

## Dermatomyositis with Calcifications of the Periodontal Ligament: A Rare Oral Finding

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### Abstract

**Aim:** Dermatomyositis is a rare, idiopathic inflammatory myopathy that affects multiple organ systems and may ultimately be fatal. Classical signs and symptoms include progressive muscle weakness, cutaneous rashes, and calcinosis; however, the clinical presentation of the disease is variable and may be mistaken for other autoimmune disorders. We present the first published case of a patient diagnosed with dermatomyositis who developed calcifications within the periodontal ligaments of multiple teeth.

**Material and methods:** A 46-year old female previously diagnosed with dermatomyositis and generalized severe periodontitis was found to have numerous calcifications within the periodontal ligaments of mandibular and maxillary teeth.

**Results:** The histopathology of biopsied lesions showed spherical calcifications within the periodontal ligaments with minimal inflammatory response. The calcifications were radiopaque lesions distinct from dental calculus.

**Conclusion:** Dental professionals who detect periodontal ligament calcifications or other signs of dermatomyositis through clinical or radiographic examination should refer the patient for further diagnostic testing to prevent severe morbidity and possible mortality.

**Keywords:** Autoimmune; Calcinosis; Dermatomyositis; Dystrophic calcification; Idiopathic inflammatory; Myopathy; Periodontal ligament

**Abbreviations:** DM: Dermatomyositis; PDL: Periodontal Ligament; SLE: Systemic Lupus Erythematosus

### Introduction

Dermatomyositis [DM] is a rare autoimmune disorder found in fewer than 10 per 1 million persons. First described in 1863 [1], the disease consists of a subset of the idiopathic inflammatory myopathies, outlined in (Table 1). DM can affect multiple organ systems including the skin, joints, muscles, lungs, heart, and blood vessels (Table 2). In addition to the classic findings of progressive muscle weakness, cutaneous rashes and calcinosis, a variety of oral and systemic manifestations have been described (Table 3).

In this article we describe a case in which a woman with known DM developed calcifications [calcinosis] of the periodontal ligaments [PDL] around teeth in the setting of severe periodontitis. To our knowledge, no other cases of calcinosis within the PDL have been documented in patients with DM, although tongue and pulp chamber calcinosis have been recognized as a component of the DM disease process [2].

### Case Report

An otherwise healthy female was diagnosed with generalized severe

Inclusion body myositis
Polymyositis
Dermatomyositis
- Classical DM
- Premyopathic DM
- Hypomyopathic DM
- Amyopathic/clinically amyopathic DM
- Post-myopathic DM

**Table 1:** Classification of idiopathic inflammatory myopathies.

Aspiration pneumonia
Calcinosis of the skin and muscles
Cardiac disease
Gastroesophageal reflux disease
Gottron's papules
Heliotrope malar rash
Interstitial lung disease
Myositis
Periungual capillary changes
Progressive muscle weakness
Raynaud's phenomenon
Telangiectasia of the skin
Thoracic myopathy

**Table 2:** Systemic signs and symptoms of dermatomyositis.

periodontitis and oral lichen planes at the age of 27. Six years later, the patient developed muscular weakness, myositis, arthralgia, weight loss, and cutaneous rash of the eyelids, dorsal hands, knees, hips, and scalp. The patient complained of difficulty climbing stairs due to weakness and joint pain. The patient was ultimately seen by a rheumatologist and tests for lupus erythematosus, rheumatoid arthritis, dermatomyositis and polymyositis were undertaken. Serological studies for myositis specific antibodies (anti-mi-2 antibodies) were positive as was the

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patient's antinuclear antibody (ANA) assay. A skin biopsy was not performed. The patient was diagnosed with dermatomyositis. The patient began a treatment regimen including systemic steroids and methotrexate. Over time, the patient developed calcinosis cutis of the lower back, buttocks, groin and arms. The largest documented

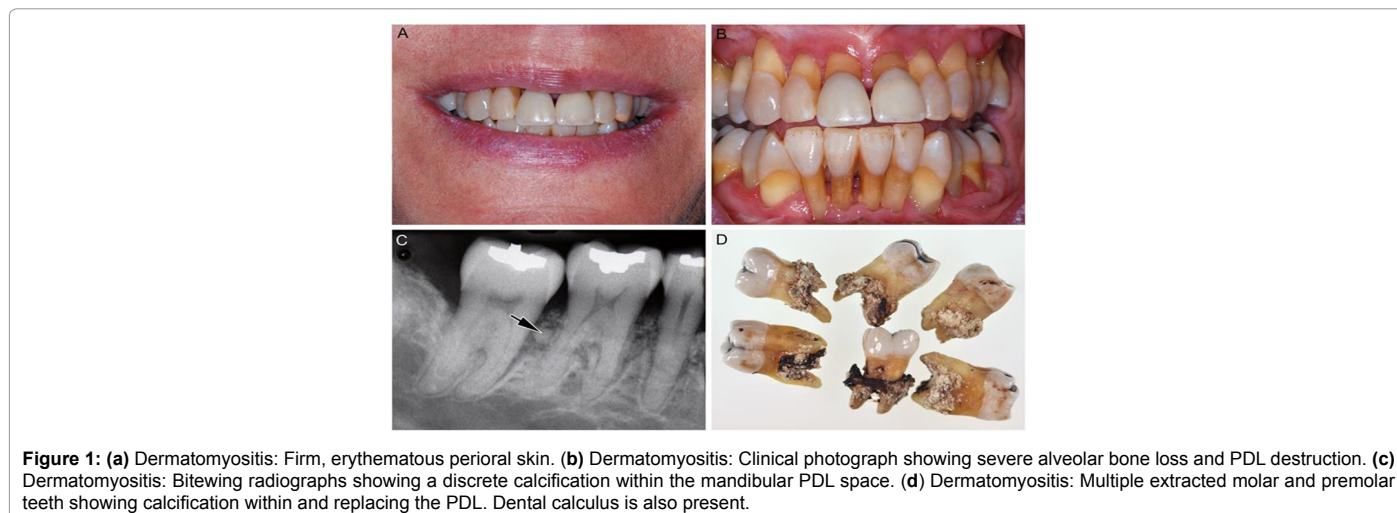
skin lesion measured approximately 30 centimetres in diameter. The perioral skin remained tight, firm and erythematous (Figure 1a).

The patient's periodontitis initially managed her periodontitis non-surgically, but stabilizing gingival grafts were ultimately placed. At the age of 46, the patient's periodontal disease had worsened and the teeth were deemed hopeless (Figure 1b).

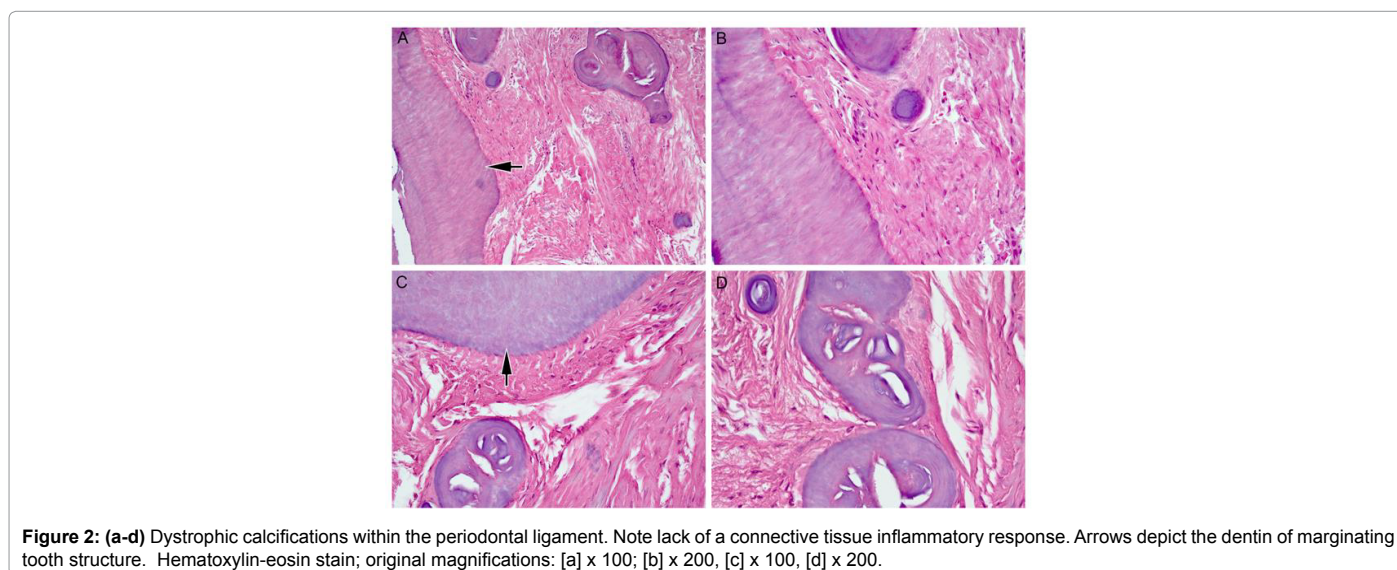
Radiographic examination revealed severe bone loss and radiopacities associated multiple tooth roots (Figure 1c). At the time of extraction, the periodontics noted grossly evident calcifications attached to the tooth roots and bony defects of the alveolar ridge. The calcifications were clinically distinct from dental calculus. Multiple mandibular and maxillary teeth were submitted for pathologic examination with associated soft and hard tissue structures (Figure 1d). Decalcified sections demonstrated anatomically normal appearing teeth and associated hard tissue structures. Nodular dystrophic calcified aggregates were identified with the PDL of many teeth (Figures 2a-2d). These dystrophic calcifications were randomly distributed throughout the PDL and rarely attached to the teeth. The collagen making up the PDL showed little inflammatory response. Aggregates of dental calculus and plaque were noted.

Calcification and obliteration of tooth pulp chambers and canals
Calcinosis of the tongue
Decreased masticatory force
Dysphagia
Edema of lips
Extramedullary plasmacytoma on the tongue
Flaccid vocal cords
Leukoplakia
Macroglossia
Mandibular condyle resorption
Mucosal atrophy
Squamous cell carcinoma of the tongue
Telangiectasia of the oral mucosa and gingiva
Ulceration of the oral mucosa, tongue and gingiva
Xerostomia

**Table 3:** Intraoral and perioral signs and symptoms of dermatomyositis.



**Figure 1:** (a) Dermatomyositis: Firm, erythematous perioral skin. (b) Dermatomyositis: Clinical photograph showing severe alveolar bone loss and PDL destruction. (c) Dermatomyositis: Bitewing radiographs showing a discrete calcification within the mandibular PDL space. (d) Dermatomyositis: Multiple extracted molar and premolar teeth showing calcification within and replacing the PDL. Dental calculus is also present.



**Figure 2:** (a-d) Dystrophic calcifications within the periodontal ligament. Note lack of a connective tissue inflammatory response. Arrows depict the dentin of marginating tooth structure. Hematoxylin-eosin stain; original magnifications: [a] x 100; [b] x 200, [c] x 100, [d] x 200.

## Discussion

The clinical presentation of DM is variable. When muscular weakness is absent, the disease can mimic other conditions such as systemic lupus erythematosus, scleroderma, rheumatoid arthritis, or Sjogren's syndrome. Alternatively, the disease may be concomitant with autoimmune or connective tissue diseases [3,4]. Early diagnosis and treatment is important due to the potential for severe morbidity and mortality. The degree of muscle weakness ranges from clinically undetectable to severely disabling. Mortality from DM is associated with malignancies and respiratory failure due to interstitial lung disease and aspiration pneumonia [5-7]. About 20% to 25% of patients with adult-onset classic dermatomyositis has an occult malignancy. Oral cancer associated with dermatomyositis has been reported in the literature.

Both juvenile and adult forms of DM have been described. Although the disease may present at any age, a bimodal distribution of patients under the age of 18 and between the ages of 45 and 65 is most common. Females are more commonly affected than males. While the exact etiology of the disease remains uncertain, several biologic mechanisms have been proposed, including complement-mediated microangiopathy, overexpression of interferon alpha/beta, and elevated tumor necrosis factor alpha. Predisposing factors that may contribute to the development of DM include ultraviolet light exposure, medications such as statins, genetic mutations, and the Epstein-Barr virus [8,9]. Accurate diagnosis of DM depends on clinical findings, serum markers such as elevated creatinine kinase, and skin or muscle biopsy results. Serum autoantibody levels including anti-Jo-1 and anti-Mi-2 correlate with disease progression but the sensitivity of these tests is low [10,11].

Compared to people with DM, patients with systemic lupus erythematosus typically present with higher antinuclear antibody titres and positive nuclear antigen. Skin rashes associated with SLE tend to appear over the malar eminences more than the periorbital area and the dorsal surface of the fingers instead of the superficial surfaces over the joints. The rash associated with SLE will also be more pruritic and red to pink in color instead of violaceous as in DM [12]. Tissue specimens subjected to histologic evaluation will typically demonstrate C5b-9 membrane attack complexes in DM, but not in SLE [13].

There is no known cure for DM, but treatment modalities can slow the disease progression, improve function, and relieve symptoms. Treatments include steroids, immunosuppressants, intravenous immunoglobulins, metabolic inhibitors, and aminoquinolines. The use of sunscreen and avoidance of direct sunlight is recommended for all DM patients. Antipruritics can alleviate discomfort, while surgery may be indicated to remove bothersome dermal calcifications and periodontally involved teeth.

This case report documents one patient with a novel manifestation of a rare disease. Further research is needed to determine the prevalence of PDL calcinosis in patients afflicted with dermatomyositis, as well as the relationship between the formation of the calcifications and the systemic progression and severity of the disease. If PDL calcifications are an early manifestation of DM then their detection through

dental examinations may be an important indicator for additional rheumatologic testing. Further research is also needed to establish the relationship between periodontal health and the development of PDL calcifications.

## Summary

Dermatomyositis is a rare, idiopathic inflammatory myopathy that can progress from benign to debilitating to lethal. The disease can affect people across a wide age range and is often confused with other autoimmune or connective tissue diseases. Early recognition and accurate diagnosis of the disease is important to initiate therapies that slow the progression and alleviate its symptoms. Calcifications of the periodontal ligament are a novel sign of DM that can be used by health care and dental professionals to screen patients for definitive diagnostic testing, and follow-up, especially in light of the fact that cancer associated dermatomyositis does occur, and a wide range of cancers, most often ovarian, lung, pancreatic and stomach cancer have been documented in association with the disease.

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