A case of CREST syndrome.

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Patient: 58 years, female

Clinical History:

A case of CREST syndrome in a 58 woman.

Imaging Findings:

A 58 year old patient came to our attention with a remote history of Sjogren syndrome, calcinosis, Raynaud phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia (CREST syndrome). At the age of 28 during her second pregnancy she suffered from gestational diabetes and, a year later, she was diagnosed type 2 diabetes for which she started insulin therapy. At the age of 42 after experiencing hands and knees arthralgia, she was diagnosed with scleroderma and treated with steroids. Two years later in 1994, she was diagnosed with Hashimoto's thyroiditis resulting in hypothyroidism treated with Eutyrrox until now. In 2007, she was hospitalized for phlegmon of the left foot's first toe due to chronic microvascular disease. During hospitalization she underwent bilateral arteriography of the lower limbs that indicated obstruction of the posterior tibial and pedidia arteries and therefore started treatment with prostaglandin. A chest radiograph documented bilateral diffuse peri-articular soft tissue calcifications and bibasilar pulmonary fibrosis. Respiratory Function Test and the diffusion capacity of the lung for carbon monoxide (DLCO) were performed showing severe restrictive respiratory alterations and also a marked reduction of the CO diffusing capacity. This year because of the worsening of the respiratory condition, the patient received a repeat chest radiograph (Fig.1), that was similar to the one from 2007, so she underwent a thoracic high resolution computed tomography (HRCT) that pointed out the presence of a severe bilateral interstitial fibrosis (especially of the posterior-basal segment), thickening of the peribronchial-subpleuric interstitial tissues and several peri-articular soft tissue calcifications (Fig.2).

Discussion:

CREST syndrome (calcinosis, Raynaud phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia) is a member of the heterogeneous group of scleroderma, or systemic sclerosis, a chronic connective tissue disease classified as one of the autoimmune rheumatic diseases. Three primary pathologic features are found in scleroderma and include increased collagen deposition, perivascular mononuclear cell infiltration, and vascular abnormalities. Mononuclear infiltration probably precedes fibrosis of tissues. While this inflammatory infiltrate can accompany fibrosis in tissues, it can also be present without fibrosis, suggesting that it is an early event in the pathogenesis of scleroderma. Ischemia is an important contributor to end organ damage in scleroderma patients. While the primary trigger for CREST syndrome is not known, a reasonable speculation is that vascular endothelial cell abnormalities incite mononuclear infiltration, resulting in abnormal fibroblast activity and increased collagen deposition. The incidence of systemic sclerosis in the USA is approximately 3-19 new cases per million adults p.a. The prevalence is 250-290 cases per million persons. Females have a greater incidence of scleroderma than males. The usual age of onset of scleroderma is 30-65 years. In the typical course of limited scleroderma, the patient first notices Raynaud phenomenon. Over the years, fingers become puffy, then the skin thickens slowly. Internal organ manifestations are delayed for many years. Calcinosis may be subclinical, but when symptomatic, tissues become
tender and painful. Patients find that the advancement of skin disease occurs slowly, over many years. While the entire intestine can be involved in scleroderma, esophageal involvement is most common and clinically relevant. Barrett oesophagitis and oesophageal adenocarcinoma have been found in scleroderma patients, perhaps at a higher rate. In CREST patients, telangiectases occur on the face, upper trunk, hands and also on mucosal surfaces (e.g. lips), throughout the GI tract in association with recurrent GI bleeding that can be chronic and cause anaemia. Autonomic dysfunction of the GI tract is also described. Other manifestations are: pulmonary hypertension in the absence of interstitial fibrosis, as a very late event; myocardial involvement; entrapment neurologic syndromes; sicca symptoms in approximately 35% of patients. CT scan can be performed in the prone position. The initial alterations on plain thoracic radiograph may be subtle with only a fine reticulation in the lower lobes, then it becomes thicker and diffuse. High Resolution Computed Tomography (HRCT) shows radiographical signs of interstitial disease, also in asymptomatic patients, for example, parenchymal and subpleural micronodules, intralobular linear opacities, ground glass and honey comb pattern lesions. CT scan also demonstrates the dilatation of the esophagus due to atrophy and atonia and air-fluid level.
The differential diagnosis includes systemic lupus erythematosus, polymyositis, and rheumatoid arthritis. Lupus erythematosus is rarely associated with chronic pulmonary fibrosis; up to 20% of patients with rheumatoid arthritis and 10% of those with polymyositis develop pulmonary fibrosis superimposable of the one in CREST. Scleroderma is associated with an increased risk of cancer, in particular lung cancer.
The survival rate from time of diagnosis was computed to be 78% at 5 years, 55% at 10 years, 37% at 15 years, and 26% at 20 years.

**Differential Diagnosis List:** Pulmonary fibrotic disease in a CREST syndrome.

**Final Diagnosis:** Pulmonary fibrotic disease in a CREST syndrome.

**References:**

High-Resolution CT of the Lungs
Ella A. Kazerooni1
1 Department of Radiology, 2910 Taubman Center, University of Michigan Medical Center, 1500 E. Medical Center Dr., Ann Arbor, MI 48109-0326.
K. VEERAPEN, I. WATT, and P. DIEPPE
SEVERE SUBCUTANEOUS CALCIFICATION IN THE ‘CREST’ SYNDROME: EVIDENCE OF HIGH TURNOVER OF CALCIFIC DEPOSITS
Rheumatology, April 1987; 26: 89 - 92.
Description: Chest radiograph shows diffuse subcutaneous eggshell calcifications, located in the infraclavicular region, widespread on the thoracic wall and surrounding the shoulders. Origin:
Description: The radiograph also shows interstitial fibrosis especially of the lower and posterior lobes, with emphysema. Origin:
Figure 2

Description: High resolution computed tomography (HRCT) rules out severe bilateral interstitial fibrosis with thickening of the peribronchial-subpleuric interstitial tissues, bronchiectasies and cystic lesions especially of the posterior and basal regions. The exam shows the diffuse pattern of soft tissues calcifications located around the thoracic wall and shoulders and also the dilatated esophagus. **Origin:**
Description: Origin:

b

c

description: origin: