

## Letters to the Editor

### Hemoptysis as the first symptom of limited cutaneous systemic sclerosis

Sirs,

A widely used classification scheme divides systemic sclerosis (SSc) patients according to skin and internal organ involvement into two major categories: limited cutaneous SSc (lSSc) and diffuse cutaneous SSc (dSSc). In some cases though patients are classified in other subsets of SSc such as pre-scleroderma and scleroderma *sine* scleroderma (1). We describe a patient who presented with hemoptysis due to oral telangiectasias as the first symptom of lSSc.

A 62-year-old man was admitted to the hospital on July 2001 because of hemoptysis. Just before admission he had consumed large quantities of food and alcohol at a party where he also exerted his vocal cords singing. On admission the patient was well. Physical examination, including ENT examination, chest x-ray and EKG did not reveal any abnormalities. No skin lesions or changes in skin thickening were noticed. The patient had a history of asymptomatic nephrolithiasis which was diagnosed 3 years earlier by ultrasound. He had been smoking 20 cigarettes/day for about 30 years. He also mentioned that one year earlier he noticed that his hands changed color when exposed to cold. Laboratory tests including biochemical tests, urinary analysis, blood gases and pulmonary functional tests were within normal limits.

A tuberculin test showed a positive reaction at 30 mm. C/T scan of the chest revealed only a few emphysematic areas. Bronchoscopy revealed lesions of anthracosis of the right upper lobe. Cytology, as well as cultures of the sputum and bronchoalveolar lavage for common bacteria and B. Koch, were negative. Endoscopic examination of the esophagus and stomach revealed no evidence of recent hemorrhage. A ventilation-perfusion lung scan was normal. Digital angiography of the thoracic aorta and its branches showed no abnormalities. Laryngoscopy revealed multiple telangiectasias in the soft palate, bilateral false vocal cords and buccal mucosa (Fig 1). An immunofluorescence test for antinuclear antibodies revealed a centromere pattern in a 1/1280 titer, while anti-cenp B antibody was detected by dot blot.

The patient was diagnosed as having lSSc. Since we did not carry out nailfold capillaroscopy we cannot classify this patient as suffering from a pre-scleroderma condition



Fig. 1. Telangiectasias in soft palate (arrow).

even he were to fulfill all the other criteria, i.e.: Raynaud's phenomenon, no internal organ involvement and anticentromere autoantibodies (1). Eight months after the diagnosis he is not receiving any treatment and is free of symptoms.

It is well known that limited scleroderma patients often have a long history of Raynaud's phenomenon before the appearance of other symptoms (2). In this case the patient had Raynaud's phenomenon one year before his admission to hospital, which was overlooked probably because it did not cause him any major problems. Anticentromere antibodies are associated with a range of scleroderma syndromes, usually the milder CREST variant (80%). Rarely if ever is the antibody seen in dSSc, which has a poorer prognosis than the antibody limited form (3, 4).

Although there have been several reports of hemorrhage due to telangiectasias in the gastrointestinal tract (5), respiratory tract (6) and even the nasal mucosa (7), to our knowledge this is the first published case of hemoptysis due to telangiectasias of the oral and throat mucosa which furthermore was not visible on the first, outpatient ENT examination and led to hospital admission. In this case it is quite possible that the patient's previous activities, which included hot meals, alcohol and singing, triggered the bleeding from these lesions of the microvasculature.

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### References

1. SYSTEMIC SCLEROSIS: Current pathogenetic concepts and future prospects for targeted therapy. *Lancet* 1996; 347: 1453-8.
2. STEEN VD: Clinical manifestations of systemic sclerosis. *Semin Cutan Med Surg* 1998; 17: 48-54.
3. KALLENBERG CG, WOUDE AA, HOET MH, VAN VENROOIJWJ: Development of connective tissue disease in patients presenting with Raynaud's phenomenon. *Ann Rheum Dis* 1988; 47: 634-41.
4. KYNDT X, LAUNAY D, HEBBAR M *et al.*: Influence of age on the clinical and biological characteristics of systemic scleroderma. *Rev Med Interne* 1999; 20: 1088-92.
5. DUCHINI A, SESSOMS SL: Gastrointestinal hemorrhage in patient with systemic sclerosis and CREST syndrome. *Am J Gastroenterol* 1998; 93: 1453-6.
6. NEWMAN ED, HARRINGTON TM, AMOROSO A: Hemoptysis secondary to respiratory tract telangiectasias in CREST syndrome. *J Rheumatol* 1998; 15: 1874-6.
7. CHEN DA, JOHNSON JT: Epistaxis secondary to CREST syndrome. *Trans Pa Acad Ophthalmol Otolaryngol* 1985; 37: 194-5.