Spontaneous resolution of dermatomyositis associated with fallopian-tube carcinoma following staging surgery

A case report

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**Abstract**

**Rationale:** Paraneoplastic dermatomyositis (DM) is an inflammatory disease of the connective tissue caused by immunologic events in the presence of malignant tumors, which are typically related to ovarian, pancreatic, stomach, and colon cancer. Traditional treatment of paraneoplastic DM includes combination therapy for the underlying malignancy with systemic steroids.

**Patient concerns:** A 41-year-old woman presented with facial erythema and myalgia of the extremities.

**Diagnosis:** The patient was diagnosed with DM associated with a fallopian-tube carcinoma.

**Interventions:** The cancer staging surgery was performed via multi-port laparoscope and administered 6 cycles of adjuvant chemotherapy with paclitaxel (210 mg) and carboplatin (600 mg) right ovary and the left fallopian tube were removed laparoscopically.

**Outcomes:** The DM healed spontaneously without the use of general glucocorticoids after the cancer staging surgery. During the 9-month follow-up, no recurrence of DM or neoplasm was observed.

**Lessons:** This case highlights the fact that paraneoplastic DM can heal spontaneously after therapy for the underlying neoplasm, thereby avoiding the use of systemic steroids and their side effects. Moreover, DM can be an initial symptom for gynecological cancer such as fallopian-tube cancer. Thus, if DM is refractory to standard treatment, gynecological neoplasms should be considered.

**Abbreviations:** CA = carbohydrate antigen, CK = creatine kinase, CT = computed tomography, DM = dermatomyositis, FIGO = Federation International of Gynecology and Obstetrics.

**Keywords:** dermatomyositis, fallopian-tube carcinoma, general glucocorticoids

1. Introduction

Dermatomyositis (DM) is a rare, autoimmune, idiopathic, inflammatory myopathy involving the skin and muscles. It presents with a spectrum of manifestations and has an incidence of 0.5 to 0.89 per 100,000 in the general population.<sup>[1]</sup> In 18% to 32% of patients, especially those who are 40 years or older, DM is closely associated with underlying neoplasms, usually of the ovary, breast, lung, pancreas, stomach, colon, rectum, and non-Hodgkin lymphomas, presenting as paraneoplastic symptoms.<sup>[2,3]</sup> However, its association with fallopian-tube carcinoma is rare. The traditional treatment for paraneoplastic DM involves a high dose of glucocorticoids along with management of the malignancy.<sup>[4,5]</sup> Here we report an interesting case wherein DM was the first manifestation of a fallopian-tube carcinoma, which healed spontaneously without the use of general glucocorticoids following the malignancy staging surgery.

2. Case presentation

The patient was a 41-year-old multiparous woman in good health. Her medical history included a cesarean section and a laparoscopic right fallopian-tubectomy for an ectopic pregnancy. Her mother had died of ovarian cancer. She had a 2-month history of facial erythema, progressive soreness in the upper and lower limbs, and asthenia for roughly 1 month. The significant findings of the physical examinations were violaceous erythemas on the malar, dorsal, and the knuckles. Her laboratory examinations revealed negative autoimmune disease profiles, creatine kinase (CK): 303U/L (normal 30–135U/L), and CK-MB: 4.37 μg/L (normal 0–3.38 μg/L). After being diagnosed with DM, she was treated with Plaquenil (2g, ivgtt, bid), loratadine (10 mg, oral, qd), ketotifen (1 mg, oral, bid), and topical hydrocortisone (bid). One week later, the symptoms persisted, indicating that...
DM was refractory to treatment, which was considered a paraneoplastic sign. A whole body enhanced computed tomography (CT) revealed a 2.5 × 2.9 × 2.9 cm mass in the right ovary. The level of tumor marker carbohydrate antigen-125 (CA-125) was 48.2 U/mL (normal <35 U/mL), while alpha-fetoprotein (AFP) and carbohydrate antigen-19–9 (CA19–9) levels were normal. Based on these findings, laparoscopy was performed. The laparoscopic examination revealed a solid mass in the right ovary as well as a hard, thickened left fallopian tube with atresia fimbria. The right ovary and the left tube were removed. Examination of the fast-frozen section showed an adenocarcinoma, and staging surgery was performed. After the staging surgery, the DM healed spontaneously without the use of general glucocorticoids. The final histopathology indicated a high-grade serous adenocarcinoma in the left tube (Fig. 1A and B). Immunohistochemistry revealed CK7++, CA125+, ER+, P16+, P53+++ (Fig. 1C), WT-1++, and Ki67 positivity in 80% of the cells and absence of CK20 and PR. The neoplasm FIGO staging was IIIc. The patient was administered 6 cycles of chemotherapy with paclitaxel (210mg) and carboplatin (600mg) and was followed-up every 3 months to survey both the malignancy and DM. During the 9 months of follow up there was no recurrence of either the DM or the neoplasm.

3. Discussion

DM is a rare, idiopathic, inflammatory myopathy that with heliotrope rashes, typically involving the malar, forehead, and back, as well as the violaceous papules over the knuckles and symmetric proximal muscle weakness. Elevated CK, which is released from damaged muscle, is common but not required for diagnosis. Other myositis-specific autoantibodies including anti-Mi-2, anti-Jo-1, anti-U1-RNP, and anti-155/140 are associated with DM. Anti-p155/140 increases the risk of an underlying malignancy by 18-fold and some reports have shown that 71% of patients with this antibody develop cancer. Incidence of DM shows a bimodal distribution, typically occurring as either juvenile or adult DM. Very few cases of juvenile DM associated with cancer have been published, as it is not typically a paraneoplastic disease. In contrast, women who are 40 years or older and diagnosed with DM should be assessed for underlying malignancies since 15% to 25% of adult DM cases.
involve paraneoplastic conditions.[9,10] In particular, when DM does not overlap with other collagen vascular diseases, such as polymyositis or scleroderma, lacks auto-antibodies, and is refractory to treatment, it is often associated with an underlying malignancy.[11]

Paraneoplastic DM can predate, coincide with, or develop after the diagnosis of cancer. The risk of cancer is highest within the first year of DM diagnosis and drops thereafter. After being diagnosed with DM, patients are advised to undergo screening for malignancy during the first 3 years, with the exception of ovarian cancer, which may present during the next 5 years. Therefore, DM patients should be followed-up for the underlying malignancy, even if primary screening is negative. It is also advisable that screening is repeated after 3 to 6 months.[12,13]

The mechanism of DM is that of a microangiopathy, characterized by complement deposition in the microvasculature of the muscle and antibody-mediated capillary destruction. However, the pathogenesis of DM in malignancy is poorly understood, but it is thought to be caused by an altered immune system. Traditional treatment for paraneoplastic DM includes combination therapy for the underlying malignancy with systemic steroids.[4,5] There is a report of DM symptom regression with definitive treatment of the underlying malignancy.[4] In most cases, the recurrence of muscle weakness and skin rashes should be assessed because they may be associated with the relapse of malignancy.

According to previous reports, as many as 15% to 25% of DM cases are associated with an occult neoplasm, one of the most common ones being ovarian (13.3–26%).[12,15] The association with fallopian-tube carcinoma is rare. However, recent reports have suggested serous tubal intraepithelial carcinoma as the precursor for serous ovarian or peritoneal carcinoma with a P53 signature.[14,15] Our case met the clinical features of paraneoplastic DM, which healed spontaneously when definitive treatment of the underlying malignancy was administered. However, this case has some limitations, including the lack of a skin or muscle biopsy as well as the lack of testing to establish the presence/absence of the autoantibodies anti-p155/140.

In summary, integrating the findings of this case with the existing literature, there are some lessons to be learned. First, it is possible for paraneoplastic DM to heal spontaneously when the associated malignancy is treated. Therefore, the use of systemic steroids and their side effects may be avoided in some cases. Second, dermatologists and gynecologists should screen for underlying gynecologic malignancies if a female patient, 40 years or older, is diagnosed with DM, particularly in high-risk cases, as a third of paraneoplastic DM cases are associated with ovaries. The involvement of the ovary or fallopian-tube carcinoma does not usually manifest in the early stages, and therefore staging is an important factor for prognosis. Finally, in general, DM responds well to steroids. However, if it is refractory, the patient should be screened for an occult malignancy.

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