**Dermatomyositis: an association of gingival telangiectases and anti Jo-1 antibody in the adult**

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**Background:** The presence of gingival telangiectases is an unusual clinical finding in adults with dermamyositis (DM). Patients with aminoacyl-tRNA synthetase autoantibodies express one or more of the following features: myositis, interstitial lung disease, “mechanic’s hands,” and capillary abnormalities (facial telangiectases and Raynaud's phenomenon).

**Case report:** A 45-year-old woman with a classic form of DM of ten years’ duration was evaluated. Clinical investigation revealed periorbital edema and violaceous erythema of the eyelids, Gottron’s papules of the fingers, Gottron’s sign on the elbows and malleoli, a plantar fissured, hyperkeratotic, and scaling eruption (“calloused feet”), ragged cuticles with dilated nail-fold telangiectasia, and gingival telangiectases. The patient fulfilled Bohan and Peter’s criteria for the clinical, histological, EMG, and biochemical diagnosis of DM. Elevated titers of ANA (1:320) with a speckled pattern and anti Jo-1 antibodies were found in her sera by ELISA and Western blot.

**Conclusion:** The recognition of subsets within the spectrum of DM characterized by certain clinical and serological features may be important. Because facial telangiectases are a recognized finding in this subset of patients, we suggest that gingival telangiectases might be a marker for the antisynthetase syndrome.

**KEY WORDS**

Dermatomyositis, gingival telangiectases, antisynthetase syndrome

**A B S T R A C T**

**Introduction**

Dermatomyositis (DM) is a heterogeneous group of acquired, multisystem, inflammatory diseases that affect mainly skeletal muscles and skin. It has variable clinical features and laboratory characteristics (1, 2). The typical features of oral mucosa in DM are erythema and edema, hemorrhage, vesicles, erosions or ulcers, leukokeratosis (white plaques), and a net of dilated superficial vessels (3, 4). Capillary abnormalities in the gingiva have been proposed as an important diagnostic marker of juvenile DM (3). Antibodies to the Jo-1 antigen (cytoplasmic histidyl-tRNA synthetase) were described and characterized over 25 years ago (5, 6). The Jo-1 antibody was introduced as classification criteria for polymyositis (PM) or DM after epidemiological investigation in Japan (7). In this case report, we describe an adult DM patient that had prominent gingi-
Dermatomyositis with gingival telangiectases  

**Case report**

A 45-year-old Caucasian woman was admitted in December 1997 with a heliotrope rash of the face, and pain, stiffness, and weakness of muscles in the upper and lower extremities. The proximal muscle weakness of her arms and legs progressed rapidly. She had difficulty elevating her arms above her head, combing her hair, raising her body from a sitting position, climbing stairs, or exiting a car. She had no lung or joint complaints, fever, or signs or symptoms suggestive of Raynaud’s phenomenon. There was no family history of connective tissue disease. The patient had been treated for a long period for tinea pedis without effect. Clinical examination revealed periorbital edema and violaceous erythema of the eyelids, saturated wine-red colored flat-topped papules overlying the skin covering metacarpophalangeal, proximal interphalangeal, and distal interphalangeal joints of the phalanges (Fig. 1), hyperkeratotic erythematosus patches on elbows, medial and lateral malleoli (Fig. 2), and ragged cuticles with dilated nail-fold telangiectasia (Fig. 3). The plantar aspect of the feet was rough with hyperkeratosis and scaling. Physical examination of the oral cavity revealed edema of the gingiva and single dilated telangiectases (Fig. 4). The patient fulfilled all of Bohan and Peter’s criteria for confirming the clinical, histological, EMG, and biochemical diagnosis of DM in adults (7, 8). Cuticular changes were prominent in our patient. In 1939, Keining (9) first described hyperkeratotic, thickened, roughness-distorted cuticles in a DM patient. In 1958, Gottron (10) noted that the “Keining sign” was a “specific symptom” of disease (11). The cuticles have a disorderly, picked appearance characterized by irregular overgrowth and thickening often associated with periungual erythema, telangiectases, and hemorrhages.

A DM patient may also have hyperkeratotic erythematous patches on the elbows, and a planar fissured, hyperkeratotic, and scaling eruption with the appearance of “calloused feet.” Usually they are diagnosed incorrectly as having pityriasis rubra pilaris (PRP). In 1953, O’Leary (12) first described a patient with DM with generalized erythema and hyperkeratosis of the soles, in whom “a biopsy confirmed the clinical impression of pityriasis rubra pilaris.” Later, Christianson et al. (13) reported 270 patients with DM, including O’Leary’s patient and another with cutaneous changes clinically resembling PRP but without histological confirmation. In 1969, Wong (14) described 11 patients with general-
ized hyperkeratotic follicular papules and hyperkerato-
tic papules on the palms and soles in a series of 23
DM patients from Southeast Asia, and suggested the
influence of racial factors. In 1976, Dupre et al. (15)
described generalized spinulosis and hyperkeratotic
papules on palms and soles in a 12-year-old child and
found features of arrector pili myositis in the cutaneous
biopsy specimen. In 1997, Requena et al. reported an
18-year-old woman with DM displaying generalized
follicular hyperkeratosis and a yellowish diffuse
palmoplantar keratoderma, which histologically proved
to be with compact orthohyperkeratosis without evi-
dence of epidermolytic hyperkeratosis (16). Lupton et
al. (17) described a 53-year-old woman with DM and
clinical findings of PRP, also lacking palmoplantar al-
terations.

We report on a patient with a classic form of DM of
ten years’ duration, with oral gingival telangiectases
and the presence of circulating anti-Jo-1 antibodies.

Few reports have provided a complete description of
oral lesions in DM patients (3, 4, 18–21). Oppenheimer
(18) first described mucous membrane involvement in
DM in 1903. Later, in a study of 263 DM patients,
Schuerman (19) noted that mucosal involvement of
pharynx, larynx, and conjunctiva occurred in about
20% of DM patients. In 1942, Keil (4) published details
of mucous membrane involvement in DM and de-
scribed 6 main clinical features: (i) erythema and
edema, (ii) hemorrhage, (iii) vesicles, (iv) erosions or
ulcers, (v) leukokeratosis (white plaques), and (vi) a

Figure 1: Gottron’s papules overlying dorsal
metacarpophalangeal, proximal
interphalangeal, and distal interphalangeal
joints of the fingers.

Figure 2: Hyperkeratotic erythematous patches
on elbows (a) and erythematous rash on the
lateral malleolus, heel, and foot with rough
hyperkeratosis and scaling (b).

Figure 3: Dilated periungual telangiectases.

Figure 4: Dilated telangiectases on the lower
gingiva and on neighboring mucous
membranes.
network of dilated superficial vessels (4). He also focused on the prominent dusky red or bluish erythema on various portions of the oral mucous membrane, including gingiva, where there were closely set telangiectatic vessels. Edema of the mouth, near the gingival margins, is frequently observed; however, edema without erythema was also noted. Vesicles, erosions, and ulcerations of the oral mucous membrane were occasionally found. It has been suggested that leukokeratosis, mainly affecting the buccal mucosa, tongue, and palate, is one of the most important features in the oral cavity DM (3, 4, 20, 21). Leukokeratosis can reflect oral lichen planus, or be seen with mixed connective tissue diseases.

Capillary abnormalities in the gingiva have been proposed as an important diagnostic marker of juvenile DM (3), similar to cutaneous periungual telangiectases in older patients with chronic DM (22, 23). The development of telangiectatic blood vessels scattered over the trunk has also been described in the late stages of juvenile DM (24). Histopathological investigations of the skin and muscles in DM patients have suggested that these findings represented a systemic angiopathy (25–27), and that microvascular injury plays an important role in the pathogenesis of cutaneous lesions of DM (28). One of the earliest changes in DM was the focal depletion of muscle capillaries (26). The DM lesions showed a significant degree of endothelial injury, vascular ectasia, and vascular fibrin deposition (27). C5b-9 complement membrane attack complex deposits were found on the vessel walls of the dermis in approximately 80% of biopsy specimens of DM patients (28). Myositis-specific autoantibodies (MSA) are commonly found in the sera of PM/DM patients (29). It became apparent that MSA defined a group of myositis patients with distinctive clinical features (30). The patients of Ghali et al. (3), with prominent dilated capillaries along the marginal gingiva, were associated with positive findings for ANA in 2 of 5 (40%) cases. The oral vascular lesions and “calloused feet” in our DM patient are associated with the presence of antibodies to histidyl-tRNA synthetase (HisRS). The patients with antibodies to HisRS and clinical manifestations of myositis, interstitial lung disease, symmetrical polyarthritis, and Raynaud’s phenomenon were described as “Jo-1 syndrome” or “antisynthetase syndrome” (30, 31). The terms “antisynthetase syndrome” and “Jo-1 antibody syndrome” were introduced by Marguerie and Ray (32, 33). During the course of the syndrome, the patients with anti-Jo-1 autoantibodies expressed one or usually more signs of a characteristic spectrum of various organ manifestations including myositis, interstitial lung disease, arthritis, carpal tunnel syndrome, sclerodactyly, Sjögren syndrome, calcinosis, “mechanic’s hands,” and capillary abnormalities (facial telangiectases and Raynaud’s phenomenon) (30–36).

Some have suggested that gingival telangiectases and “mechanic’s hands” in patients with DM do not reflect the fundamental histopathological changes of DM (37). Our case could represent a coincidental association of gingival telangiectasia with Jo-1 antibody positive dermatomyositis. However, it may be presumed that telangiectases on the gingiva, face, and trunk are due to systemic microangiopathy, as far as they are evident in DM patients, and are analogs of cuticular vascular abnormalities in the disease.

Recognition of subsets within the spectrum of DM characterized by the aforementioned clinical and serological features is important in establishing a diagnosis and planning treatment, and is of prognostic significance. Oral lesions may be significant not only in identifying subsets of DM (38), but also as an initial manifestation of dermatomyositis with occult malignancy (39). Clinicians should be aware that gingival telangiectases in DM patients have recently been associated with “antisynthetase syndrome.”

REFERENCES


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