Letter to the Editor

Amyopathic dermatomyositis associated with an endometrial adenocarcinoma

Giuseppe Famularo

Department of Emergency, Internal Medicine, San Camillo Hospital, Rome, Italy

Corresponding author: Dr. Giuseppe Famularo, E-mail: gfamularo@scamilloforlanini.rm.it

Sir,

Dermatomyositis (DM) is an inflammatory muscle disease with an associated increased risk of malignancy [1]. We report a patient with amyopathic DM and an adenocarcinoma of the uterus.

A 65-year-old Asian woman presented with a heliotrope rash, red marks, pain and swelling of the face, neck and upper chest with an accompanying periorbital swelling (Fig. 1). The patient fully moved her neck and did not complain of muscle pain or reduced strength. Past medical history was unrevealing, she was not taking any medications, and there was no family history of allergy or muscle, skin, and connective tissue disorders. Physical examination was otherwise normal with no weakness in any muscle groups. Blood muscle enzyme levels and the other laboratory findings were within normal limits and an autoimmune screening was negative. A clinical diagnosis of DM was done. We considered the possibility of a paraneoplastic syndrome so we performed a total-body computed tomography that revealed an intrauterine 4 x 5 cm contrast-enhanced mass. Histopathological examination confirmed it was an endometrial adenocarcinoma. Surgical resection was planned, however the patient did not consent to surgery and was lost to follow-up.

This woman presented with *de novo* onset of cutaneous changes pathognomonic of DM, no clinical evidence of proximal motor weakness, and normal blood levels of skeletal muscle enzymes. Taken altogether, these findings suggest the amyopathic pattern of DM, i.e. a subtype of DM that represents an estimated 20% of all DM cases and has cutaneous manifestations such as Gottron's sign, heliotrope rash, and poikiloderma that

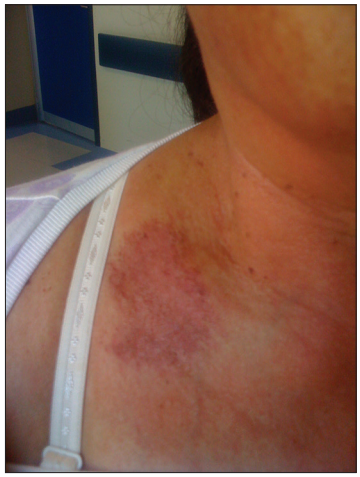

Figure 1: A heliotrope rash, red marks, and swelling of the neck and upper chest consistent with the clinical diagnosis of dermatomyositis.

overlap with those of classic DM but lacks any clinical or laboratory findings consistent with the involvement of skeletal muscles [2]. In our case, the outbreak of the uncommon amyopathic subtype of DM was the obscure presentation that led to the diagnosis of a common malignancy, i.e., an adenocarcinoma of the uterus.

This is the first report of the association of amyopathic DM with an adenocarcinoma of the uterus. Studies have consistently demonstrated a DM-associated risk of malignancy, especially in the elderly and within three years of the first DM diagnosis [1]. Patients with DM including the amyopathic sybtype in association with and as a non-metastatic effect of ovarian cancer or other solid tumors of the female reproductive tract have been described but very rarely with either a carcinoma or a sarcoma of the uterus [3,4]. It is unclear if amyopathic DM patients have as a comparable risk of an associated malignancy

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as those with classic DM. A recent review has found 79 reported cases of amyopathic DM associated with solid tumors or hematologic malignancies with three patients having more than one malignancy. Similar to classic DM, ovarian and nasopharyngeal cancers were common in amyopathic DM [5]. However, in contrast to lung cancer, which is the most frequent malignancy associated with classic DM, breast cancer was the most common type of malignancy reported in patients with amyopathic DM [5]. Features such as photosensitivity and periungueal erythema seem to predict the lowest probability of an associated malignancy in amyopathic DM patients [6].

The mechanistic relationship underlying the association of DM including the amyopathic subtype with a malignancy of the female reproductive tract is poorly understood. Cross-reactivity of skeletal muscle-specific autoantigens and autoantigens of breast, lung and liver cancer cells has been demonstrated [7]. The over-expression of the transcriptional intermediary factor 1γ in tumor cells has been shown to trigger the development of anti-p155/140 autoantibody and the resultant occurrence of DM in a patient with an endometrial carcinoma [8].

Screening for a malignancy of the uterus could be advised in older women presenting with *de novo* onset of amyopathic DM.

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