

Pregnancy Triggered Onset Diffuse Cutaneous Scleroderma

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Systemic sclerosis (SSc), is a rare autoimmune disease that causes vasculopathy and extensive fibrosis of the internal organs and skin. The disease can present in different forms. Diffuse cutaneous scleroderma (DcSSc) is the most aggressive type that has a relatively poor prognosis. It is characterized by progressive diffuse thickening of the skin, fibrosis of the lungs, heart, and kidneys.

A 40 year old Haitian female with a history of obesity, uncontrolled hypertension, presents with fever, shortness of breath, orthopnea, and productive cough with yellow sputum over the last two days. Patient is concerned about skin tightness that started shortly after the birth of her second child two years ago.

On admission to ED the patient was tachypneic (RR 42), tachycardic (HR 116), febrile (102.1F), normotensive (BP 136/82), and hypoxemic (O2 saturation 60's). On a physical exam, the patient was noted to have fine bibasilar crackles on auscultation. Diffuse thickening of the skin along with hyperpigmentation was noted on the areas proximal to elbows, knees, anterior trunk, and face. Assessment for scleroderma, pneumonia, and heart failure was initiated.

Significant laboratory findings included: WBC 15.63, Eosinophils 8.5, fibrinogen 700, ESR 100, CRP 14, LDH 428. Sputum culture was positive. Serology was positive for antibodies: ANA, RF, anti-DNA topoisomerase I (scl70), anti-dsDNA, and anti-U3-RNP (fibrillarin). Chest x-ray showed diffuse bilateral infiltrates, and enlarged cardiac silhouette. Pulmonology was consulted and they suspected pulmonary hypertension related to diastolic dysfunction which was confirmed on echocardiogram. High resolution CT revealed extensive ground glass opacities, diffused reticulations, and extensive traction bronchiectasis with areas of interlobular septal thickening.

The diagnostic criteria was met for usual interstitial pneumonia with idiopathic pulmonary fibrosis (ILD Rads 4). In light of these findings, diagnosis of diffuse cutaneous scleroderma was confirmed. Methylprednisolone was started. Patient was cleared for discharge with close outpatient follow-up.

Learning Objectives:

- Discuss diagnostic criteria of diffuse cutaneous scleroderma.
- Describe the most effective way in management of the disease.