

# DERMATOPATHOLOGY UNIT

## UPP | Department of Dermatology

### UPMC Dermatopathology “Case of the Month” Presentations

#### UPP - Department of Dermatology, Dermatopathology Unit

5230 Centre Avenue (412) 623-2614

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*Peggy Lin MD, Muammar Arida MD, Thaddeus W. Mully MD, Matthew J. Zirwas MD*

### OCTOBER 2006 CASES OF THE MONTH

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#### CLINICAL FINDINGS

#### CLINICAL HISTORY

A 40 year old white female initially presented to an outside dermatologist complaining of pruritus and discomfort, making her so uncomfortable that she could not sleep. Review of systems was significant for fatigue and weakness in her arms. She also complained of numbness and tingling in her fingers and hands. She denied bladder or bowel complaints.

Physical exam revealed mild erythema and edema of the upper eyelids (Fig 1). There were violaceous erythematous papules on the upper back in a “shawl sign” distribution (Fig 2), upper chest, and extensor arms (Fig 3). There was cuticular dystrophy with capillary dropout and dilation (Fig 4). There was equivocal weakness on physical examination. Labs showed normal CPKs. Malignancy workup was negative.

#### HISTOPATHOLOGY

H&E sections revealed a perivascular infiltrate of lymphocytes (Fig 5). Lymphocytes extended to the epidermis, where there were scattered necrotic keratinocytes at the dermoepidermal junction and focally above it (Fig 6, 7). There were foci of slight epidermal atrophy. PASD stain highlighted a focally thickened basement membrane (Fig 8). Colloidal iron stain highlighted some deposits of connective tissue mucin. (Fig 9.)

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### Figures & Images

1. Click on the *Figure number* you wish to review.
2. Click on the *image* to enlarge



FIG 1



FIG 2



FIG 3



FIG 4

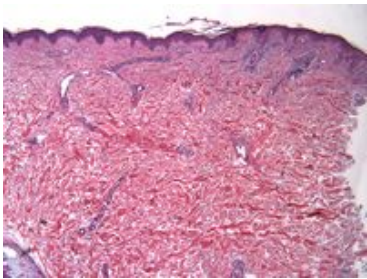


FIG 5

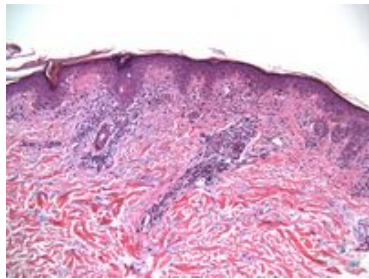


FIG 6

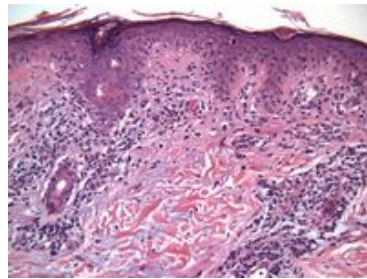


FIG 7

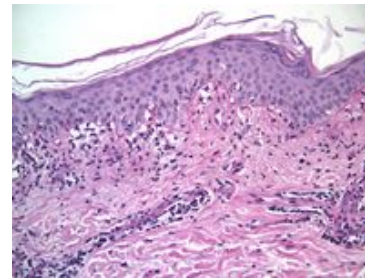
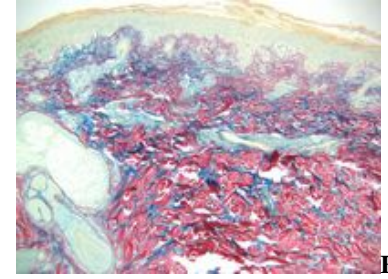


FIG 8



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FIG

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## DISCUSSION & DIAGNOSIS

### Diagnosis

### *DERMATOMYOSITIS*

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### Discussion

Dermatomyositis (DM) and polymyositis (PM) are considered to be opposing ends of a spectrum of inflammatory disease of striated muscles.[A] In addition to muscle involvement, DM has distinctive cutaneous changes. In contrast to DM, PM affects only muscle and does not involve skin.[A] Importantly, both DM and PM have systemic manifestations beyond skin and muscle. Both diseases affect adults and children.

Diagnosis of PM can be made if the patient presents with proximal muscle weakness, elevated skeletal muscle enzyme levels, electromyographic evidence of myopathy, and muscle biopsy consistent with myositis. No cutaneous manifestations occur in this disease. In general, DM requires distinctive cutaneous changes in addition to the preceding criteria. However, there is a variant of DM with normal muscle enzyme concentrations (DM sine myositis or amyopathic DM). It has been well documented that cutaneous lesions may be antecedent to clinical myositis and may exist independent of evidence of inflammatory myopathy. [B,C]

## Epidemiology

DM and PM can affect any age and individual, however, adults are affected by PM more commonly than by DM.[A] The typical affected DM patient is in the fifth or sixth decade. In Caucasians, females are affected more frequently than males. [A] Children more commonly are affected by DM than PM.[A]

## Adult PM and DM

Usually skin disease occurs before muscle disease in DM. [A] Characteristic skin involvement in classic DM includes an intense erythematous to a violaceous coloration with scaling and edema. Poikilodermatous changes may be present. The violaceous hue is especially distinctive in the periorbital regions (most prominently on the upper eyelids), but not pathognomonic. [A] The upper trunk is often involved as well, particularly over the "V" of the neck, upper chest, and back in a "shawl" distribution. Gottron's papules are violaceous papules located over the interphalangeal and /or metacarpophalangeal joints. Gottron's papules in the setting of prominent nailfold capillary alterations are considered to be pathognomonic for DM. Violaceous discoloration affecting elbows and/or knees is referred to as Gottron's sign. [A] The rash of dermatomyositis is pruritic. Another typical cutaneous finding is "Mechanic's hands," which resemble calluses. Mechanic's hands describes hyperkeratotic, scaly, and fissured skin changes located on fingers (radial aspect, extending onto the palms) and the thumbs (ulnar aspect). Although the scalp is characteristically involved, it is frequently misdiagnosed or overlooked. [D]

Proximal inflammatory myositis most commonly involves shoulder, neck flexors, and pelvic areas and may be painful or painless. Patients may present with an abnormal gait, trouble climbing steps, or complain of difficulty arising from a seated position. Cricopharyngeal and esophageal muscles may also be involved, which may present as chewing impairment and dysphagia.

Malignancy is associated with adult- type DM and some feel that amyopathic DM actually represents a paraneoplastic syndrome. [E] Malignancy is associated with 20% to 25% of older patients with new-onset classic DM.[A, F] The tumors have been reported to occur before, concomitant, and after the diagnosis of DM was made. Ovarian cancer [A] is reported to have a high relative risk. Therefore, age appropriate malignancy screening, directed by patient's history and physical examination is recommended. There is a lower risk of associated internal malignancy in PM. [A]

Systemic manifestations include interstitial lung disease, arthritis, cardiomyopathy, conduction defects, and vasculopathy.[A] However, a diagnosis of DM may be made without any systemic manifestations at all. [G]

## **Juvenile DM**

Childhood disease has the same diagnostic criteria of adult disease. Although mortality rate is lower in childhood disease (since there is a lower association with malignancy), there is a distinctively higher incidence of morbidity in the form of vasculopathy and consequent dystrophic calcification.[A] Juvenile DM is reported to follow upper respiratory infection. [A]

Vasculopathy can present as gastrointestinal infarction/perforation, retinopathy, central nervous system involvement and ischemic skin lesions. Dystrophic calcification occurs secondarily. Contractures can occur secondary to muscle infarction, which can occur in the last stages of juvenile disease. Hypertrichosis and lipodystrophy are also seen more frequently in the childhood disease. In addition to the above stated differences in comparison to adult disease, juvenile onset DM shows less overlap with other connective tissue disease.

### **Prognosis:**

Poor prognostic indicators in adult disease include cardiac or pulmonary involvement, dysphagia, severe muscle weakness, older age, delay of therapy, and associated malignancy. Children who develop muscle contractures and widespread soft tissue damage with subsequent dystrophic calcification also suffer higher morbidity, though association with internal malignancy is not reported.[A]

### **Histology:**

Skin biopsies of DM show interface changes consisting of epidermal atrophy, vacuolar alteration of basal keratinocytes, and basement membrane degeneration. Dermis often shows increased interstitial mucin and infiltrates are sparse and composed mainly of lymphocytes. Histological findings are identical to those of lupus erythematosus. Immunofluorescent studies may be helpful to distinguish DM from lupus erythematosus.[G]

Muscle biopsies show type II muscle fiber atrophic changes with necrosis, regeneration, and hypertrophy with central sarcolemmal nuclei. Lymphocytes are distributed in a perifascicular and perivascular array. [G] Preferred muscle biopsy site is the triceps, since it is affected late in the disease process. [G]

### **Differential diagnosis:**

The differential diagnosis for DM includes systemic lupus erythematosus (SLE), scleroderma, psoriasis, airborne or allergic contact dermatitis, photodrug eruption, CTCL, atopic dermatitis, and trichinosis. [G] Though SLE and scleroderma both may occur as overlap syndromes with DM, SLE typically does not exhibit the muscle weakness present in DM. Scleroderma does not exhibit the same pigmentary changes as DM. Edema in airborne disease is typically more

pronounced. Photodrug would exhibit a photodistribution. Atopic dermatitis exhibits more eczematous changes, rather than the characteristic violaceous hue. CTCL shows poikiloderma in intertriginous areas, rather than on the face, extensor surfaces, and scalp. Trichinosis exhibits periorbital edema and painful muscles, but not the cutaneous features exhibited in DM.

### **Treatment:**

Systemic therapy includes prednisone at a dose of 1 mg/kg tapered to 50% over 6 months to none over 2-3 years in alternate day, split, or pulse dosing regimens. Calcium replacement is recommended if systemic corticosteroid therapy is used. Hydroxychloroquine, low dose methotrexate, azathioprine, or retinoids are alternative steroid sparing therapies.[G] Additional systemic therapeutic options include intravenous gamma globulin, cyclophosphamide, chlorambucil, cyclosporine, dapsone, thalidomide, and mycophenolate mofetil.[G] Topical treatments include sunscreen and topical corticosteroids. [G] margins, but the spongiform pustulation is not as pronounced as that seen in pustular psoriasis (Weedon). There is some exocytosis of neutrophils and scattered apoptotic keratinocytes. The papillary dermis is edematous with a heavy mixed infiltrate with eosinophils; eosinophils are rarely seen in pustular psoriasis (Weedon), helping to distinguish these two entities.

### **References:**

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[F] Callen JP. The value of malignancy evaluation in patients with dermatomyositis. J Am Acad Dermatol. 1982; 6: 253-9.

[G] Jorizzo JL. Dermatomyositis. In: Dermatology. Spain: Elsevier, 2003:615-23.

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