Endobronchial Telangiectasias and Hemoptysis in Scleroderma

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Hemoptysis is considered a rare event in scleroderma and to date only two previous cases could be identified. The occurrence of hemoptysis with bleeding and friable telangiectasias is reported in a patient with rapidly progressing systemic sclerosis. This represents the first report of this association, although bleeding telangiectasias have been reported in other systems. A brief review of the relevant literature is included.

Cutaneous telangiectasias are a common clinical sign in scleroderma. These vascular malformations have also been identified in the gastrointestinal and genitourinary tracts and have been reported to cause both melena and hematuria [1,2]. Although involvement of the respiratory system is common in scleroderma, hemoptysis is rarely encountered and only two cases have been reported to our knowledge [3,4]. This report describes the occurrence of hemoptysis in a patient with scleroderma who was discovered to have endobronchial telangiectasias.

CASE REPORT

A 55-year-old white woman with diffuse progressive scleroderma was admitted with worsening dyspnea, hemoptysis, hypertension, and azotemia. Nine months prior to admission, she had complained of increasing joint stiffness and thickening of the skin over the joints of her knees, ankles, and small joints of her hands. Serologic demonstration of a positive fluorescent antinuclear antibody at 1:2,560 (speckled/nucleolar pattern), a negative latex rheumatoid factor, negative anti-DNA antibodies, negative rapid plasma reagin, negative direct Coombs' test, and normal total hemolytic complement levels contributed to the diagnosis of scleroderma. Enteric-coated aspirin was initially prescribed, with the addition of amitriptyline and short courses of prednisone for continued pain. Seven months prior to admission, she began to experience a morning cough, intermittently productive of "pink" sputum. Delayed skin tests for fungi and mycobacteria were consistent with cutaneous anergy. Sputum cytology and mycobacterial studies were unrevealing. Two months prior to admission, penicillamine was administered but was discontinued soon after due to pruritus. In the month prior to admission, the patient experienced daily hemoptysis with increasing dyspnea on exertion, fatigue, and orthopnea. Facial and truncal telangiectasias appeared. Prednisone was prescribed at 5 mg per day. The patient had no fevers, chills, or sweats. There was no history of smoking or known exposure to tuberculosis.

On admission, she was febrile and hypertensive with blood pressure 200/100 mm Hg. Physical examination was notable for sclerodactyly, telangiectasias of the face and arms, bilateral chest rales, and bilateral lower extremity edema. Admitting laboratory values included blood urea nitrogen 26 mg/100 dl, creatinine 2.1, mg/dl, hemoglobin 12.7 g/100 dl, platelet count 207,000/mm^3, white blood count 8,300/mm^3, prothrombin time 9.4 seconds (ratio 0.9), partial thromboplastin time 29 seconds (nor-

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mal, 23 to 34 seconds), bleeding time 13 minutes, and room air arterial pH 7.40, partial carbon dioxide pressure 33 mm Hg, and oxygen partial pressure 49 mm Hg. A chest radiogram revealed an alveolar filling process in the left lower lobe. Fiberoptic bronchoscopy was performed (Olympus BF-1T10) with examination of the tracheobronchial tree to the first subsegmental level of all lobes. Old blood was present in the trachea and airways of both lungs without evidence of active bleeding. There were, however, multiple telangiectasias present in the mucosal lining of the right and left mainstem bronchi and the bronchus of the left upper lobe. The lesions were 3- to 5-mm networks of small vessels, red in color, flat, friable, nonpulsatile, and surrounded by normal-appearing mucosa. Bronchoscopic biopsies were not performed. Bronchial washings grew normal respiratory flora and coagulase-positive staphylococci, thought to be a nasal contaminant. No antibiotic therapy was given and the patient remained afebrile. Aspirin therapy was discontinued on admission and the patient's hemoptysis abated spontaneously over two to three days. Her oxygenation simultaneously improved and a follow-up chest radiogram showed resolution of the alveolar filling process.

COMMENTS

Scleroderma has been associated with pulmonary arteriovenous shunts that are thought to arise secondary to hypoxic vasoconstriction [5], but we have no knowledge of a previous description of endobronchial telangiectasias in this disease. Hemoptysis is rarely reported in scleroderma [1,3]. Endobronchial telangiectasias have not been described despite the performance of fiberoptic bronchoscopy in patients with scleroderma [6]. Similarly, a list of the causes of hemoptysis does not include scleroderma [7]. A case report of hemoptysis in scleroderma attributed the bleeding to alveolar hemorrhage, and bronchoscopy did not disclose endobronchial telangiectasias [4]. Telangiectasias and, more generally, pulmonary arteriovenous malformations, have been previously reported to occur in the bronchial tree [8–10], but never in association with scleroderma. There was little indication that another etiology might explain this patient's hemoptysis. There was no suggestion of acute bronchitis, and the patient remained afebrile during her course. Discontinuation of aspirin therapy was associated wtih cessation of hemoptysis and resolution of her chest radiographic abnormalities. Bronchoscopic specimens did not demonstrate abnormal cytology, evidence of fungal or mycobacterial infection, or other pathogens considered to be significant. The telangiectasias seen in Osler-Weber-Rendu syndrome may appear similar to the lesions described in this patient [2]. However, the recent onset of cutaneous lesions in this patient and a lack of family history suggest that this is an unlikely explanation. We have attributed the hemoptysis in this patient with scleroderma to the presence of endobronchial telangiectasias, predisposed to bleeding by aspirin therapy and mild azotemia.

REFERENCES

1. Rocco VK, Hurd ER: Scleroderma and scleroderma-like disorders. Semin Arthritis Rheum 1986; 16: 22–69.
2. Braverman IM: Connective tissue (rheumatic) diseases: In: Braverman IM. Skin signs of systemic disease, 2nd ed. Philadelphia: WB Saunders, 1981; 155–377.
3. Weaver AL, Divertie MB, Titus JL: Pulmonary scleroderma. Dis Chest 1968; 54: 4–12.
4. Kallenbach J, Prinsloo I, Zwi S: Progressive systemic sclerosis complicated by diffuse pulmonary hemorrhage. Thorax 1977; 32: 767–770.
5. D'Angelo WA, Fried JK, Masi AT, Shulman LE: Pathologic observations in systemic sclerosis (scleroderma). Am J Med 1969; 46: 428–440.
6. Owens GR, Follansbee WP: Cardiopulmonary manifestations of systemic sclerosis. Chest 1987; 91:118–127.
7. American Thoracic Society: The management of hemoptysis: a statement by the committee on therapy. Am Rev Respir Dis 1966; 93: 471–474.
8. Jackson CL, Diamond S: Hemorrhage from the trachea, bronchi and lungs of nontuberculous origin. Am Rev Tuberc 1942; 46:126–138.
9. Masson RG, Altose MD, Mayock RL: Isolated bronchial telangiectasia. Chest 1974; 65: 450–452
10. Burke CM, Safai C, Nelson DP, Raffin TA: Pulmonary arteriovenous malformations: a critical update. Am Rev Respir Dis 1986; 134: 334–339.