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Thoracic Calcinosis Cutis in a Case of Dermatomyositis

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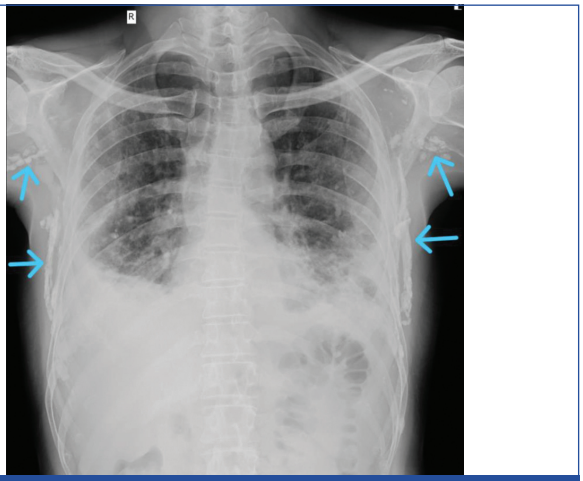
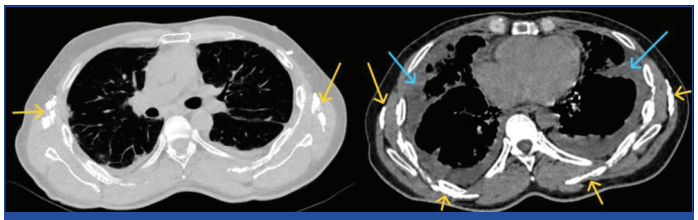
A 42-year-old man presented at the outpatient department with a two-month history of cough accompanied by mucoid expectoration, as well as a gradual onset of breathlessness rated at Modified Medical Research Council (MMRC) Grade 1 to 2 over the past five years, progressing to Grade 3 in the last two months. Additionally, he reported intermittent joint pain, loss of appetite, and a progressive weight loss of approximately 20 kilograms over the course of five years. The cough tended to worsen in the mornings but improved throughout the rest of the day. He experienced stiffness in the small joints of his hands, which persisted throughout the day. He also mentioned a previous occurrence of maculopapular rash, a month earlier to the onset of respiratory symptoms, on his hands and face, although no lesions were currently present. Despite seeking treatment in the past including irregular use of both allopathic and ayurvedic medicines, his symptoms had not been alleviated. He denied any seasonal variation in symptoms, allergies, comorbidities, or past known diseases and reported being a non-smoker. Upon examination, the patient was found to be underweight with a Body Mass Index (BMI) of 17 kg/m^2. Vital signs were within normal limits, with a pulse rate of 82/min, blood pressure of 130/80 mmHg, respiratory rate of 22 breaths/min, and oxygen saturation of 94% on room air. Auscultation revealed bilateral crackles in the infrascapular, infra-axillary, and mammary areas. No other significant abnormalities were noted.

Blood tests revealed elevated Lactate Dehyrogenase (LDH) levels (300 U/L), while Antinuclear Antibody (ANA) test was negative. Serum calcium, phosphorous, and creatine phosphokinase levels were within normal range. All other serological panels with respect to connective tissue, autoimmune diseases, HIV, Hepatitis-B and Hepatitis-C were negative. Chest X-ray showed bilateral patchy infiltrates in the lower and mid-zones with blunting of the costophrenic angles and bilateral linear subcutaneous calcifications in the thoracic wall and proximal part of the upper limbs [Table/Fig-1]. Spirometry revealed a restrictive pattern with Forced Vital Capacity at 64% of the predicted value. High-Resolution Computed Tomography (HRCT) of the thorax demonstrated peripheral, bilateral lower lobe subpleural reticular shadows and septal thickening, with additional findings of ground glass opacities, predominantly in the right middle and lower lobes. Fascial calcifications were noted along the posterolateral chest wall [Table/Fig-2]. Abdominal and pelvic CT scans showed no significant abnormalities besides soft-tissue and muscle calcifications. X-rays of bilateral femurs showed soft-tissue calcifications. The above-mentioned radiological scans did not show any evidence suggestive of malignancy. Echocardiography results were within normal limits. Skin biopsy confirmed Dermatomyositis (DM), showing lymphocytic infiltrate with mucin deposition in the reticular dermis, mild irregular epidermal hyperplasia, and smudged dermo-epidermal junction. Magnetic Resonance Imaging (MRI) of the thighs and pelvis revealed muscle signal alterations and intermuscular fluid. Subsequent therapy with oral prednisolone, methotrexate, and hydroxychloroquine showed no improvement, leading to initiation of Mycophenolate Mofetil (MMF) at 1 g/day, resulting in reduced joint pain and symptom frequency. The patient was discharged on maintenance therapy with MMF, which led to symptomatic improvement, and he remained stable at the one-month follow-up.



[Table/Fig-1]: Chest X-ray showing bilateral patchy infiltrates in the lower and mid-zone with blunting of bilateral costo-phrenic angles; there is also a linear margin of calcification (blue arrows) in the subcutaneous plane on both sides depicting calcinosis cutis.



[Table/Fig-2]: High-resolution computed tomography of the thorax showing peripheral, bilateral lower lobes, subpleural reticular shadows, and septal thickening along with bilateral pleural effusion (blue arrows). Bilateral diffuse fascial calcification (yellow arrows) predominantly along the posterolateral chest wall deep to the chest wall muscles, along the fascia between the muscles and subcutaneous fat inferior to the axilla.

DM, an autoimmune inflammatory disease affecting skin and muscles, is characterised by progressive symmetrical proximal muscle weakness and a typical skin rash, varying in intensity. Systemic manifestations often involve the pulmonary and gastrointestinal systems, and there's some association with internal malignancy too [1]. Calcinosis cutis is a persistent condition characterised by the accumulation of insoluble calcium deposits in the skin and subcutaneous tissue. This condition affects 20-40% of individuals with Juvenile Dermatomyositis (JDM) [2,3] and up to 20% of those with adult DM. Currently, there are no universally effective treatments for calcinosis; however, immunosuppressive therapies might be used to prevent its development and progression [4-6].

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