Our Dermatology Online

Case Report

Dermatomyositis revealing an endometrial papillary serous carcinoma associated with a peritoneal carcinomatosis: A case report

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ABSTRACT

Dermatomyositis is a rare inflammatory disease of connective skeletal muscle tissues. Patients with dermatomyositis have a predisposition for the development of malignant tumors. We report the case of a 69-year-old female who presented herself with two-month face and limb erythemas, muscle weakness, followed by fifteen days of intermittent metrorrhagia. A physical examination revealed typical skin lesions, including erythema, in photo-exposed areas, ragged cuticles, and numerous ulcerations. A blood analysis revealed high muscle enzyme levels. A muscle biopsy revealed myositis and positive anti-TF1 antibodies. An endometrial papillary serous carcinoma was diagnosed by an etiological investigation and classified as stage IV by the FIGO classification. The patient was started on 2 mg/kg/day of prednisolone with concomitant palliative chemotherapy.

Key words: Dermatomyositis; Cancer; Paraneoplastic syndrome

INTRODUCTION

Dermatomyositis is a rare disease characterized by inflammatory myopathy (progressive, symmetric, proximal weakness) and typical skin lesions (Gottron's papules, heliotrope eruptions, periungual telangiectasias, and ragged cuticles). Adults diagnosed with dermatomyositis have an increased risk of malignancy, with the most common tumors being ovarian, cervical, uterine, gastric, and colorectal [1].

We report a case of dermatomyositis revealing an endometrial carcinoma in a Moroccan female.

CASE REPORT

A 69-year-old female consulted for face and limb erythemas associated with muscular weakness present for the last fifteen days and intermittent metrorrhagia present for the last two months.

A physical examination revealed an erythema in photo-exposed areas, hypertrophy, and pain of the cuticles on palpation, as well as numerous ulcerations at the lateral sides of the thighs, lateral sides of the arms, and at the finger pads (Figs. 1 – 3). Blood tests revealed high levels of muscular enzymes, and a muscle biopsy revealed myositis and positive anti-TF1 antibodies. Other examinations revealed endometrial papillary serous carcinoma. Local and general staging revealed multiple diffuse intraperitoneal nodular lesions and masses with an infiltration of mesenteric fat. Hence, the disease was classified as stage IV by the FIGO classification. The patient was started on 2 mg/kg/day of prednisolone with concomitant palliative chemotherapy.

DISCUSSION

The association between dermatomyositis and cancer has been recognized since the report of a case of dermatomyositis and cancer in 1916 by Stertz [2].

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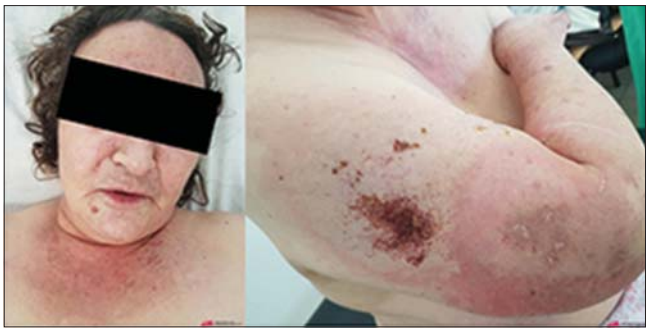

Figure 1: Erythema on the face and upper limbs as well as ulcerations on the arms.

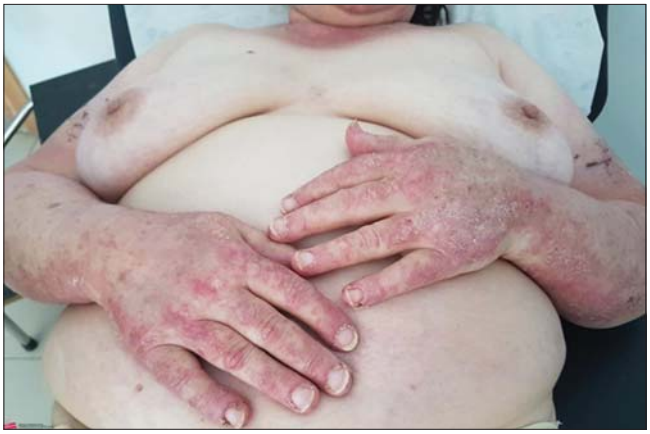

Figure 2: Erythema on the hands with ulceration (Gottron's papules).

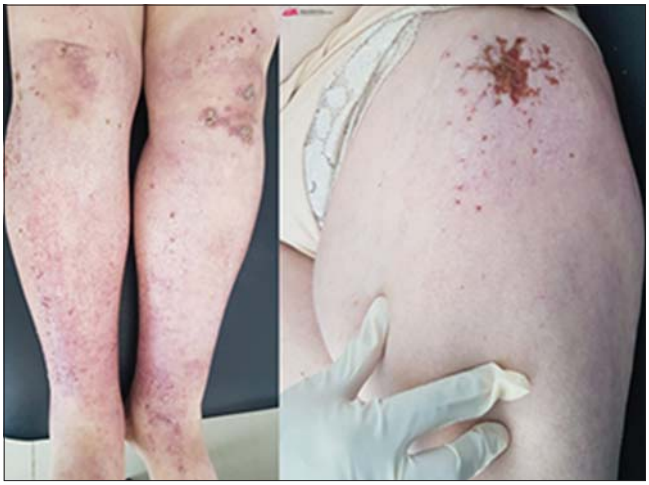

Figure 3: Erythema on the lower limbs with erosive lesions surmounted by hemorrhagic crusts.

While the pathogenesis of dermatomyositis and cancer is incompletely understood, it is thought to be caused by altered humoral and cellular immunity. Myositis-specific autoantigens are expressed at high levels in several cancers known to be associated with the development of inflammatory myopathy. Neoplasia may be detected before, during, or after the diagnosis of dermatomyositis [3].

Currently, the association with lung, pancreatic, stomach, and colon cancers as well as non-Hodgkin lymphomas is well established. Ovarian cancer appears to bear the highest association with dermatomyositis (13.3–26%); breast cancer is less common (13.5%); and the association of dermatomyositis with other gynecologic malignancies, such as endometrial cancer, is relatively rare (1.7%) [2]. One patient out of ten patients with gynecological cancer had endometrial carcinoma associated with dermatomyositis [3,4].

Paraneoplastic dermatomyositis is statistically related with some clinical and biological criteria, such as the sudden onset of symptoms, necrotic lesions or periungual erythema, an age beyond 50 years, and some serum antibodies, such as antibodies to transcription intermediary factor (TIF)-gamma, anti-p155, and antibodies to nuclear matrix protein (NXP)-2, anti-MJ, and anti-p140. Conversely, the presence of myositis-specific (anti-synthetase antibodies, anti-Mi2, and anti-SRP) and myositis-associated antibodies (anti-RNP, anti-PM/Scl) appears to be associated with a decreased risk of malignancy [5].

CONCLUSION

In female patients with DM, a systematic evaluation for a possible gynecologic malignancy should be performed as it may be associated with severe clinical and biological presentations. Thorough history taking and a physical examination, including a rectal examination and breast and pelvic examinations, in females should be performed, as well as a further investigation, including computed tomography, a scan of the chest, abdomen, and pelvis, colonoscopy, mammography, and a Pap smear [1].

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

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