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A novel presentation of calcinosis cutis of the scalp in adult dermatomyositis

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Abstract

Dystrophic calcinosis cutis is the aberrant deposition of insoluble calcium in cutaneous tissue generally secondary to inflammatory connective tissue disease. Although calcinosis cutis is commonly seen in juvenile dermatomyositis, it is a relatively rare occurrence in adult disease. Herein, we discuss an 82-year-old woman with extensive history of dermatomyositis of the scalp who presented with new-onset calcinosis cutis of the scalp.

Keywords: calcinosis cutis, dermatomyositis

Introduction

Calcinosis cutis is characterized by the deposition of insoluble calcium salts in the skin and subcutaneous tissue. The syndrome is separated into 5 subtypes: dystrophic calcification, metastatic calcification, idiopathic calcification, iatrogenic calcification, and calciphylaxis. Dystrophic calcification appears with aberrant tissue architecture as a result of local tissue damage and serum calcium and phosphate levels are normal. Metastatic calcification is characterized by an abnormal calcium and/or phosphate metabolism. Idiopathic calcification occurs without any underlying tissue damage or metabolic disorder. Skin calcification in iatrogenic calcinosis cutis is often a side effect of therapy. Calciphylaxis presents with small vessel calcification primarily of the dermis or subcutaneous fat [1,2]. Of these subtypes, dystrophic calcinosis cutis is the most common, and it is most frequently seen in association with underlying autoimmune connective tissue disease, particularly systemic sclerosis and dermatomyositis [1,3].

Dermatomyositis is an inflammatory myopathy with characteristic cutaneous manifestations. These manifestations include heliotrope rash, Gottron papules, cuticular vascular changes, a photodistributed erythema or poikiloderma, and a scaly erythematous alopecia [4]. The adult and juvenile subtypes of dermatomyositis share the hallmark features of pathognomic skin rash and muscle inflammation. However, the frequency of several clinical features varies markedly between adult and juvenile disease. Perhaps most prominently, calcinosis cutis is significantly more common in juvenile dermatomyositis patients when compared to adult disease [5]. We report an unusual case of calcinosis cutis of the scalp in an adult dermatomyositis patient.

Case Synopsis

An 82-year-old woman with a decade long history of amyopathic dermatomyositis presented to our clinic with several subcentimeter, firm, yellow-white papules on the occipital scalp (**Figure 1**). She was being managed with oral methotrexate

(10mg/weekly) and topical clobetasol 0.05% solution. She reported no pain or irritation of the affected region. She had a history of breast cancer that was treated at the age of 46, as well as multiple basal cell carcinomas of the ear, face, and scalp that were previously excised. Given concern for a neoplastic process, a prominent papule was removed in clinic via shave biopsy at the base of the lesion. After completion of the removal, a chalky white material was noted at the site of biopsy. The biopsy was submitted for histopathological analysis.

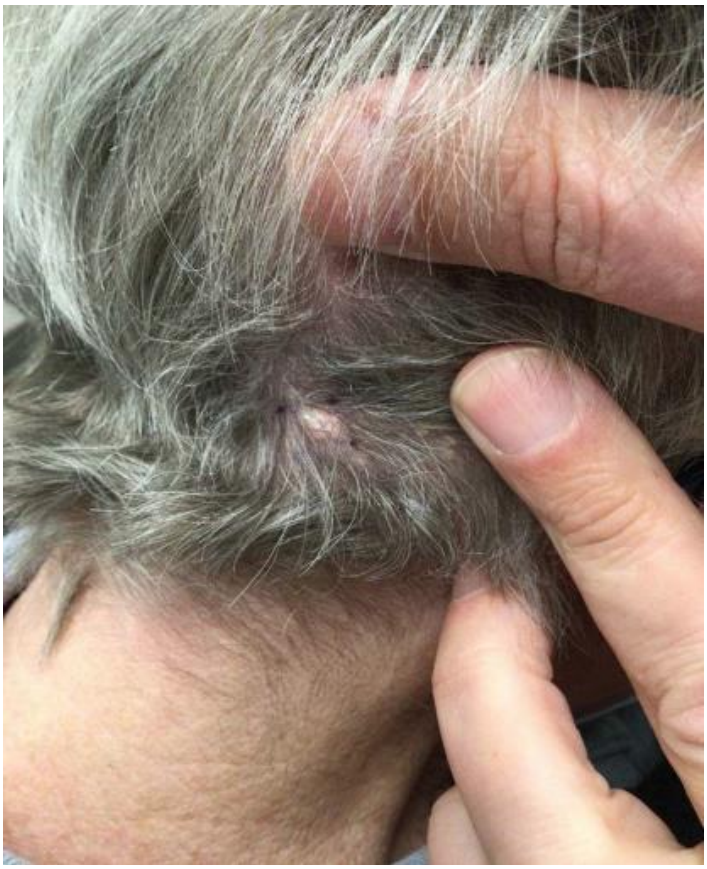

Figure 1. *Right occipital scalp demonstrating a 6mm, yellow-white crusted papule.*

Microscopic findings revealed aggregations of homogenous amorphous basophilic material consistent with calcium (**Figure 2**). Given this finding, accompanied by the gross appearance of the lesions and extensive history of dermatomyositis in the affected region, a diagnosis of calcinosis cutis was made.

The patient had a history of breast cancer that was treated 40 years prior to presentation, recent basal cell carcinoma of the face, prurigo nodularis, and hypothyroidism. At the time of this visit, her scalp irritation was improving on a treatment regimen of methotrexate (10mg/weekly), folic acid (5mg/daily), and clobetasol 0.05% applied 5 times weekly. Given the improvement of her dermatomyositis overall, serologies were not performed. The patient was offered treatment of her calcinosis cutis with intralesional sodium thiosulfate, but she chose to decline.

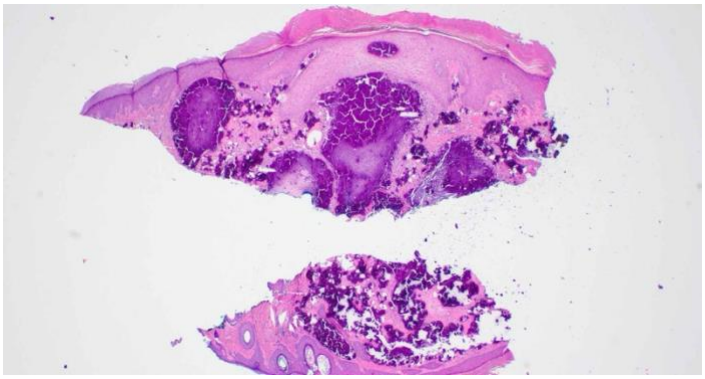

Figure 2A. *Histopathological findings with hematoxylin and eosin staining. Aggregations of homogenous amorphous basophilic material consistent with calcium.*

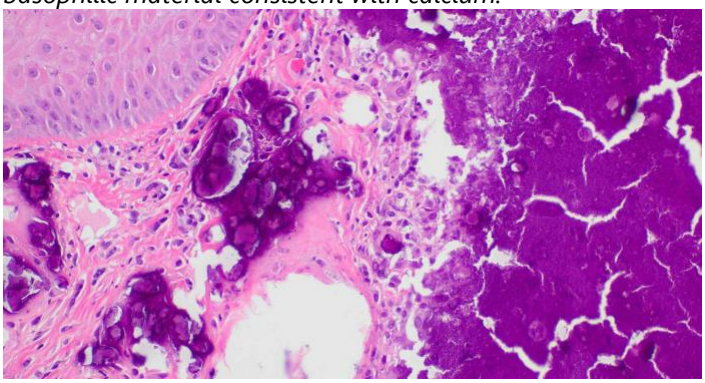

Figure 2B. *Focal calcium deposits in the dermis.*

Case Discussion

Both adult and juvenile forms of dermatomyositis are complicated by calcinosis cutis, though generally, calcinosis of the skin related to dermatomyositis is thought to be unusual in adults and quite common in children. The current literature suggests that calcinosis cutis manifests in 40% to 70% of juvenile dermatomyositis patients and 20% of adult dermatomyositis patients [3,4]. Reports of calcinosis cutis in adult dermatomyositis patients typically involve painful discrete lesions of the

extremities and sometimes of the trunk [6]. Lesions in the scalp in juvenile cases are relatively uncommon but well documented [1,7]. Whereas, to our knowledge, this is the first reported instance of calcinosis cutis of the scalp in a patient with adult dermatomyositis. Rare cases of dystrophic calcinosis cutis of the scalp have been documented in patients with scalp discoid lupus erythematosus, indicating some precedence for calcinosis cutis in patients with autoimmune connective tissue disease directly affecting the scalp [8].

The diagnosis of calcinosis cutis is typically initially suspected on the basis of physical findings, firm, yellow, or flesh-colored nodules or papules, often over bony prominences. Lesions may extrude chalky yellow-white material. Occasionally, these lesions can extrude through the surface of the skin and potentially lead to a secondary infection. In severe forms, calcinosis can cause loss of function and rarely, bone formation is possible [2,4]. Diagnosis should be confirmed via skin biopsy and will demonstrate calcium in the dermis or subcutis manifesting as intensely basophilic deposits on hematoxylin and eosin-staining.

Once established, dystrophic calcinosis cutis is often challenging to treat. Although possible, spontaneous regression is uncommon [4]. Treatment of the underlying autoimmune connective tissue disease should be a priority. Although there is no standardized protocol for direct treatment of calcified lesions, several studies have demonstrated diltiazem and bisphosphonates to be useful [9]. Alternatively, topical or intralesional sodium thiosulfate, an inorganic salt shown to increase calcium solubility, has been used with some success in the treatment of calcinosis cutis. This approach is particularly useful when a localized effect is desired [3]. Although generally not a primary treatment modality, surgical excision was found to be generally effective in over 80% of patients with discrete lesions [1].

Conclusion

We report a case of calcinosis cutis of the scalp in an adult patient with long-standing dermatomyositis of the scalp. Although relatively rare, adult dermatomyositis has been reported to cause dystrophic calcinosis cutis. To the best of our knowledge, this is the first case described that exhibits calcinosis of the scalp in an adult dermatomyositis patient.

Potential conflicts of interest

The authors declare no conflicts of interest.

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