Hemoptysis, History of Oral Ulcers, and Abnormal Chest Radiograph in a 64-Year-Old Man*

Marvin I. Schwarz, MD, FCCP

A 64-year-old man presents with 2 months of intermittent blood-tinged sputum and cough. He also noted mild dyspnea and subjective fevers. For the past 30 years, he has noted self-limiting clusters of oral ulcers. During these periods, he also has difficulty swallowing and was found to have esophageal ulcers. He has had one episode of nose swelling and two episodes of ear swelling and was treated with corticosteroids. Four years ago, he had bloody diarrhea, and colonoscopy revealed colonic ulcerations. He also reports an episode of iritis, intermittent nonpruritic rashes, and arthralgias.

At the present time, he denies sinus symptoms, ear or throat pain, epistaxis, or Raynaud phenomenon. He is receiving prednisone, 10 mg/d. He has not traveled recently; he works as a plumber and has no HIV risk factors. His son and daughter have oral ulcers. The patient is in no distress, is afebrile, and the remainder of his vital signs are normal. Examination of the ears, eyes, nose, and throat are normal, as is the remainder of the physical examination. His chest radiograph (Fig 1), CT scan (Fig 2), and lung biopsy (Fig 3) are shown. All of the following statements about this patient’s disease are true EXCEPT:

A. There is a tendency for hypercoagulability and venous thrombosis.
B. Serum antineutrophil cytoplasmic antibody positivity confirms the diagnosis.
C. There is a familial predisposition.
D. A positive pathergy test result is often present.
E. Male subjects are more commonly affected.

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Answer: B. Serum antineutrophil cytoplasmic antibody positivity confirms the diagnosis.

This patient has Behcet disease (BD), which is a multisystem vasculitis involving vessels of all sizes in both the venous and arterial systems. It is complicated by hypercoagulability and recurrent venous thrombosis and embolization. Antineutrophil cytoplasmic antibodies are usually negative, and when positive are neither directed against cytoplasmic serine protease (classic pattern antineutrophil cytoplasmic antibody) or myeloperoxidases (perinuclear antineutrophil cytoplasmic antibody), but rather other antigens in the neutrophil cytoplasm that are nonspecific and seen in other inflammatory diseases.

There is a clear familial tendency (11% of cases). Whether this is due to genetic or environmental influences is not known. The highest incidence of this disease is found in Turkey (300/100,000 cases). Male subjects are more commonly affected, have more severe multisystem disease, and are more apt to have pulmonary involvement.

BD is a clinical diagnosis requiring at least two of the following in combination with recurrent painful oral aphthous ulcerations: recurrent genital ulcerations, eye involvement (uveitis, retinal vasculitis), skin lesions (erythema nodosum, pseudofolliculitis), or a positive pathergy test result (a 2- to 3-mm papule or pustule appearing 24 to 48 h following a sterile pin prick). Other findings include arthritis, esophageal ulcerations, colonic ulcerations that may mimic inflammatory bowel disease, aseptic meningitis, superficial thrombophlebitis, deep venous thrombosis, arterial occlusion and/or aneurysmal dilatation anywhere, glomerulonephritis, and cardiac involvement.

The histology is nonspecific as demonstrated in Figure 3. It shows a neutrophilic vasculitis with a mixed cellular perivascular infiltrate. With involvement of larger vessels, this can result in aneurysmal dilatation. Lung involvement consists of vasculitis causing infarction with cavitation, as shown in the chest radiograph (Fig 1) and CT scan (Fig 2), or infarction secondary to thromboembolism. Pulmonary artery aneurysm with rupture can be catastrophic. A diffuse alveolar hemorrhage syndrome with underlying pulmonary capillaritis can also occur. The lungs are primarily involved in 30% of cases and, when this is the case, survival is < 2 years. Treatment with varying combinations of corticosteroids, cytotoxic agents (cyclophosphamide and azathioprine), cyclosporin, and tacrolimus have been employed.

SELECTED READINGS