# Severe Calcinosis Cutis with Cutaneous Ulceration in Juvenile Dermatomyositis

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**Background:** Calcinosis cutis is usually seen in long standing and untreated cases of juvenile dermatomyositis. **Case characteristics:** 7-year-old girl with severe calcinosis cutis who developed cutaneous ulceration, rash and myopathy. **Observation:** Myopathic changes in EMG, muscle edema in MRI, elevated muscle enzymes and Jo-1 positive antibodies. **Outcome:** Treatment with prednisolone and methotrexate resulted in improvement of the lesion. **Message:** Calcinosis cutis may be a presenting feature of juvenile dermatomyositis even in the absence of characteristic findings of rash and weakness.

**Keywords**: *Calcification, Dermatomyositis, Diagnosis*.

Juvenile dermatomyositis (JDM) is a rare multisystem autoimmune disorder with an incidence of 3 per million population [1]. It is characterized early in its course by perivascular inflammation of striated muscles and skin, and later by development of calcinosis. Classic JDM manifests with an insidious progression of malaise, easy fatigue, muscle weakness, fever and rash that may appear before diagnosis by 3 to 6 months [2]. Lipodystrophy and calcinosis are well described in JDM and are more commonly seen in long standing or neglected disease. We report a case of juvenile dermatomyositis who presented with extensive calcinosis cutis with ulceration long before development of characteristics rash and weakness.

## CASE REPORT

A 7-year-old girl born out of non-consanguineous marriage presented to us with multiple tender nodular swelling at the back of both thigh with overlying deep ulcers at the back of right thigh. At 6 years of age, she had pain in both thighs more on right side – for which she had consulted physicians, neurologist, rheumatologist and dermatologist. The pain was dull aching in nature, not related to physical activity, and was associated with limping on the right side. She had multiple nodular swelling on thighs and arms. Investigations revealed normal blood counts, high erythrocyte sedimentation rate (20 mm in 1$^{st}$ hr.), raised c-reactive protein (25 mg/dL), low vitamin D3 (17.95 ng/mL), normal calcium and phosphorus, normal thyroid function tests, normal creatine kinase (94 U/L), normal C3 (126 mg/dL), normal C4 (40 mg/dL), negative antinuclear antibody by EIA (0.13) and negative anti-ds DNA (2 U/mL). She was treated conservatively and started vitamin D supplementation (60000 IU/week). The patient did not seek any medical advice for the next 6 months.

On presentation to our hospital, the patient had multiple tender nodular swellings with reddish brown pigmentation on the back of both thighs with ulcerations on right side ***(Fig.* 1**). Due to pain and restriction of movements, muscle weakness in the lower limbs could not be elicited. Violaceous rash suggestive of heliotrope rash was visible on the eyelids, and multiple bright pink shiny thickened plaques on the metacarpophalangeal joints and proximal interphalangeal joints suggestive of Gottron papules were seen. Provisional diagnosis of juvenile dermatomyositis was made and wound debridement was done. Pus culture was positive for *Staphylococcus* and histopathology revealed necrotic granulomatous inflammation with foci of calcifications. Appropriate antibiotic was started with regular dressing.

Her investigations revealed normal blood counts, raised erythrocyte sedimentation rate (44 mm in 1 hr), raised aspartate transaminase (92U/L), raised serum LDH (681U/L, normal 313-618), normal alanine transaminase (44 U/L), normal serum creatine kinase (75 U/L), normal anti-phospholipid antibody (IgG 2.4 GPLU/mL, IgM 2.5 MPLU/mL), normal renal function tests and normal ionized calcium (1.25mmol/L), phosphate (5.6 mg/dL) and alkaline phosphate (600 U/L). Doppler ultrasound of limbs showed normal arterial and venous blood flow and multiple echogenic lesions in subcutaneous tissue of both thighs and legs. Electromyography (EMG) showed features of inflammatory myopathy. Plain radiograph of limbs showed multiple linear and stippled calcifications in the subcutaneous tissue in all limbs, more marked in the lower limbs ***(Fig.* 1**). Magnetic Resonance Imaging (MRI) revealed ill-defined high signal in Short Tau inversion recovery (STIR) sequences suggestive of muscle edema in bilateral thigh regions and few low signal foci suggestive

of calcifications (***Fig.* 2**). Antinuclear antibody (ANA) was positive: speckled 1 and ANA profile (ELISA) revealed strongly positive for Anti-Jo-1 antibody. Chest radiograph and high resolution computed tomograhy (HRCT) thorax did not show any features of interstitial lung disease. A diagnosis of Juvenile dermatomyositis was considered in the presence of pathognomic rash, myopathic changes in electromyogram (EMG), muscle edema in magnetic resonance imaging (MRI), elevated muscle enzymes and supported by calcinosis cutis and Jo-1 positive antibodies.

Prednisolone was started at a dose of 1 mg/kg/day. Follow-up after one month of treatment revealed healing of one ulcer with presence of well vascularized granulation tissue in the base of another ulcer and disappearance of the nodules. Radiograph showed partial resolution of the calcinosis. Oral methotrexate 10 mg/ week along with folate supplementation was started in addition to prednisolone.

## DISCUSSION

Subcutaneous calcifications may occur due to several rheumatological conditions (Juvenile dermatomyositis, Systemic lupus erythematosus, and Systemic sclerosis), infections or local tissue traumas [3,4]. In JDM, calcinosis is less frequently present at diagnosis, reported in only 3% to 23% of patients [5]. When calcinosis develops before weakness, pain and stiffness due to calcinosis cutis masks the weakness on clinical examination, but EMG and MRI can differentiate both. In our case, myopathy in EMG was detected after 7 months of calcinosis. Calcinosis cutis in JDM may be related to severity of disease, delayed initiation of treatment and potentially to genetic polymorphisms of TNF-$\alpha$-308. It is thought to be associated with longstanding or undertreated disease [1]. Calcium deposition may occur in subcutaneous plaques or nodules, as large tumorous deposits in muscle groups, as calcification within fascial planes, bridging joints, or as an extensive subcutaneous exoskeleton [6]. Some ulcerate through the skin and drain a soft calcific liquid, and others manifest as hard nodules along extensor surfaces or embedded along muscle [1]. Our case presented with indurated nodule and extensive subcutaneous calcification which subsequently developed ulceration in the back of thigh, typical rash (Gottron papules and heliotrope rash) and EMG feature of myopathy suggestive of JDM. Similar observation was made by Wananukul, *et al*. who reported calcinosis cutis in two cases years before appearance of other clinical manifestations of JDM [7].

Calcinosis cutis may be a presenting feature of juvenile dermatomyositis and should be thought of even in the absence of characteristic findings of rash and weakness so that early and effective therapy can be instituted to prevent progression.

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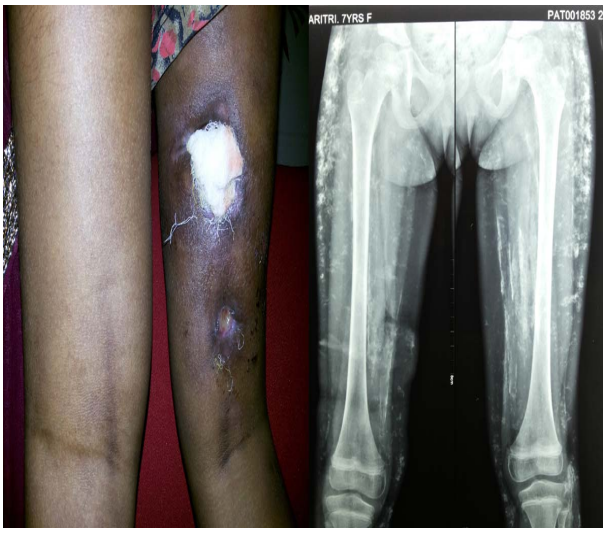


**FIG. 1** *Non-healing ulcer on lower limbs and X-ray showing extensive subcutaneous calcifications.*

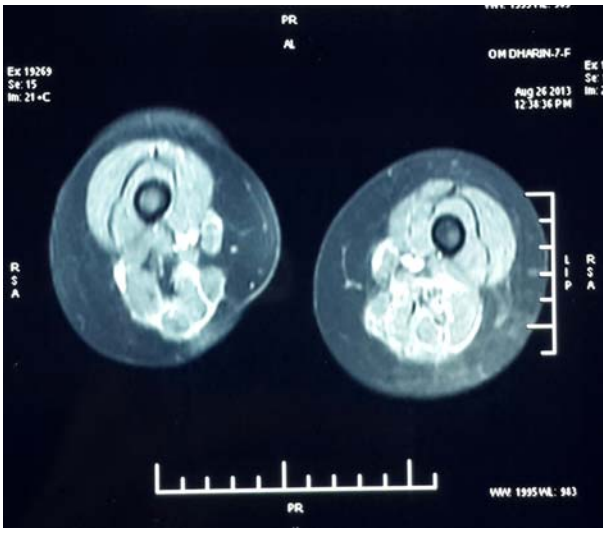


**FIG. 2** *MRI of thighs showing ill-defined high signal in STIR sequences suggestive of muscle edema in both thighs and few low signal foci suggestive of calcifications.*

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