

A Fatal Case of Dermatomyositis With Underlying Metastatic Esophageal Adenocarcinoma

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Dermatomyositis (DM) is known to be associated with underlying malignancy, though the strength of this relationship and its predisposing factors are not clearly defined. We present a case of a patient who was first diagnosed with DM and, subsequently, metastatic esophageal adenocarcinoma. Despite aggressive immunosuppressive therapy, the patient's cutaneous eruption failed to resolve and his muscle weakness progressed. He had respiratory failure and died less than 2 months after his initial presentation. To our knowledge, this is only the second case of metastatic esophageal adenocarcinoma associated with DM reported in the English language literature.

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Dermatomyositis (DM) has been associated with various underlying malignancies, most notably lung, ovarian, colorectal, breast, and nasopharyngeal cancers.¹⁻³ Generally, clinical signs and symptoms, as well as age-appropriate cancer screening, are used to help detect an underlying cancer in a patient with DM.^{2,4,5}

Case Report

A 58-year-old man presented to the emergency department with a 2-week history of a worsening photodistributed erythematous cutaneous eruption. The eruption began 2 days after a cardiac catheterization, initially arising on the posterior neck and quickly spreading to involve the face, anterior neck, and chest. The patient was suspected of having an allergic reaction to the contrast dye used during the catheterization. After the eruption failed to respond

to high doses of prednisone, the patient was admitted to the hospital for further evaluation of his progressive skin disease.

At the time of admission, the patient's only complaints were his cutaneous eruption and minimal dyspnea. Recent episodes of chest pain had prompted a thallium stress test and subsequent cardiac catheterization that did not indicate heart disease as the source of his chest pain. Prior surgeries included cholecystectomy, tonsillectomy, and adenoidectomy. There was a family history of diabetes mellitus, though the patient was not affected. His father died at 58 years of age with the cause of death unknown, and his sister died of complications from systemic lupus erythematosus. The patient had no personal history of any skin or connective tissue diseases. Medications at the time of admission included prednisone and cetirizine. He had recently received 2 doses of simvastatin, but this medication was discontinued after the cutaneous eruption developed.

A dermatology consultation was obtained upon the patient's admission to the hospital. Cutaneous examination was positive for photodistributed erythema on the face, anterior neck, and chest. In addition to this eruption, there was marked edema of the periocular area (Figure 1) and erythematous, mildly scaly papules over the extensor aspects of the fingers (Figure 2).

Skin biopsies were obtained from representative areas. The results of the biopsies showed basal vacuolization, rare dyskeratotic cells, increased dermal mucin deposition, and sparse superficial perivascular lymphocytic inflammation (Figure 3). Muscle biopsies were performed and results showed scattered atrophic and degenerated myofibers. These histologic findings were consistent with a diagnosis of DM.

Laboratory studies revealed an elevated creatine phosphokinase level of 1818 U/L (reference range, 38–174 U/L for men) with normal cardiac indices. The result of an antinuclear antibody test titer

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**Figure not
available online**

Figure 1. Cutaneous erythema and periorbital edema.

of 1:640 was positive, with a speckled pattern. Tests for antibody levels were negative for anti-Jo-1, SS-A (Sjögren syndrome antigen A), SS-B (Sjögren syndrome antigen B), and anti-Mi-2. The findings from a chest x-ray were normal.

An aggressive screening for malignancy was undertaken. A computed tomographic scan without contrast of the chest, abdomen, and pelvis showed extensive lymphadenopathy in the mediastinal, hilar, retroperitoneal, and gastrohepatic areas. An upper gastrointestinal endoscopy was performed and a biopsy of an esophageal ulcer was obtained. The results of the biopsy revealed poorly differentiated adenocarcinoma. A positron emission tomographic scan showed diffuse metastasis, revealing inoperable stage IV cancer.

Intravenous dexamethasone was initiated on admission to treat the cutaneous eruption and it was continued throughout the patient's hospital stay. Intravenous immunoglobulin at 2 mg/kg daily was given for 3 days as an adjunctive therapy for DM, with no benefit. After the diagnosis of metastatic esophageal adenocarcinoma was made, the patient was started on a regimen of palliative chemotherapy and radiation. The patient's condition rapidly deteriorated. He required insertion of a feeding tube because of dysphagia and was eventually intubated secondary to respiratory failure. After extensive discussion with his family, the patient was extubated and died shortly thereafter, less than 2 months after his initial presentation.

Comment

DM is an inflammatory myopathy that presents with distinctive cutaneous manifestations, as well as progressive, symmetric, proximal muscle weakness and atrophy. Cutaneous signs usually present before



Figure 2. Erythema, swelling, and mild scaling of fingers.

muscular deficits are evident, and approximately 10% of patients with DM have an amyopathic form of the disease, presenting with cutaneous signs only.⁶ The cutaneous discoloration, telangiectasia, atrophy, and lichenification seen in DM is called *poikiloderma*. The pattern of skin involvement in DM includes the face, a V-sign rash on the neck and chest, and a shawl sign across the back and shoulders. Other cutaneous hallmarks of DM include purple discoloration and edema of the upper eyelids called *heliotrope rash*, and a violaceous scaly eruption on the knuckles called the *Gottron sign*. Patients also may complain of pruritus in predominantly sun-exposed areas.³

DM has a recognized association with malignancy; however, the strength of this association has yet to be clearly defined. Various reports have suggested that anywhere from 6% to 60% of patients diagnosed with DM will have an underlying malignancy.^{1,2,5,7} Most investigators agree that a good estimate of this relationship is approximately 20%.⁷⁻¹⁰ Reasons for the differences in association rates of DM and malignancy include both the criteria used for diagnosing DM and variable aggressiveness of screening for cancer in patients with DM. Males, though less commonly diagnosed with DM overall, are more likely to have DM with an underlying malignancy than females.⁶ In childhood DM, an association with malignancy has not been reported.

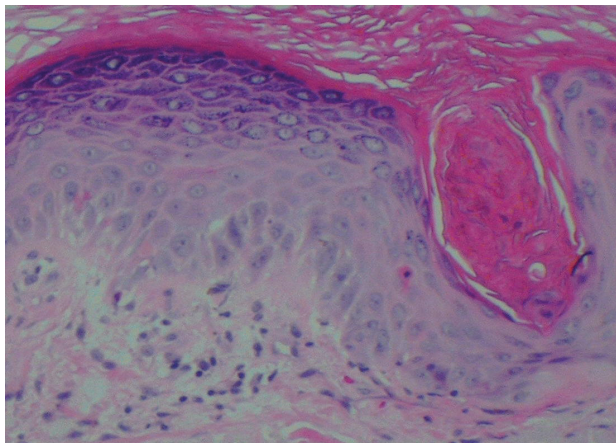


Figure 3. Skin biopsy from neck showing interfacial dermatitis with basal vacuolization, dyskeratosis, and sparse superficial perivascular lymphocytic inflammation (H&E, original magnification $\times 200$).

Patients have the greatest risk of a coexisting malignancy at the time of diagnosis of DM or within the first year after diagnosis.² Because most malignancies occur early in the course of DM, the increased rate of malignancy is unlikely to be secondary to the immunosuppressive therapy that these patients receive.⁴ It has been postulated that DM arises as a paraneoplastic event rather than the cause of a concurrent malignancy.² Clinical evidence supporting this theory includes the usual improvement of DM symptoms after cancer treatment and the recurrence of symptoms among patients with cancer who relapse.^{2,9,11,12}

Malignancies associated with DM include lung, ovarian, colorectal, breast, and nasopharyngeal cancers, as well as non-Hodgkin lymphoma and melanoma.¹⁻³ While adenocarcinoma is the most common, DM is associated with an increased incidence of all histologic types of cancers.² To our knowledge, this is only the second case of metastatic esophageal adenocarcinoma in a patient with DM described in the English language literature.¹³

Metastatic esophageal adenocarcinoma is a rare cancer with a poor prognosis. In the United States, 14,250 individuals were diagnosed with esophageal cancer in 2004, and 13,300 of those patients died.³ The diagnosis of metastatic esophageal adenocarcinoma in a patient with DM is complicated by the fact that dysphagia, its most common presenting symptom, also is a common symptom in DM, especially among those patients resistant to immunosuppressive therapy.¹⁴ Therefore, when physicians evaluate symptoms to identify a malignancy, dysphagia easily may be attributed to purely being a result of DM and overlooked as a sign of an underlying esophageal malignancy.

Conclusion

In this case report, we describe a patient with DM and metastatic esophageal adenocarcinoma. To our knowledge, only one other similar case has been reported in the English language literature. Although metastatic esophageal adenocarcinoma is uncommonly associated with DM, it must be considered in evaluating a patient with sudden onset of this cutaneous disease, especially in cases refractory to immunosuppressive therapy.

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