CREST Syndrome with Pulmonary Arterial Hypertension

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A 79-year-old woman was admitted to our hospital with effort dyspnea. Plain radiographs of the chest showed an enlargement of the heart and pleural fluid (Picture 1). Echocardiography showed remarkable right heart loading (Picture 2). Pulmonary arterial hypertension (PAH) and right heart failure manifesting hepatomegaly, ascites, jugular venous congestion, and pretibial pitting edema were observed. PAH was characterized by remarkable pulmonary hypertension with a normal pulmonary capillary wedge pressure assessed by Swan-Ganz catheterization. She had Raynaud’s phenomenon, sclerodactyly, telangiectasia about 10 years previously. Calcinosis circumscripta was found around the fingers (Picture 3, arrows). Antinuclear and anti-centromere antibodies were revealed to be 320 fold. We diagnosed the patient as CREST (calcinosis, Raynaud phenomenon, esophageal motility disorders, sclerodactyly, and telangiecta-
sia) syndrome resulting in PAH. PAH has a very poor prognosis, though CREST syndrome without PAH is a benign variant of progressive systemic sclerosis (1).

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Reference

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