Case of the Month:
October’s Diagnosis
Dermatomyositis
by Thomas T. Provost, MD

The first 3 respondents in each time zone to identify July/August’s Case of the Month correctly are:

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<th>EASTERN</th>
<th>CENTRAL</th>
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<td>Charles Grabiak, M.D. - NJ</td>
<td>Savita Chander, M.D. - WI</td>
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<td>David Pankin, M.D. - NY</td>
<td>Mark Schaten, M.D. - WI</td>
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<td>Deepa Subramani, M.D. - VA</td>
<td>Shuja Yousuf, M.D. - MS</td>
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<th>PACIFIC</th>
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<td>George Ma, M.D. - CA</td>
<td>Harold Jacobs, M.D. - CO</td>
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<td>Eugene Ocampo, M.D. - WA</td>
<td>Rolf Paulson, M.D. - ND</td>
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<td>Mohinder Cheema, M.D. - WA</td>
<td>Howard Schwartz, M.D. - NM</td>
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Dermatomyositis is a cutaneous inflammatory disease process that may or may not be associated with clinical evidence of muscle disease. The latter condition is termed “dermatomyositis sine myositis” or “amyopathic dermatomyositis.” Dermatomyositis occurring after the age of 40 years may be a paraneoplastic event. In 1980, Callen called attention to the fact that approximately 25% of dermatomyositis patients had an associated malignancy. In recent years, 3 Scandinavian national registry studies from Denmark, Sweden, and Finland confirmed this association and have demonstrated an increased frequency of ovarian cancer in female dermatomyositis patients. In addition, carcinoma of the lung, breast, and gastrointestinal tract have also been reported in patients with dermatomyositis.

Dermatomyositis also occurs as an autoimmune disease process not associated with malignancy. In children, widespread cutaneous and muscle involvement can be especially crippling. Involvement of blood vessels along the gastrointestinal tract in children may produce bleeding. Dystrophic calcification of muscles can be a severe problem as well.

Muscle biopsies in patients with dermatomyositis demonstrate a mononuclear infiltrate composed predominantly of CD4 positive lymphocytes in a perivascular distribution. (In contrast, patients with polymyositis have a disease process characterized by CD8 positive lymphocytes distributed in a perifascicular pattern around individual muscle fibers. The association, if any, of polymyositis with malignancy is rare.)

The cutaneous manifestations of dermatomyositis include a photosensitive facial dermatitis with erythematous violaceous macular lesions most prominent in the periorbital area (heliotrope). In addition, these patients also develop very prominent erythematous lesions in the “V” of the neck and over the shoulders. The latter is termed the “shawl sign” and both features represent cutaneous distribution of a photosensitive disease process.

Patients with dermatomyositis also characteristically demonstrate cuticle nail fold injection, and prominent erythematous lesions over the dorsal surface of the joints of the hands, knees, and elbows. This is termed “Gottron’s sign,” named for the physician who initially described these lesions. With time, the dermatitis produces plaque-like lesions over the dorsal surface of the joints (Gottron’s papules).

At times, patients with dermatomyositis may demonstrate linear-like swirls or streaks termed “centripetal flagette dermatitis,” or individual dermatomyositis lesions may demonstrate hypo- and hyperpigmentation, telangiectasia, or cutaneous atrophy (poikiloderma). The atrophy may be extensive resulting in ulcer formation on the sides of the neck and in axillary regions. Intractable pruritus unresponsive to topical and systemic therapy is common.

Serologic studies have demonstrated that the Mi-2 antibody system, which may be found in as many as 20% of dermatomyositis patients, is specific. On unusual occasions in the author’s experience, antisynthetase antibodies, which are characteristic of patients with polymyositis who have recalcitrant muscle disease, pulmonary involvement, arthritis and Raynaud’s phenomenon, may be detected in patients with dermatomyositis.

Patients with dermatomyositis at risk for the development of an associated malignancy are those who fail to demonstrate autoantibody formation, lack Raynaud’s phenomenon, and are older than 40 years. As noted above, approximately 25% of all patients with dermatomyositis have an associated malignancy. Evaluating and following 33 patients with only the cutaneous features of dermatomyositis, the author’s group found that approximately 25% of them had an associated underlying malignancy or later developed one. The studies summarized above indicate that dermatomyositis, whether associated with clinical evidence of muscle disease or not, is frequently a paraneoplastic disease process.

Based upon this and other investigator’s experience, it is recommended that patients with dermatomyositis who are over the age of 40 years be evaluated routinely every 6 to 12 months with a chest x-ray, mammogram, and a test for the presence of occult blood in the gastrointestinal tract.

Given the increased frequency of ovarian carcinoma in patients with dermatomyositis, the author also recommends pelvic and intravaginal sonography and a CA125 determination every 6 months. Because of the incidence of ovarian car-

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cinoma in female patients with dermatomyositis who are over the age of 40 years, it is conceivable that in the future prophyllactic oophorectomy may be a therapeutic approach in this high risk patient population.

On the one hand, the treatment of the myositic component of dermatomyositis involves a combination of oral steroids and methotrexate, and generally is very successful. On the other hand, cutaneous manifestations of dermatomyositis may be recalcitrant to therapy. As noted above, photosensitivity is a major component of the cutaneous features of dermatomyositis. In fact, biopsies of cutaneous lesions of dermatomyositis demonstrate interface dermatitis with mucin deposition similar to that seen in lupus erythematosus. Under the microscope, the pathologic features of cutaneous dermatomyositis cannot be differentiated from those of lupus erythematosus.

Hydrochloroquine 200 mg twice a day, as well as sun protective clothing, sunscreens, and generally avoiding the sun, are generally at least partially successful in the treatment of cutaneous dermatomyositis. Antihistamines as well as tranquilizers have met with varying success in suppressing the pruritus. The author also has had positive experience with the use of intravenous IgG (IVIG), which may produce a remission of the very prominent cutaneous features of dermatomyositis resistant to therapy with methotrexate and steroids.7

References