 3 years ago and had a 20 pack-year history of smoking.

He was a thin man in no distress. His vital signs were stable and temperature was 38.6°C (101.6°F). Bilateral enlarged firm but nontender axillary and inguinal lymph nodes were noted. Chest wall expansion was limited, but otherwise the thoracic examination was unremarkable. There was kyphosis of the lumbar spine with limited forward bending. The range of motion of the cervical spine was severely limited, and there was no lateral motion of the neck. The pelvis was anteverted, and there was a flexion deformity of both knees.

Laboratory findings were as follows: WBC count, 11,300 cells/μL; and erythrocyte sedimentation rate, 30 mm/h. There was a positive human leukocyte antigen-B27 antigen. Sputum smears and cultures for acid-fast bacilli were negative. Smears of BAL fluid and bronchial washings revealed rare acid-fast bacilli. The purified protein derivative skin test was negative.

A chest radiograph (Fig 1) revealed bilateral upper lobe cavitary lesions. The remainder of the lungs were hyperinflated. There was mild upward retraction of the hilar structures indicating upper lobe volume loss. Contrast-enhanced chest CT (Fig 2) showed distortion of the lung architecture with bilateral upper lobe cavitation and bronchial thickening. A radiograph of the cervical spine (Fig 3) demonstrated straightening of the cervical spine with squaring of the vertebral bodies and calcification of the anterior longitudinal ligaments. There was ankylosis of both sacroiliac joints (Fig 4).

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Figure 1. Posteroanterior chest radiograph reveals bilateral upper lobe cavitary lesions. The lungs are hyperinflated. Mild upward retraction of the hilar structures suggests upper lobe volume loss related to fibrosis.
What is the diagnosis?

Figure 2. Contrast-enhanced chest CT shows distortion of the lung architecture with bilateral upper lobe cavitating and bronchial thickening.

Figure 3. Oblique radiograph of the cervical spine demonstrates straightening of the cervical spine with squaring of the vertebral bodies and calcification of the anterior longitudinal ligaments (arrowheads).

Figure 4. Anteroposterior radiograph of the pelvis shows complete obliteration of bilateral sacroiliac joint spaces with bony ankylosis (arrowheads).
Diagnosis: Apical fibrobulous disease in a patient with ankylosing spondylitis

Discussion

The association of ankylosing spondylitis and pulmonary disease has been known since the 1940s.1,2 Thoracic manifestations of ankylosing spondylitis are of two types: upper lobe fibrobulous disease and chest wall restriction.3,4 The true incidence of fibrobulous lung disease, the more common manifestation of ankylosing spondylitis, is not known, but reports range from 1 to 30%.5,6 It is predominantly seen in adult male subjects with the male-to-female ratio of 50:1.8 The human leukocyte antigen-B27 antigen result is positive in up to 95% of patients with ankylosing spondylitis.3,4,6,7 The cause of apical fibrobulous changes is unknown, but several theories have been suggested. These include diminished upper lobe ventilation due to chest wall rigidity,9 altered apical mechanical stress due to rigid thoracic spine,3 recurrent pulmonary infection due to impaired cough, and respiratory mechanics as a result of thoracic rigidity.2 Other theories include prior thoracic irradiation, repeated aspiration pneumonitis secondary to esophageal muscle dysfunction,10 and cricoarytenoid joint disease.11

Unless extensive, apical fibrosis is clinically silent. Symptoms may include cough, hemoptysis, increased sputum production, and dyspnea. Patients with fibrocystic disease may develop mycetomas and other infections, such as atypical mycobacterium.

High-resolution CT12 is frequently helpful to evaluate the lung apices for the presence of infection and especially mycetoma.

References