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Clinical Aspects and Treatment of Pityriasis Lichenoides Et Varioliformis Acuta: A Retrospective Vietnamese Study

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Abstract

BACKGROUND: Pityriasis lichenoides et varioliformis acuta (PLEVA) is an uncommon condition which presents acutely with papulo-vesicles that may develop necrotic, ulcerative, or hemorrhagic changes.

AIM: We studied clinical, and treatment characteristics of PLEVA hospitalised patients at our hospital from September 2009 to December 2014.

METHODS: The records of 15 PLEVA patients were retrospectively reviewed.

RESULTS: The median age of onset was 21.8 ± 18.81 (from 1 to 68), male to female ratio was 2/1. The common area of onset was trunk (60.0%) and extremities (33.3%). Clinical features were purpuric papules (100%), hemorrhagic crusted papules (46.7%), pustular purpuric papules (40%), and necrotic ulcerating lesions (13.3%).

CONCLUSION: All patients were received systemic antibiotics (macrolides: 53.3%, others: 46.7%), 2 patients were added immunosuppressive drugs. A 1-year-old patient died, others had a good response.

Introduction

Pityriasis lichenoides is a group of rare skin diseases, including Pityriasis lichenoides chronic (PLC) and Pityriasis lichenoides et varioliformis acuta (PLEVA) [1]. PLEVA may occur at any age, but most frequently occurs in children and young adults. The average age of disease in our study was 7.75 for children and 37.9 for adults. Survey of the gender distribution depicted that males had approximately 2 times more frequently than females; this data is similar to the study of Jandrei Rogério Markus [2].

Lesions can occur on any area of the body, except palms and soles, and are characterised by the sudden onset of red papules that quickly develop into scaling papules. The papules may erode into crusted red-brown spots and then turn into a central necrotic ulcer, leaving scars after healing — this condition responses well to erythromycin and UV therapy [3].

This study aims to describe the clinical features and treatment methods of PLEVA at National hospital of dermatology and Venereology (Vietnam).

Methods

A total of 15 patients were enrolled from September 2009 to December 2014. The age of onset

was 21.8 years (range: 1-68 years). The majority of patients were males (67.7%). The lesions located on many parts of the body, except palms and soles, but trunk and extremities were most frequently affected areas of onset, which accounted for 60%.

Results

Clinical features were purpuric papules (100%), hemorrhagic crusted papules (46.7%), pustular purpuric papules (40%), necrotic ulcerating lesions (13.3%). The treatment proposed was erythromycin or other macrolide antibiotics in 8 patients (53.3%) and not macrolide in 7 patients (46.7%); 1 patient was treated by phototherapy combined with erythromycin, and 2 patients have also received immunosuppressive drugs. A 1-year-old patient died, others had a good response as shown in Table 1.

Table 1: Patient demographics, clinical and treatment characteristics

Parameter	Value
Age (years), mean (SD), min/max	21.8 (18.8); 1/68
Children (n/age)	8/7.75 (3.2)
Adults (n/age)	7/37.9 (16.2)
Sex men, n (%)	10 (67.7)
Area of onset, n (%)	
Chest – Abdomen	8 (53.3)
Back – Buttocks	1 (6.7)
Upper limbs (except palms)	3 (20.0)
Lower limbs (except soles)	2 (13.3)
Armpits - Groin – Genitals	0 (0.0)
Scalp - Face – Neck	1 (6.7)
Rates of skin lesion, n (%)	
Purpuric papules	15 (100.0)
Hemorrhagic crusted papules	7 (46.7)
Pustular purpuric papules	6 (40.0)
Necrotic ulcerating lesions	2 (13.3)
Other symptoms	
Itching	8 (53.3)
Fever	7 (46.7)
Pneumonia	2 (13.3)
Anaemia	3 (20.0)
Leucocytosis	7 (46.7)
Dead	1 (6.7)
Oral drugs	
Erythromycin	6 (40.0)
Other Macrolide antibiotics	2 (13.3)
Other antibiotics	7 (46.7)
Phototherapy	1 (6.7)
Immunosuppressive drugs	2 (13.3)

N = 15.

Discussion

In our study, trunk area and extremities accounted for 60.0% and 33.3% respectively. These data are constant to other studies in the literature [3], [4]. The skin lesions are divided into 4 types: purpuric papules (100%), hemorrhagic crusted papules (46.7%), pustular purpuric papules (40%), necrotic ulcerating lesions (13.3%). According to Pradeen et al., [5], 100% PLEVA patients had purpuric papules while the necrotic ulcerating lesions were quite rare than demonstrated Wahie S et al., (24.6%) [3].

Patients with PLEVA may have a fever which can be considered as a sign of infection. In this study, fever and infections occurred in 46.7% and 20.0% of patients, respectively. Two patients suffered from pneumonia. In literature, it was reported that internal organs could be involved in many cases with abdominal or joint pain and even central nervous system could be damaged [6]. In our study, all patients received oral antibiotics (erythromycin: 40%), immunosuppressive drugs (methotrexate, corticosteroids) were given in severe cases and a single case using phototherapy (PUVA). A 1-year-old patient died. She was hospitalised for high fever, widespread hemorrhagic crusted papules and necrotic ulcerating lesions, pneumonia, anemia, leukocytosis and diagnosed with Febrile Ulceronecrotic Mucha-Habermann disease (a severe form of PLEVA). Other patients had good response.

The experience in National Hospital of Dermatology and Venereology (Vietnam) from September 2009 to December 2014 demonstrated that PLEVA occurred at any age but was most common in children and young adults, clinical manifestations were acute, diverse and most patients had good response to treatment with macrolide. Only one patient died for Ulceronecrotic Mucha-Habermann disease.

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