Raynaud's Phenomenon with Incomplete Features of Scleroderma; A Wolf in Sheep's Clothing and a Possible New Indication for Pulmonary/Heart Graft

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Raynaud's phenomenon associated with incomplete features of scleroderma is generally agreed to be a minor benign form of systemic sclerosis, into which it may evolve during the long-term follow-up [1,2]. We report here 2 recent cases with fatal pulmonary hypertension of sudden onset.

Case 1

The first patient was a 49-year-old female. She had complained of Raynaud's phenomenon for 35 years and had minor features of scleroderma consisting of a few telangiectasias, one episode of small fingertip necrosis, giant capillaries at the nail fold and antinuclear auto-antibodies. She had no sclerodactyly. The patient developed pulmonary hypertension with systolic pressure of the pulmonary artery of 80 mm Hg. A pulmonary/heart graft was proposed, but she refused it and died in 1990 despite nifedipine therapy.

Case 2

The second patient was a 53-year-old female who had complained of Raynaud's phenomenon for 7 years and had minor features of scleroderma consisting of telangiectasias, puffy hands, small fingertip necrosis, oesophagitis, antinuclear auto-antibodies and giant capillaries at the nail fold. She had no sclerodactyly. In 1988, she developed pulmonary fibrosis and was treated with D-penicillamine. In 1991, she developed pulmonary hypertension of sudden onset with systolic pressure of the pulmonary artery of 80 mm Hg. She was strongly encouraged to accept a pulmonary/heart graft, but her family refused and she died 3 months later. It has been reported that Raynaud's phenomenon is associated with poor survival in patients classified as cases of primary pulmonary hypertension [3], and in this respect, the 2 present cases have several implications: (1) vital prognosis of scleroderma is not always connected with the extent of the sclerodermatous skin changes; (2) Raynaud's phenomenon with incomplete features of scleroderma has the same prognosis as the CREST syndrome variant of systemic sclerosis as regards the risk of severe pulmonary hypertension [4]; (3) patients with primary pulmonary hypertension should be screened for features of scleroderma; (4) pulmonary hypertension should be routinely evaluated using echography coupled with Doppler techniques in patients with Raynaud's phenomenon and incomplete features of scleroderma; (5) when severe pulmonary hypertension occurs in patients with Raynaud's phenomenon and incomplete features of scleroderma, they should be encouraged to accept a pulmonary/heart

graft, and (6) pulmonary/heart graft departments should be encouraged to consider this pathology as an indication for these grafts.

References

Priollet P, Vayssairat M, Housset E: How to classify Raynaud's phenomenon. *Am J Med* 1987;83:494–498.

Fitzgerald O, Hess EV, O'Connors GT, Spencer Green G: Prospective study of the evolution of Raynaud's phenomenon. *Am J Med* 1988;84:718–726.

D'Alonzo GE, Barst RJ, Ayres SM, et al: Survival in patients with primary pulmonary hypertension: Results from a national prospective registry. *Ann Intern Med* 1991;115: 343–349.

Stupi AM, Steen VD, Owens GR, Barnes EL, Rodnan GP, Medsger TA Jr: Pulmonary hypertension in the CREST variant of systemic sclerosis. *Arthritis Rheum* 1986;29:515–524.