Case 21

Multiple ring-shaped brown patches on the abdomen

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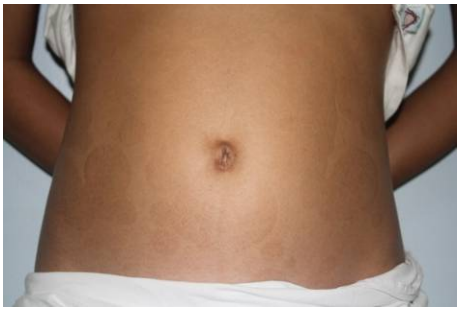
Patient: A 12-year-old girl from Nonthaburi

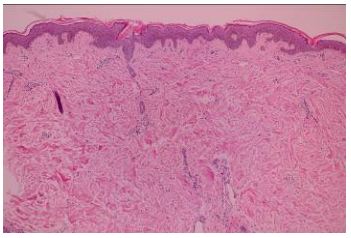
Chief complaint:
Multiple brownish patches on trunk and extremities for 5 years

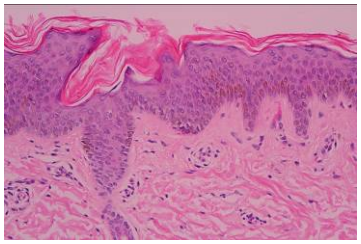
Present illness:
The patient noticed multiple asymptomatic brownish patches on trunk, which were progressive to extremities for 5 years. Her father also had the same lesion on the arm. None of her siblings were affected.

Past history: No underlying disease

Skin examination:
- Multiple well-defined brownish round and oval scaly patches, diameter 5-10 cm on trunk and extremities.







Histopathology: (S14-6265, left arm)
There is mild hyperkeratosis and vary sparse superficial cells infiltrate.

Investigation: 10% KOH preparation was negative.

Diagnosis: **Pityriasis rotunda**

Treatment:
- 0.025% Tretinoin cream apply at bedtime
- Cold cream apply twice daily

Discussion:

Pityriasis rotunda (PR) is a rare skin disease that was initially described in Japan by Toyama in 1906 as "pityriasis circinata" and was later named pityriasis rotunda by Matsuura.[1, 2] It is characterized by well-defined circular, slightly scaly hypo- or hyperpigmented patches over the trunk and proximal extremities. PR is considered to be a disorder of keratinization and is common among the Japanese, West Indian blacks, South African Bantu, but is less common in Caucasians.[3-6] PR is most commonly observed between the ages of 25 and 45 years (7 and 76 years are the reported extremes), except among the Sardinian cohort in which patients presented during childhood.[6, 7] The incidence is equal among men and women. PR is typically asymptomatic, large well-circumscribed circular hyper- or hypopigmented scaly patches that are often 10 cm (and may be up to 30 cm) in diameter, favors on trunk and proximal extremities, without inflammation.[6]

The etiology of PR is still unknown but it is thought to be a form of acquired ichthyosis (possibly a paraneoplastic condition) or a late-onset congenital ichthyosis.[4, 5, 8] It has been associated with many conditions included tuberculosis, cirrhosis, malignancy, and malnutrition.[3, 9] It can be classified in 2 types. Type I occurs in blacks or Asians, without family history, with fewer than 30 hyperpigmented lesions, possibly in association with an internal malignancy or systemic diseases in 30% of cases. Type II occurs more commonly in white or Caucasians and the lesions are usually hypopigmented, numerous (> 30). Patients may have a family history of PR but not associated with internal malignancy.[3-6]

The differential diagnosis of PR includes tinea versicolor, tinea corporis, nummular eczema, erythrasma, leprosy, fixed-drug eruption, pityriasis rosea, sarcoidosis, and pityriasis alba.[5] Potassium hydroxide examination, fungal culture and routine histology can excludes some diseases in the differential diagnosis.

PR has histopathological features of ichthyosis vulgaris. There is an absent or diminished granular layer with hyperkeratosis, increased pigmentation of the basal layer, pigment incontinence, and a mild superficial perivascular lymphocytic infiltrate. In some cases, the histopathology may be normal.[3-5]

PR is relatively difficult to treat. Lesions usually improve or resolve with treatment of the underlying internal disease or malignancy. Symptomatic treatment using topical glucocorticoids, antifungal agents, salicylic acid, topical retinoids, lactic acid, tar, and emollients has been used, but improvement has been reported with lactic acid lotion and oral vitamin A.[4, 5]

Familial PR have been reported mostly in Caucasians, and 2 Nigerians.[3, 7, 9] There are 2 case series reported from Italy suggested a strong familial tendency with one group reporting 69% (29/42) and the other 83% (39/47).[7, 9] These findings suggest that PR may be a genetic disease. In most of these cases the disease tended to affect father and child or siblings but the pattern of inheritance could not be established. In southern Italy, an association with glucose 6-phosphate dehydrogenase (G6PD) deficiency and favism has been reported.[10] This report and a later report by Grimalt Ramon, et al., the pattern of inheritance appeared to be autosomal dominant.[10, 11] In the first report of familial PR in black-skinned patients occurred in female (the patient and her mother), but their siblings were not affected, thus it cannot determine a mode of inheritance.[3]

Interestingly, our patient has a familial PR, a first report in Asian skin, has no association with malignancy or other systemic diseases, and has never

been reported in Asian skin. She was treated with 0.025% tretinoin cream and emollients with mild clinical response.

References:

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