

CONTINUING MEDICAL EDUCATION ΣΥΝΕΧΙΖΟΜΕΝΗ ΙΑΤΡΙΚΗ ΕΚΠΑΙΔΕΥΣΗ

Medical Imaging Quiz – Case 29

A 45-year-old woman with a four year history of scleroderma under immunosuppressive treatment presented to our department due to shortness of breath and feeling of heartburn. She denied any respiratory symptoms before this episode. Patient was known diabetic for 3 years, and hypertensive for 2 years. She mentioned no imaging workup in the last three years. Her vital signs upon admission were: heart rate 80 bpm, blood pressure 120/50 mm/Hg, respiratory rate 20/min. She was afebrile and oxygen saturation was 98% on room air. Physical examination revealed mild wheezing in basal lung, sclerodactyly, tightness and thickness of the skin on her hands and telangiectasia. Breast palpation revealed multiple, non painful hard lumps with no axillary lymphadenopathy. Her total blood count test showed WBC 4.800/ μ L, HCT 30%, platelets 148,000/ μ L, ESR 135 mm/hour. Chest X-ray showed few bilateral lower lobe interstitial infiltrates. Incidental and conspicuous finding though, were coarse diffuse calcifications in both breasts (figures 1–3). Chest CT scan was performed after intravenous administration of contrast media. Findings included areas of ground glass attenuation with interlobular septal thickening in lung bases, multiple diffuse dense breast calcifications with relatively smooth margins (fig. 4), and dilatation of the inferior third of the esophagus (fig. 5).

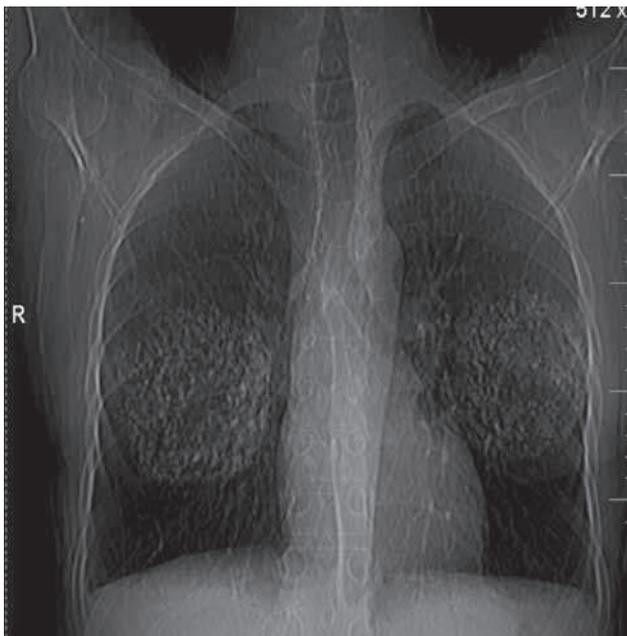


Figure 1. Chest X-ray shows diffuse calcifications in both breasts.

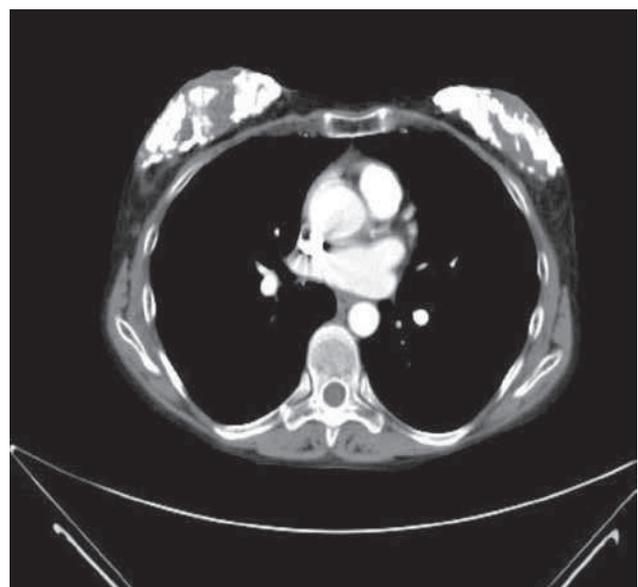


Figure 2. Chest CT scan shows dense dystrophic breast calcifications.

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Comment

Dystrophic calcification (DC) is the calcification occurring in degenerated or necrotic tissue as a reaction to tissue damage. It can occur even if the amount of calcium in the blood is not elevated. Basophilic calcium salt deposits aggregate, initially in the mitochondria and progressively throughout the cell. These calcifications concern an indication of previous microscopic cell injury. Dystrophic calcification is associated with a various range of disorders, including connective tissue diseases, inherited disorders, cutaneous neoplasms, and infections. In collagen vascular diseases such as scleroderma, dermatomyositis, and systemic lupus erythematosus, diffuse dystrophic calcifications of the subcutaneous fat may be seen that progressively coarsen over time.

Scleroderma is an autoimmune connective tissue disease whose hallmark is excessive collagen deposited around capillaries and in affected tissues, such as the skin, lungs, kidneys, and esophagus. The etiology is unknown, but its pathogenesis involves an activated immune system, abnormal vascular endothelium, and an exagger-



Figure 3. Breast ultrasonography shows diffuse coarse irregular calcifications in the subcutaneous fat of the breast.



Figure 4. Chest CT scan shows basal ground glass infiltrates with interlobular septal thickening.

ated production of fibroblasts that results in abnormal collagen build up. This disease is found among all races worldwide, but women are four times more likely to develop scleroderma than men. Most adults are diagnosed after their 30th birthday and before age of 50 years. Limited systemic sclerosis/scleroderma involves cutaneous manifestations that mainly affect the hands, arms, and face. It was previously called CREST syndrome in reference to the following complications: Calcinosis, Raynaud’s phenomenon, esophageal dysfunction, sclerodactyly, and telangiectasias. Diffuse systemic sclerosis/scleroderma is rapidly progressing and affects a large area of the skin and one or more internal organs, frequently the kidneys,

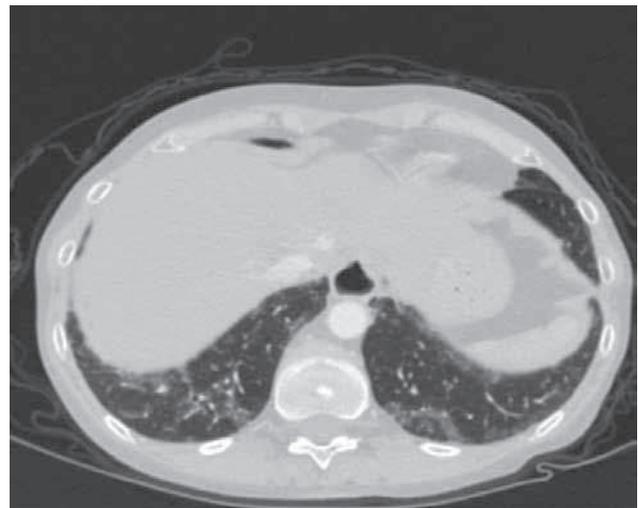


Figure 5. Chest CT scan shows dilatation of the esophagus.

esophagus, heart, and lungs. This form of scleroderma can be quite disabling. Other forms of scleroderma include systemic sine scleroderma (which lacks skin changes but has systemic manifestations) and two localized forms, morphea and linear scleroderma, which affect the skin but not the internal organs. There is no direct cure for scleroderma. Because the exact cause is unknown, any treatment is patient-specific and aimed at treating complications of individual organ system and ameliorating symptoms of the disease. Since it is an autoimmune disease, one of the major pillars of treatment involves the use of immunosuppressive agents. These drugs include methotrexate, cyclophosphamide, azathioprine, and mycophenolate. The prognosis is generally good for limited cutaneous scleroderma patients who escape pulmonary complications, but is worse for those with the diffuse cutaneous disease, particularly in older age, and for males. Death occurs most often from pulmonary, heart, and kidney complications. In diffuse cutaneous disease, five-year survival is 70%, and 10-year survival is 55%.

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Diagnosis: Dystrophic breast calcification in scleroderma