

# Calcinosis Cutis with Occult Abscess Complicating Juvenile Dermatomyositis

## Juvenil Dermatomiyozi Komplikasyonu Edinen Kalsinosis Kutis ile Beraber Apse

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#### Abstract

Juvenile dermatomyositis is a chronic inflammatory disorder of unknown etiology that affects primarily skin and muscle. Calcinosis is a common and debilitating complication of juvenile dermatomyositis, with an incidence of 30 % to 70 %, contributing morbidity and mortality of disease. The well-known risk factors for calcinosis include delayed treatment and severe disease. Calcium deposits were first noticed at a median of 3.4 years after onset of juvenile dermatomyositis. In this case report we present a child with early development of calcinosis and accompanying multifocal abscess worsening the clinical situation. We want to emphasize with our case the importance to recall occult infections, when there is increment of the patient's complaints even the classical infectious findings are lacking.

Key Words: **Abscess; Calcinosis; Dermatomyositis.**

#### Özet

Juvenil dermatomiyozi cildi ve kasları tutan, sebebi bilinmeyen kronik bir inflamatuvar hastalıktır. Kalsifik depozitler hastaların % 30 ile % 70'inde görülerek hastalığın morbidite ve mortalitesine katkıda bulunmaktadır. Gecikmiş tedavi ve şiddetli hastalık kalsinoz için iyi bilinen risk faktörlerindedir. Kalsiyum birikiminin juvenil dermatomiyozi başladıktan ortalama 3.4 yıl sonra geliştiği bildirilmiştir. Biz bu vakada erken kalsinozis bulgularına eşlik eden ve primer hastalığı ağırlaştırıcı multifokal absesleri olan bir hastayı sunduk. Bu vaka sunumuyla, enfeksiyonun klasik bulguları olmasa da hastaların semptomlarında artış olduğunda, gizli bir enfeksiyon odağının araştırılması gerektiğini hatırlatmak istedik.

Anahtar Sözcükler: **Apse; Dermatomiyozi; Kalsinozis.**

## Introduction

Juvenile dermatomyositis (JDM) is a chronic inflammatory disorder of unknown etiology that affects primarily skin and muscle. It is the most common pediatric myopathy, affecting approximately 3.1 children/million (1). Calcinosis is a common and debilitating complication of juvenile dermatomyositis, with an incidence of 30% to 70% (2). It is also a hallmark of the disease, occurring mainly in pediatric patients. The known risk factors for calcinosis include delayed treatment and severe disease (3). Cutaneous calcinosis is frequently located on the elbows, knees and other acral parts, and may cause significant debility with severe pain, joint contracture, skin ulcers and muscle atrophy (4,5).

In this case report there is relatively early developed cutaneous calcification and widespread staphylococcal abscesses. The complaints disappeared when the infection was treated. With this case report, we want to emphasize the importance to recall occult infection, when there is increment of the patient's complaints even the classical infectious findings are lacking.

## Case report

A fourteen year old boy was referred to our center with a four year history of weakness and morning stiffness of about 30 minutes. His initial symptoms were misdiagnosed as rheumatoid arthritis and therapy with methotrexate, corticosteroids and chloroquinone failed. Color change

in his tip of fingers as firstly white and then purple in cold weather, and cutaneous findings over his left knee joint were the following complaints. He had difficulty in walking, for two years. Two months before this presentation, his skin biopsy over his knee revealed calcinosis cutis and he was diagnosed as dermatomyositis. His temperature was 37 °C on the admission to hospital. He had desquamated ulcerations over the joints for a week. In his physical examination, he had abscess formation behind his left knee, white shiny scars over elbows due to the old ulcerations, he had joint contractures over his knees, elbows and ankles and other systemic examination was normal. Initial studies revealed a total white blood count of  $9.9 \times 10^9/L$ , hemoglobin level of 9.9 g/dL, platelet count of  $269 \times 10^9/L$ , erythrocyte sedimentation rate of 77 mm/hour, C reactive protein level of 7.6 mg/dL. On X-rays of the extremities (Figure 1 and 2), there was calcified material in connective tissues all over the body, his calcium and phosphate levels were normal. Skin biopsy from white scars revealed 'calcinosis cutis'. Magnetic resonance imaging (MRI) of the left knee showed a huge abscess formation (280x30mm) (Figure 3).

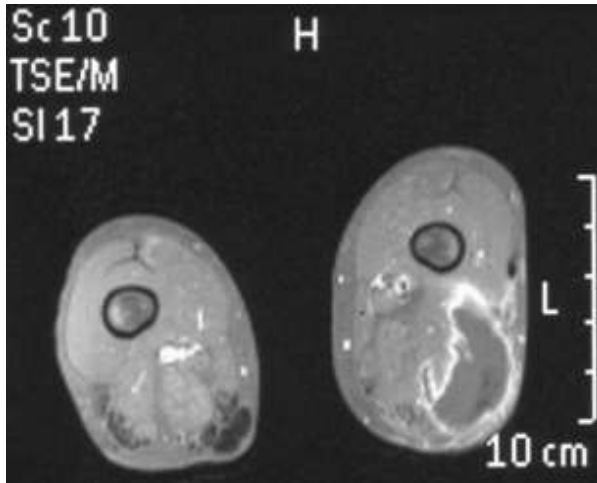
From the pus material *Staphylococcus aureus* was cultured. Under the combined therapy of sulbactam-ampicillin and clindamycin for 21 days, his abscess was drained on the third day. After the operation he gained his normal activity with physiotherapy.



**Figure 1.** Cutaneous calcification in thigh region(X-ray)



**Figure 2.** Lateral view of cutaneous calcification in thigh region (X-ray)



**Figure 3.** Abscess formation in left knee region (MRI)

### Discussion

Juvenile dermatomyositis is frequently complicated by cutaneous calcinosis. Ectopic calcification in JDM is thought to develop through a dystrophic mechanism, whereby damaged muscle releases mitochondrial calcium into matrix vesicles, which then promote mineralization (6). Another suggestion for the mechanism of calcification is that denaturated proteins preferentially bind phosphate ions, which in turn react with calcium ions to form a precipitate of calcium phosphate (7). Histological study of the lesions shows hydroxyapatite accumulation rather than bone (8). Serum calcium and phosphate levels are reported to be normal. Calcification associated with DM has been categorized into five different subtypes: Small and hard plaques or nodules that can be felt just below the skin surface; Large tumorous deposits of calcium, which often appear 'popcorn like' on X-ray examination; Deposits in the intermuscular fascia with limitation of movement in the involved muscle group; A severe form of dystrophic calcification, which resembles an exoskeleton; and a mixed form of calcinosis (9).

Patients with JDM are on increased risk for developing infections (10). In case series with JDM infectious complications have been described in up to %30 (11-14). The increased risk for developing infections is the result of immune abnormalities and organ system manifestation associated with this disease and treatment with immunosuppressive medications (15).

Calcinosis frequently described in the childhood form of JDM, represents as a predisposing factor for the development of staphylococcal soft tissue and dermal

infections due to *S. aureus* in the area of calcinotic region (16). Panniculitis and fasciitis caused by *S. aureus* is very rare, and it is strongly advised that the possibility of infection must be ruled out via biopsy and tissue culture before increasing the immunosuppressive regimen in children with presumed autoimmune panniculitis (17). Classical findings of infectious disease such as fever, fatigue, etc. might be absent in these patients because of their treatment or relative immunosuppression so, index of suspicion must be high in them especially. In our case, laboratory findings are not enough for differentiation between the reactivation of JDM and infection, so invasive procedures are necessary for exact diagnosis in suspicious cases. In cases of increment of patient's complaints, occult infection spots should be investigated. Also diffuse cutaneous calcification and pubertal age of the patient might have facilitated multiple abscess formation due to staphylococcus aureus.

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