

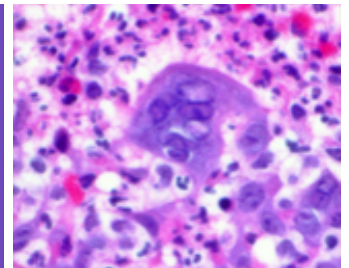


# eLITERATURE REVIEW

## eMedicalDermatology Review

Presented by  
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## June 2010: VOLUME 2, NUMBER 5

### *Dermatomyositis – Association with malignancy and patient evaluation*



#### In this Issue...

Dermatomyositis (DM) is a disease with characteristic cutaneous findings that is often observed in conjunction with muscle disease. DM in adults has been associated with malignancy: recent papers revisiting the frequency of malignancy in patients with inflammatory myopathies have noted clinical and serologic features that may be predictive of a greater risk. While there is a need to evaluate patients for malignancy, the exact tests that should be ordered and the timing of when they should be repeated are controversial.

In this issue, we review current publications that shed light on the relationship of DM to malignancy and the appropriate evaluation of such patients.

#### Program Information

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#### Length of Activity

1 hour  
1 contact hour Nurses

#### Release Date

June 22, 2010

#### Expiration Date

June 21, 2012

#### Next Issue

September 21, 2010

## LEARNING OBJECTIVES

At the conclusion of this activity, participants should be able to:

- Explain why the incidence of dermatomyositis and amyopathic dermatomyositis seem to be rising
- Describe the relationship between dermatomyositis and malignancy as the basis for evaluating potential malignancies in patients with dermatomyositis
- Discuss pulmonary disease as a complication of dermatomyositis and evaluate management therapies

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- **Bernard A. Cohen, MD**, has indicated he has received grants for studies from Novartis Pharmaceuticals and Astellas Pharma Inc.
- **Susan Matra Rabizadeh, MD, MBA** has disclosed no relationship with commercial supporters.
- **Mark Lebwohl, MD** has disclosed that he has received grants for clinical research, Advisory Board, speaker honorarium for/from Abbott, Amgen/Wyeth, Astellas, Centocor, Galderma, Genentech, Novartis, GlaxoSmithKline, Triax, Warner Chilcott. Serving as a consultant and receiving honorarium for/from Actelion, Cerexa, DermiPsor, Electro Optical Sciences, Helix BioMedix, Magen Biosciences, NeoStrata, Peplin, Sanofi-Aventis, Taro, Graceway and Pharmaderm. Advisory Board and receiving honorarium for/from Medicis, Nycomed and Pfizer. Speaker honorarium from Ranbaxy.
- **Elizabeth Sloand, PhD, CRNP** has disclosed no relationships with commercial supporters.

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### **Guest Faculty Disclosures**

**Jeffrey P. Callen, MD** has disclosed that he has no financial relationship with commercial supporters.

### **Unlabeled/Unapproved Uses**

The author has indicated that there will be references to unlabeled or unapproved uses of drugs or products in this presentation, including immunosuppressive agents used for treatment of pulmonary disease in patients with dermatomyositis. There are no agents approved for this indication. The discussion might mention mycophenolate mofetil, cyclosporin, cyclophosphamide and others.

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## COMMENTARY

Dermatomyositis (DM) is a systemic disease with prominent skin and muscle features. Some patients have been described who appear to have disease localized to their skin for long periods of time and (in some instances) never seem to develop muscle disease. These patients have been labeled amyopathic dermatomyositis (ADM, previously known as dermatomyositis-sine-myositis). Classic DM has been linked to a potential for internal manifestations in addition to the muscle disease, including, but not limited to, pulmonary

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disease and malignancy. The risk of these DM features in patients with ADM has been controversial; while some clinicians who specialize in this disease have felt that its incidence was rising, until recently there have been no epidemiologic studies that have detailed this increase.

As reviewed herein, Bendenwald et al<sup>1</sup> used the Rochester Epidemiology Project (REP) to examine the incidence of DM and ADM in residents of Olmstead county, MN. They found that the incidence of ADM was roughly 20% of all DM cases, appeared to have risen over the past 30 years, and that both DM and ADM patients were at risk of having pulmonary disease or malignancy.

The possibility that patients with dermatomyositis may harbor a malignancy dates back to the early 1900s. By the mid-70s, when I began as a resident in dermatology, the relationship of skin findings to an increased risk of malignancy was considered controversial. My study comparing patients with dermatomyositis to those with polymyositis (PM) was perhaps one of the first to document this propensity (*Arch Dermatol* 1980);<sup>2</sup> in a further report I suggested that the evaluation of patients should be more focused (*J Am Acad Dermatol* 1982).<sup>3</sup> By the early 2000s, this controversy had been further resolved by findings from multiple population-based studies, primarily from Scandinavia (Sigurgeirsson et al. 1992;<sup>4</sup> Chow et al. 1995;<sup>5</sup> Ario et al. 1995;<sup>6</sup> Hill et al. 2001<sup>7</sup>). Further, several reports have attempted to determine the clinical and serologic factors that might be effective in predicting the chance of an associated malignancy. In addition, a study from Dartmouth University has shown demonstrated that the risk of cancer, even in a US population, is linked to skin associated disease (DM).<sup>10</sup>

Overall, the reported frequency of malignancy in dermatomyositis has varied from 6% to 60%, with most large population-based cohort studies revealing a frequency of about 20 to 25%. Fardet et al<sup>9</sup> reported that 29 of their 121 patients with DM had a malignancy. As reviewed herein, Antichos et al<sup>10</sup> found malignancy in 32 patients overall—24 with classic DM, 3 with ADM, 3 with PM, and 1 with overlapping features of another connective tissue disorder. Their analysis demonstrated not only that malignancy was significantly associated with DM, but that the incidence was much above that expected in the US population. In the recent population-based study from Mayo Clinic,<sup>1</sup> malignancy was present in 28% of patients, strengthening the best previously existing data that 18-32% of dermatomyositis patients have or will develop a malignancy.

Malignancies may occur prior to the onset of myositis, concurrently with myositis, or after the onset of DM. In addition, the myositis may follow the course of the malignancy (a paraneoplastic course) or may follow its own course independent of the treatment of the malignancy. Studies demonstrating the benefits of cancer surgery on myositis, as well as those showing no relationship of the myositis to the malignancy have been reported. In one retrospective study, independent factors associated with an underlying malignancy in dermatomyositis included age at diagnosis, rapid onset of skin and/or muscle symptoms, the presence of skin necrosis or periungal erythema, and a low baseline level of complement factor C4—whereas a low baseline lymphocyte count was a protective factor for associated malignancy.<sup>9</sup> In addition, one recent study (Trallero-Araguás et al,<sup>11</sup> reviewed herein) suggested that those with an anti-p155 antibody were more frequently linked to cancer associated disease, however there were patients with malignancy who did not have anti-p155 and there were patients in whom the antibody was positive but there was no associated malignancy.

A wide variety of malignancies have been reported in patients with DM. Gynecologic malignancy, in particular ovarian carcinoma, may be overrepresented in DM. Malignancy is more common in older patients (>50 years), but reports of young adults and children with DM and associated malignancy have appeared (Morris & Dare,<sup>12</sup> reviewed herein), suggesting that age alone should not dissuade the physician from a careful evaluation. The site of malignancy can be predicted by the patient's age (eg malignancy in a young man is more often testicular cancer, whereas, in an elderly male, colon or prostate cancer would be more common). In addition, the ethnicity of the patient is a factor in the sites of malignancies as noted in a recent national cohort study from Taiwan.<sup>13</sup>

In the past, there was concern about whether the use of immunosuppressive therapies would predispose the patient to an excess cancer risk. This has not proven to be the case, with most cancers being reported within the first 3 years following diagnosis. There have been, however, cases of Epstein-Barr virus-associated lymphomas arising in

patients with rheumatic diseases, including dermatomyositis, on methotrexate or other immunosuppressive agents, which have resolved on discontinuation of immunosuppressive therapy without requiring additional therapy.<sup>14</sup>

The potential for lung disease in dermatomyositis was reported initially in Asian patients<sup>16</sup>, but the epidemiologic study from Mayo Clinic suggested that 17% of patients with ADM have pulmonary disease.<sup>1</sup> In most cases patients have had interstitial fibrosis, although some patients with severe disease, including bronchiolitis obliterans-organized pneumonia (BOOP), have been reported as well. Treatment of pulmonary involvement has traditionally included high dose corticosteroids and cyclophosphamide. However, in a recent study in this issue, Morganroth et al<sup>15</sup> reported control and resolution of pulmonary disease in patients treated with mycophenolate mofetil.

Although this disease is relatively rare, these recently published papers provide practical utility in evaluating and managing patients with dermatomyositis.

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## RISING INCIDENCE OF DERMATOMYOSITIS AND AMYOPATHIC DERMATOMYOSITIS

Bendewald MJ, Wetter DA, Li X, Davis MD. **Incidence of dermatomyositis and clinically amyopathic dermatomyositis: a population-based study in Olmsted County, Minnesota.** *Arch Dermatol.* 2010 Jan;146(1):26-30

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This study used the Rochester Epidemiology Project to identify new and existing cases of dermatomyositis and its subtypes in Olmsted County, Minnesota, between 1976 and 2007. Its primary purpose was to establish a population-based estimate of the incidence and prevalence of dermatomyositis and amyopathic dermatomyositis; in addition, the authors wished to assess whether there was a change in the incidence over time, and to compare the frequency of associated conditions (in particular malignancy and pulmonary involvement) in these patients.

The investigators identified 29 patients, 6 (21%) of whom had clinical amyopathic dermatomyositis. Female predominance, regardless of subset, was noted in 22/29 patients (76%). Overall, the age- and sex-adjusted incidence of dermatomyositis, including all subtype, was 9.63 (95% confidence interval [CI], 6.09-13.17) per 1 million persons; the incidence of amyopathic dermatomyositis was found to be 2.08 (95% CI, 0.39-3.77) per 1 million persons. The incidence rose from the first decade of measurement to the last decade, from 4.4/million to 12.5/million persons. Eight patients (28%) had a malignant condition during the study period; the risk of malignancy for classic dermatomyositis compared to amyopathic dermatomyositis was 4.61 (not statistically significant). Pulmonary disease occurred in 5 (17%) patients, and was present in both classic DM and ADM. Death occurred in 8 of the 29 patients, with 6 deaths due to cancer. The risk of death for patients without cancer or lung disease did not appear to differ from that of the expected rate in the general population.

This is one of the first reports to demonstrate an increasing incidence of dermatomyositis over the past 3 decades. The investigators demonstrated that roughly 1/5 of patients with DM have an amyopathic variant. This study further shows that malignancy and pulmonary disease are not uncommon in both DM and ADM patients, and that, aside from malignancy, the prognosis appears to be not too different from that expected in the general population.

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## SKIN DISEASE IN PATIENTS WITH INFLAMMATORY MYOPATHY IS ASSOCIATED WITH THE RISK OF MALIGNANCY

Antiochos BB, Brown LA, Li Z, Tosteson TD, Wortmann RL, Rigby WF. **Malignancy is associated with dermatomyositis but not polymyositis in Northern New England, USA.** *J Rheumatol.* 2009 Dec;36(12):2704-2710.

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The authors performed a retrospective analysis of 198 patients seen at Dartmouth-Hitchcock Medical center to evaluate the association of idiopathic inflammatory myopathy (IIM) and malignancy in patients over a 23-year period. They compared the presence of malignancy to standardized incidence ratios for their region (Vermont and New Hampshire) and validated the diagnoses of the patients included in their cohort. Of the



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198 patients with IIM, 61 had DM, 63 polymyositis (PM), 23 ADM, 22 with mixed connective tissue disease (MCTD), 11 with inclusion body myositis (IBM), and 18 with juvenile dermatomyositis (JD).

Malignancy associated with myositis was defined as occurring within 2 years prior to diagnosis or 3 years following diagnosis. Using these criteria, the investigators found that malignancy occurred in 32 patients (16.2%), including 24 DM patients, 3 ADM patients, 3 PM patients, and one case each in the MCTD and IBM groups. These authors findings demonstrate a statistically significant increase occurrence of cancer in patients with DM in comparison to any of the other groups of patients in their study (OR 10.46, 95% CI 4.34–25.21).]. In addition, the investigators evaluated the lifetime risk of cancer and similarly found that DM patients had a significant increase. This study found that cancer seemed more prevalent in men, and age greater than 45 years. Cancer was inversely related to the presence of interstitial lung disease. The authors did not comment upon evaluation of patients and how the cancers in their patients were discovered.

*Dr. Callen notes: These authors separated ADM from DM. I have found a number of cancers in my patients who seem to have only skin changes without weakness or muscle-derived enzyme abnormalities. I believe that these patients should have been included in their analysis as DM patients, and that the finding of skin changes in myositis is important as opposed to myositis without skin changes. However, the importance of this paper is that it clearly documents that the presence of cutaneous disease is important as a marker of malignancy in myositis patients.*

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## IS THERE A SEROLOGIC MARKER OF MALIGNANCY IN PATIENTS WITH DERMATOMYOSITIS?

Trallero-Araguás E, Labrador-Horrillo M, Selva-O'Callaghan A, et al. **Cancer-associated myositis and anti-p155 autoantibody in a series of 85 patients with idiopathic inflammatory myopathy.** *Medicine (Baltimore)*. 2010 Jan;89(1):47-52.

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An autoantibody directed against a 155-kDa protein has been described in patients with myositis. These authors conducted a study to determine the incidence and types of cancer occurring in a cohort of patients with polymyositis (PM) or dermatomyositis (DM), and analyzed the value of this autoantibody as a serologic marker of cancer-associated myositis (CAM).

Eighty-five patients with myositis (20 PM and 65 DM) were included in the study. CAM was detected in 16 patients (19%), of whom 14 had DM. The shawl sign (a rash on the upper back) was significantly ( $p < 0.01$ ). more frequent in patients with CAM than in those without cancer; otherwise no other clinical markers of malignancy were identified in this group of patients. Adenocarcinoma was the most frequent type of cancer (87.5%), and included cancers of the breast ( $n = 4$ ), ovary ( $n = 3$ ), lung ( $n = 2$ ), colon ( $n = 2$ ), cervix ( $n = 1$ ), stomach ( $n = 1$ ), and pancreas ( $n = 1$ ). Anti-p155 autoantibody was found in 1 of the 20 (5%) patients with PM and in 15 of the 65 (23%) patients with DM. A relationship between anti-p155 and CAM was found in DM patients (OR 23; 95% CI 5.23-101.2). Among these 16 anti-p155-positive patients, cancer was diagnosed in 11 patients, with 10 cases classified as CAM (62.5%). Other myositis-associated antibodies were generally absent in patients with CAM. When the authors combined the serologic findings in their patients with DM, the negative and positive predictive value of presence of the anti-p155 autoantibody with the absence of other myositis-associated autoantibodies suggested a strong association of anti-p155 with cancer. This finding suggests that we might be more aggressive in our search in those patients with this antibody.

*Dr. Callen notes: This autoantibody is not readily available, and in general I have felt that the use of myositis-associated antibodies was not clinically useful. I recommend that patients with DM have an appropriate evaluation for malignancy at diagnosis and annually for roughly 3 years following diagnosis.*

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## DO YOUNG PATIENTS WITH DERMATOMYOSITIS NEED TO BE EVALUATED FOR MALIGNANCY?

Morris P, Dare J. **Juvenile dermatomyositis as a paraneoplastic phenomenon: an update.** *J Pediatr Hematol Oncol.* 2010;32:189–191.

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In adults, there is a known association between DM and malignancy. However, this phenomenon has rarely been noted in pediatric patients, and therefore extensive workup for malignancy is not indicated in pediatric patients. In 1993, six patients with malignancy and juvenile dermatomyositis/polymyositis (JDM/PM) that appeared to be part of a paraneoplastic phenomenon were reported. The authors sought to update the literature for reported cases of malignancy associated with JDM/PM; they reviewed the literature over the last 15 years and located 6 additional cases.

The researchers found 12 JDM patients with malignancy, of whom 9 were found to have unexpected physical findings (splenomegaly and/or unusual rash) at the time of diagnosis. In these 9 patients, a malignancy was diagnosed within a mean of 5 months of the diagnosis of JDM. It is also noteworthy that 5 of the 9 patients in this group had lymphadenopathy to the extent that the reporting physician felt it was worthy of comment (more than expected). These patients seem to fit the profile of having a paraneoplastic syndrome.

Even though the number of cases reported over a period of 45 years is quite small, it is still important for clinicians to be aware that there is a possibility of malignancy in patients with JDM, especially in those with an unusual presentation.

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## IS THERE A STEROID-SPARING THERAPY AVAILABLE FOR TREATMENT OF PULMONARY DISEASE IN PATIENTS WITH DERMATOMYOSITIS?

Morganroth PA, Krieder ME, Werth VP. **Mycophenolate mofetil for interstitial lung disease in dermatomyositis.** *Arthritis Care Res (Hoboken).* 2010 Apr 9.

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In this retrospective case series, the authors report on their experience using mycophenolate mofetil as a treatment for dermatomyositis-associated interstitial lung disease (ILD), describing the clinical course of 4 patients with definitive evidence of interstitial lung disease on radiologic imaging. All patients also received prednisone. Of the 4 patients, 3 had at least one year of follow-up on mycophenolate mofetil, and experienced complete normalization of pulmonary function tests (including diffusing capacity for carbon monoxide) and resolution of dyspnea. They were also able to reduce their prednisone dosage. One patient who had both pre- and post-treatment chest computed tomography imaging showed complete resolution of interstitial opacities. The fourth patient included in the report had only 5 months of post-treatment follow-up, but experienced an improvement in diffusing capacity for carbon monoxide from 44% to 77% predicted (but no change in dyspnea).

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The treatment of dermatomyositis-associated ILD has included corticosteroids alone or in combination with one or more immunosuppressive agents, including cyclosporine, tacrolimus, and cyclophosphamide. However, these therapies are all aggressive and are associated with a variety of serious side effects. This paper offers hope for patients that a less toxic agent might be effective for the treatment of pulmonary disease associated with inflammatory myopathy.

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