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Congenital Form of Isolated Benign Primary Cutaneous Plasmacytosis in a Child

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Dear Editor:

Lymphoplasmacytic proliferation in the skin can be seen in various disorders from secondary syphilis to malignancies including cutaneoues involvement of multiple myeloma[1]. Among various conditions, isolated benign primary cutaneous plamacytosis (PCP) is known as a rare entity and is characterized by the infiltration of plasma cells in the dermis without systemic involvement. Isolated benign PCP, a rare disease itself, in childhood or at birth is extremely rare, with only four reported cases in the literature[2,3]. Herein, we present a congenital form of isolated benign PCP. A 1-year-old female visited Ajou University Hospital for a skin rash on her right lower leg noted at birth. The skin rash was a small plaque at birth but became insidiously larger in size. She had been using a topical steroid on the rash before visiting us, but claimed there was no improvement. Physical examination revealed a solitary asymptomatic erythematous plaque on the right calf (Fig. 1). Her general appearance was good, and no enlarged lymph nodes were palpated. She had no previous surgicomedical history or traumatic episodes. Laboratory tests including a complete blood count and serum chemistry were within the references ranges. The result of the venereal disease research laboratory (VDRL) test was negative and serum protein and serum immunoeletrophoresis were within the normal limits. A skin biopsy revealed psoriasiform hyperplasia with dense lymphoplasma cells infiltration in the superficial and deep dermis without atypia (Fig. 2A). Through immunopero-

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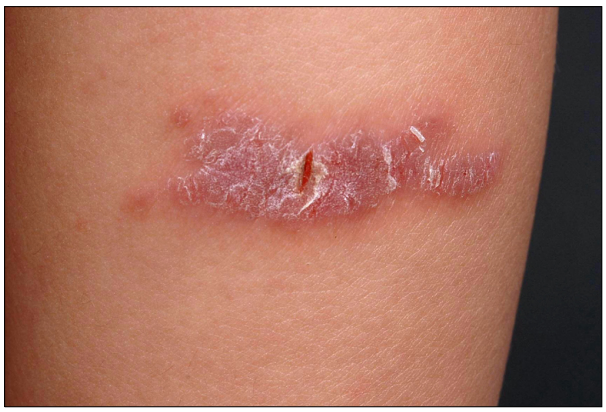

Fig. 1. A solitary asymptomatic erythematous plaque on the right lower leg.

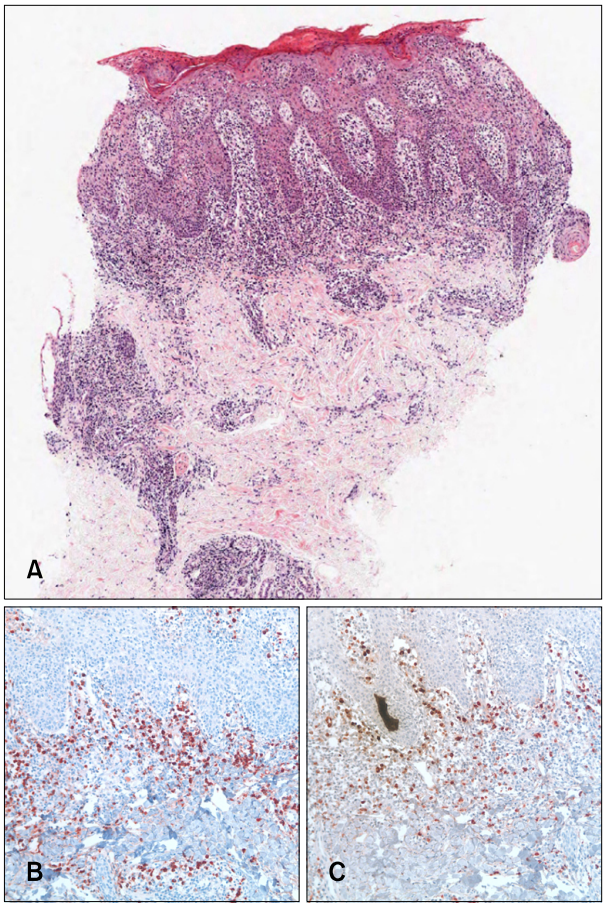

Fig. 2. (A) Psoriasiform hyperplasia with dense lymphoplasma cells infiltration in the superficial and deep dermis without atypia (H&E, ×10). Polyclonal hypergammaglobulinemia was proven with Kappa light chain stain (B) and lambda light chain (C) (immunoperoxidase stain, ×200).

xidase stains, dense expressions of both kappa and lambda light chains were observed (Fig. 2B, C). Based on these clinicopathologic and laboratory findings, we diagnosed her with isolated benign PCP. We prescribed methylprednisolone aceponate cream. One month later, the lesion was slightly improved showing a mild erythematous patch but was not yet fully recovered. She is now undergoing regular follow-up for 3 months, and the skin lesion remains on her leg. Primary and systemic cutaneous plasmacytosis are uncommon[4]. They show erythematous plaques or nodules on the head, neck, and trunk with or without lymphadenopathy. Histopathologically, they are characterized by infiltration of mature plasma cells with polyclonal hypergammaglobulinemia localized within the skin[4]. Among these conditions, isolated benign PCP is extremely rare, especially in children. Our case is isolated benign PCP in a child and thought to be a rare case showing a congenital form of this disorder. Although interleukin-6, human herpes virus-8 is suggested to be related to PCP, the exact pathomechanism is unknown[3]. Further studies will be necessary to elucidate the exact pathogenesis of PCP. The treatment of systemic cutaneous plasmacytosis is performed with steroids and cyclophosphamide[1]. While there is no definite treatment guideline for PCP, especially for isolated benign PCP, various trials can be carried out regarding the treatment of PCP including oral, topical corticosteroids, and intra-lesional injection of corticosteroids[5]. Herein, we report a case of congenital skin rash on the leg characterized by an erythematous asymptomatic plaque without systemic involvement and infiltration of mature plasma cells but with polyclonal hypergammaglobulinemia restricted to the skin. We believe this is the first time this congenital form has been reported in the literature, and thus warrants further investigation.

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