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Pruritic Urticarial Papules and Plaques of Pregnancy with Unique Distribution Developing in Postpartum Period

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Dear Editor:

Pruritic urticarial papules and plaques of pregnancy (PUPPP) is one of the most common diseases associated with pregnancy, and is characterized by urticarial papules and plaques with pruritus on the abdomen, buttocks and thighs[1]. In most cases, the skin lesions develop in the third trimester of primigravida and disappear within 7 to 10 days after labor[1]. Lesions mostly appear first on the abdomen, and then spread to the proximal extremities. Therefore, the abdomen is involved in most cases, especially the stria distensae. A 30-year-old female patient visited our department due to pruritic erythematous papules and plaques on both arms and both legs (Fig. 1A～C). She complained that the erythematous skin lesions had first developed on both legs and were very pruritic. The lesions then spread to both arms. The patient's abdomen was spared. She went through labor seven days before the lesions developed in both thighs. She was in a postpartum period, which is the period beginning immediately after the birth of a child and extending for about six weeks. It was her first labor and a single pregnancy. She had no specific medical or dermatological history. We did laboratory studies including complete blood counts, liver function tests, renal function tests, thyroid function tests urinalysis and autoimmune study. She has no specific abnormal findings during these studies. We performed a biopsy of the lower leg for an exact diagnosis. Histopathological findings showed spongiosis of the epidermis, edema of the papillary dermis and perivascular infiltration of lymphocytes and eosinophils (Fig. 1D, E). The results of direct immunofluorescence were negative. The patient began to take prednisolone 20 mg daily for 4 days and then tapered to 5 mg per week. She was also treated with an oral antihistamine and a topical corticosteroid. After two weeks of treatment, the patient's symptoms of pruritus and erythematous skin rash were improved. In most cases, this disorder develops in the third trimester of pregnancy. Tiny pruritic erythematous papules first appear in the striae distensae of the abdomen, and then spread to the buttocks and legs. In our case, multiple pruritic erythematous papules and plaques occurred after labor, and the lesions were limited to the legs and arms, sparing the abdomen. Postpartum PUPPP is very rare (Table 1)[2-5]. In previous cases, the lesions first developed on the abdomen, and then spread to other parts of the body. In our patient, the pruritic skin lesions were limited to the extremities, while the abdomen was spared. Some previous cases also exhibited unique distributions. In contrast to all these cases, our case spared the abdomen and involved only the extremities. Generally, histological findings of PUPPP showed dyskeratosis, spongiosis of the epidermis, edema of the papillary dermis and perivascular lymphocytic infiltrations[1]. Direct immunofluorescence studies are negative. We also noted these histological features in our case. Based on these histopathological and clinical findings, we diagnosed the case as PUPPP developed in postpartum

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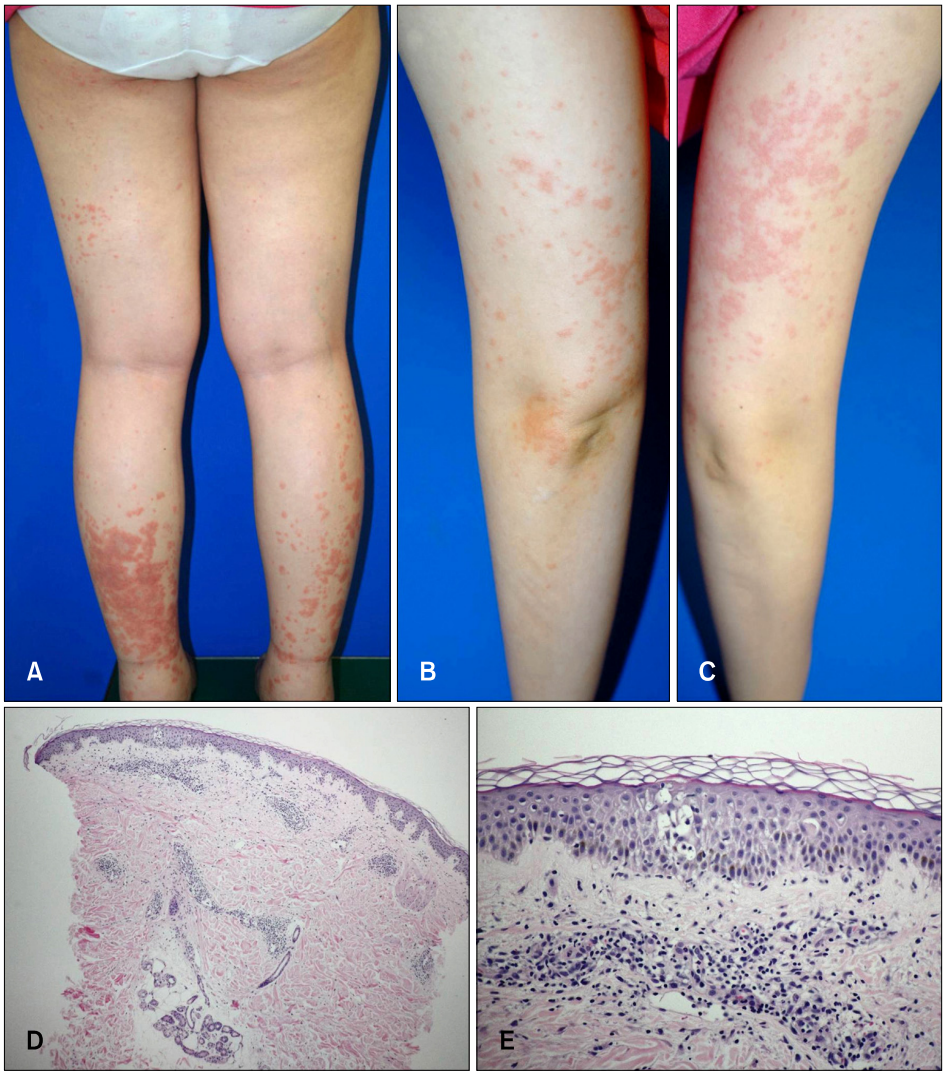

Fig. 1. Multiple itchy erythematous papules and plaques were observed on both the arms and legs. (A) Lesions on the lower extremities. (B, C) Lesions on the upper extremity. Histopathology of the skin lesions. (D) There were mild epidermal spongiosis and perivascular inflammatory cell infiltration (H&E, ×40). (E) At high power magnification, the epidermal focal spongiotic vesicle and perivascular infiltration of many lymphoid cells and few eosinophils were observed (H&E, ×100).

Table 1. Summary of postpartum pruritic urticarial papules and plaques of pregnancy cases

Patient No.	Age (yr)/ gender	Period after labor	Distribution	Treatment	Author (year)
1	23/F	4 weeks	Abdomen, thighs, face	A	Kirkup and Dunnill[2] (2002)
2	25/F	10 days	Abdomen, buttock, all extremities	C	Buccolo and Viera[3] (2005)
3	34/F	10 days	Abdomen, thigh	A, B, C	Byun et al.[4] (2010)
4	31/F	4 weeks	Abdomen, buttock, breast, arms	A, B, C	Byun et al.[4] (2010)
5	20/F	3 days	Abdomen, thighs, forearms, palmoplantar regions	A, B	Özcan et al.[5] (2011)
6	30/F	7 days	All extremities	A, B, C	Current case

F: female, A: topical corticosteroid, B: oral prednisolone, C: oral antihistamine.

period. Although our case was similar to urticarial vasculitis, clinically, the specimen did not show findings of leukocytoclastic vasculitis, and she improved without hyperpigmentation. Thus, we can rule out the urticarial vasculitis. Our case characterized itself by postpartum-period developments which simultaneously show unique distributions of the disease that only limits the lesions to the extremities and sparing abdomen. This pattern of the disease has never been reported, and thus, display the strength of our case report. In conclusion, we report a case of PUPPP in which the lesions developed after labor and were limited to both the legs and arms.

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A Novel Missense Mutation of Keratin 17 Gene in a Chinese Family with Steatocystoma Multiplex

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Dear Editor:

Steatocystoma multiplex (SM; OMIM184500) is a rare disorder of the pilosebaceous unit which is characterized by multiple sebum-containing dermal cysts. This disorder inherits an autosomal dominant mode, however, most sporadic cases have also been described. It may be associated with pachyonychia congenita, hypertrophic lichen planus, acrokeratosis verruciformis and so on[1]. Mutations in the keratin 17 gene (*KRT17*) underlie SM as well as pachyonychia congenita type 2. Thus, it is likely that these two conditions are phenotypic variants of the same disorder for some patients[2,3].

We reported a rare mutation of *KRT17* in two patients from a Chinese SM family. The proband, a 22-year-old male, presented with many small cysts for 7 years. The cysts originally occurred in his chest, and then gradually involved other parts of his body and grew larger. On examination, numerous cysts with 0.1 to 0.5 cm in diameter were diffusely distributed around his entire body (Fig. 1A, B). All his fingernails and toenails were normal. His father also shared similar clinical features (Fig. 1C). The histopathology of biopsy taken from his upper chest showed flattened sebaceous lobules close to the cystic wall which consists of stratified squamous epithelia without a granular layer (Fig. 1D). Based on the clinical and histopathological features, the diagnosis of SM was established.

We collected blood samples and extracted genome DNA, and then carried out mutation analysis of *KRT17* by direct sequencing when using the previous primers[4] in all

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