A Case of Disseminated Perforating Granuloma Annulare in a Child

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Granuloma annulare is a common, benign, inflammatory, usually self-limited dermatosis. Disseminated perforating granuloma annulare is a rare variant of granuloma annulare.

A 23-month-old female patient had asymptomatic, multiple umbilicated papules on the face, limbs including palms and soles, buttock and trunk for about 5 months. The new lesions have developed with mild fever or symptoms of upper respiratory infection. Histopathological examination revealed transepidermal elimination of mucinous degenerated collagen fibers and surrounding palisading lymphohistiocytic granuloma. After she was treated with prednisolone and antibiotics, the lesions slightly resolved, but thereafter, new lesions have developed frequently with mild fever.

We herein reported a rare case of disseminated perforating granuloma annulare in the youngest patient yet reported. (Ann Dermatol 8:(3)223~226, 1996).

Key Words: Disseminated perforating granuloma annulare, Transepidermal elimination.

Disseminated (or generalized) perforating granuloma annulare (DPGA) is a rare variant of granuloma annulare (GA) which was first described by Duncan et al. in 1973. It is characterized by central umbilicated papules on the whole body, and is most commonly seen in children and young adults. Transepidermal elimination of mucinous, degenerating collagen fibers and surrounding palisading lymphohistiocytic granulomas are important histologic features.

We herein reported a rare case of DPGA in the youngest patient yet recorded in the literature.

REPORT OF A CASE

A 23-month-old female patient had intermittently pruritic, multiple papules on the face, limbs including palms and soles, buttock, and lower part of the trunk for about 5 months (Fig. 1). Physical examination revealed multiple, 2 to 5 mm-sized, erythe matous papules that gradually developed atrophic centers covered with some scales or crusts, and several healed depressed scars. New lesions have developed with mild fever or symptoms of upper respiratory infection.

In the laboratory study, except for WBC count (11,040/mm³), the results of complete blood cell count, liver function test, chest roentgenogram, and urine analysis were within normal limits, and surface antibody of hepatitis B virus was positive.

Histopathological examination revealed epidermal perforation, collagen degeneration surrounded by palisading epitheloid and lymphohistiocytic cells in the dermis, and transepidermal elimination of degenerative collagen and cell debris through the perforated epidermis (Fig. 2, 3, 4). Mucin was demonstrated within degenerated collagen bundles by alcian blue stain. Immunohistochemical staining of α1-antichy-motrypsin revealed a positive reaction of activated macrophages in the granuloma (Fig. 5), but S-100 protein and α1-antitrypsin stain were negative.
Fig. 1. Multiple 2-5 mm sized erythematous non-perforating or perforating papules and healed depressed scars on the face, buttock, and forearm.

After she was treated by oral prednisolone for 7 days, the lesions slightly resolved. However, new lesions have developed 7 times in the last 9 months with mild fever, and some previous lesions have become erythematous. Subsequent lesions have been controlled by antibiotics and antihistamines.

Fig. 2. There is an epidermal perforation (arrow head) communicated with areas of degenerated collagen surrounded by granuloma in the dermis (H&E, × 40).

Fig. 3. Degenerated collagen and peripheral palisading of epitheloid and lymphohistiocytic cells (H&E, × 200).

Fig. 4. α1-antichymotrypsin staining revealed activated macrophage in the granuloma (ABC staining, × 40).
DISCUSSION

Granuloma annulare (GA) is a common, benign inflammatory, usually self-limited dermatosis. Unusual clinical variants of GA include generalized GA, perforating GA, erythematous GA, and subcutaneous GA. All types of GA share the similar histologic findings of palisading granulomas, and the epidermis is usually normal except in the perforating lesion.

Perforating GA is a rare clinical variant of GA first described by Civatte in 1952. It is characterized by the presence of asymptomatic, small, firm papules usually located over the distal part of limbs, although localized and generalized forms have been reported, and it can be associated with mild pruritus or pain. Females are affected twice as frequently as males, and seasonal variations have been described with the clearing of lesions during the winter and exacerbation during the summer. Generalized GA has been reported with diabetes, a later age of onset, rare spontaneous resolution, and poor response to therapy.

Perforating GA is of unknown etiology, although the basic pathogenesis of GA has been postulated to involve immune complexes with participation of the helper-inducer T cell subset. Necrotizing changes suggesting vasculitis are present in the dermal vessel, but obvious acute leukocytoclastic vasculitis is uncommon. The nature of the antigen eliciting the vasculitis is unknown, but several pathogenic or precipitating factors have been implicated, including ultraviolet light, insect bite, trauma, viral infections, thyroiditis, and vitamin D administration. We suggest that the necrotizing changes in our case are hypersensitivity vasculitis following a viral infection, because the lesions showed generalized distribution, and they have recurred with fever and other symptoms of the upper respiratory infection.

Histopathological findings of the well-developed papules are epidermal perforations that communicated with areas of necrobiosis in the dermis. The necrobioitic areas are surrounded by palisading lymphohistiocyte. The perforation of epidermis contains a plug of cellular debris, necrotic tissue, and mucinous material. In the adjacent epidermis, there were mild acanthosis and hyperkeratosis.

In some cases, the elimination is transfollicular, in other cases, there is merely a central ulcer rather than true transepidermal elimination.

The initial clinical differential diagnosis of the findings in our case were papulonecrotic tuberculid, molluscum contagiosum, PLEVA (pityriasis lichenoides et varioliformis acuta), Gianotti-Crosti syndrome and sarcoidosis which we ruled out by the clinical and biopsy findings.

GA has a spontaneous remission in about 50% of patients in two years, and recurrences in the same site in about 40% of patients. However, various forms of treatment have been tried and some variants of GA, particulary DPGA, are often difficult to treat. Intradermal injections of triamcinolone suspension are the treatment of choice for the more localized type. In generalized form, systemic steroids may be very effective, but side effects of high doses are dangerous. Schleicher et al. reported successful therapy for six cases of disseminated GA, and Ratnavel et al. reported in 1995 for 2 cases of perforating GA with isotretinoin. Other forms of treatment are topical steroid, dapsone, niacinamide, potassium iodide, chloroquine, alkylating agents, cryotherapy, surgery, electrocautery, and X-ray therapy.

To our knowledge, the earliest onset of DPGA was previously reported by Izumi in a 2-year-old female patient, thus our report of DPGA is the earliest onset reported in the world.

REFERENCES