

Pityriasis Rotunda: A Case Report of Familial Disease in an American-Born Black Patient

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Key Words

Pityriasis rotunda · Ichthyosis · Hyperpigmentation

Abstract

Pityriasis rotunda is an uncommon dermatosis with an unusual geographic and racial distribution. The skin disorder is characterized by sharply defined, perfectly circular, scaly patches with no inflammatory changes. Notably, it may be associated with underlying malignancy or chronic infection. We report an uncommon familial case in an American-born female.

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Introduction

Pityriasis rotunda is an unusual skin disease that is characterized by hyper- or hypopigmented scaly plaques that involve the trunk and proximal extremities. The lesions are notable for being asymptomatic or mildly symptomatic, sharply defined and typically perfectly circular with no evidence of induration or inflammation [1]. The etiology of pityriasis rotunda remains unknown, and the diagnosis is usually based on clinical examination. Importantly, the condition is often associated with underlying malignancies and chronic diseases or infections. Treatment of the disease can be challenging, and several different pharmacological treatments have been tried with limited success. However, for patients with an underlying disease, successful treatment of the underlying disease often leads to remission of the skin lesions [2].

A striking aspect of pityriasis rotunda is its unusual geographic and racial distribution. First described in 1906 by Toyama [3] in Japan, and subsequently reported among South

African [4, 5] and West Indian [6] blacks, it is not uncommon in these groups and may be associated with underlying disease. In contrast, pityriasis rotunda is rare and not associated with underlying disease among Caucasians [1]. There has only been one previous report of pityriasis rotunda occurring in American-born blacks [7].

Case Report

A 49-year-old American-born black female presented with a 3–5-year history of asymptomatic skin lesions on the trunk. She was in generally good health with no other known significant medical history. Her family history was notable for a sister with similar skin lesions, who is otherwise in good health. Cutaneous examination revealed three hyperpigmented well-defined perfectly circular patches varying in size from 3–8 cm in diameter on the abdomen. The lesions were dry and scaly. Erythema and induration were absent (fig. 1). Routine investigations including complete blood count, liver function tests, urine analysis and chest X-ray were all normal. The biopsy report showed a sparse infiltrate of small lymphocytes and a few melanophages. There is epidermis with orthokeratosis and a suggestion of a slightly diminished granular layer in the epidermis. A PAS stain was negative for fungus.

The patient was treated with Tazorac cream 0.05% once daily for 3 weeks followed by Tazorac cream 0.1% once daily for an additional 3 weeks. There was some improvement of her lesions but not full resolution.

Discussion

Pityriasis rotunda is an idiopathic disorder of keratinization characterized by hyper- or hypopigmented circular sharply defined scaly patches. It is thought by some to be a form of acquired ichthyosis because of histologic features similar to ichthyosis vulgaris. The lesions are most commonly situated on the buttocks, thighs, abdomen, back or upper arms. The hands, feet and face are usually spared [1, 5]. The number of lesions may range from 1 to rarely greater than 100, with a typical diameter of 2–3 cm that may exceed 20 cm in some cases. Both genders are affected with a slight female to male predominance (1.5:1) and lesions commonly develop between the ages of 25 and 45, although cases have been reported from 7 to 87 years of age [1, 5, 8].

Diagnosis of pityriasis rotunda is typically a clinical one. The age of onset, distribution, distinct circular appearance, and lack of inflammatory changes usually suggest the diagnosis. Histologically, findings are often subtle, but hyperkeratosis, a thinned granular layer, pigmented basal layer, and mild perivascular lymphocytic infiltrate are commonly reported [8]. Of note, pityriasis rotunda may be found as a paraneoplastic phenomenon in a range of malignancies, and thus should be recognized as a potentially important clinical sign [9].

The first description of this skin disorder was by Toyama [3] in 1906 who used the term pityriasis circinata. In the same year, Matsura [10] described the same skin disorder referring to it as pityriasis rotunda, a term which has subsequently been adopted for this idiopathic disorder characterized by well-demarcated circular lesions. In 1960, an extensive review of the literature by Ito and Tanaka [8] of 182 cases from Japan and Korea was followed by reports among South African and West Indian blacks [4–6], where the disorder is not uncommon. Subsequently, there have been case reports among Caucasians, including a familial occurrence in Sardinia, Italy [11, 12].

Based on these reports and others, Grimalt et al. [1] in 1994 proposed a classification system. Type I pityriasis rotunda is characterized by hyperpigmented lesions in Asian and black patients, less than 30 lesions, a nonfamilial pattern and an association with malignancies (stomach, liver, esophagus, lung, prostate, leukemia, multiple myeloma among others), systemic diseases (most commonly liver, female genital tract and malnutrition), and chronic infections (most commonly tuberculosis). Type II consists of hypopigmented lesions in Caucasians, typically involves more than 30 lesions, a familial pattern and has no association with underlying disease. While useful, there have been exceptions to this classification scheme.

We are aware of only one prior report of pityriasis rotunda in American-born blacks. In 1986, Rubin and Mathes [7] reported an 87-year-old black male with underlying metastatic carcinoma and a 67-year-old black man with recurrent renal calculi and diabetes mellitus. In 2007, Friedman et al. [13] reported the first familial case in blacks: a Nigerian-born mother and her British-born daughter who were both otherwise well with the exception of asthma in the mother. Our case of a 49-year-old American-born black female does not fit neatly into the classification system; black race and presence of few lesions fit into type I, but the familial history and lack of association with underlying disease are characteristic of type II.

Treatment of pityriasis rotunda can be difficult, as topical glucocorticoids, antifungal agents, salicylic acid and tars have shown limited benefit. However, improvement in skin lesions has been noted after treatment with vitamin A [14]. Our patient had a partial response to a topical retinoid. Noteworthy, successful treatment of the underlying disease, if present, often leads to remission of the skin lesions.

Conclusion

Pityriasis rotunda is a skin disorder of unknown etiology with an unusual geographic and racial distribution. Common in South African and West Indian blacks, its prevalence in American-born blacks has not been well described. While not present in our patient, the association of pityriasis rotunda with underlying malignancies makes the skin disorder a potentially important clinical sign.

Statement of Ethics

The authors have no ethical conflicts to disclose. The patient has given her consent, including for the use of the picture.

Disclosure Statement

The authors have no conflicts of interest to disclose. No funding support was obtained for this work.

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Fig. 1. Hyperpigmented, well-demarcated, perfectly round, dry, and ichthyosis-form scaly lesion on the lower abdomen.