Generalized Erythematous to Violaceous Papules and Confluent Patches with Vesiculobullous Lesions in a 53-year-old Man


CASE REPORT

A 53-year-old man, an alcoholic, developed itchy, scaling erythematous patches on large areas of the face, scalp, V area of the anterior neck and extensor extremities. Three months later, many tense vesicles and bullae followed by ulcers developed over his neck, abdomen (Fig. 1) and upper limbs. Besides, a widespread erythematous to violaceous papules and confluencing patches on the trunk, Gottron's papule-like lesions on the extensor surface of finger joints (Fig. 2), elbows and knees, poikiloderma over the neck, and periungual telangiectasia were noted. However, neither weakness nor tenderness of his proximal muscles was found.

Laboratory examination revealed elevation in serum LDH level of 141 IU/L (50-110), AST 26 IU/L (0-23 ), and ALT 33 IU/L (0-23). A test for ANA was negative and the serum levels of CPK and complements were normal. Systemic examinations were all negative for internal malignancy. Neither electromyography nor muscle biopsy showed definite evidence of myositis.

Fig. 1
Numerous tense vesicles and bullae on the abdomen.

Fig. 2
Gottron's papules over dorsal metacarpophalangeal and proximal interphalangeal joints.

Fig. 3
Photomicrograph reveals cell-poor subepidermal vesicle with marked dermal edema (H & E, X100).
DIAGNOSIS: Bullous Dermatomyositis

HISTOPATHOLOGICAL FINDING

A skin biopsy specimen from the vesicular lesion on the abdomen revealed cell-poor subepidermal vesicles, liquefactive degeneration of the epidermal basal layer with marked edema of the dermis, and a sparse perivascular infiltrate of lymphocytes in the upper dermis (Fig. 3). Alcian blue stain revealed only slight deposits of mucin in the upper dermis. The direct immunofluorescence (DIF) study did not show deposition of any immunoreactant in the basement membrane zone.

DISCUSSION

Dermatomyositis (DM) is a systemic disease characterized by inflammatory myopathy and variable cutaneous eruptions. Typical cutaneous manifestations of DM included Gottron’s papules of the skin overlying the metacarpal or interphalangeal zones, which are pathognomonic for DM, cuticular telangiectasis, peri orbital edema and heliotrope, erythematous papulosquamous eruption of the face and trunk, photosensitivity, and poikilodermia. Only a few references concerning vesicular and bullous lesions in DM can be found. Because of the rarity of this complication, its pathophysiology remains obscure. Although edema and mucin deposition were thought to be the major factors in the development of bullae in lesions of DM, it was not an absolute requirement. In our patient, the histopathologic findings revealed cell-poor subepidermal vesicles and marked edema with little mucin in the upper dermis, which suggested that the vesicles and bullae were caused by severe edema in the dermis. The DIF did not show the deposition of immunoreactants in the basement membrane zone. All these results made the diagnosis of other subepidermal blistering disorders unlikely.

The association of DM with internal malignant diseases has been known since 1960s. The incidence of internal malignancy in DM is 5% to 30%. It has been suggested that when DM is associated with unusual lesions such as vesicles and bullae, the presence of an internal malignancy and worse prognoses may be more likely. Kubo et al. pointed a strong association between DM with vesicles or bullae and internal malignant diseases, especially gynecologic malignant diseases. Furthermore, the appearance of vesicles and bullae may be a sign of a severe flare of the condition.

Besides, our patient manifested pathognomonic cutaneous lesions of DM in the absence of clinical evidence of muscle disease for longer than 6 months. During the 11-month treatment course, the laboratory data of our case showed persistently elevated levels of LDH, AST, ALT and TG despite marked improvement of the skin lesions, while no CPK elevation nor muscle weakness had been noted. Moreover, EMG and muscle biopsy revealed negative findings. Thus, our patient probably represents a case of amyopathic bullous DM.

REFERENCES