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Dermatomyositis following the diagnosis of ovarian cancer

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Abstract. Ben-Zvi N, Shani A, Ben-Baruch G, Agmon-Levin N, Sthoeger Z, Huszar M, Ben-Arie A, Dgani R. Dermatomyositis following the diagnosis of ovarian cancer. *Int J Gynecol Cancer* 2005;**15:**1124–1126.

We present a case history of a woman who developed dermatomyositis following the diagnosis of stage IV ovarian cancer. Dermatomyositis is a rare paraneoplastic syndrome that usually precedes the diagnosis of ovarian cancer by several months or years. Ours is the fifth reported case of dermatomyositis after an established diagnosis of ovarian cancer in the literature.

KEYWORDS: dermatomyositis, ovarian cancer.

Dermatomyositis is a rare paraneoplastic syndrome that may precede the development of malignancies such as lung, breast, or ovarian cancer by several months and even years[1]. In a retrospective population-based cohort study, various malignancies were diagnosed in 50 out of 707 patients with dermatomyositis. Risk was elevated in both sexes, however, more significantly for females in the age group of 45–74 years at the time of diagnosis[2].

Dermatomyositis after an established diagnosis of ovarian cancer is very rare, and we like to report another case of this unique entity.

Case report

A 69-year-old woman was diagnosed with stage IV ovarian cancer in July 2000. She had huge pelvic masses, ascites, and pleural effusion at presentation, with marked CA125 elevation 3200 U/mL (normal < 35 U/mL). Diagnosis was established with the presence of malignant epithelial cells in the ascitic fluid. In August 2000, the first course of neoadjuvant chemotherapy with carboplatin AUC-6 and paclitaxel 175 mg/m^2 was given by 3-h intravenous infusion every 3 weeks was given. Seven days later, an erythematous rash appeared over her chest, back, and forehead. After dermatological consultation, the differential diagnosis was between photosensitivity drug eruption and paraneoplastic syndrome, dermatomyositis. Treatment with prednisone and antihistamine was started.

A few days later, she complained of difficulty in climbing stairs and muscle soreness. Proximal muscle weakness was noted upon examination and serum creatine kinase level was found to be significantly elevated at 1148 IU/L (normal range 0–200 IU/L). The diagnosis of dermatomyositis was suspected, and skin biopsy was obtained since the patient refused to have muscle biopsy. The skin biopsy revealed a predominantly perivascular lymphocytic infiltrate and increased mucin deposits in the reticular dermis.

These features were consistent with a histologic diagnosis of dermatomyositis (Fig. 1).

The patient was placed on prednisone. After two additional courses of chemotherapy, she developed severe dysphagia, hoarseness, exacerbation of proximal muscle weakness, neutropenia, and severe exacerbation of skin rash with vesicle formation. There was no symptomatic improvement with steroids, but creatine-phospho-kinase (CPK) level declined from 1148 to 357 IU/L and CA125 decreased from 3200 to 62 U/mL. Treatment with intravenous immunoglobulin 0.5 g/kg every 5 days was started due to severity of dermatomyositis symptoms. After three additional courses of carboplatin–paclitaxel with dose reduction of 25% due to neutropenic fever, complete clinical and radiologic

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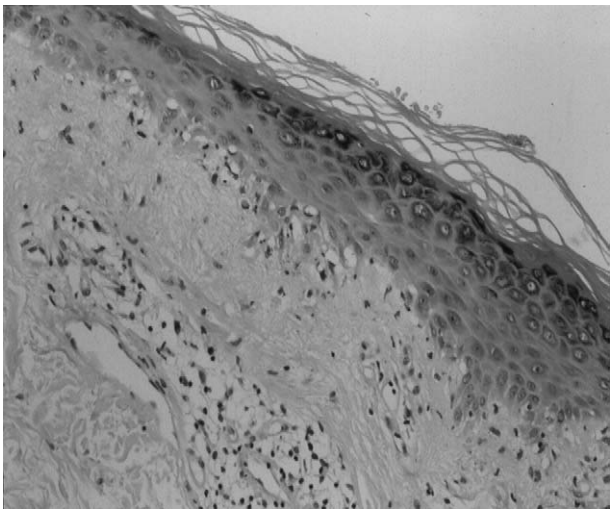

Figure 1. Skin biopsy showing vacuolar alteration of basilar epidermis, few necrotic keratinocytes, flattening of basilar keratinocytes, and perivascular lymphocytic infiltrate around the small vessels.

response was achieved: normalization of the marker CA125 was noted; computed tomography (CT) scan of the chest, abdomen, and pelvis did not reveal any measurable disease; and all the signs and symptoms of dermatomyositis disappeared. Patient was referred for explorative laparotomy. In January 2001, explorative laparotomy was performed and no macroscopic tumor was found. On pathologic examination in the omentum, single cells of moderately to poorly differentiated carcinoma were found, with no tumor in both ovaries, fallopian tubes, and uterus. Patient was placed on close follow-up with CA125 marker tests, physical examination, and serial CT scans. Four months after the explorative laparotomy, she experienced severe skin rash associated with moderate ascites and elevated tumor marker CA125. Recurrent disease was confirmed radiologically (CT scan of chest, abdomen, and pelvis). Second-line chemotherapy was started using weekly docetaxel 40 mg/m^2. After four courses, there was an insignificant decline in CA125 as well as temporary improvement in her skin rash.

Despite the treatment, her disease progressed and she was switched to a third-line chemotherapy with weekly gemcitabine 1000 mg/m^2. No response was achieved. Her dermatomyositis was partially controlled with immunosuppressant azathioprine (Imuran), 1.5 mg/kg daily, and immunoglobulin (IVIg) therapy.

In November 2001, at 16 months since her primary diagnosis the patient died from progressive disease.

Discussion

The incidence of ovarian cancer developing in the setting of dermatomyositis was reported to be as high as 13.3% in one series[1] and usually precedes the diagnosis of ovarian cancer. Ours is the fifth case of dermatomyositis presenting after the diagnosis of ovarian cancer.

Some authors suggest that serum CA125 screening for ovarian cancer in patients with dermatomyositis may be warranted with sensitivity of 50% and specificity of 100%[3]. However, the general consensus is that early detection methods for ovarian cancer are still lacking.

Most common symptoms of dermatomyositis are rash in a shawl-like distribution, Gottron's papules, cuticular changes (periungual teleangiectasia, photodistributed erythema or poikiloderma, scaly alopecia), proximal muscle weakness, and serum CPK elevation. The diagnosis of dermatomyositis is usually established by muscle biopsy that demonstrates the typical pathologic changes of myositis.

The principal changes in muscle consist of infiltrates of inflammatory cells (lymphocytes, macrophages, plasma cells, and rare eosinophils and neutrophils) and destruction of muscle fibers with a phagocytic reaction. Perivascular (usually perivenular) inflammatory cell infiltration is the hallmark of polymyositis. Evidence of muscle fiber degeneration and regeneration is almost invariably present. Many of the residual muscle fibers are small, with increased numbers of sarcolemmal nuclei. Either the degeneration of muscle fibers or the infiltration of inflammatory cells may predominate in any given biopsy specimen. Perifascicular atrophy of muscle fibers, muscle fiber atrophy, and muscle infarcts may also be found.

Some authors suggest that vesicle formation[4] and crycopharyngeal achalasia[5], as was demonstrated in our patient, are strongly related to the presence of malignancy, especially of gynecological origin. In most cases, symptoms of dermatomyositis regress after treatment of the underlying cancer and the manifestations progress in synchrony with tumor spread. Corticosteroids and intravenous immunoglobulins may be effective in some cases[6].

Development of dermatomyositis after an established diagnosis of ovarian cancer, as demonstrated by our patient, seems to be less common, and only four such cases have been previously reported[7,8].

Our patient demonstrates that dermatomyositis may develop in the setting of preexistent ovarian cancer and that skin manifestations may precede clinically obvious muscle symptoms.

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