A 45-year-old Woman with Erythroderma and Bullous Eruption: Erythrodermic Bullous Pemphigoid

INTRODUCTION

Erythrodermic Bullous Pemphigoid, first described by Tappeiner et al in 1982, is a rare variant of Bullous Pemphigoid characterized by erythroderma along with blister formation with only eight patients reported so far. The present case is the youngest to develop erythrodermic bullous pemphigoid.

CASE DESCRIPTION

45-year-old Indian woman presented with erythroderma for 3 1/2 months and bullous eruption for 2 weeks. It started with erythema and scaling in the neck accompanied with bullous eruption which ruptured in 2–3 days.

Histopathological examination showed sub-epidermal blistering. Direct immunofluorescence (DIF) of perilesional skin demonstrated linear deposition of C3 at dermo-epidermal junction. DIF for IgG and IgA was negative.

The patient was treated with 2.5 mg/kg body weight/day prednisolone and 2 mg/kg body weight/day azathioprine orally along with symptomatic treatment. She was also given three daily intravenous pulses of 100 mg dexamethasone to combat new lesions and resistant nature of the disease. The treatment led to almost complete disappearance of lesions. Despite this improvement, the patient expired following septicemia.

DISCUSSION

Erythrodermic BP is a rare variant of BP characterized by erythroderma along with blister formation. Clinical presentation of the present patient differed in respect of age (45 years) from most of the other reported cases.

It was also observed that onset of erythrodermic bullous pemphigoid at an early age may be associated with resistance to treatment.

KEYWORDS: Bullous pemphigoid, erythroderma, autoimmune blistering disorder

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INTRODUCTION

A 45-year-old Indian woman presented with erythroderma for 3 1/2 months and bullous eruption for 2 weeks. Erythema and scaling had started from neck and subsequently involved trunk and limbs in less than a week. Bullous eruption started on hands and feet and the bullae used to rupture in 2–3 days without subsequent peripheral extension. All lesions A were associated with itching and burning sensations. On examination, there was generalized erythema and fine scaling with multiple tense bullae over hands, thighs, and feet (Fig. 1). The skin surrounding the bullae was erythrodermic. Oral and genital erosions were also present. Patient’s general condition was good. Clinical diagnosis of pemphigus foliaceous was considered.

Tzanck smear was negative for acantholytic cells. Histopathological examination showed sub-epidermal blistering with numerous eosinophils and neutrophils in blister cavity. Dermis showed moderately dense superficial perivascular infiltrate mainly of lymphocytes and eosinophils. Periphery of blister showed spongiosis with eosinophils (Fig. 2). Direct immunofluorescence (DIF) of perilesional skin demonstrated linear deposition of C3 at dermo-epidermal junction (Fig. 3). DIF for IgG and IgA was negative. Final diagnosis of erythrodermic bullous pemphigoid (BP) was made.

The patient was treated with 2.5 mg/kg body weight/day prednisolone and 2 mg/kg body weight/day azathioprine orally along with symptomatic treatment on the basis of clinical suspicion of pemphigus foliaceous.
and pending the results of investigations. There was improvement in erythroderma, but bullous lesions continued to appear even after 4 weeks of this treatment. To control development of new lesions she was also given three daily intravenous pulses of 100 mg dexamethasone followed by three weekly pulses. This, along with daily prednisolone and azathioprine, led to almost complete disappearance of lesions. Despite this improvement, the patient expired following septicemia.

**DISCUSSION**

Bullous pemphigoid (BP) is an autoimmune blistering disease characterized histologically by sub-epidermal blister without epidermal necrosis and with a superficial infiltrate consisting of lymphocytes, histiocytes and eosinophils and immunopathologically by linear deposition of C3 (almost all cases) and/or IgG (about 80% cases) at the dermal-epidermal junction. The circulating IgG autoantibodies are directed against two epidermal hemi-desmosomal glycoproteins (BP Ag1 of 230 kDa; BP Ag2 of 180 kDa). Erythrodermic BP, first described by Tappeiner et al., is a rare variant of BP characterized by erythroderma along with blister formation. Only eight patients with this condition have been reported so far (Table 1). The present case is the youngest to develop erythrodermic bullous pemphigoid.

Clinical presentation of the present patient differed in respect of age (45 years) from most of the other reported cases. All previously reported cases, except one, had onset of disease in the eighth decade or later. Other causes of erythroderma and associations of pemphigoid were excluded by clinical examination and investigations in the present case.

Bullous pemphigoid is usually believed to be easily controlled by lower doses of glucocorticoids. There is some evidence to show equal efficacy of topical application of clobetasol propionate (0.05%) and oral

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**Fig. 1** Several small tense blisters with diffuse erythema and scaling over thigh (a) and forearm (b).

**Fig. 2** Photomicrograph showing subepidermal blistering with numerous eosinophils and neutrophils in the blister cavity. Dermis shows moderately dense superficial perivascular mixed infiltrate of lymphocytes and eosinophils. Papillary dermis shows edema and numerous eosinophils some of which are present at dermoepidermal junction (H&E ×400).

**Fig. 3** Linear deposition of C3 at the dermo-epidermal junction (direct immunofluorescence) (×400).
However, in the present case there was insufficient response to even high doses of oral prednisolone along with azathioprine. Of all cases of erythrodermic bullous pemphigoid reported so far (Table 1), the two cases who developed the disease earlier (in the fifth decade) had poor response to conventional treatment in contrast to good response obtained in the elderly patients. This suggests that onset of erythrodermic bullous pemphigoid at an early age may be associated with resistance to treatment.

REFERENCES


Table 1: Reported patients with erythrodermic bullous pemphigoid.*

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Gender (F/M)**</th>
<th>Age at onset</th>
<th>Blisters before or after erythroderma</th>
<th>DIF</th>
<th>IIF</th>
<th>IB</th>
<th>Response to conventional treatment</th>
<th>Reference</th>
</tr>
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<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>89</td>
<td>Before</td>
<td>Linear deposition of C3 in the BMZ</td>
<td>IgG (1:1280 anti-BMZ)</td>
<td>Not done</td>
<td>Good</td>
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<td>2</td>
<td>F</td>
<td>76</td>
<td>Before</td>
<td>Linear deposition of IgG and C3 in the BMZ</td>
<td>IgG (1:80 anti-BMZ)</td>
<td>180 kDa (BP Ag2) 230 kDa (BP Ag1)</td>
<td>Good</td>
<td>4</td>
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<tr>
<td>3</td>
<td>F</td>
<td>73</td>
<td>After</td>
<td>Linear deposition of C3 in the BMZ</td>
<td>IgG (1:20 anti-BMZ antibodies)</td>
<td>180 kDa (BP Ag2) 230 kDa (BP Ag1)</td>
<td>Good</td>
<td>5</td>
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<tr>
<td>4</td>
<td>F</td>
<td>79</td>
<td>Before</td>
<td>Linear deposition in the BMZ</td>
<td>Antibodies (1:200) anti-BMZ</td>
<td>Not done</td>
<td>Good</td>
<td>6</td>
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<tr>
<td>5</td>
<td>M</td>
<td>85</td>
<td>After</td>
<td>Linear IgG and C3 in the BMZ</td>
<td>IgG (1:20 anti-BMZ)</td>
<td>Not done</td>
<td>Good</td>
<td>7</td>
</tr>
<tr>
<td>6</td>
<td>M</td>
<td>85</td>
<td>Before</td>
<td>Linear IgG and C3 in the BMZ</td>
<td>IgG (1:20 anti-BMZ)</td>
<td>180 kDa (BP Ag2)</td>
<td>Good</td>
<td>7</td>
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<tr>
<td>7</td>
<td>M</td>
<td>87</td>
<td>Not mentioned</td>
<td>Linear IgG and C3 in the BMZ</td>
<td>Not done</td>
<td>Not done</td>
<td>Good</td>
<td>8</td>
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<tr>
<td>8</td>
<td>M</td>
<td>46</td>
<td>Before</td>
<td>Consistent with BP***</td>
<td>Consistent with BP***</td>
<td>Consistent with BP***</td>
<td>Poor</td>
<td>9</td>
</tr>
<tr>
<td>9</td>
<td>F</td>
<td>45</td>
<td>After</td>
<td>Linear deposition of C3 in the BMZ</td>
<td>Not done</td>
<td>Not done</td>
<td>Poor</td>
<td>Present case</td>
</tr>
</tbody>
</table>

*Two patients 6,10, who presented as erythroderma, had no bullous eruption despite having DIF findings suggestive of BP are not included. These patients had non-bullous or sine bulla pemphigoid, which is considered to be a distinct entity 11.

**F, Female; M, Male.

***We could not find the entire text of this paper, which is in Japanese. These statements are based on the abstract.
Woman with erythroderma and bullous eruption


