ISSN 2307-8960 (online)

World Journal of *Clinical Cases*

World J Clin Cases 2017 June 16; 5(6): 191-257

Published by Baishideng Publishing Group Inc

World Journal of Clinical Cases

Contents

Monthly Volume 5 Number 6 June 16, 2017

World Journal of Clinical Cases
Volume 5 Number 6 June 16, 2017

Contents

NAME OF JOURNAL
World Journal of Clinical Cases

ISSN
ISSN 2307-8960 (online)

LAUNCH DATE
April 16, 2013

FREQUENCY
Monthly

EDITORIAL OFFICE
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World Journal of Clinical Cases
Baishideng Publishing Group Inc
7901 Stoneridge Drive, Suite 501, Pleasanton, CA 94588, USA
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E-mail: editorialoffice@wjgnet.com
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7901 Stoneridge Drive,
Suite 501, Pleasanton, CA 94588, USA
Telephone: +1-925-2238242
Fax: +1-925-2238243
E-mail: bpgoffice@wjgnet.com
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PUBLICATION DATE
June 16, 2017

INSTRUCTIONS TO AUTHORS
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ONLINE SUBMISSION
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World Journal of Clinical Cases

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World J Clin Cases 2017 June 16; 5(6): 203-211

DOI: 10.12998/wjcc.v5.i6.203

ISSN 2307-8960 (online)

MINIREVIEWS

Clinical variants of pityriasis rosea

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Author contributions: All the authors have contributed to the preparation of the manuscript and collection of pictures.

Conflict-of-interest statement: Authors declare no conflict of interests for this article.

Manuscript source: Invited manuscript

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Received: January 28, 2017
Peer-review started: February 9, 2017
First decision: March 7, 2017
Revised: March 21, 2017
Accepted: April 18, 2017
Article in press: April 19, 2017
Published online: June 16, 2017

Abstract

Pityriasis rosea (PR) is a common erythemato-squamous dermatosis which almost always, is easily diagnosed. Mostly the disease presents in its classical form. However, clinical dermatology is all about variations and PR is not an exception. Variants of the disease in some cases may be troublesome to diagnose and confuse clinicians. Prompt diagnosis and treatment of the condition becomes necessary to avoid unnecessary investigations. We hereby review and illustrate atypical presentations of the disease, including diverse forms of location and morphology of the lesions, the course of the eruption, and its differential diagnoses.

Key words: Pityriasis; Pityriasis rosea; Pityriasis rosea of Gibert; Herald patch; Papulo-squamous dermatosis

Core tip: Pityriasis rosea (PR) is a common, self-limited disease which in its typical form should not raise diagnostic doubts. Atypical forms represent 20% of cases, with diverse variants with respect to morphology and location of lesions, and evolution of the disease. Recognition of these forms may avoid unnecessary procedures. Drug ingestion may simulate PR in some cases.

Urbina F, Das A, Sudy E. Clinical variants of pityriasis rosea. *World J Clin Cases* 2017; 5(6): 203-211 Available from: URL: http://www.wjgnet.com/2307-8960/full/v5/i6/203.htm DOI: http://dx.doi.org/10.12998/wjcc.v5.i6.203

INTRODUCTION

Pityriasis rosea (PR) is a relatively common, self-limited papulo-squamous dermatosis of unknown origin, which mainly appears in adolescents and young adults (10-35 years), slightly more common in females. It has a sudden onset, and in its typical presentation, the eruption is preceeded by a solitary patch termed "herald patch", mainly located on the trunk. Few days later, a secondary eruption appears, with little pink, oval macules, with a grayish peripheral scaling collarette around them. The secondary lesions adopt a

Table 1 Clinical classification of pityriasis rosea

Classical adult PR and pediatric PR
Based on herald patch
No herald patch
Only herald patch (absence of secondary lesions)
Multiple herald patches
Herald patch in atypical location
Based on location of lesions
Limited to scalp
Limited to trunk
Limited to limbs-girdle (pityriasis circinata et marginata of Vidal)
Limited to flexures (inverse type)
Limited to the extremities
Acral type
Along the lines of Blaschko
Unilateral
Based on morphology of lesions
Purpuric or hemorrhagic
Urticarial
Erythema multiforme-like
Papular
Follicular
Vesicular
Giant
Hypopigmented
Irritated
Based on course of the disease
Relapsing
Recurrent
Persisting
Relapsing and persisting
PR-like rashes (drug-induced)

PR: Pityriasis rosea.

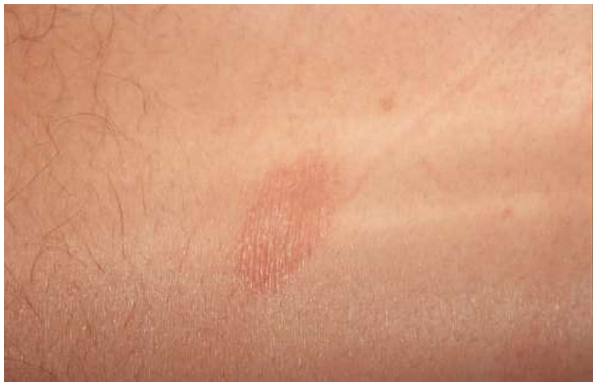

Figure 1 Herald patch. Solitary erythemato-squamous lesion, sharply defined, round or oval, mainly located on the trunk or proximal extremities.

characteristic distribution along the cleavage lines of the trunk, with a configuration of a "Christmas tree". In most cases, the eruption lasts for 6 to 8 wk. Its incidence has been estimated to be 0.68% of dermatologic patients[1], varying from 0.39%[2] to 4.8%[3].

Not so rarely (20%)[4,5], an atypical eruption may develop, concerning several aspects about the morphology or distribution of the lesions, their symptomatology and evolution.

The purpose of this article is to review and illustrate the diverse clinical presentations of PR (Table 1), which may vary in morphology, symmetry, duration, size and distribution of lesions, mucosal involvement and symptomatology.

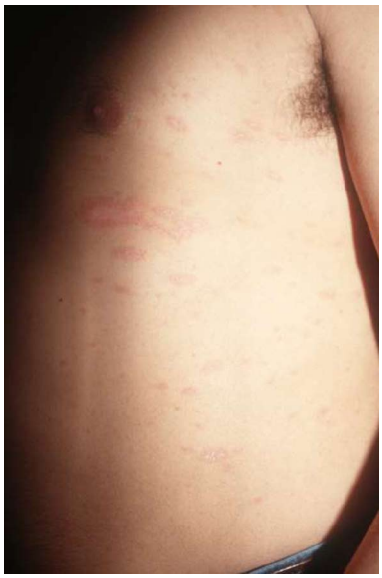

Figure 2 Classical pityriasis rosea. Exanthematous eruption with erythemato-squamous lesions following cleavage lines on the trunk.

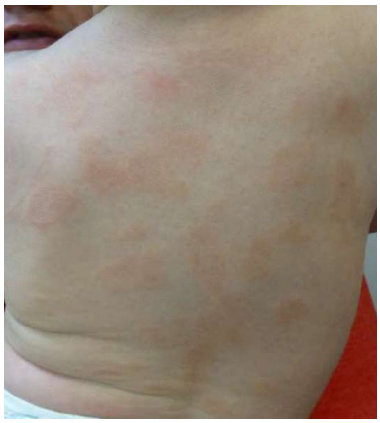

Figure 3 Pediatric pityriasis rosea. Typical lesions of PR affecting an 8-mo-old boy. PR: Pityriasis rosea.

Classical PR

A classical PR is preceeded by the herald patch, an erythematous round or oval lesion, 2-5 cm in diameter, ocassionally covered by fine scales (Figure 1). Prodromal symptoms, consisting of headache, general malaise, or flu-like symptoms are ocassionally encountered. Few days later (5-15 d), a secondary rash appears, consisting of similar, but smaller lesions, mainly located on the trunk (Figure 2). Pruritus is usually mild or absent, but can vary in intensity. The eruption lasts for 4-6 wk and fades, leaving no sequelae. Generally, it only appears once throughout life. In 75% of patients the lesions appear between the ages of 10-35 years[6].

Pediatric PR

Infrequently PR may affect children (Figure 3), with a prevalence between 8%[7] to 12%[6] below 10 years

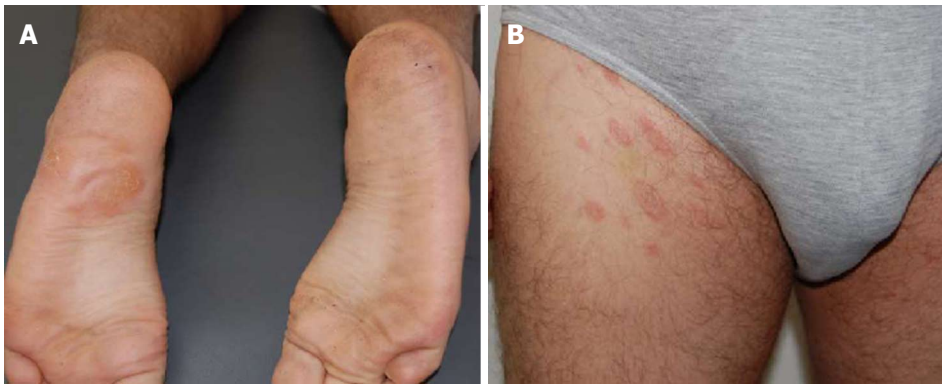

Figure 4 Herald patch in atypical location. Herald patch on a sole (A) and (B) typical PR eruption affecting trunk and proximal thighs. PR: Pityriasis rosea.

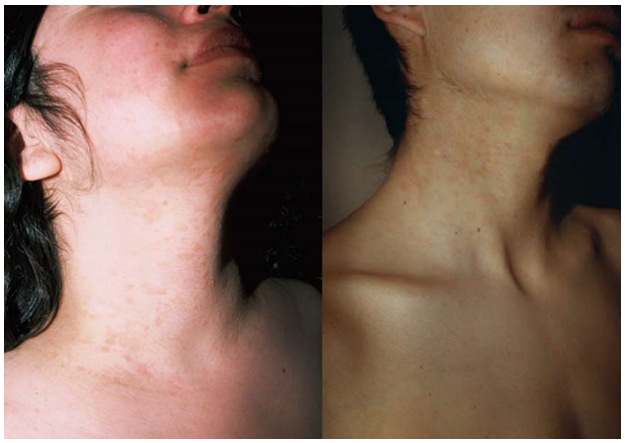

Figure 5 Inversus pityriasis rosea. Lesions distributed on face and neck in two patients; the trunk is not affected.

and 4% below 4 years of age[6] in Caucasians, whereas in dark-skinned children it increases to 26%[8]. Papular lesions prevail in them, with a short period between the herald patch and the general eruption (4 d *vs* 14 d in adults), and a shorter duration of the exanthema (16 d *vs* 45 d). The majority of cases have been described in children with ages between 3 to 9 years old, contrasting with the illustrated case of 8-mo, showing a classical variant. About half of the cases show prodromal symptoms[7].

BRIEF DESCRIPTION OF CLINICAL VARIANTS OF PR

Herald patch in atypical location

Although not mentioned in the literature, we had the opportunity to come across a patient who presented with a herald patch on a sole, and a secondary classical eruption on the trunk and proximal aspect of the extremities (Figure 4).

Circinata and marginata PR

Seen mainly in adults with few and large lesions only located on limbs-girdle, hips, shoulders, axillae or inguinal regions[9-11].

Inversus PR

The lesions are located on flexural areas (axillae, groins), face, neck (Figure 5), and acral areas (palms and soles), without affecting the trunk[12].

PR of extremities

In this variant, the lesions are confined to the extremities, with typical squamous plaques (Figure 6). The trunk is not affected.

Acral PR

The lesions are exclusively located on palms, wrists, soles[13] (Figure 7), without involvement of the flexures (axillae, groins and face), opposite to inversus PR.

Purpuric or hemorrhagic PR

Macular purpuric lesions and petechiae may appear over different locations (Figure 8) including the palate. Purpuric lesions have also appeared bilaterally on the legs in a man with a typical rash on the trunk, affecting the lines of cleavage and with collarette scaling[4].

Urticarial PR

Palpable itchy wheals-like lesions with peripheral collarette scaling (Figure 9) following the lines of skin cleavage[4,10].

Erythema multiforme-like PR

In some cases, classical lesions of PR may be accompanied by targetoid lesions resembling erythema multiforme (Figure 10). It presents with papulo-squamous lesions, admixed with few targetoid lesions distributed on the trunk, face, neck or arms[14,15]. There is no history of herpes simplex infection.

Papular PR

Multiple small papular lesions, 1-3 mm in diameter with peripheral collarette, located on the trunk and proximal extremities, along the skin cleavage lines (Figure 11). It appears predominantly in young patients[4].

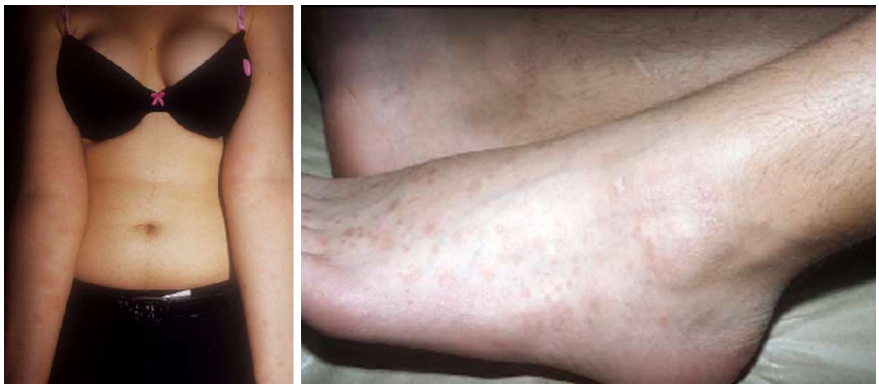

Figure 6 Pityriasis rosea of the extremities. Lesions affecting only the extremities in two different cases, without trunk involvement.

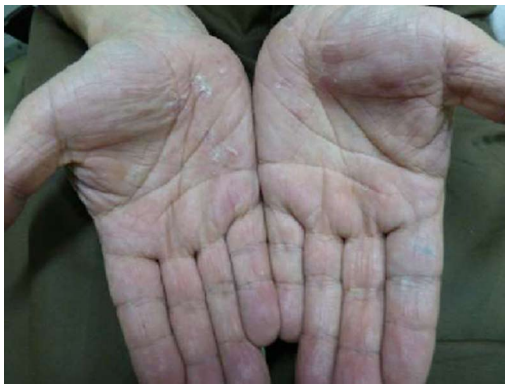

Figure 7 Acral pityriasis rosea. Desquamation affecting the palms.

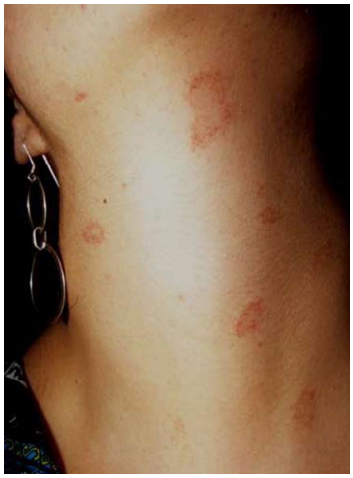

Figure 8 Purpuric pityriasis rosea. Round and oval purpuric lesions affecting the neck of a young woman.

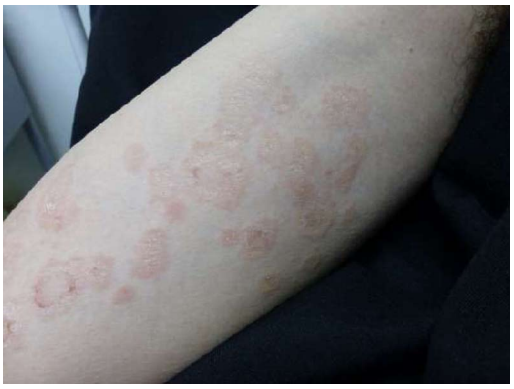

Figure 9 Urticarial pityriasis rosea. Palpable edematous, erythematous lesions with collarette scaling.

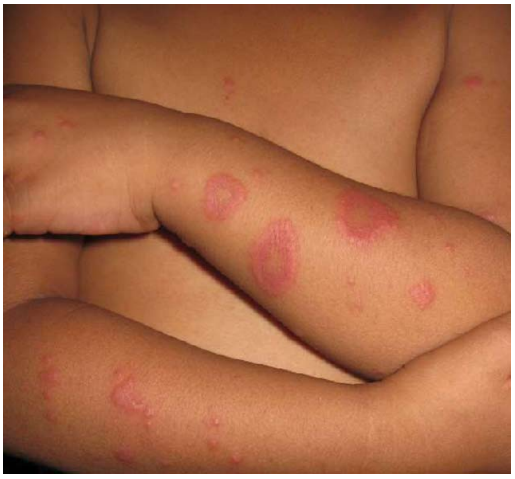

Figure 10 Erythema multiforme-like pityriasis rosea. Annular and papular lesions resembling erythema multiforme.

Follicular PR

It has been described in a 9-year-old boy with predominantly follicular scaly lesions, arranged in annular configuration[16]. The initial lesions consisted of pruritic plaques mainly located on the abdomen, thighs and groins; five days later, a striking follicular eruption - with central clearing and a peripheral collarette- developed on the posterior trunk. Prodromal symptoms included sore throat, malaise and low grade fever (Figure 12).

Vesicular PR

Generalized itchy eruption of vesicles of 2-6 mm in diameter with a rosette scaling has been described in young adults and children[17-21] (Figure 13).

Gigantea PR of darier

The dimensions of the herald patch is greater than usual, being described with the size and shape of a

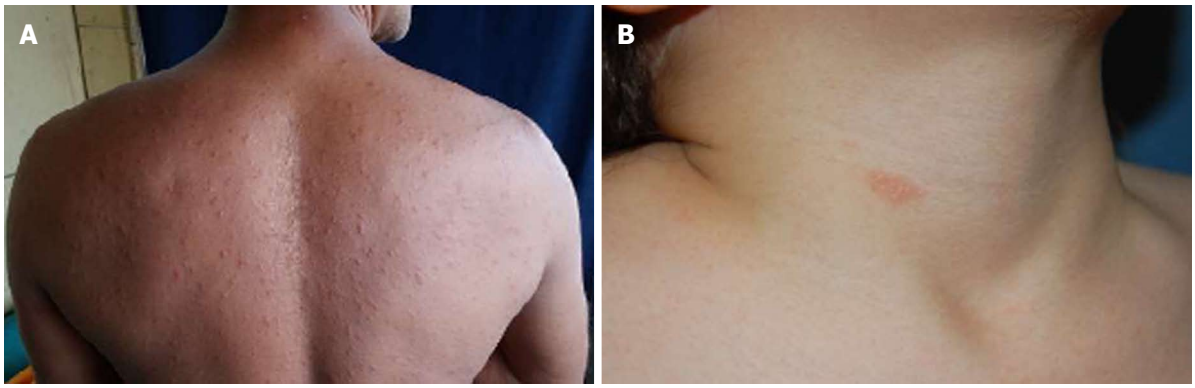

Figure 11 Papular pityriasis rosea. A: Papular lesions with peripheral collarette (Courtesy of Priyankar Misra, Junior Resident, Dermatology, Burdwan Medical College, West Bengal, India); B: Herald patch on the neck and disseminated discrete papular eruption in a girl.

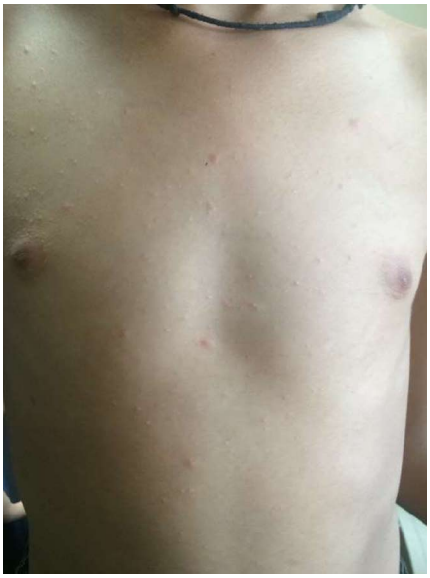

Figure 12 Follicular pityriasis rosea. Follicular lesions with scaling (Courtesy of Shankila Mittal, Junior Resident, Dermatology, Maulana Azad Medical College, New Delhi, India).

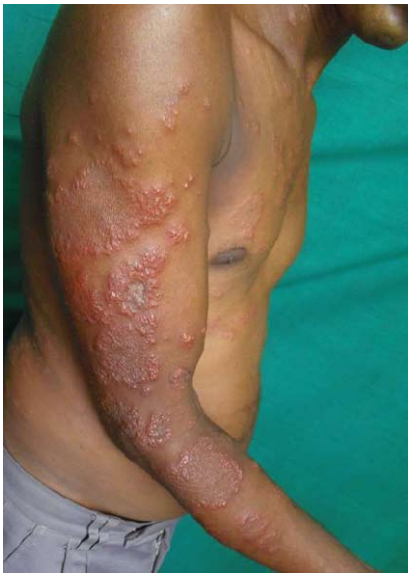

Figure 13 Vesicular pityriasis rosea. Vesicular lesions surrounding round to oval plaques (Courtesy of Dibyendu Basu, Junior Resident, Dermatology, Medical College and Hospital, Kolkata, West Bengal, India).

pear[22] (Figure 14).

Hypopigmented PR

It is essentially similar to the classic PR, with a preceding herald patch and a secondary eruption, but with hypopigmented lesions from the beginning, mainly distributed on the trunk (Figure 15). It is more frequent in dark-skinned individuals. It should not be confused with secondary hypopigmentation after a common PR.

Irritated PR

A PR with severe itch, pain and burning sensation on contact with sweat[5,23] (Figure 16).

Relapsing PR

It usually recurs within one year of the first episode, among 2.8%-3.7% of patients[8,24]. Relapses usually show absence of herald patch, and the size and number of secondary lesions are smaller. The duration of this episode is shorter and with less constitutional symptoms. Multiple relapses - though rare - have been described[25,26].

Persistent PR

By definition it lasts more than 3 mo. Its incidence in a series was 2%[1]. Most patients (75%) show a herald patch[1] and complain of systemic symptoms (most commonly fatigue, or headache, insomnia, irritability). The eruption persists for 12-24 wk. Oral lesions are common (75%), principally strawberry tongue, erythematous macules, vesicular lesions and petechiae.

Recurrent PR

Rarely, there can be multiple episodes of PR in a lifetime[25-27].

Relapsing and persisting PR

It has been described in a young man with three

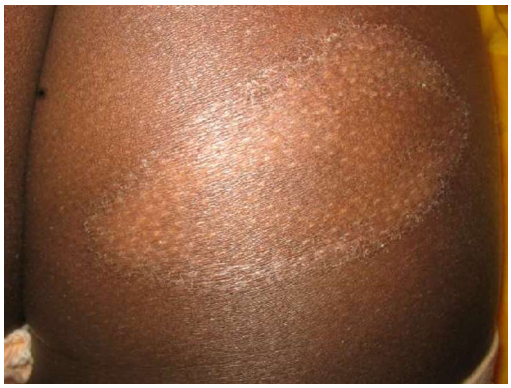

Figure 14 Giant pityriasis rosea. Large herald patch (Courtesy of Soumya Jagadeesan, Assistant Professor, Dermatology, Amrita Institute of Medical Sciences, Kochi, Kerala, India).

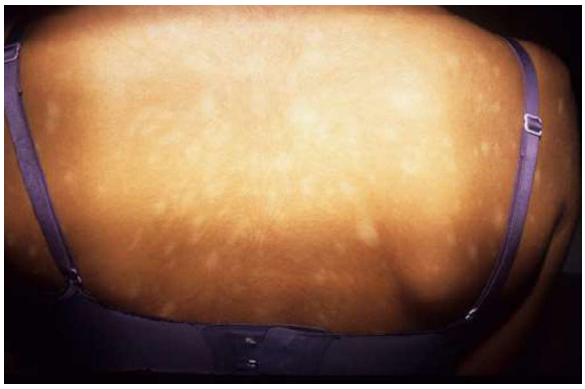

Figure 15 Hypopigmented pityriasis rosea. Round to oval hypopigmented lesions during the whole course of the eruption.

episodes of PR within one year-fulfilling the criteria for relapsing PR, and the last episode during 7 mo-consistent with persistent PR. Noteworthy, the patient presented with multiple oral ulcers[28].

Oral involvement in PR

Oral lesions in PR are more common in dark skinned people[29]. The lesions are difficult to differentiate from aphthous ulcers. Its appearance should coincide with a generalized eruption with the characteristics of PR[4]. The lesions may be punctate, erosive, bullous or hemorrhagic. They disappear concomitantly as the skin eruption fades.

PR-like rashes

They consist of exanthematous rashes which appear following the intake of several drugs: ACE inhibitors[30-32], gold[33-36], isotretinoin[37], non-steroidal anti-inflammatory agents[38,39], omeprazole[40], terbinafine[41], and tyrosine-kinase inhibitors[42]. Many of them resemble PR vaguely (Figure 17), so it may be considered as a separate condition. There is no previous herald patch and the eruption is monomorphous.

DISCUSSION

PR is a self-limited, acute inflammatory dermatosis,

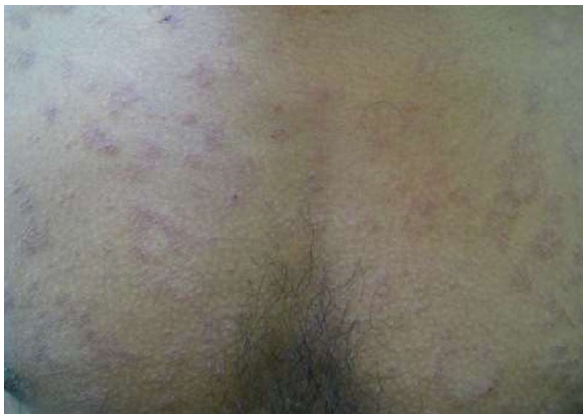

Figure 16 Irritated pityriasis rosea. Symptomatic eczematous lesions (Courtesy of Dipti Das, Consultant Dermatologist, Dr Marwah's Skin Clinic, Mumbai, Maharashtra, India).

Table 2 Diagnostic criteria of pityriasis rosea[47]

Mandatory clinical features
Discrete circular or oval lesions
Scaling within most lesions
Peripheral collarette scaling
Optional clinical features
Trunk and proximal limb distribution
Distribution along cutaneous cleavage lines
Previous herald patch

which occasionally could be persistent or recurrent. In rare situations, the symptoms or presentation may be troublesome, thus making difficulty in diagnosis or having a significant impact on the patient's quality-of-life. Its etiology has not been clearly established, but a viral origin has been suspected for years.

Recently, there are increasing evidences to suggest the role of human herpes virus (HHV) in the etiopathogenesis of PR[43,44]. Additional evidences suggest that PR is associated with reactivation of HHV 6-7[44]. Diminished levels of natural killer cells and B-cell activity in the lesions of PR has been observed. This suggests the role of a T-cell mediated immunity. Besides, increased amounts of CD4 T cells and Langerhans cells have been found in the dermis, which possibly points towards viral antigen processing and presentation. However, this matter is still debated since some individuals are infected with HHV 6-7 and do not develop the disease. PR has been also reported following vaccinations as well (B*acillus* Calmette-Guerin, influenza, H1N1, diphtheria, smallpox, hepatitis B, pneumococcus, *etc.*)[45,46].

The diagnosis of PR is essentially clinical (Table 2)[47], and in rare circumstances a biopsy may be required. Histological features are not specific and include focal parakeratosis, hypogranulosis, spongiosis, papillary dermal edema, mild perivascular lymphohistiocytic infiltrate, exocytosis and extravasated erythrocytes in the papillary dermis.

Differential diagnosis[48]

Secondary syphilis: Meticulous history taking, previous history of chancre, lymphadenopathy, positive VDRL

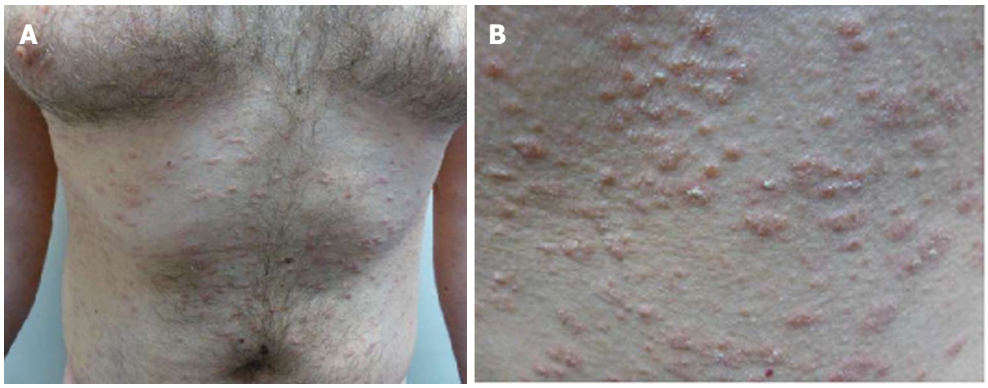

Figure 17 Pityriasis rosea-like rash. A: The eruption in this case was probably related to the ingestion of levothyroxine in a 33-year-old man, extensively affecting the trunk; B: The lesions are small and monomorphous (Courtesy of Dr. Elizabeth Rendic).

test, histology showing plasma cells and endarteritis obliterans are suggestive. Lesions of secondary syphilis are monomorphous and always asymptomatic; they almost always affect palms and soles.

Dermatophytosis: It may be troublesome to differentiate when the only lesion of PR is the herald patch. However, a mycotic lesion expands progressively and shows a clear center, whereas herald patch remains inalterable. Positive KOH mount is the pointer.

Guttate psoriasis: History of sore throat, presence of rain-drop pattern and histology are important clues. Scales are thicker and silvery-white.

Subacute cutaneous lupus erythematosus: Photosensitivity is the rule. Besides, histology shows epidermal atrophy and basal layer degeneration.

Rarely, primary HIV infection, seborrhoeic dermatitis, drug rash, erythema multiforme and cutaneous T cell lymphoma may also be confused with PR. Hypopigmented variant may be confused with pityriasis alba (lesions are mainly located on the face or arms and it is usually associated with atopic dermatitis), hypopigmented mycosis fungoides (lesions are large, persistent, and mainly distributed on buttocks and lower trunk), and progressive macular hypomelanosis of the trunk (lesions are slowly progressive, tend to coalesce, and do not show desquamation).

Therapeutic options

Many cases require no treatment at all, only reassurance directed to the patients, underlying the benign nature and self-limited duration of the disease, which do not leave sequelae and that other members of their family or friends will be not affected. Therapeutic options when needed (in the case of many or symptomatic lesions) include the use of emollients and topical corticosteroids, and antihistamines when itching.

The use of oral macrolides (erythromycin and azithromycin) have shown controversial results[49,50]. Initially, these were found to be beneficial but recent studies show that macrolides are ineffective in the management of PR.

Since the current concepts of etiopathogenesis may imply the role of HHV-7 and HHV-6 in the causation of PR, antivirals like acyclovir have been found to show good response[51-54]. The effectiveness of phototherapy is debated and further studies need to be conducted[55,56]. A statement about the management of PR has been recently raised[57]. Main conclusions include an adequate diagnosis, impact of the eruption in the quality of life since many patients do not necessitate any treatment, and use of oral acyclovir 400 mg three times daily for seven days, when not contraindicated or possible adverse effects are suspected.

CONCLUSION

The diagnosis of typical PR should not be difficult for any dermatologist. Nevertheless, its atypical presentations - as defined here - can be a challenge for the clinician. We hope the article will be helpful to the clinicians, in identifying numerous variants of this common disease.

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P- Reviewer: Firooz A, Hu SCS, Sadighha A, Vasconcellos C
S- Editor: Ji FF **L- Editor**: A **E- Editor**: Wu HL

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