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A Case of Thymoma-Associated Paraneoplastic Pemphigus Presenting as Psoriasis

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Clinical Image

Thymoma was reported to be frequently associated with autoimmune diseases, including not only Myasthenia Gravis (MG), but also a variety of dermatoses, such as lichen planus, pemphigus vulgaris, pemphigus foliaceous, graft-vs-host disease, and Paraneoplastic Pemphigus (PNP) [1-3]. Here, we report a rare case of thymoma-associated pemphigus presenting as psoriasis.

A 48-year-old man presented with pruritic skin rash all over the body for 2 months. He underwent thymectomy (type B2 thymoma) 9 years ago, followed by postoperative chemotherapy and radiotherapy, but 2 years ago, pleural metastasis was found and he developed MG, which was controlled by pyridostigmine, prednisone and plasmapheresis. Two months before, he developed pruritic skin eruptions. Physical examination revealed annular erythemas with white scales on the trunk and extremities, no oral lesion was found (Figure 1a and 1b). Laboratory examination revealed an elevated SCC antigen level of >100ng/ml (normal 0-2.5 ng/ml). ANA, anti-SSA, anti-SSB, anti-dsDNA, and anti-Sm antibodies were all negative. Histopathology showed parakeratosis, psoriasis-like epidermal hyperplasia, necrotic keratinocytes, basal cell edema and lichenoid lymphocytes infiltration (Figure 1c). DIF showed positive intercellular IgG and C3 deposition. IIF identified positive intercellular IgG on rat bladder epithelium (titer 1: 80). Anti-Dsg1 and anti-Dsg3 antibodies were normal. CT scan revealed pleural metastasis of thymoma (Figure 1d). The patient was diagnosed as PNP based on the skin eruption, histopathology, IIF of rat bladder epithelium and pleural metastasis of the thymoma [4]. The lesions relieved partly with oral prednisone 40mg/d. The patient finally died from respiratory failure within half a year.

PNP is an autoimmune multi-organ syndrome with a diverse spectrum of clinical and immunopathological features. Clinical presentations of lesions associated with PNP include pemphigus, pemphigoid, erythema multiforme, graft-vs-host disease, and lichen planus [5,6]. However, PNP presenting psoriasis-like erythema without oral lesions as seen in our patient is rare. Pleural metastasis was found 2 years ago, with an elevated serum SCC level and no evidence of other tumors. The skin lesions of PNP and MG in this patient were considered to be associated with the pleural metastasis of the type B2 thymoma, which is invasive. The absence of anti-Dsg1 and anti-Dsg3 antibodies in our patient suggested the possibility of other target antigens [7].

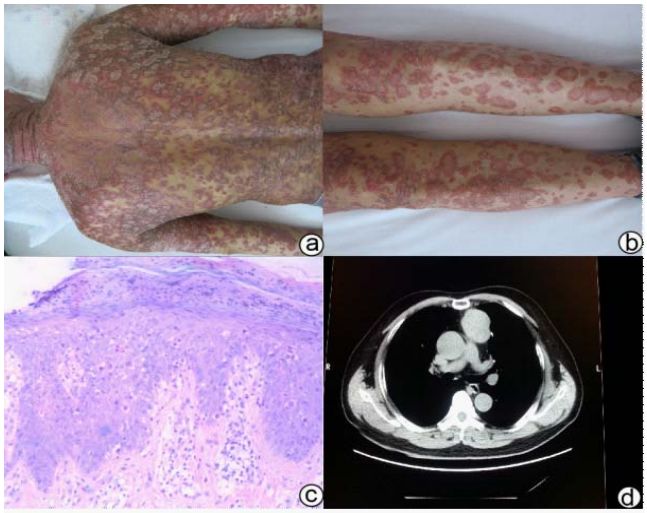

Figure 1: (a) Annular erythemas with white scales on the trunk; (b) annular erythemas with scales on the lower limbs; (c) histopathology of the lesion on the trunk showing hyperkeratosis, parakeratosis, psoriasis-like epidermal hyperplasia, necrotic keratinocytes, basal cell edema, and lichenoid lymphocytes infiltration (×100); (d) CT scan showing pleural metastasis (arrow).

Together, our case added new information to PNP and mucocutaneous manifestations associated with thymoma. Prednisone showed partially effect on skin lesions, and the prognosis of the patient might depend on the condition of the underlying thymoma.

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