Paraneoplastic dermatomyositis associated with testicular cancer: a case report and literature review

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SUMMARY

Dermatomyositis (DM) is frequently associated with neoplastic disorders, mainly carcinomas. However, the association of DM with testicular cancer appears to be very rare. A 37-year-old male was treated for a skin rash on the face and arms with pronounced itching. In December 2005, a testicular swelling and ulceration appeared and a unilateral orchiectomy was performed after a diagnosis of seminoma testis was made. However, the skin rash worsened and symmetric muscle weakness appeared in February 2006. The patient was admitted to the Department of Dermatology, Medical University of Sofia, with characteristic heliotrope erythema, eyelid edema, erythematous plaques on the upper chest, and Gottron's papules on the wrists. Severe muscle weakness was found in the proximal limb muscles. The patient's creatine kinase level was significantly elevated, whereas ASAT and ALAT were within normal ranges. Electromyography and skin biopsy supported a diagnosis of DM. Treatment with moderate corticosteroids and a short course of azathioprine markedly improved the skin lesions and muscle weakness. Even in young patients with DM, the risk of neoplasm is increased. Early recognition of the characteristic skin rash may provide a clue to the diagnosis, and screening for neoplasm may improve the prognosis.

KEY WORDS

dermatomyositis, seminoma testis

Introduction

Dermatomyositis (DM) is a disease of unknown etiology characterized by the association of progressive, proximal muscle weakness and pathognomonic or specific cutaneous findings. The first two cases of malignancy-associated DM were reported in 1916 by Stertz (1), who described a patient with gastric carcinoma, and Kankeleit (2), who reported on a female with DM associated with breast carcinoma. Since then, many case reports and multicenter, retrospective, and case-control studies have been published in the literature focusing on the incidence, type of tumors, possible predictive factors, and prognosis in paraneoplastic DM (3–7). Malignancy was associated with DM in 23% to 43% of patients, highly supporting the paraneoplastic nature of the disease (7). We present a rare case of a patient that developed clinical signs of DM 1 month before being diagnosed with testicular cancer.

Case report

A 37-year-old male was treated in November 2005 for acute dermatitis because of a skin rash on the face and arms with pronounced itching. One month later, testicular swelling and ulceration appeared, seminoma testis (T_1 N_0 M_x) was diagnosed, and a unilateral right orchidectomy was performed in January 2006. A CT of the pelvis revealed no indications of metastasis to the lymph nodes. However, the cutaneous lesions worsened, symmetric muscle weakness of the upper and lower extremities appeared, and the patient was admitted to the Department of Dermatology, Medical University of Sofia.

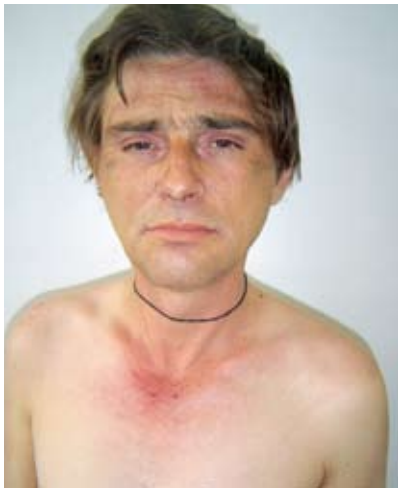

Figure 1. Periorbital violaceous erythema and V-shaped chest exanthema of a patient with paraneoplastic dermatomyositis associated with seminoma testis.

Clinical examination revealed periorbital edema and violaceous erythema of the eyelids (Fig. 1), erythematous plaques on the upper chest, and Gottron's papules on the wrists (Fig. 2). Severe muscle weakness involving the proximal limb muscles was noted; the patient could not raise his hands above his head. The patient's creatine kinase level was significantly elevated at 1,360 U/l, his ESR was slightly elevated (28 mm/h Westergreen), and his ASAT and ALAT were within normal ranges. Indirect immunofluorescence for ANA on HEp-2 cells and the anti-Jo-1 antibody were negative. Electromyography demonstrated multiple polyphasic low-amplitude potentials with sharp edges and short duration typical of myogenic injury. A skin biopsy from a lesion revealed atrophic epidermis with focal basal degeneration and perivascular lymphoid infiltration in the dermis.

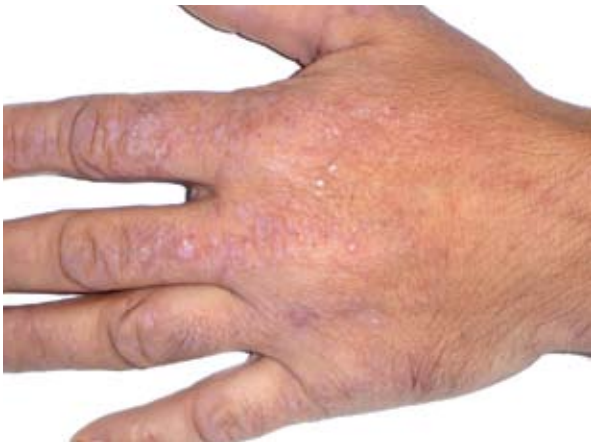

Figure 2. Gottron's papules on the wrist of the same patient.

The patient was initially treated with oral methylprednisolone 60 mg daily with decreasing doses, and a short course of azathioprine 100 mg/ daily, which markedly improved the skin lesions and muscle weakness. The patient has been in complete remission since September 2006 and has been maintained on 8 mg methylprednisolone daily and topical sun protection. At his last visit in May 2009, no clinical or laboratory sings of disease activity were evident.

Discussion

Although the association between DM and cancer is well recognized, only a limited number of reports of DM associated with testicular cancer have been reported.

DM is associated with many different types of malignant tumors, mainly carcinomas rather than sarcomas. Breast (3), colon, and lung carcinomas (6) were reported as the most frequent tumors, and ovarian carcinoma was found much more frequently in white women with DM than would be expected in the general population (9). Lymphomas, Hodgkin disease, myeloma, and thymoma are much rarer (4). The association of DM with testicular cancer appears to be very rare. Our review of the literature suggests that this is the eleventh case of such an association and only the second case of DM associated with seminoma (9–17) (Table 1). The reason could be the relative rareness of both disorders; nevertheless, in some cases, a strong relationship of testicular cancer and DM was evident, as in our patient.

Seminoma is a tumor considered to be relatively homogenous and developmentally close to the germ cells from which it derived during fetal development (18). As a histologic subtype of germ cell, seminoma tumors usually present in males between 15 and 35

Table 1. Case reports of paraneoplastic dermatomyositis (DM) associated with testicular tumors published in the literature.

Author	Year	Case	Malignancy
Fife RS et al. (9)	1984	DM after cancer treatment	Metastatic non-seminomatous germ cell tumor
Barker RA et al. (10)	1987	30-year-old man, DM precedes malignancy	Metastatic non-seminomatous germ cell tumor
Clayton A et al. (11)	1998	28-year-old man, DM 2 months after BEP chemotherapy; 30-year-old man, DM precedes malignancy	Malignant teratoma
Hayami S et al. (12)	1998	24-year-old man, DM precedes malignancy	Malignant teratoma
Ishizawa et al. (13)	1999	24-year-old man, DM precedes malignancy	Intratubular GCT
Di Stasi SM et al. (14)	2000	31-year-old man, DM 3 years after orchiectomy	Nonseminomatous GCT
von Heyden B et al. (15)	2000	46-year-old man, DM precedes malignancy	Seminoma
Yoshinaga A et al. (16)	2005	31-year-old man, DM precedes malignancy	Nonseminomatous testicular cancer
Curiel RV et al. (17)	2005	34-year-old man, DM 3 months after orchiectomy	Nonseminomatous testicular cancer

years old as a painless, homogenous, and enlarging testicular nodule (19). This observation underscores the substantial prevalence of patients with DM and testicular cancer presenting in the third decade of life, which contrasts with the age distribution in other paraneoplastic DM patients (20). Seminoma is confined to the testicles in about 70% of cases and metastasizes to the draining lymph nodes in the retroperitoneum in about 25% of cases (19). Metastatic disease is present in 5% of cases and spreads hematogenously to the lung parenchyma, bone, liver, or brain (21).

The malignancy can precede, occur concurrently with, or follow the appearance of DM (20). Thus the definition of clinical and laboratory markers for association with malignancy are of great importance for disease prognosis. Several predictive factors, such as the patient's age, male gender, cutaneous necroses, an increased ESR level, and very high or, alternately, normal serum levels of creatine kinase (CK) have been proposed (22–24). Our patient, however, fit only two of these criteria: a highly elevated CK serum level and male gender.

Recently Ishizawa et al. (13) focused on the clinical usefulness of LDH-1 as a marker of germ cell tumor appearance. LDH is frequently found to be elevated in patients with DM and polymyositis. However, an increased serum level of LDH-1 in a patient with DM might raise suspicion of a concomitant malignant germ cell tumor from either

the testis or ovary (13), and may have practical significance.

In conclusion, the risk of neoplasm is increased even in younger patients with DM. Early recognition of the characteristic skin rash may provide a clue to the diagnosis, and screening for neoplasm may improve the prognosis in patients with inflammatory myopathies.

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