Background: MEDLINE searches (1966-June 1969) failed to identify references that give detailed descriptions of the oral manifestations of dermatomyositis (DM). However, several reports predating MEDLINE provided more complete descriptions of oral lesions associated with DM.

Observations: We describe 5 cases of juvenile DM with oral manifestations, primarily in the form of gingival telangiectases. These findings are compared with those descriptions found in earlier reports.

Conclusions: Oral lesions in juvenile DM have rarely been reported. Mucous membrane involvement associated with DM may include telangiectases, edema, erosions, ulcers, and leukoplakia-like areas. In cases of DM, gingival telangiectases likely represent an underappreciated diagnostic finding analogous to nail-fold telangiectases.

Arch Dermatol. 1999;135:1370-1374

Several early reports have focused on the oral manifestations of connective tissue diseases, but the recent literature has rarely described the oral findings of dermatomyositis (DM). While a few authors have commented on the gingival involvement in DM, we propose that gingival telangiectases are a diagnostic sign similar to nail-fold telangiectases.

CASE REPORTS

CASE 1

An 8-year-old white boy previously in good health initially presented to his dentist for evaluation of gingival tenderness and bleeding. Shortly thereafter, the patient developed a facial eruption that prompted a referral to our dermatology clinic. In addition to a 6-week history of gingival discomfort, his medical history was remarkable for erythema over the face and extremities accompanied by arthralgias and muscle weakness during the last 4 weeks.

One week prior to dermatologic consultation, his referring physician, in an effort to control his oral and cutaneous symptoms, prescribed a 2-week tapering dose of prednisone beginning at 1 mg/kg per day. Despite this intervention, his oral, cutaneous, and muscular symptoms continued to worsen. He had no history of photosensitivity, dysphagia, or gastrointestinal problems.

Results of physical examination were remarkable for a heliotrope facial eruption with bilateral eyelid edema, erythematous papules, and plaques over the elbows, knees, and metacarpal joints of both hands (ie, Gottron papules). He also had ragged cuticles and nail-fold telangiectases on the fingers.

The results of oral examination revealed prominent erythema of the gingiva and telangiectases along the gingival margin abutting both the dentition and interdental region (Figure 1). The patient had marked muscle weakness of the neck and abdominal flexors and the proximal upper and lower extremities. Laboratory evidence was remarkable for elevated muscle enzyme levels, with a creatinine phosphokinase level of 1560 U/L (normal <185 U/L) and an aldolase level of 23 U/L (normal <7.6 U/L), a negative finding for antinuclear antibodies, and electromyelographic findings consistent with a myopathic process.

On further evaluation, a diagnosis of juvenile DM was confirmed by a pediatric rheumatologist. The patient's prednisone dose was increased to 60 mg/d (2 mg/kg per day), with prompt improvement of the patient's muscle weakness, cutaneous eruptions, and gingival disease.
However, gingival telangiectases and dilated capillary nail loops remained at 1-month follow-up. Two months after the beginning of treatment, the muscular and cutaneous findings, erythema, and swelling of the gingiva had substantially improved. His prednisone dose was tapered to 1 mg/kg per day over 3 months. At 1-year follow-up, while on a tapering prednisone regimen (0.5 mg/kg per day), he continued to improve, regaining proximal muscle strength of his shoulders and hips with good strength of his neck flexors.

Regarding his cutaneous signs, the heliotrope rash and Gottron papules had cleared, but nail-fold telangiectases were still present. Although prominent gingival erythema with dilated vasculature was present at 1-year follow-up, the patient had no oral complaints. In addition to the gingival findings, the patient had intermittent episodes of asymptomatic depapillated atrophic patches with thick white margins over the dorsum of the tongue that clinically resembled geographic tongue.

CASE 2

An 11-year-old white girl presented to the emergency department for evaluation after a few-week history of a facial rash, myalgias, weakness, and difficulty rising from a sitting position. The patient denied oral tenderness, difficulty swallowing, or gastrointestinal discomfort. Results of physical examination were remarkable, with several cutaneous findings, including a heliotrope rash on the eyelids, a scaly erythematous rash on the extensor surfaces of the arms and legs, Gottron papules over the extensor surfaces of the metacarpal and proximal interphalangeal joints, and dilated proximal nail-fold telangiectases. Marked muscle weakness involved the proximal muscle groups of the upper and lower extremities, the neck flexors, and the abdominal flexors.

Results of oral examination did not reveal any ulcerations, but were remarkable for a vivid, reddish hue on the lower gingiva and dilated gingival telangiectases best seen under 2- to 4-fold magnification. Diminished muscular strength involved the shoulder and hip girdle muscles, with marked difficulty performing leg lifts and, with squatting maneuvers. Laboratory values were remarkable, with a creatine kinase level of 23 229 U/L; aldolase, 208 U/L; and a positive finding for antinuclear antibodies (1:640, speckled pattern).

The patient was admitted to the hospital for intravenous methylprednisolone pulse therapy followed by outpatient oral prednisone treatments at 2 mg/kg per day. After a month of prednisone therapy, there was substantial improvement in muscular strength. Similarly, her cutaneous involvement vastly improved over the face and extremities. Dilated nail-fold telangiectases were less pronounced but still present. Methotrexate (7.5 mg/wk) was added to the treatment regimen, and while maintaining...
a tapering prednisone schedule (0.5 mg/kg per day), she had no oral complaints despite the evidence of dilated gingival telangiectases.

CASE 4

A 5-year-old girl with a 1-year history of moderately severe DM and evidence of recent esophageal dysmotility was referred to a pediatric dentist for evaluation of a 2-month history of oral discomfort. A few months after she began prednisone and methotrexate treatment, the patient’s rheumatologist attempted to replace the latter drug with azathioprine; however, worsening cutaneous and muscular disease developed, and the patient had to resume treatment with both prednisone and methotrexate (dosage not available). She had moderately active proximal muscle disease with prominent cutaneous findings, including a facial “butterfly” rash. Oral discomfort prompted a pediatric dental evaluation, the results of which revealed severe gingival erythema and inflammation of unknown cause. Although methotrexate was considered a possible source of the patient’s oral complaints, it was not thought to be a likely cause, and the methotrexate treatment was continued by the referring rheumatology team. Results of closer examination revealed the gingival erythema to be composed of individual dilated telangiectases most visible on the anterior gingival margins (Figure 3).

Over the next few months, she continued to have moderate oral discomfort that finally resolved, as did most of her cutaneous and muscular symptoms. From age 6 to 7 years, the signs and symptoms of her disease remained well controlled. The patient’s methotrexate therapy was discontinued, and she was treated solely with oral prednisone (dosage not available). At age 7 years, the results of her follow-up dental examinations revealed, in addition to inflamed and erythematous gingiva, whitish, reticulated patches on both the buccal mucosa and tongue (Figure 4). The patient’s aforementioned mucous membrane manifestations were once again accompanied by an exacerbation of both cutaneous and muscular involvement. Immediate improvement of all symptoms occurred following the combination of an increase in prednisone dose and the reintroduction of methotrexate (dosage not available). At a follow-up visit at age 9 years, her pediatric dentist found whitish, reticulated intraoral plaques. At this time, the patient was doing relatively well on combination prednisone and methotrexate treatment, although asymptomatic, gingival erythema and inflammation were once again evident.

By age 10 years, her disease was well controlled; prednisone therapy was discontinued, and methotrexate (dosage not available) became her sole treatment. She continued to remain asymptomatic, and at age 14 years, she was no longer taking any medication. At several follow-up dental visits, her oral examination results were completely normal.

CASE 5

A 10-year-old white girl with a diagnosis of DM was referred to our hospital for evaluation of progressively worsening disease despite multiple immunosuppressant therapies. The DM diagnosis had been made approximately 2 years earlier when she presented with an erythematous facial rash and severe muscle weakness. Elevated muscle enzyme levels and electromyogram and magnetic resonance imaging findings were consistent with a myopathic process. A computed tomography–guided needle biopsy of the left deltoid revealed atrophy and mild myopathic change; however, the small specimen size precluded a definitive histopathologic diagnosis. Despite multiple forms of treatment, she developed worsening skin and muscle symptoms as well as symptoms of dysphagia and oral discomfort. Just prior to our evaluation, the patient was treated with a combination regimen that included monthly intravenous immunoglobulin, intravenous methylprednisolone (1 g/wk), oral methotrexate (12.5 mg/wk), and oral prednisone (0.5 mg/kg per day).

Results of physical examination were remarkable for a cushingoid appearance, with pronounced muscle weakness of the proximal extremities and neck flexors. Cutaneous findings included a diffuse, erythematous scaling eruption over the trunk, face, and extremities; Gottron papules over the extensor surfaces of the metacarpal joints of both hands; and ragged cuticles with dilated nail-fold telangiectases (Figure 5). Results of oral examination revealed severe erythema composed of multiple dilated telangiectases on both the upper and lower gingival mar-
She also had a few tender erosions on the left buccal mucosa. Her admission laboratory results were remarkable for a positive finding for antinuclear antibodies (speckled, 1:640), negative results on extractable nuclear antigen panel (eg, Smith antibodies [Sm], ribonuclear protein, Ro, La, antihistidyl–transfer RNA synthetase [Jo-1] antibodies), a slightly elevated aldolase level (11.2 U/L), and a normal creatine kinase level (43 U/L). Secondary to worsening muscle weakness, a muscle biopsy was performed, the results of which were remarkable for an inflammatory process with areas of necrosis. This pathologic finding may be found in either steroid-induced myopathy or with a flare of the patient’s current myositis; however, clinical correlation was more suggestive of steroid-induced myopathy. Consequently, her prednisone dose was tapered to 0.3 mg/kg per day. She was discharged on monthly cyclophosphamide therapy (500 mg/m²) in conjunction with a tapering prednisone schedule. A summary of the pertinent details of each case is provided in the Table.

**CASE PRESENTATIONS**

<table>
<thead>
<tr>
<th>Case/Sex/Age, y</th>
<th>Onset of DM</th>
<th>Laboratory Findings and Test Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/M/8</td>
<td>New</td>
<td>Neg</td>
</tr>
<tr>
<td>2/F/11</td>
<td>New</td>
<td>Neg</td>
</tr>
<tr>
<td>3/F/10</td>
<td>New</td>
<td>(1:640)†</td>
</tr>
<tr>
<td>4/F/5</td>
<td>1 y prior</td>
<td>Elevated</td>
</tr>
<tr>
<td>5/F/10</td>
<td>2 y prior</td>
<td>(&gt;1:640)†</td>
</tr>
</tbody>
</table>

*DM indicates dermatomyositis; ANA, antinuclear antibodies; EMG, electromyogram; MRI, magnetic resonance imaging; Neg, negative findings for the given test; Pos, findings consistent with myopathic process; 0, patient’s family refused the test; and ellipses, data not available (records regarding specific laboratory data from this case that presented 20 years prior were not accessible).

†Extractable nuclear antigen (ie, Smith antibodies [Sm], ribonuclear protein, Ro, La, and antihistidyl–transfer RNA synthetase [Jo-1] antibodies) panel findings were negative.

‡Inflammatory infiltrate with myofiber necrosis and regeneration along perivascular zones.

§Original muscle biopsy results prior to our visit documented atrophy and mild myopathic changes. Results of muscle biopsy performed during our most recent evaluation showed areas of inflammation and necrosis. Clinico-pathologic correlation of the latter biopsy results was thought to be more consistent with steroid-induced myopathy.

**COMMENT**

Our cases illustrate the association of gingival telangiectases with DM. With the exception of case 1, where there were gingival findings prior to obvious systemic involvement, these affected children already had other cutaneous and muscular manifestations of DM when their gingival lesions were noted. Gingival findings were most prominent at the time of the initial diagnosis of DM and during exacerbations of disease. Oral lesions were less noticeable when other manifestations of DM disease were quiescent.

Prominent dilated capillaries along the facial anterior marginal gingiva were found in all 5 cases. Their appearance was similar to the dilated capillary loops seen at the nail folds of patients with DM. Some authors have suggested that these capillary formations may represent an attempt at revascularization in areas of ischemic tissue. Interestingly, all of our patients who presented with these gingival telangiectases also had similar nail-fold changes. Nail-fold capillaroscopy studies in juvenile DM may offer important diagnostic information. In one study, the presence of enlarged nail-fold capillaries correlated with increased severity of disease and may serve as a potentially useful diagnostic marker in patients with a more chronic course. Similarly, in our 5 cases, when the muscular and other cutaneous involvement of the DM was better controlled, both the gingival and nail manifestations were less prominent. This suggests that a similar
pathophysiologic process may give rise to the gingival and nail-fold telangiectases.

Much of the literature on oral manifestations of DM predates those articles currently available by MEDLINE searches (1966-June 1999). In 1903, Oppenheim4 wrote the initial report describing mucous membrane involvement in DM. Later, in 1939, Schuerman,6 in a study of 263 cases of DM, reported that mucosal membrane involvement (ie, pharynx, larynx, and conjunctiva) occurred in approximately 20% of patients. In 1945, Keil1 published details of mucous membrane involvement in DM. Although several reports have more recently been written on the oral manifestations of rheumatic disorders,2,5,6 few have focused on DM. Moreover, these articles have lacked detailed descriptions of the gingival findings.

Keil's article1 provides an excellent discussion of the oral findings in DM. However, he did not mention whether the oral manifestations of DM were more common in children than adults. His descriptions are similar to those of the patients we have presented. He described 6 main oral features: erythema, edema without an erythematous component, hemorrhage, vesicles, erosions and ulcers, and leukoplakia-like areas.

Keil focused on the prominent erythema on various portions of the oral mucous membrane, including the gingiva. Hurwitz,2 who similarly commented on the vivid reddish hue of the gingiva in patients with DM, thought that it may represent an initial diagnostic clue. Keil1 describes the color as dusky red or bluish red, and he often discovered closely set telangiectatic vessels after detailed examination. He mentioned that the bluish red mucous membrane appearance was largely due to dilatation of the superficial capillaries; consequently, it did not represent true hemorrhage. Furthermore, Keil thought it likely that nonspecific local vascular damage played a role in the development of these telangiectases.

Keil also mentioned an edematous component of the mouth proper, particularly near the gingival margins, with an increased tendency to bleed. This was a presenting feature in case 1 reported here, and may be akin to the cuticular hemorrhage that can occur in DM. Keil made the important observation that, although it bears some resemblance to scurvy or acute leukemia, there is no special predilection for gingival involvement of the interdental papillae as there is in scurvy.

Keil observed edema without an erythematous component on the tongue, gingiva, palate, or other intraoral sites. He compared this edema with that seen in the skin and other body sites, and he occasionally found vesicles, erosions, and ulcerations. He mentioned that leukoplakia-like lesions were one of the most important alterations found in the oral cavity in DM. Other authors have made similar discoveries in patients with DM.10,11 Keil noted that these lesions, which mainly affected the buccal mucosa, tongue, and palate, may resemble oral lesions similarly encountered in lichen planus or lupus erythematosus. Reticulated leukoplakia was noted in case 4 of our series, but to our knowledge, no histologic information on the leukoplakia-like areas found in these patients with DM has been published. Keil concluded that these whitish lesions might represent simply areas of localized hyperkeratosis similar to those seen in relation to proximal nail folds. Leukoplakia-like areas have been documented in the oral cavities of patients with other mixed connective tissue diseases, including lupus erythematosus.8,10 Other valuable information gathered from Keil's report includes data on lip swelling and intraoral pain associated with DM.

Mucosal changes are found in other connective tissue diseases. Although rarely reported in the literature, similar gingival findings (ie, a dilated venous pattern, erythema, and inflammation) have been documented in scleroderma and lupus erythematosus.8,10 Morphologically, the dilated gingival telangiectases in our patients clinically resemble the nail-fold telangiectases in patients with connective tissue diseases. Dilated nail-fold capillary loops are a well-recognized diagnostic feature of lupus erythematosus, scleroderma, and DM.11,12 In our 5 patients with juvenile DM, gingival telangiectases were found consistently in conjunction with dilated telangiectases of the nail fold.

In summary, mucous membrane involvement is underappreciated in DM. The frequency of its occurrence, as well as its natural history and correlation with muscle involvement in both juvenile and adult DM, needs to be further defined. We believe the presence of gingival telangiectases in our 5 cases of DM may have diagnostic use akin to nail-fold telangiectases.

Accepted for publication June 23, 1999.

This study was supported in part by the Department of Dermatology, University of North Carolina, Chapel Hill.

This manuscript was presented as a poster presentation at the Society for Pediatric Dermatology Annual Meeting, July 1999, Thompsonville, Mich.

We are grateful to Jasper Lewis, Jr, DDS, for his contribution of case 4 to our manuscript.

Corresponding Author: Fred E. Ghali, MD, Pediatric Dermatology, Cook Children's Physician Network, 801 7th Ave, Fort Worth, TX 76104-2976 (e-mail: fredghali@pol.net).

REFERENCES