Bullous Dermatomyositis Associated with Gastric Carcinoma
— A Case Report —

Tsung-Hua Tasi     Pa-Fan Hsiao     Hsin-Yi Su     Yang-Chih Lin

Cutaneous manifestations of dermatomyositis commonly include Gottron's papules, periorbital heliotrope rash, periungual erythema or telangiectasia, and poikiloderma. Vesicles or bullae have rarely been reported. We describe a patient with dermatomyositis who presented with vesicles and was subsequently diagnosed as having gastric carcinoma. It is important to recognize this vesiculo-bullous variant in order to avoid a delay in diagnosis. Vesiculo-bullous dermatomyositis may be associated with a higher incidence of internal malignancy than typical dermatomyositis and portend a poorer prognosis. (Dermatol Sinica 22 : 243-247, 2004)

Key words: Dermatomyositis, Vesiculo-bullous dermatomyositis, Gastric carcinoma

皮肌炎的皮膚表現通常包括Gottron氏丘疹，眼週圍的heliotrope紅疹，指甲週圍紅斑或血管擴張，以及多形皮膚萎縮。水泡的產生較少被報告過。我們報告一例以水泡表現的皮肌炎患者，她後來被診斷出有胃癌。早期辨認出這種以水泡表現的皮肌炎能避免延誤診斷。水泡型的皮肌炎可能有比典型皮肌炎更高的合併癌症比例和較不好的預後。（中華皮誌22 : 243-247, 2004）
INTRODUCTION
Dermatomyositis is a connective tissue disease that may present with a variety of cutaneous manifestations. Vesiculo-bullous dermatomyositis is a rare variant of dermatomyositis.1-12 A number of cases reported were associated with internal malignancy.1-4, 7-10 We describe a woman with vesiculo-bullous dermatomyositis associated with gastric cancer.

CASE REPORT
An 86-year-old woman had an erythematous eruption on the face, scalp, neck, chest and back for 2 months before she sought treatment. Her medical history was unremarkable other than for mild hypertension. She was not taking any medication. She denied friction or scratching the lesions. Three months after onset, she developed scattered vesicles and pustules on her chest and back (Fig. 1). There was no Gottron's papule, heliotrope rash or periungual telangiectasia at initial presentation. Major diagnostic considerations at that time included vesicular bullous pemphigoid, dermatitis herpetiformis, linear IgA dermatosis, epidermolysis bullosa acquisita, pemphigus foliaceus, bullous lupus erythematosus, and bullous dermatomyositis. A biopsy of a vesicle on her back showed subepidermal vesicles (Fig. 2), adjacent focal hydropic degeneration of basilar keratinocytes and colloid bodies, marked edema of the papillary dermis, and a mild superficial perivascular mononuclear cell infiltrate. A small amount of mucin in the upper dermis was demonstrated by alcian blue stain (pH 2.5). Direct immunofluorescence studies (DIF) showed no immunoglobulin deposits on the basement membrane and around blood vessels.

After admission, the number of vesicles and pustules on her chest and back increased; they soon ruptured and fused into large ulcerations (Fig. 3). One week later, the erythematous patches on the forehead, scalp, and chest turned deep red to purplish. The signs of heliotrope rash, Gottron's papules, and periungual telangiectasia became obvious and easily recognized. She also began to suffer from progressive dys-
phagia, and weakness of the proximal muscles. Her laboratory data were as follows: creatine kinase, 1038 U/L (normal 50 to 232 U/L), CK-MM, 99%; AST, 114 U/L (normal 5 to 35 U/L); antinuclear antibody, positive at 1:640 in a speckled pattern; WBC count, 10400 with 82% neutrophils; hemoglobin, 7.4 g/dL; and stool occult blood, 3+. Electromyography showed a myopathic pattern. Based on these findings, a diagnosis of vesiculo-bullous dermatomyositis was made. Further investigation of esophagogastroduodenoscopy with biopsy revealed advanced poorly-differentiated gastric adenocarcinoma. Although chest radiography was normal, computed tomography of the chest and abdomen showed regional lymphadenopathy and adrenal gland metastasis. Culture of aspiration from the pustules and ulcers grew heavy amounts of Pseudomonas aeruginosa.

Treatment for the skin infection was initiated with intravenous gentamicin (120 mg/d), and the ulcers dried and healed progressively. Once the diagnosis of dermatomyositis was confirmed, she was given intravenous dexamethasone (3.3 mg/d). No new vesicle appeared and the creatine kinase level also declined. However, she had several episodes of melena and coffee-ground vomitus, and her hemoglobin remained at low level despite several transfusions. Eighteen days after admission, she suddenly developed wheezing, hypoxemia, and aspiration pneumonia. She died of septic shock 6 days later.

**DISCUSSION**

Dermatomyositis is a connective tissue disease that may manifest with cutaneous eruptions, proximal muscle weakness, and abnormal laboratory values. Classical cutaneous findings include Gottron's papules, a peri orbital heliotrope rash, periungual erythema or telangiectasia, and poikiloderma. However, these features are seen in almost all cases, other skin findings have been observed on occasion. Less common

<table>
<thead>
<tr>
<th>Authors</th>
<th>Year</th>
<th>Age</th>
<th>Sex</th>
<th>Histopathologic findings</th>
<th>DIF</th>
<th>Site of malignancy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Niizke et al.²</td>
<td>1990</td>
<td>49</td>
<td>F</td>
<td>Subepidermal vesicle</td>
<td>Negative</td>
<td>Uterine tube</td>
</tr>
<tr>
<td>Ishikoh et al.³</td>
<td>1991</td>
<td>60</td>
<td>M</td>
<td>Subepidermal vesicle</td>
<td>Negative</td>
<td>Lung</td>
</tr>
<tr>
<td>Goto et al.³</td>
<td>1992</td>
<td>46</td>
<td>F</td>
<td>Subepidermal vesicle</td>
<td>Negative</td>
<td>Ovary</td>
</tr>
<tr>
<td>Jin et al.⁴</td>
<td>1992</td>
<td>34</td>
<td>F</td>
<td>Subepidermal vesicle</td>
<td>Negative</td>
<td>Mammary</td>
</tr>
<tr>
<td>Aranami et al.⁴</td>
<td>1993</td>
<td>60</td>
<td>M</td>
<td>Subepidermal vesicle</td>
<td>Negative</td>
<td>Not found</td>
</tr>
<tr>
<td>Komine et al.⁵</td>
<td>1993</td>
<td>66</td>
<td>F</td>
<td>Subepidermal vesicle</td>
<td>Negative</td>
<td>Not found</td>
</tr>
<tr>
<td>Kubo et al.(case 1)⁵</td>
<td>1996</td>
<td>58</td>
<td>F</td>
<td>Subepidermal vesicle</td>
<td>Negative</td>
<td>Ovary</td>
</tr>
<tr>
<td>Kubo et al.(case 2)⁵</td>
<td>1996</td>
<td>46</td>
<td>F</td>
<td>Subepidermal vesicle</td>
<td>Negative</td>
<td>Ovary</td>
</tr>
<tr>
<td>McCollough et al.⁶</td>
<td>1998</td>
<td>56</td>
<td>F</td>
<td>Subepidermal vesicle</td>
<td>Positive*</td>
<td>Ovary</td>
</tr>
<tr>
<td>(case 1)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>McCollough et al.⁶</td>
<td>1998</td>
<td>45</td>
<td>F</td>
<td>Subepidermal vesicle</td>
<td>Positive**</td>
<td>Not found</td>
</tr>
<tr>
<td>(case 2)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ang et al.⁹</td>
<td>1999</td>
<td>48</td>
<td>M</td>
<td>Dermal edema with perivascular lymphocytic infiltrate#</td>
<td>Positive***</td>
<td>Nasopharyngeal carcinoma</td>
</tr>
<tr>
<td>Lin et al.¹¹</td>
<td>2000</td>
<td>47</td>
<td>F</td>
<td>Subepidermal vesicle</td>
<td>Negative</td>
<td>Not found</td>
</tr>
<tr>
<td>Peng et al.¹²</td>
<td>2002</td>
<td>53</td>
<td>M</td>
<td>Subepidermal vesicle</td>
<td>Negative</td>
<td>Not found</td>
</tr>
<tr>
<td>Fujimoto et al.¹³</td>
<td>2002</td>
<td>56</td>
<td>M</td>
<td>Subepidermal vesicle</td>
<td>Negative</td>
<td>Stomach</td>
</tr>
<tr>
<td>Current case</td>
<td>86</td>
<td>F</td>
<td></td>
<td>Subepidermal vesicle</td>
<td>Negative</td>
<td>Stomach</td>
</tr>
</tbody>
</table>

DIF: Direct immunofluorescence test

* scattered cytoid staining for IgM and C3.
** slight deposition of C3 around vessels.
*** linear C3 and C1q along the dermoeidermal junction.
# biopsy from an erythematous patch, not from vesicles.

Table I. A summary of reported cases of vesiculo-bullous dermatomyositis

245 Dermatol Sinica, September 2004
cutaneous manifestations include cutaneous mucinosis, lichen planus, panniculitis, urticaria, cutaneous vasculitis, pityriasis rubra pilaris-like lesions (i.e., type Wong dermatomyositis) and vesiculo-bullous dermatomyositis.

There have been several reports of vesiculo-bullous dermatomyositis, which occurred predominantly in Eastern Asians. We reviewed previously reported cases of vesiculo-bullous dermatomyositis excluding those with other coexistent bullous diseases or without DIF reports. A summary of these 15 cases, including our case, is shown in Table I. In addition to two Caucasian cases, nine cases were Japanese, one case was Singaporean-Chinese, and the other three cases were Taiwanese. There were 5 males and 10 females. The ages of the patients ranged from 34 to 86 years, with a mean age of 54 years. Blisters and erosions have been described over the usual areas affected by dermatomyositis such as the extensors of the arms and upper chest and back. The histopathologic findings may not be specific. Usually a subepidermal vesicle or bulla is seen along with dermal edema, mucin deposition and superficial perivascular inflammation. DIF studies are usually negative, as seen in our patient, with few exceptions. The pathophysiology is thought to be due to marked dermal edema, as seen in our patient, or cell-mediated epidermal damage and necrosis. In two cases, vesicle or bulla arose as a consequence of abrupt discontinuation of corticosteroids, which induced an acute exacerbation of the disease. Vesicle or bulla formation in dermatomyositis has occasionally been reported occurring in conjunction with other immunobullous diseases, such as bullous pemphigoid, linear IgA bullous dermatosis, and dermatitis herpetiformis. These patients with concurrent immunobullous disorders all had DIF findings compatible with their specific bullous diseases.

The association of dermatomyositis with internal malignancy is well recognized and reported to occur in 15% to 34% of patients. In our review of previously reported cases of vesiculo-bullous dermatomyositis excluding those with other coexistent bullous diseases or without DIF reports (Table I), the incidence of malignancy in vesiculo-bullous dermatomyositis was 10 of 15 (67%), including our patient. Of the 7 women with malignancy, five had a gynecological cancer, one had a mammary carcinoma, and our patient had gastric carcinoma. This suggests that vesiculo-bullous dermatomyositis may be associated with a higher risk of malignancy than typical dermatomyositis, especially gynaecological cancers in female patients, and hence, a worse prognosis. Therefore, a detailed and comprehensive investigation for underlying malignancy is required in patients with vesiculo-bullous dermatomyositis.

Patients suffering from vesiculo-bullous dermatomyositis may also be more susceptible to wound infection because of immunosuppressed status caused by immunosuppressive therapy or underlying malignancy. Appropriate wound care, antibiotics, and corticosteroid therapy are essential to the control of both the vesicles and secondary infection.

In conclusion, we remind dermatologists should consider vesiculo-bullous dermatomyositis in the differential diagnosis of blistering disorders, especially in the presence of other suggestive clinical features. A subepidermal bulla with marked dermal edema, mucin deposition and negative DIF result raise the possibility of dermatomyositis. Furthermore, vesiculo-bullous dermatomyositis appears to be associated with a higher risk of malignancy than typical dermatomyositis. Early diagnosis of vesiculo-bullous dermatomyositis may prevent unnecessary delay in the institution of appropriate treatment and search for an underlying tumor.

REFERENCES
2. Ishikoh A, Kurihara S: [Two cases of dermatomyositis with unusual eruptions]. Jpn J Dermatol Sinica, September 2004 246


11. 林煥然，王莉芳，薑正祥: 皮肌炎伴隨水泡形成。台灣皮膚科醫學會第二十六屆年會暨學術討論會知識 103。

12. 彭成華，李建寬，蘇琳惠等: 皮肌炎合併水泡表現。台灣皮膚科醫學會第二十八屆年會暨學術討論會知識 140。


