

Pityriasis Rotunda with Familial Occurrence

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Pityriasis rotunda is an uncommon chronic dermatosis characterized by multiple, round or oval, hyperpigmented or hypopigmented patches that have a fine scale on the trunk and extremities.

Most of the cases reported predominantly occurred in Oriental and black patients in association with internal disease. However, in Caucasians it has been documented in healthy persons usually as a familial tendency.

We report a case of pityriasis rotunda which showed familial occurrence and had no underlying disease. (Ann Dermatol 9:(3) 194~196, 1997).

Key Words : Pityriasis rotunda, Familial occurrence

Pityriasis rotunda is an uncommon dermatosis characterized by mostly asymptomatic, multiple, perfectly round or oval, hyperpigmented or hypopigmented scaly patches located mainly on the trunk and extremities¹.

Since the first description of this disorder in 1906², the majority of the cases reported occurred in Oriental and black patients in association with underlying diseases. However, in white people it may rarely occur and has been documented in healthy persons usually as a familial tendency.

In 1994, Grimalt et al³ suggested that this dermatosis has a familial occurrence in white people while in Oriental or black patients it is associated with internal malignancy or systemic disease and is non-familial.

Recently we experienced a case of familial pityriasis rotunda.

CASE REPORT

A 24-year-old Korean girl had multiple, brownish, hyperpigmented, scaly, patches that had been present on the left leg and back for 2 years. The lesions

were asymptomatic, oval and 1.5-3 cm in diameter. They gradually enlarged and increased in number and coalesced with each other. They tended to be exacerbated and distinct during the winter and improved and pale in the summer.

Her maternal grandfather had a solitary, 30 cm-sized, circular, brownish patch on the back and suffered from transitional cell carcinoma of the bladder. Her mother also had similar cutaneous patches on the left leg and the back.

The patient had no history of underlying disease such as infection or internal malignancy.

On KOH examination of the lesion, fungi were not discovered.

A biopsy specimen revealed slight hyperkeratosis and hyperpigmentation in the basal layer, but no other specific abnormalities were found.

Topical application with 10% urea ointment and 0.025% tretinoin ointment for 1 month resulted in disappearance of the cutaneous lesions. For the following 2 months, there was no evidence of recurrence.

DISCUSSION

Pityriasis rotunda is a chronic cutaneous disorder of keratinization manifesting as perfectly circular or oval, hyperpigmented or hypopigmented, well-circumscribed patches of variable number and diameter that are slightly scaly^{1,3}.

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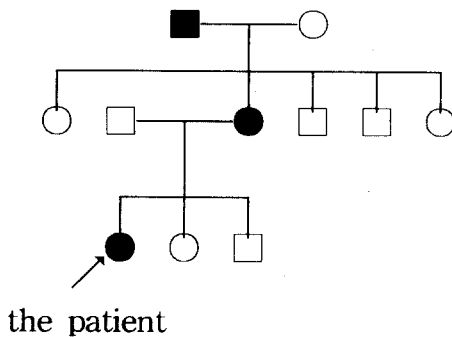


Fig. 1. Family pedigree for case: the patient, her mother and her maternal grandfather had pityriasis rotunda.

Fig. 2. The patient had oval, hyperpigmented, scaly patches which coalesced with each other on the back.

Fig. 3. A biopsy specimen revealed slight hyperkeratosis and increased melanin pigments in the basal layer (H & E stain, $\times 100$).

This dermatosis may occur all over the world. However, it usually occurred in certain ethnic groups. It was originally described by Toyama in 1906². Thereafter, most of the cases reported were predominantly from the Far East particularly from Japan^{2,4} or in South African black people^{5,6,7} and West Indians⁸ usually in association with underlying diseases. It was also described in Koreans^{9,10} and black Americans¹¹. In white people the occurrence of pityriasis rotunda is extremely rare and it has been documented in healthy persons either as a familial tendency in most cases or sporadically in some cases³.

Pityriasis rotunda affects both males and females but it may affect women more than men in the ratio of 1.5 to 1¹¹. The majority of patients are from 20 to 45 years of age⁴.

Lesions are generally multiple and 1 to 28 in number¹. However, it may be usually over 30 le-

sions in white patients¹. The size of individual patches may vary from 1 to 30 cm in diameter¹². Clinically they are usually asymptomatic but occasionally pruritic. They tend to enlarge and join together with adjacent lesions. This condition usually involves the trunk and the extensor surfaces of the extremities¹².

This dermatosis seems to be characterized by its chronicity and the lesions may last for many years¹². It is exacerbated during the winter but is partially clear in the summer¹.

The histologic appearances of pityriasis rotunda resemble those of ichthyosis vulgaris^{4,12}. A lesional biopsy specimen reveals moderate hyperkeratosis, a thinned or absent granular layer, increased pigmentation of the basal layer, sparse lymphohistiocytic perivascular infiltrates in the upper and mid dermis.

Differential diagnoses include tinea corporis, tinea versicolor, pityriasis alba and extraflexural erythrasma^{1,8,12}.

There are no effective therapies to treat pityriasis rotunda. Topical therapies using corticosteroids, tar, salicylic acid and retinoids have been of little benefit¹. Effective vitamin A or etretinate therapy was reported in some patients^{13,14}. In cases of pityriasis rotunda associated with internal malignancy or infection, treatment of these underlying disease has resulted in disappearance of the cutaneous lesions^{15,16}. In our case topical urea and retinoid was effective but these therapies failed to completely clear the lesions.

The etiology of pityriasis rotunda is still un-

known. A number of authors have agreed the hypothesis suggested by Ito and Tanaka⁴ that pityriasis rotunda is a variant of acquired ichthyosis on the basis of the histological, histochemical and morphological findings.

This dermatosis has been considered as a cutaneous sign of systemic disease. Many reported cases have revealed the association of pityriasis rotunda with other diseases such as infection, malignancy and chronic debilitating diseases.

Pityriasis rotunda was found in certain families or genetically predisposed patients. The first case of a familial occurrence was described by Zina et al¹⁷ in 1986. Grimalt et al¹ suggested that pityriasis rotunda is more closely related to congenital ichthyosis on the basis of the ultrastructural findings and its familial occurrence. They classified this disorder into two groups, type I and Type II. Type I is characterized by occurrence in Oriental and black people without familial tendency, mostly hyperpigmented patches (less than 30 in number) and is often associated with underlying disease. On the other hand, type II is characterized by occurrence in white people with a familial tendency, mostly hypopigmented patches (more than 30 in number) and is not associated with internal disease.

However, we recently experienced a case of pityriasis rotunda which showed familial occurrence and had no underlying disease. In addition, the Korean literature reports two familial cases one in 1973⁹ and one in 1983¹⁰. Three out of four sisters had pityriasis rotunda in one family and one representative case (a 45 year-old female), her mother (74 years old) and her son (17 years old) had pityriasis rotunda in the other family. All these cases were not associated with internal disorders.

Consequently familial cases of pityriasis rotunda can occur in Oriental as well as Caucasian people.

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