Childhood *Pityriasis rosea inversa* without Herald Patch Mimicking Cutaneous Mastocytosis

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Abstract

Background: Pityriasis rosea is a self-limited inflammatory condition of the skin that mostly affects young adults. Several less common atypical presentations have been reported.

Case Presentation: A 6-year old girl with red-brown maculopapular eruption sized 0, 5-1 cm in diameter localized on neck, trunk and popliteal region visited our general pediatric outpatient clinic. The eruption was wide spread especially on flexural areas. After consulting dermatologist skin biopsy was performed. According to clinical and histopathological findings as inverse (flexural) pityriasis rosea was diagnosed. For treatment, systemic antihistamine, topical corticosteroid cream and emollient were applied. The lesions healed in two months. Spontaneous healing of the eruption also confirmed the diagnosis of pityriasis rosea.

Conclusion: We present this interesting pediatric case to show and discuss pityriasis rosea, atypical presentations, differential diagnosis and the importance of dermatological examination and importance of dermatologic consultation for pediatric patients with skin eruption.

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Introduction

Pityriasis rosea is an acute, self limited papulosquamous dermatosis character-rized by the development of a large erythematous scaling 'herald patch' and the subsequent eruption of multiple similar, smaller, salmon-coloured oval patches in the classic 'fir tree' or 'Christmas tree' distribution. The disease occurs most commonly between ages 10-40 years^[1].

The lesions are most heavily localized on the trunk and proximal extremities but may extend

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onto the distal extremities and face^[2]. Several less common clinical presentations have been reported. Inverse pityriasis rosea presents with more lesions on the extremities, flexural areas and face^[1,3]. Localized variants limited to a small area, unilateral variant, pityriasis circinata et marginata of Vidal, vesicular variant, purpuric (haemorrhagic) variants have been identified^[1].

Since it can be mistaken for several skin diseases, clinical diagnosis of pityriasis rosea may be sometimes difficult, especially in atypical variants.

Case Presentation

A 6-year old girl applied to our general pediatric outpatient clinic with the complaint of nonpruritic pink-red eruption beginning on the trunk for ten days. The lesions became generalized to the whole body and were crusted.

There was no history of drug intake or vaccination. After consulting dermatologist skin biopsy was taken. The dermatological examination revealed red-brown maculopapular eruption sized 0,5-1 cm in diameter, some of them with scaling, localized on neck, trunk and popliteal region. The eruption was wide spred especially on flexural areas (Figs. 1 and 2). Cutaneous mastocytosis, psoriasis guttata, and atypical pityriasis rosea were taken into consideration as differential diagnoses.

Histopathological examination of the skin superficial orthokeratosis parakeratosis, minimal irregular acanthosis and papillomatosis in epidermis, and chronic inflammatory cell infiltration in dermis (Fig. 3). According to clinical and histopathological findings inverse (flexural) pityriasis rosea was diagnosed. Physical examination, count of blood cells, biochemistry and urine analysis were in normal ranges. Intestinal parasites were not detected on Gaita examination. For the treatment. systemic antihistamine. topical corticosteroid cream and emollient were applied. The lesions healed in two months. Spontaneous healing of the eruptions also suggested the diagnosis of pityriasis rosea.



Fig 1: Red-brown maculopapular eruption localized on neck and trunk, some of them with scaling



Fig 2: Red-brown maculopapular eruption localized on popliteal region

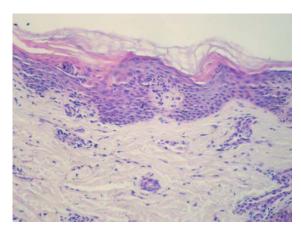


Fig 3: Superficial orthokeratosis and parakeratosis, minimal irregular acantosis and papillomatosis in epidermis, and chronic inflammatory cell infiltration in dermis

This pediatric case is interesting, because there was no herald patch and her lesions were commonly localized on flexural areas.

Discussion

Pityriasis rosea is a self-limited inflammatory condition of the skin that mostly affects young adults[1,4]. The overall incidence of it is 6,8 per 1000 dermatological patients^[5]. The etiology of pityriasis rosea is unknown. Viral and bacterial causes have been sought, but convincing answers have not yet been found[6]. More recently, attention has been focused on the human herpesvirus group (HHV-6 and HHV-7), with conflicting results^[1,7]. It has been postulated that pityriasis rosea may be due to reactivation of a latent virus rather than a primary viral infection. Further studies to investigate the question of primary infection or reactivation of pathogens are strongly warranted[8].

Pityriasis rosea classically presents with a singular, large scaling plaque that commonly appears on the trunk and gradually enlarges over a few days. This initial plaque is referred to as herald patch or mother patch, and is often misdiagnosed as tinea corporis^[1,4]. Herald patches vary from 1-10 cm in diameter; they are annular in configuration and have a raised border with fine, adherent scales^[2]. The herald patch is estimated to occur in 80% of cases^[1].

Approximately 5-10 days after the appearance of the herald patch, a widespread, symmetric eruption becomes evident involving mainly the trunk and proximal limbs. Typical lesions are oval or round, less than 1 cm in diameter, slightly raised, and pink to brown. The developed lesion is covered by a fine scale that gives the skin a crinkly appearance; some lesions clear centrally, producing a collarette of scale that is attached only at periphery. The long axis of each lesion is usually aligned with the cutaneous cleavage lines, a feature that creates the so called Christmas tree pattern on the back. Duration of the eruption varies from 2-12 weeks. The lesions may be asymptomatic or mildly to

severely pruritic. Fever, malaise, arthralgia, and pharyngitis can be seen as a prodrome. Children rarely complain of such symptoms ^[2].

Several less common clinical presentations have been reported. A simple classification for atypic pityriasis rosea has been proposed by Chuh, et al ^[5].

Atypical morphology of lesions: Atypical rash morphology includes rashes in vesicular, purpuric, haemorrhagic and urticarial forms. Vesicular pityriasis rosea is commoner in children and young adults, and may be severely pruritic and extensive^[5]. Skin lesions are with the characteristic arrangement and distribution but with vesicles surmounting the papules^[9].

Pityriasis rosea with erythema multiformelike lesions and purpuric (hemorrhagic) variants have also been identified^[1]. Urticarial and annular lesions are the other unusual variants^[2]. Papular pityriasis rosea is more often seen in children. Numerous small papules 1-2 mm in diameter may be seen together with classical pityriasis rosea patches^[5].

Atypical size of lesions: Pityriasis rosea gigantea of Darier that has enormous plaques is rare. The clinical course is similar to typical pityriasis rosea^[5].

Atypical distribution of lesions: Pityriasis rosea inversa presents with more lesions on the extremities, flexural areas and face^[1,3]. In the limb-girdle type, the eruption is restricted to the shoulders or hips^[5]. Unilateral involvements have been reported. Localized variants limited to a small area, such as the axilla or breast, have been reported^[10].

Atypical number of lesions: Pityriasis circinata et marginata is sometimes considered a special form of pityriasis rosea. This is mainly seen in adults, with fewer and larger lesions often localized to the axillae or inguinal region^[5].

Atypical site of lesions: Involvements of the face, scalp, hands and feet are not rare in pityriasis rosea. Involvements of finger and toe tips, eyelids, penis and oral cavity have been reported^[5]. Oral lesions have been described in several small case series^[1].

Atypical severity of symptoms: Pityriasis rosea is usually nonpruritic. The term pityriasis rosea irritata is sometimes coined to describe patients complaining of severe itch, pain and a

burning sensation^[5].

Atypical course of the eruption: Recurrent or relapsed cases have been reported. Recurrent episodes have been estimated to occur in 1,8-3,5% of cases [11].

Drug-induced pityriasis rosea-like rashes: Many drugs, including captopril, gold, isotretinoin, nonsteroidal anti-inflammatory agents, omeprazole, terbinafine and tyrosine kinase inhibitor have been implicated in causing pityriasis rosea-like rashes^[5].

The diagnosis of pityriasis rosea is clinical^[2]. Biopsy usually is not indicated in the evaluation of patients with suspected pityriasis rosea^[6]. The generalized eruption resembles a number of other diseases; of these secondary syphilis is the most important. Drug eruptions, viral exanthems. guttate psoriasis, pityriasis lichenoides chronica, tinea versicolor and nummular eczema can also be confused with pityriasis rosea^[1,2,12]. Some of the atypical morphological variants and clinical presentations may suggest other diagnoses. The inverse form has been reported to mimic papular acrodermatitis of childhood (Gianotti-Crosti syndrome). Papular lesions may resemble lichen planus or a lichenoid drug eruption. The vesicular variant may be mistaken for varicella or scabies. Cases of purpuric pityriasis rosea may be confused with the pigmented purpuric dermatoses, Kaposi's sarcoma and vasculitis, including Henoch-Schönlein purpura. presentation closely resembles unilateral asymmetric periflexural exanthem of childhood (APEC) [1,13].

Pityriasis rosea may be confused with cutaneous mastocytosis, especially urticaria pigmentosa. Urticaria pigmentosa is characterized by monomorphic pigmented maculopapular or nodular lesions mainly on the trunk and with a widespread symmetrical distribution^[14]. Our patient's lesions have been mimicking cutaneous mastocytosis with the clinical appearance of the lesions. So, differential diagnosis of pityriasis rosea can be sometimes difficult, especially in atypic forms.

Histopathological findings in pityriasis rosea are not pathognomonic for the disorder. Biopsies usually reveal a lymphocytic, perivascular, primarily superficial infiltrate associated with exocytosis. Epidermal spongiosis with focal parakeratosis, an absence or decrease in the granular layer and extravasation of erythrocytes into the papillary dermis are also noted^[1,6].

Therapy is unnecessary for asymptomatic patients. If scaling is prominent, a bland emollient may suffice. Pruritus may be suppressed by a lubricating lotion containing menthol and camphor or by an oral antihistamine for sedation, when itching may be troublesome. Occasionally, a nonfluorinated topical corticosteroid preparation may be necessary to alleviate pruritus^[2]. Our patient healed with systemic antihistamine, topical corticosteroid cream and emollient within two months. Spontaneous healing of the eruption also suggested the diagnosis of pityriasis rosea.

Conclusion

It is important that clinicians are aware of the wide spectrum of pityriasis rosea variants so that appropriate management and reassurance can be offered. Especially in children the differential diagnosis of skin eruption is more difficult than in adults. Referral to a dermatologist is warranted if the diagnosis is in doubt, symptoms are severe, or the rash is not following the typical course of pityriasis rosea. We suggest that besides psoriasis, eczema, syphilis, drug eruptions, etc., cutaneous mastocytosis should also be placed in the differential diagnosis of atypical pityriasis rosea.

For atypical eruptions without a definite diagnosis, it is safer to consider lesional skin biopsy and other investigations so that important differential diagnoses will not be missed.

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