

Case 13

A 2-year-old girl from Bangkok

Chief complaint: Asymptomatic multiple papules on face, trunk and all extremities for 3 months

Present illness: The patient gradually developed asymptomatic multiple flesh- to red-colored papules on the face, trunk and all extremities which healed with hypopigmented scar over the course of 3 months. She had no systemic symptoms.

Past history: She had no underlying disease or fever before skin eruption.

Family history: There was no family history of cutaneous diseases or metabolic diseases.

Physical examination:

HEENT: no pale conjunctiva, anicteric sclera, no lymphadenopathy

Heart: normal S1, S2, no murmur

Lungs: clear on both sides

Abdomen: soft, no distension, no hepatosplenomegaly

Extremities: no edema

Skin examination:

Generalized flesh- to red-colored, 1-5 mm with central crusted papules on face, trunk and all extremities interspersed with hypopigmented scar. No lesion on palms and soles. (Fig. 13.1, 13.2)

Histopathology: (S08-21095) (Fig. 13.3, 13.4)

There are scale-crusts, mild epidermal hyperplasia in association with superficial perivascular and nodular infiltrate of lymphohistiocytes and a hint of central degenerated collagens.



Fig. 13.1



Fig. 13.2

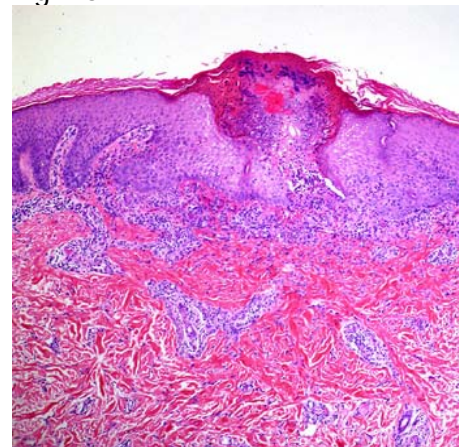


Fig. 13.3

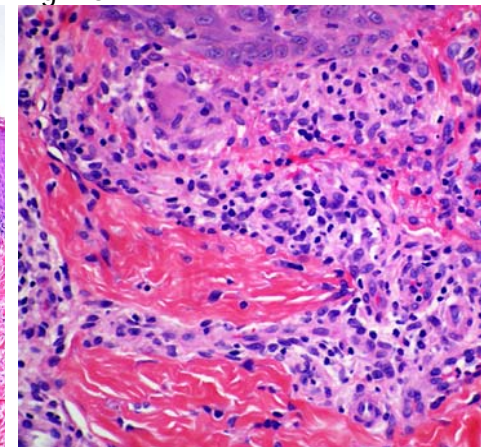


Fig. 13.4

Diagnosis: Generalized perforating granuloma annulare

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Treatment: Topical mometasone cream

Discussion:

Granuloma annulare (GA) is a benign, usually self-limited dermatosis, characterized by necrobiotic dermal papules that often assume an annular configuration. Clinically GA can be divided into a number of distinct types: localized, generalized, subcutaneous, perforating, macular and erythematous^{1,2}.

Generalized perforating granuloma annulare (GPGA) is an unusual variant of GA. It was first reported in 1973 by Duncan et al⁴. It characteristically presents with flesh- to red-colored, 1-5 mm papules that have a central umbilicated crust or scale, usually involving the extremities, face, neck and trunk³. These lesions pass through four stages: (1) erythematous or skin color papules which evolve into (2) pustular-like lesion that exude thick, creamy or clear, viscous material forming (3) umbilicated or crusted papular lesions that heal leaving (4) atrophic hypo- or hyperpigmented scars. Following the criteria of Dabski and Winkelmann for GA, the generalized form was defined as affecting at least the trunk and either the upper or the lower extremities, or both, and those cases with extensive lesions over the extremities only were recorded as dissiminated⁵.

GPGA has appeared at age from 1 to 60 years, with 40% of cases appearing on children and affects mostly girls and women. All areas of the body can be affected, especially the extensor surfaces of both extremities. The GPGA subtype often proves recalcitrant to treatment and may ulcerate⁷. About 68% of patients have atrophic hypo- or hyperpigmented scars.

The cause of GPGA is unknown (similar to GA), but ultraviolet (UV) light, insect bite, trauma, viral infection, thyroiditis, diabetes and vitamin D have been implicated⁶.

Histopathologically the presence of palisading granulomas consisting of necrobiotic collagen, fibrin, and mucin deposits, partially surrounded by infiltrates of histiocytes and lymphocytes, is the

common microscopic finding. The granulomas are located in the upper dermis, and perforate it, extruding the necrobiotic material⁸.

The differential diagnosis of GPGA includes insect bite, papular urticaria, molluscum contagiosum, varicella, scabies, secondary syphilis, papulonecrotic tuberculid, sarcoidosis, drug eruptions, reactive perforating collagenosis, perforating folliculitis and pityriasis lichenoides et varioliformis acuta.

Seasonal variations were present, with exacerbation during the summer and clearing during the winter⁴. Some patients have noted spontaneous resolution. Lesions usually persist for an average of one to four years or may have a more chronic, relapsing course. Corticosteroids, chlorambucil, etretinate, isotretinoin, dapsone, PUVA and cyclosporin A have been used with some degree of success.

References

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