Paraneoplastic Dermatomyositis in a Patient with Metastatic Gastric Carcinoma

Dear Editor,

Paraneoplastic dermatomyositis is a distinct clinical variant of dermatomyositis (DM) in which the typical cutaneous features and muscle weakness appear before, simultaneously, or after the diagnosis of an internal malignancy. It occurs in approximately one-third of patients with DM, predominantly adults, after the age of 40 (1). Different neoplasms have been described in association with DM, the most common of which are lung, breast, ovarian, gastrointestinal, prostate, and bladder cancers. The gender distribution of cancer type corresponds roughly to that of the general population (1,2).

We report the case of a 58-year-old man who presented with facial heliotrope erythema, periorbital edema, Gottron's papules over the interphalangeal and metacarpophalangeal joints, and Gottron's sign on the elbows (Figure 1). The patient also exhibited some less frequent skin signs of DM, such as shawl sign on the upper back and shoulders and V-sign on the neck and chest. Apart from the rash, he complained of weight loss, adynamia, dysphagia, cough, and scant expectoration, which he reported experiencing over a 3-month period. The muscle involvement consisted of proximal muscle weakness and had appeared a month after the skin rash.

The histology of the skin lesion revealed epidermal atrophy, vacuolar degeneration of the basal keratinocytes, and perivascular and periadnexal lymphocytic infiltrate in the upper dermis (Figure 2). Laboratory examination found increased creatine kinase (2822 U/L) and liver enzymes, anemia, and leukocytosis. Screening for antinuclear antibodies and anti-Jo1 autoantibodies were both negative. The diagnosis of trichinosis was excluded via serologic examination.

The impaired general condition of the patient led to a prompt paraneoplastic screening. Abdominal sonography detected hepatomegaly. Computed tomography (CT) of the abdomen and pelvis visualized a mass in the distal part of the esophagus, narrowed lumen of the gastric cardia, enlarged gastric lymph nodes, lung and liver metastases, and ascites (Figure 3). The diagnosis of paraneoplastic DM in association with an advanced, metastatic, primary gastric carcinoma was established. Palliative surgery and chemotherapy were proposed to the patient, but he refused both. A systemic therapy with methylprednisolone

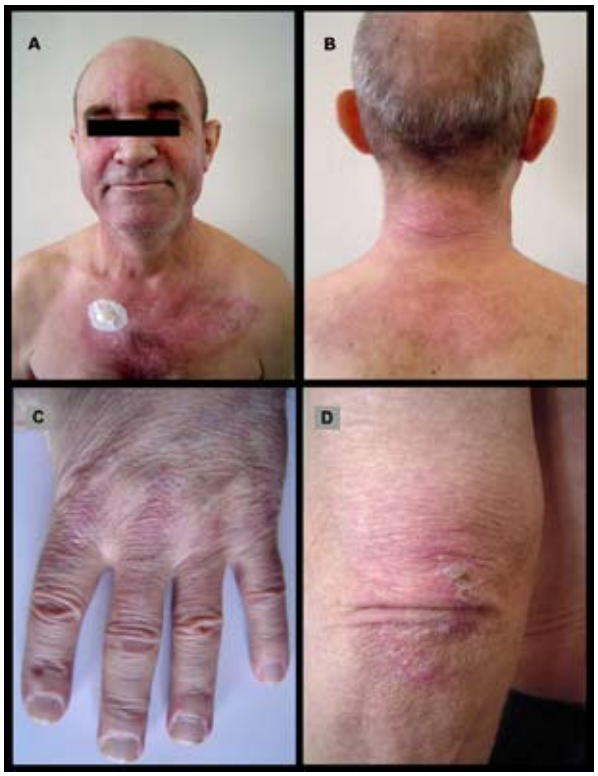

Figure 1. (A) Heliotrope erythema, periorbital edema, erythematous to livid discoloration of the face, and scaly erythema of the scalp; (B) Shawl sign on the upper back and shoulders; (C) Gottron's papules over the interphalangeal and metacarpophalangeal joints; (D) Gottron's sign over the elbows.

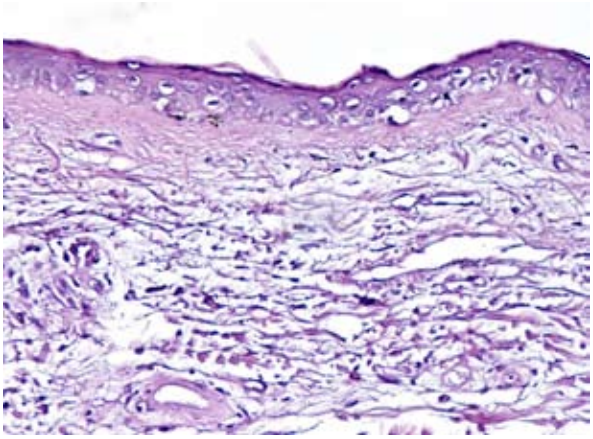

Figure 2. Hematoxylin and eosin staining (×200). Epidermal atrophy, vacuolar degeneration of the basal keratinocytes, perivascular and periadnexal lymphocytic infiltrate in the upper dermis.

60 mg/daily and azathioprine 100 mg/daily was initiated, aiming to alleviate the progressively worsening muscle weakness, but proved ineffective. The patient died two months later of combined respiratory and heart failure.

There are multiple prediction factors, such as cutaneous signs, laboratory data, and disease progression, which may direct the physician towards the possibility of paraneoplastic DM.

Some atypical cutaneous lesions, such as cutaneous necrosis or vasculitis, hyperkeratotic follicular papules, vesiculo-bullous lesions, and flagellate erythema, are seen more frequently in cancer-associated DM (3,4). None of these were present in our patient. Pruritus is also described as a paraneoplastic sign (5).

Some authors consider the increased erythrocyte sedimentation rate and C-reactive protein to be of predictive value for malignancy.

Myositis-specific autoantibodies anti-TIF1-γ and anti-NXP-2, among the numerous novel serological markers for DM, are clearly associated with the presence of neoplasia (6,7). Unfortunately, we were unable to test for those autoantibodies.

The symptom of dysphagia is a hallmark of paraneoplastic dermatomyositis and usually represents a manifestation of muscle weakness (8). In our case, it was rather a reflection of the endoluminal tumor, although it may also be a combination of both factors.

In their study, Bowerman *et al.* investigated the risk of cancer development in different DM subtypes (9). They included 201 patients with adult-onset DM, 142 of with classic DM and 59 with the clinically amyopathic type. The estimated prevalence of malignancy-associated classic and clinically amyopathic DM were 9.9% and 1.7%, respectively. The authors concluded that older age and classic DM represent independent risk factors for malignancy-associated DM within 2 years of disease onset.

Given that early diagnosis significantly impacts prognosis in patients with cancer-associated DM, recent studies support blind screening for internal malignancy (10). Leatham et al. performed a retrospective analysis of 400 patients with DM, finding a total of 53 cancers in 48 patients (some of the patients had two separate neoplasms). Among the group of paraneoplastic DM cases, 17 cancers were diagnosed via purely blind screening in patients with a lack of concerning history or physical examination. The authors

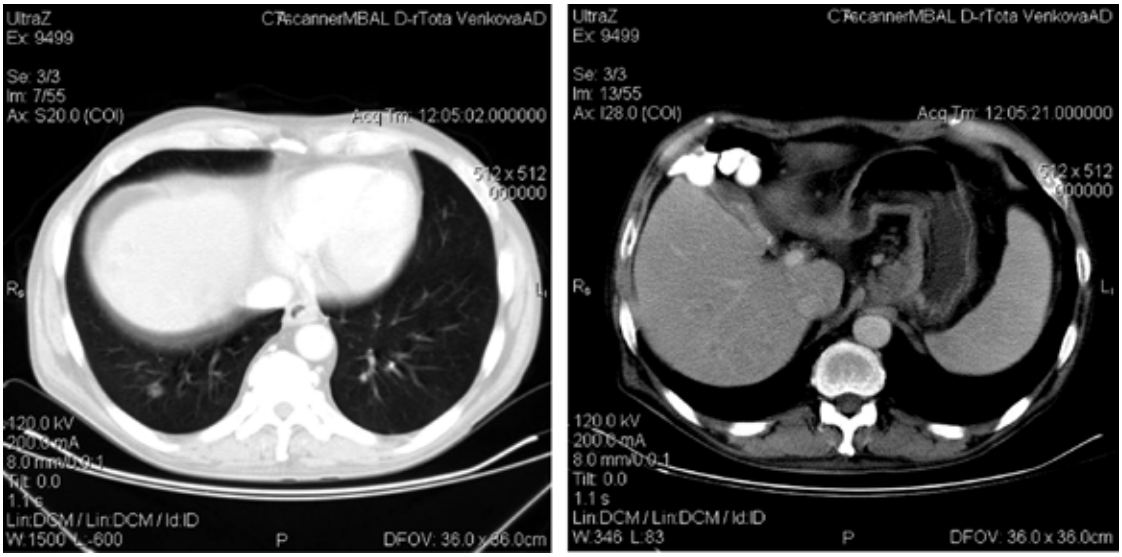

Figure 3. Abdominal CT scan visualizing a mass in the distal part of esophagus, narrowed lumen of the gastric cardia, enlarged gastric lymph nodes, and lung and liver metastases.

claimed that the most informative tests were mammography and CT scanning. The above-mentioned predictive factors for paraneoplastic DM represent a useful tool for the clinician. Although it is generally accepted that patients with DM should undergo some type of cancer screening, there is no consensus regarding methods or frequency. New data suggest that blind screening in asymptomatic patients might be of great importance for early diagnosis and treatment of patients with cancer-associated DM.

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