ORIGINAL REPORT

Pemphigoid Gestationis: A Retrospective Study in Southwest Iran

Sima Rassai[1], Nader Pazyar[1], Seyed Milad Alavi[1], Afshin Kazerouni[1], and Amir Feily[2]

[1] *Department of Dermatology, Jundishapur University of Medical Sciences, Ahvaz, Iran*

[1] *Skin and Stem Cell Research Center, Tehran University of Medical Sciences, Tehran, Iran*

Received: 26 Apr. 2012; Received in revised form: 22 Nov. 2012; Accepted: 5 Jan. 2013

Abstract- Pemphigoid gestationis (PG) is a rare autoimmune bullous dermatosis of pregnancy usually presents in the second or third trimester. It is characterized by pruritic, urticarial plaques with the development of tense vesicles and bullae within the lesions. Pathogenesis of PG is not fully established, however, most patients develop circulating autoantibodies targeting the bullous pemphigoid (BP) 180 antigen. The aim of this work is to draw a profile of the epidemiology, clinical aspects, treatment and evolution of the disease by studying hospital series. We retrospectively investigated the 13 patients who were diagnosed with PG based on hospital data at the Referral Center of Southwest Iran located in Ahvaz city between March 2002 and March 2011. The age of onset was 21 to 40 years (mean age: 27.5 years). The onset of the disease occurred in the second trimester of pregnancy in 6 patients and in the third trimester of pregnancy in 4 patients. One patient had a flare up of disease during the first trimester and two out of cases in puerperium period. In all cases, pruritus was the first symptom, followed by an erythematous vesiculobullous eruption. The diagnosis of PG was confirmed by skin biopsy. Ten out of the patients treated with oral corticosteroids (0.5-1 mg/kg/day), one of the patients underwent oral corticosteroids plus topical glucocorticoid and the last patient treated with topical glucocorticoid. PG remains a rare dermatosis of pregnancy. Our series had two particularities compared to other studies: high frequency in primigravida and the frequent involvement of the face. Additionally our study demonstrated that improvement could occur faster and provide acceptable management if the treatment of the patients would be implemented sooner.

Acta Medica Iranica, 2013; 51(6): 408-410.

Keywords: Autoimmune; Bullous dermatosis; Pemphigoid gestationis

Introduction

Pemphigoid gestationis (herpes gestationis) is a rare devastating autoimmune bullous dermatoses occurring in approximately one in 50,000 pregnancies (1-3). It usually presents during the second or third trimester, but it may be present at any stage of pregnancy or the puerperium (3). Classically, pemphigoid gestationis (PG) is characterized by pruritic, urticarial plaques and tense vesicles and bullae within the lesions (4). The periumbilical site is usually the first area involved. Pruritus is a prominent symptom associated with the onset of disease (3). Blister formation is due to a complex mechanism involving T_{h2} lymphocytes, cytokines and polymorphonuclear cells (5). In routine histological examination a subepidermal vesicle detected (6). The purpose of this study was to demonstrate the demographic data, clinico-pathologic features and therapeutic outcome of the patients with PG at the Referral Center of Southwest Iran located in the city of Ahvaz between March 2002 and March 2011.

Materials and Methods

We reviewed the clinical records of the patients who were diagnosed PG and followed at the Referral Center of Southwest Iran located in the city of Ahvaz between March 2002 and March 2011. Demographic data, clinical manifestation including the time of disease onset, site of lesions, frequency in multigravida and primigravida and treatment modalities were analyzed.

Corresponding Author: Nader Pazyar
Department of Dermatology, Azadegan Street, Imam Khomeini Hospital, Ahvaz, Iran
Tel:+98 918 8617133, E-mail: dr.pazyar@gmail.com

Results

Demographic data and clinical presentations

Thirteen patients with PG were diagnosed in our department between March 2002 and March 2011. The patients age ranged from 21 to 40 years (mean, 27.5 years). The mean gestational age was 28.5 week. Seven out of the 13 cases were primigravida and 6 cases were multigravida. The onset of the disease was reported in the second trimester of pregnancy in 6 patients and in the third trimester of pregnancy in 4 patients but one patient had a flare up of disease during the first trimester and two out of cases in puerperium period. The main complaint of the patients was itching. The lesions started from abdomen especially periumblical area in nine cases, lower extremities in two patients and upper extremities in other two cases. Mucosal involvement was not seen but facial lesions detected in the patients. They had no history of the other autoimmune diseases (such as Grave's disease) or malignancies. Six out of patients developed the disease after previous pregnancies and seven out of patients had PG during their first pregnancy.

Histopathological findings

Since immunofluorescent microscopy is not available worldwide, the diagnosis of our PG patient was done by skin biopsy and clinical measures. Histological examination revealed subepidermal blistering with inflammatory eosinophilic infiltrations.

Treatment

Oral glucocorticoid (prednisolone, initial dose: 0.5-1.0 mg/kg/day) was administered to 11out of the 13 patients. One of the patients was treated with topical corticosteroid and the last patient underwent combination of systemic and topical corticosteroids. The patients were observed for new lesions. Corticosteroids were gradually withdrawn after the patients were free of the lesions.

Discussion

PG is a rare autoimmune subepidermal bullous disease that occurs during pregnancy and postpartum (7). The patients classically show a diffuse blistering and intensely pruritic eruption that begins periumbilically and spreads to involve the rest of the body (8). Pruritic urticarial papules and plaques of pregnancy (PUPPP) are among the most common dermatoses of pregnancy (9-11). PG must be differentiated from PUPPP. Generally PG presents earlier in pregnancy and involves the lower abdomen while sparing the striae. Involvement in or near the umbilicus is common. Target, polycyclic, or vesicular lesions occasionally predominate (no bullae) (10,11). Cobo *et al.* investigated seven cases of PG with mean age 30 years (4). Disease onset was reported in the second trimester of pregnancy in four patients and in the third trimester of pregnancy in three patients (4). One patient had a flare up of symptoms during the puerperium. Main sites of involvement were lower limbs (mainly thighs), forearms, trunk, and abdomen. None of the patients had facial or mucosal lesions (4). In 2003 Boudaya *et al*. reported 15 patients with PG (12). This study demonstrated the late occurrence of PG during the course of pregnancy, the high frequency of multigravida women, however two particularities observed: the frequent involvement of the face and the efficiency of dapsone (12). Our study revealed that demographic data, clinical presentation, laboratory findings, and treatment of the patients. Our PG patients shared many similarities with many other studies performed worldwide. Our series is comparable to the literature in the occurrence of PG during the second and third trimester of pregnancy, initial lesions on the abdomen especially periumblical, the lack involvement of mucosa however two particularities were observed: high frequency in primigravida women, and the frequent involvement of the face. Regarding treatment, the majority of our patients were controlled with systemic corticosteroids however one patient received topical corticosteroid and other one underwent systemic and topical corticosteroids. Additionally, our study demonstrated that improvement could occur faster and provide acceptable management if the treatment of the patients would be implemented sooner.

Acknowledgments

The source of data which was used in this study was from general practitioner thesis of Dr Alavi and financial support was provided by Jundishapur University of Medical Sciences.

References

1. Rodrigues Cdos S, Filipe P, Solana Mdel M, Soares de Almeida L, Cirne de Castro J, Gomes MM. Persistent herpes gestationis treated with high-dose intravenous immunoglobulin. Acta Derm Venereol 2007;87(2):184-6.
2. Sachdeva S. The dermatoses of pregnancy. Indian J Dermatol 2008;53(3):103-5.

3. Daneshpazhooh M, Chams-Davatchi C, Payandemehr P, Nassiri S, Valikhani M, Safai-Naraghi Z. Spectrum of autoimmune bullous diseases in Iran: a 10-year review. Int J Dermatol 2012 ;51(1):35-41.
4. Cobo MF, Santi CG, Maruta CW, Aoki V. Pemphigoid gestationis: clinical and laboratory evaluation. Clinics (Sao Paulo) 2009;64(11):1043-7.
5. Lu PD, Ralston J, Kamino H, Stein JA. Pemphigoid gestationis. Dermatol Online J 2010;16(11):10.
6. Ingen-Housz-Oro S. Pemphigoid gestationis: a review. Ann Dermatol Venereol 2011;138(3):209-13.
7. Lipozenčić J, Ljubojevic S, Bukvić-Mokos Z. Pemphigoid gestationis. Clin Dermatol 2012;30(1):51-5.
8. Campbell SM, Balazs K, Conroy M. Pemphigoid gestationis: a case report and review of the literature. Cutis 2011;88(1):21-6.
9. Lipozenčić J, Ljubojevic S, Bukvić-Mokos Z Pemphigoid gestationis. Clin Dermatol 2012;30(1):51-5.
10. Scheinfeld N. Pruritic urticarial papules and plaques of pregnancy wholly abated with one week twice daily application of fluticasone propionate lotion: a case report and review of the literature. Dermatol Online J 2008;14(11):4.
11. Adams SP. Dermacase. Pruritic urticarial papules and plaques of pregnancy (PUPPP). Can Fam Physician 1998;44:1609-18.
12. Boudaya S, Turki H, Meziou TJ, Marrekchi S, Bouassida S, Zahaf A. Pemphigoid gestationis: a study of 15 cases . J Gynecol Obstet Biol Reprod (Paris) 2003;32(1):30-4