Title: Management of a Periodontal Patient with Dermatomyositis: A Case Report.

Authors:

Michael Wojcik DDS, MS, FACD*, †; Andrea López-Torres DMD*; Anthony Neely DDS, M.Dent.Sc, PhD*, ‡; Michael Haddow DDS, MS*, §; Bassam Kinaia DDS, MS*, ‖.

*Division of Graduate Periodontics, University of Detroit Mercy School of Dentistry, Detroit, MI; Private practice †Clinton Township, MI, ‡Southfield, MI, §Taylor, MI, ‖Sterling Heights, MI

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Corresponding author:

Michael Wojcik D.D.S., M.S., FACD

Associate Clinical Professor, Graduate Periodontics Program Department of Periodontology and Dental Hygiene, University of Detroit Mercy

2700 Martin Luther King Jr. Blvd.

Detroit, MI 48208-2576

Telephone: 1-248-953-7676

Email: docwoj@comcast.net or wojcikms@udmercy.edu

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Short running title: Management of Dermatomyositis.

Summary: Medical and dental management of patient with Dermatomyositis stabilizing periodontal involvement.

Keywords: Dermatomyositis, inflammatory myopathies, mucocutaneous, periodontitis, root planing, telangiectasia.

Abstract:

Introduction: Dermatomyositis is an uncommon inflammatory disease marked by muscle and joint weakness with skin rash. Dermatomyositis affects adults and children, with higher prevalence for females 40-60 years old. Most common oral lesions include mucosal edema, erythema and telangiectasia.

Case Presentation: A 51-year-old Caucasian female with an unremarkable medical history presented for periodontal evaluation in 2010. She reported a 6-month history of gingival inflammation and skin irritation on her hands and a rash on the center of her chest and forehead. Other complaints included fatigue, hot flashes, decreased appetite, and weight loss. Periodontal examination revealed generalized acute marginal erythema, with localized slight incipient bone loss. Oral hygiene was deemed good to fair. Oral hygiene instructions were reviewed and a prescription for chlorhexidine gluconate was given. The patient was then referred to allergist and dermatologist where a diagnosis of Dermatomyositis was made. After the initial diagnosis, localized SRP was performed using local anesthetic. The patient was managed medically using Prednisone, Mycophenolate mofetil and Methotrexate and a 3-month periodontal maintenance recall interval. The patient remained stable over eight-years.

Conclusion: Dermatomyositis is an uncommon inflammatory disease that requires medical and dental teams for proper diagnosis and management. Although the condition is chronic in nature with no definitive cure, signs and symptoms can be managed with steroids and immunosuppressants to delay progression of the disease and improve quality of life for the patient.

Keywords: Dermatomyositis, inflammatory myopathies, mucocutaneous, periodontitis, root planing, telangiectasia.

Background:

Dermatomyositis is an uncommon inflammatory disease marked by muscle weakness and skin rash.[1] The condition affects adults and children, with a higher prevalence among females.[2] Age of onset in adults occurs from the late 40s to early 60s.[1] It is called Juvenile Dermatomyositis (JDM) in cases involving children and adolescents.[3] Although oral lesions are rare in the literature, mucosal edema, erythema, and telangiectasia are the most common alterations seen.[4]

Unverricht and Wagner initially described Dermatomyositis in 1887.[5] The most common clinical signs and symptoms include: skin changes and muscle and joint weakness. Skin changes include a violet-colored or dusky red rash, most commonly on the face, eyelids, knuckles, elbows, knees and chest. Although, some of these symptoms may resemble systemic lupus erythematosus, Dermatomyositis differs by involving the metacarpophalangeal and interphalangeal joints.[6]

Progressive muscle weakness may be noted involving the muscles closest to the trunk, such as those in hips, thighs, shoulders, upper arms and neck.[1] Histopathological examination involves degeneration of the muscle fibers, and focal round-cell interstitial infiltration of the affected muscles.[7] Dermatomyositis is a collagen disease with inflammatory and degenerative changes affecting the muscles and skin with a possible link to cancer.[8] This case reports the successful medical and dental management of a patient with Dermatomyositis over an eight-year period.

Clinical Presentation:

A 51-year-old Caucasian female patient was referred by her dentist for periodontal evaluation in 2010. Both oral and written consent was given by the patient. Review of the medical history revealed hypertension and history of mitral and tricuspid regurgitation controlled with medications. The patient listed her occupation as "a house cleaner" and gave a history of latex glove and use of chemical cleaners. She reported a 6-month history of gingival inflammation and rashes on the center of her chest, forehead and hands, which were sun sensitive. Other complaints were fatigue, hot flashes, decreased appetite, and weight loss.

Prior to the periodontal evaluation, the patient saw her physician for a complete work-up including CBC and diabetes screening. Findings were unremarkable except for mild leukopenia. Periodontal clinical exam revealed generalized acute erythema of the marginal gingiva encompassing the entire dentition with an incidental finding of mandibular buccal exostosis (Fig. 1). Clinical and radiographic examinations showed signs of localized incipient bone and attachment loss (Fig. 2). Oral hygiene was deemed good to fair. Extra-orally, no neck or cervical lymphadenopathy was noted. Scaly elevated plaque lesions and rash were noted on her forehead and hands. The fingertips presented with painful fissuring and hyperkeratotic papules (Fig. 3a and 3b). This appearance has been referred to in the literature as "mechanic hands."

Case Management:

After initial periodontal evaluation, the patient was referred to an allergist to rule out contact dermatitis. In addition, oral hygiene was reviewed and a chlorhexidine gluconate ¶ prescription to be used twice daily was given. After examination with the allergist, both

contact dermatitis and allergy to latex were ruled out. The patient was referred to a dermatologist, who ran multiple tests including a 4mm punch biopsy on the chest area. Differential diagnoses included dermatitis, Dermatomyositis, Wegener's granulomatosis and mixed connective tissue disease. Histologic description showed interface dermatitis and epidermal atrophy.[6] Results of the biopsy were consistent with a diagnosis of Dermatomyositis. Initial medications prescribed included Prednisone[#] 30 mg, Mycophenolate mofetil[**] 3,000mg and Methotrexate[††] 10mg. Given the increased cancer risk associated with Dermatomyositis, a comprehensive cancer screening was performed. Cancer screening tests included: esophagogastroduodenoscopy (EGD test), bone marrow biopsy, lung x-ray, Papanicolaou test, mammography, colonoscopy, blood testing, electromyography (EGM test) for the legs and arms, magneto resonance imaging (MRI) for the hip and spine, computed tomography (CT) scan of the abdomen and pelvis, and CT scan with contrast of the lungs. She was found to be cancer free, although pulmonary nodules were noted. Pulmonary function test revealed weakness of the abdominal and diaphragm muscles.

Three weeks after the initial medical diagnosis and treatment by patient's dermatologist, localized scaling and root planing (SRP) under local anesthesia was performed. Heavy bleeding was noted during SRP. The patient was seen 3 months later for periodontal maintenance. A significant reduction in gingival inflammation was noted. Given the underlying disease and initial periodontal presentation, the marked improvement was most likely attributable to a combination of systemic medications and periodontal therapy. The use of Chlorhexidine was discontinued after initial therapy and substituted by essential oils to assist in plaque control. The patient continued to be seen on a 3-month periodontal maintenance schedule.

Clinical Outcomes:

The case has been monitored over a period of 8 years. Her periodontal condition remains stable in 2018 with probing depths of 2-3mm and reduced gingival erythema (Fig. 4 and Fig. 5). Medically, Prednisone[#] was tapered and eventually discontinued two years into treatment. Weekly periodic administration of Methotrexate[††] (22.5mg) was given and the patient continues on Mycophenolate mofetil[**] 3,000mg daily. Follow-ups with the dermatologist showed improvement and reduction in the scale-like elevated plaque lesions and rash on the hands, however, the lesions were not completely resolved (Fig. 6a and Fig. 6b). A similar reduction in peripheral skin lesions were noted. During the eight-year follow up, the patient continues to experience chronic fatigue and has had minor episodes of anemia. In addition, the patient experienced two clinical outbreaks from the Coxsackievirus (hand-foot-mouth disease), which were treated palliatively. She continues to have abdominal and diaphragm muscle weakness that is being monitored by her pulmonologist. The pulmonary nodules, though still present, have reduced in size and some have resolved. Due to long-term use of Methotrexate[††], the patient has undergone three liver biopsies to rule out any liver damage. The liver biopsies showed no significant hepatic changes. Overall, the patient continues to be followed by her physicians, with no significant medical changes.

Discussion:

Dermatomyositis is an uncommon inflammatory disease marked by muscle weakness and skin rash. Oral presentation, although rare, can mimic other periodontal pathology. The identification of gingival telangiectasia, combined with systemic manifestations is important since it will help establish an early diagnosis and treatment for Dermatomyositis. Although the condition is chronic in nature, with no definitive cure, signs and symptoms can be managed to delay progression of the disease and improve quality of life for the patient.

Summary:

Why is this case new information?	• There is a significant literature describing adult Dermatomyositis in medicine, however documentation in the dental literature is very limited. Published articles **in periodontal journals** related to adult Dermatomyositis oral manifestations are lacking. After diagnosis and treatment, this case has been followed for eight-years
What are the keys to successful management of this case?	• The key would be differential diagnosis and interdisciplinary communication, which allows appropriate case management.
What are the primary limitations to success in this case?	• Although Dermatomyositis is chronic in nature, with no definitive cure, signs and symptoms can be managed to delay progression of the disease and improve quality of life for the patient.

FOOTNOTES

Use the following symbols in the sequence shown: *, †, ‡, §, ‖, ¶, #,**, ††, etc.

¶ Chlorhexidine gluconate 0.12%, LGM Pharma, Erlanger, KY, USA.

Prednisone 30mg H.J. Harkins Company, Inc., CA, USA.

** Mycophenolate mofetil 3,000mg Genentech Manufacturer, San Francisco CA, USA.

†† Methotrexate10mg Pfizer, New York, NY, USA.

Figure Legends:

Figure 1: Initial clinical presentation showing erythematous gingival margins. **[Before]**

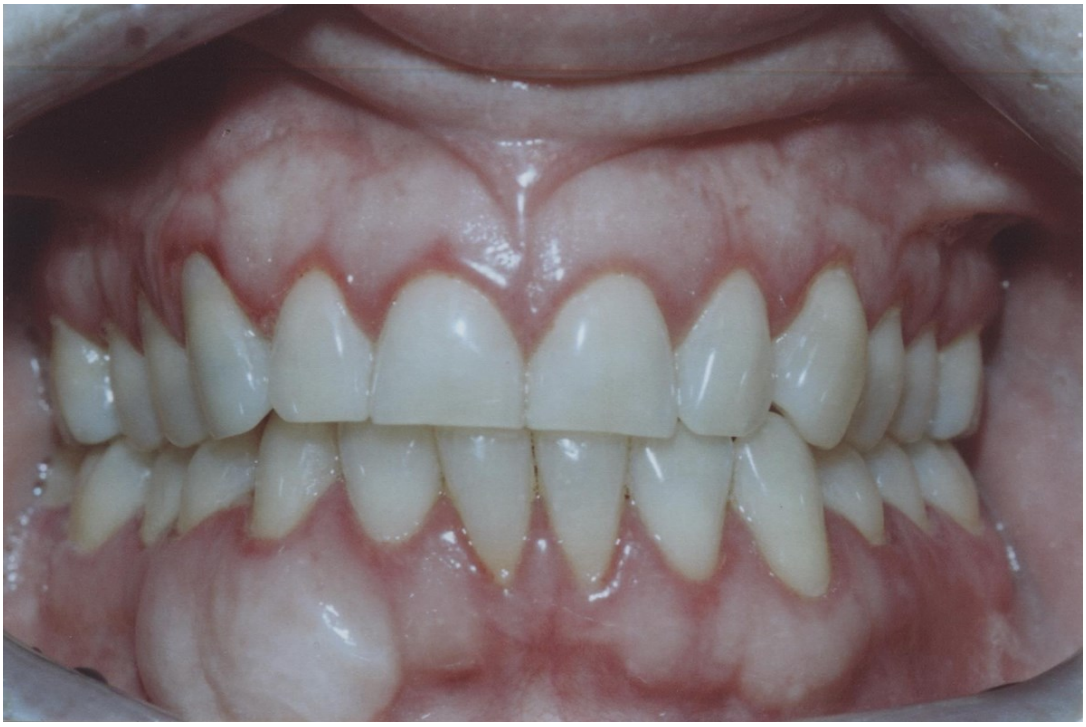


Figure 2: Initial radiographs demonstrating minimal bone loss.

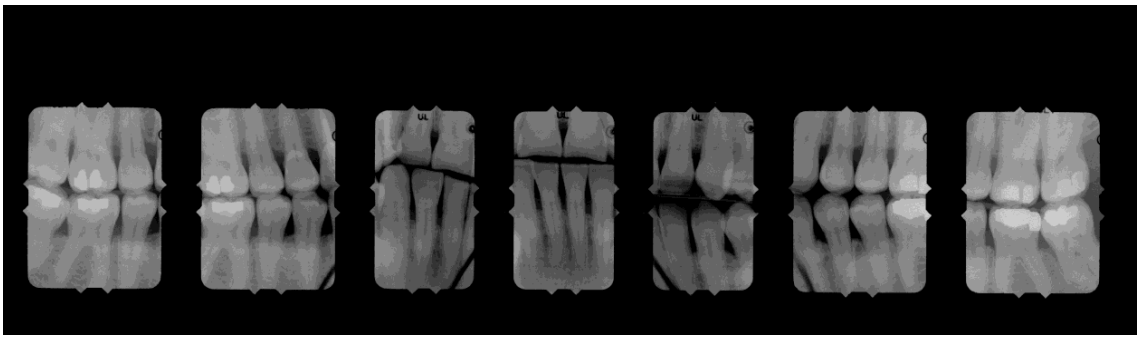


Figure 3a and 3b: Initial clinical presentation of the hands in 2010. 3a Dorsal view of hands showing reddish scale-like plaques characteristic of the disease. 3b Palms of hands show lesions similar to those on the dorsal surfaces.

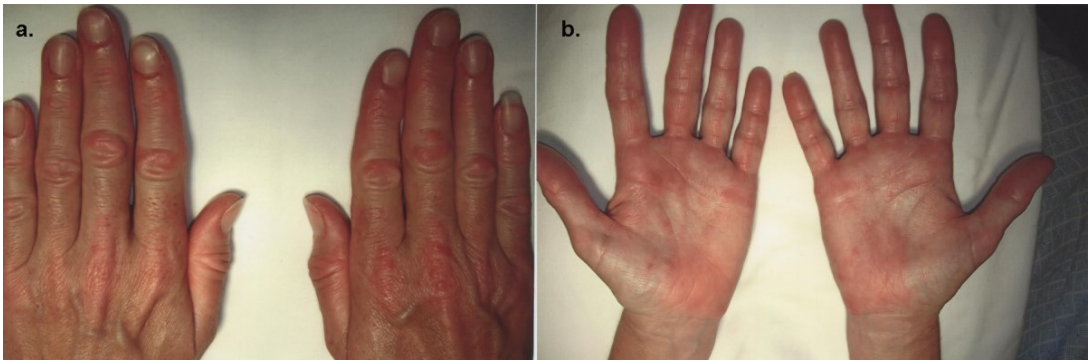


Figure 4: Eight-year post-treatment clinic appearance. Note the significant reduction in acute marginal gingival inflammation (compare to Fig 1) and gingival recession **[After]**

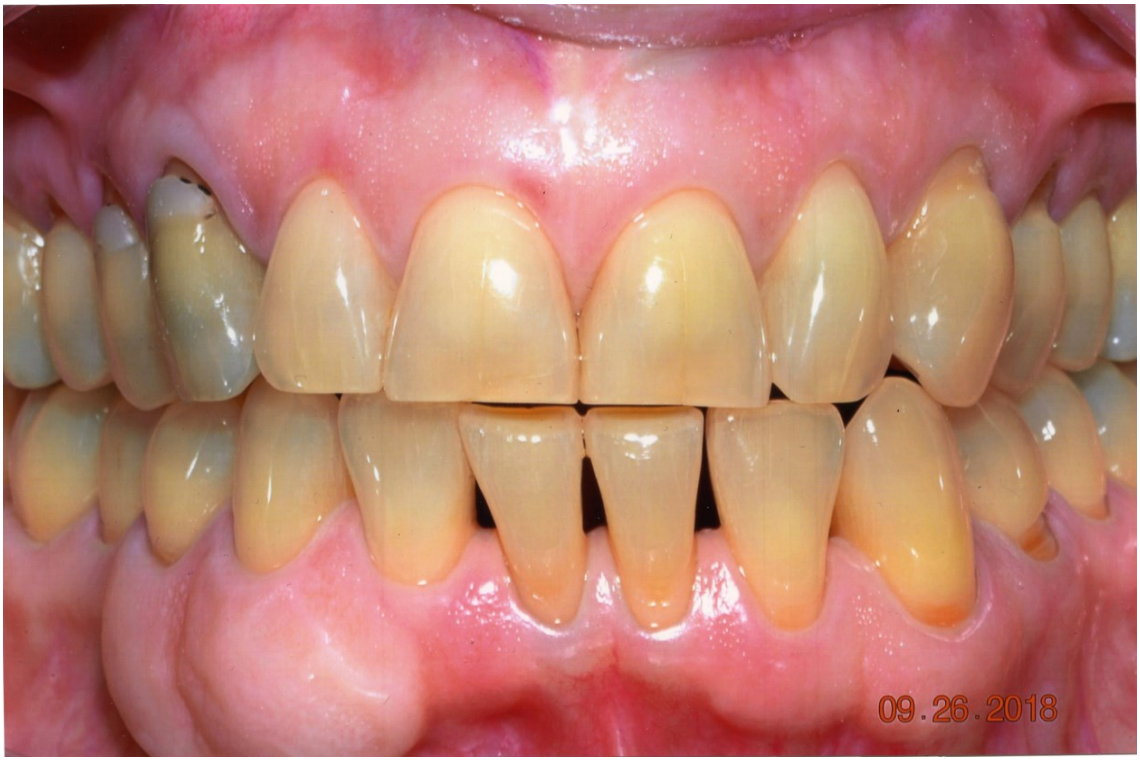


Figure 5: Eight-year post-treatment radiographs showing minimal changes over eight years (compare to Fig. 2)

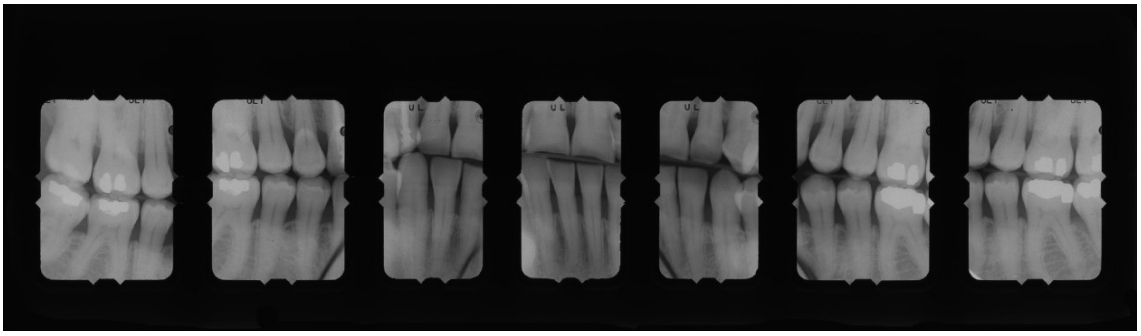
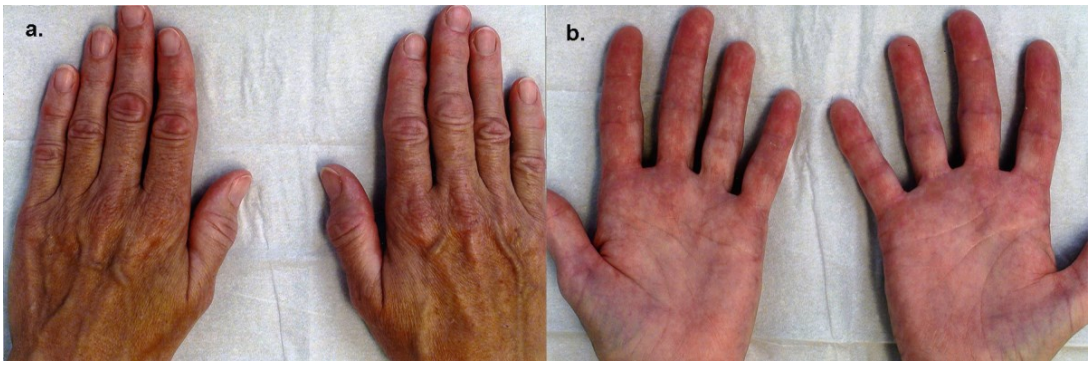


Figure 6a and 6b: Eight-year follow-up view of hands in 2018. 6a Dorsal view of hands showing noticeable decrease in redness and number of plaques compared to initial presentation (compare to Fig 3a). 6b Palms of the hands show similar reduction in redness and scales (compare to Fig 3b).



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