CREST Syndrome and Combined Pulmonary Fibrosis – Emphysema in Systemic Sclerosis (Scleroderma)

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50 y.o. male, ex-smoker (15p/y), with diffuse scleroderma under treatment with a variety of immunosupresive drugs from 2008. The patient presented with all the typical manifestations of scleroderma with multiple organ affection, pulmonary hypertension (RVSP: 45mmHg), CREST syndrome (calcinosis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly, telangiectasia) (Figure 1) and extensive telangiectasias (Figure 2 A, B, C).

The high-resolution computed tomography showed paraseptal empysema with ground glass and consolidation areas, findings that are compatible with NSIP in the limits of a combined pulmonary fibrosis – emphysema syndrome (Figure 3 A, B, C).

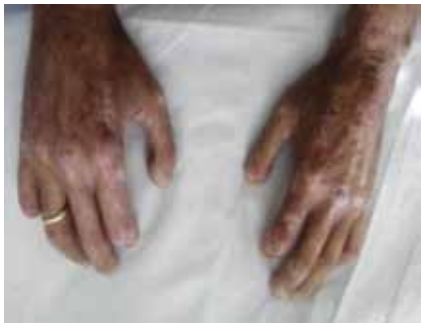

FIGURE 1.

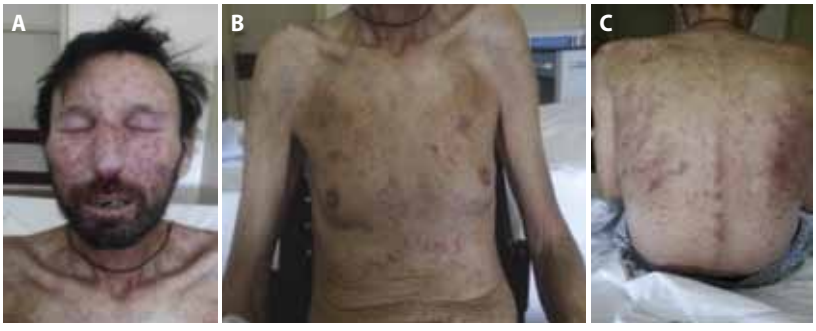

FIGURE 2.

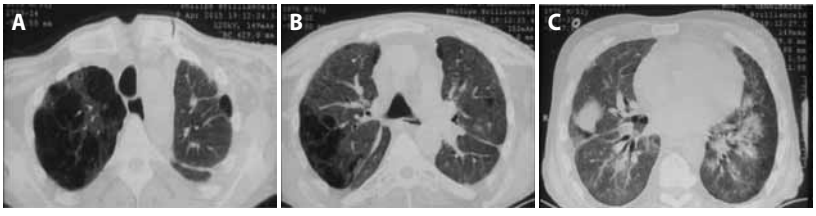

FIGURE 3.

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