CREST syndrome with severe pulmonary hypertension

Zafeiris Sardelis,
Elpida Theodorakopoulou,
Epameinondas Kosmas

3rd Department of Pneumonology Medicine,
'Sotiria' Hospital for Chest Diseases, Athens

A female patient aged 79 years, who was an ex-smoker, presented with progressive shortness of breath during physical activity for several months, severe hypoxaemia (FiO_2=21%, PaO_2= 53mmHg, $PaCO_2$= 27mmHg, pH= 7,48, HCO_3= 20mmol/lt), and severe pulmonary hypertension (PASP= 65mmHg). From her past medical history, special consideration was given to symptoms consistent with Raynaud's syndrome, and difficulty in swallowing with retrosternal burning. **Immunological assays:** Anti-nuclear Ab **(ANA**) was positive, with a high titre of anti-centromere Ab **(**1/1320).

Based on these immunological findings, along with the evidence of Raynaud's syndrome, the radiological barium swallow study of the oesophagus (Figure 1), and the plain X-ray of the hands (Figure 2), a diagnosis of **pulmonary hypertension** secondary to **CREST syndrome** (**C**alcinosis cutis, **R**aynaud phenomenon, o**E**sophageal dysmotility, **S**clerodactyly, **T**elangiectasia) was made. This patient began treatment with bosentan, and presented a significant decrease in PASP to 45mmHg at 6 months follow-up.

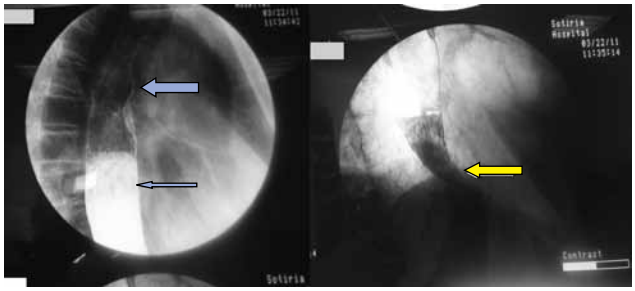

FIGURE 1. Oesophagogram with barium swallow → showing dilatation with absence of peristalsis at the lower third of the oesophagus (thin blue arrow), in contrast with the upper two thirds (thick blue arrow), and stricture of the lower oesophageal sphincter (yellow arrow).

Correspondence to:
Zafeiris Sardelis
3rd Department of Pneumonology Medicine
"Sotiria" Hospital for Chest Diseases
152 Mesogeion Avenue, 11527 Athens, Greece
e-mail: zafsard@yahoo.com

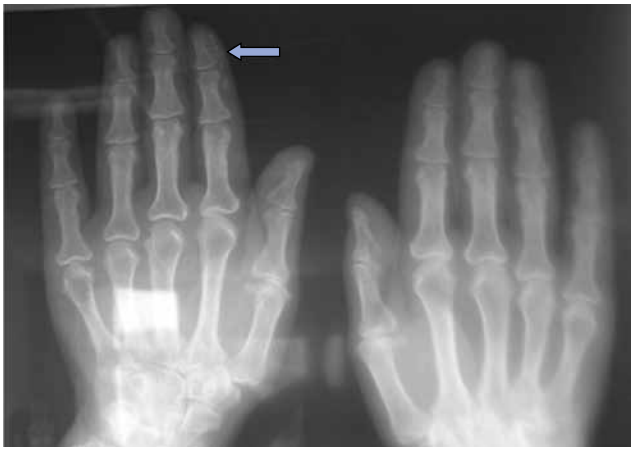

FIGURE 2. Plain X-ray of the hands → showing deposit of calcium crystals (arrow) in the skin around the distal phalanges of the fingers.