

**The Indian Journal of Dermatology,
Venereology and Leprology (IJDLV)**

is a bimonthly publication of the Indian Association of Dermatologists, Venereologists and Leprologists (IADVL) and is published for IADVL by Medknow Publications.

The Journal is **indexed/listed** with Science Citation Index Expanded, PUBMED, EMBASE, Bioline International, CAB Abstracts, Global Health, DOAJ, Health and Wellness Research Center, SCOPUS, Health Reference Center Academic, InfoTrac One File, Expanded Academic ASAP, NIWI, INIST, Uncover, JADE (Journal Article Database), IndMed, Indian Science Abstract's and PubList.

All the rights are reserved. Apart from any fair dealing for the purposes of research or private study, or criticism or review, no part of the publication can be reproduced, stored, or transmitted, in any form or by any means, without the prior permission of the Editor, IJDLV.

The information and opinions presented in the Journal reflect the views of the authors and not of the IJDLV or its Editorial Board or the IADVL. Publication does not constitute endorsement by the journal.

The IJDLV and/or its publisher cannot be held responsible for errors or for any consequences arising from the use of the information contained in this journal.

The appearance of advertising or product information in the various sections in the journal does not constitute an endorsement or approval by the journal and/or its publisher of the quality or value of the said product or of claims made for it by its manufacturer.

The journal is published and distributed by Medknow Publications. Copies are sent to subscribers directly from the publisher's address. It is illegal to acquire copies from any other source. If a copy is received for personal use as a member of the association/society, one can not resale or give-away the copy for commercial or library use.

The Journal is printed on acid free paper.

EDITOR

Uday Khopkar

ASSOCIATE EDITORS

Ameet Valia Sangeeta Amladi

ASSISTANT EDITORS

K. C. Nischal Sushil Pande Vishalakshi Viswanath

EDITORIAL BOARD

Chetan Oberai (Ex-officio)	Koushik Lahiri (Ex-officio)	Sanjeev Handa
Arun Inamdar	Joseph Sundharam	S. L. Wadhwa
Binod Khaitan	Kanthraj GR	Sharad Mutalik
D. A. Satish	M. Ramam	Shruthakirti Sheno
D. M. Thappa	Manas Chatterjee	Susmit Halder
H. R. Jerajani	Rajeev Sharma	Venkatram Mysore
	Sandipan Dhar	

EDITORIAL ADVISORY BOARD

Aditya Gupta, Canada	Jag Bhawan, USA
C. R. Srinivas, India	John McGrath, UK
Celia Moss, UK	K. Pavithran, India
Giam Yoke Chin, Singapore	R. G. Valia, India
Gurmohan Singh, India	Robert A. Schwartz, USA
Howard Libman, USA	Robin Graham-Brown, UK
J. S. Pasricha, India	V. N. Sehgal, India
Rodney Sinclair, Australia	

STATISTICAL EDITOR

S. R. Suryawanshi

OMBUDSMAN

A. K. Bajaj

IADVL NATIONAL EXECUTIVE 2006 – 2007

President

Chetan M. Oberai

Immediate Past President

Suresh Joshipura

President (Elect)

S. Sacchidanand

Vice-Presidents

Amrinder Jit Kanwar

Dilip Shah

Secretary

Koushik Lahiri

Treasurer

Arijit Coondoo

Jt. Secretaries

Rakesh Bansal

Manas Chatterjee

EDITORIAL OFFICE

Dr. Uday Khopkar

Editor, IJDLV, Department of Dermatology,
117, 1st Floor, Old OPD Building, K.E.M.
Hospital, Parel, Mumbai - 400012, India.
E-mail: editor@ijdlv.com

Published for IADVL by

MEDKNOW PUBLICATIONS

A-109, Kanara Business Centre, Off Link Road,
Ghatkopar (E), Mumbai - 400075, India.
Tel: 91-22-6649 1818 / 1816
Website: www.medknow.com

Indian Journal of Dermatology, Venereology & Leprology

Journal indexed with SCI-E, PubMed, and EMBASE

Vol 74 | Issue 1 | Jan-Feb 2008

C O N T E N T S

EDITORIAL REPORT - 2007

JDVL gets into the Science Citation Index Expanded!

Uday Khopkar 1

EDITORIAL

Registration and reporting of clinical trials

Uday Khopkar, Sushil Pande 2

SPECIALTY INTERFACE

Preventing steroid induced osteoporosis

Jyotsna Oak 5

REVIEW ARTICLE

Molecular diagnostics in genodermatoses - simplified

Ravi N. Hiremagalore, Nagendrachary Nizamabad, Vijayaraghavan Kamasamudram 8

ORIGINAL ARTICLES

A clinicoepidemiological study of polymorphic light eruption

Lata Sharma, A. Basnet 15

A clinico-epidemiological study of PLE was done for a period of one year to include 220 cases of PLE of skin type between IV and VI. The manifestation of PLE was most common in house wives on sun exposed areas. Most of the patients of PLE presented with mild symptoms and rash around neck, lower forearms and arms which was aggravated on exposure to sunlight. PLE was more prevalent in the months of March and September and the disease was recurrent in 31.36% of cases.

Comparative study of efficacy and safety of hydroxychloroquine and chloroquine in polymorphic light eruption: A randomized, double-blind, multicentric study

Anil Pareek, Uday Khopkar, S. Sacchidanand, Nitin Chandurkar, Geeta S. Naik 18

In a double-blind randomized, comparative multicentric study evaluating efficacy of antimalarials in polymorphic light eruption, a total of 117 patients of PLE were randomized to receive hydroxychloroquine and chloroquine tablets for a period of 2 months (initial twice daily dose was reduced to once daily after 1 month). A significant reduction in severity scores for burning, itching, and erythema was observed in patients treated with hydroxychloroquine as compared to chloroquine. Hydroxychloroquine was found to be a safe antimalarial in the dosage studied with lesser risk of ocular toxicity.

Many faces of cutaneous leishmaniasis

Arfan Ul Bari, Simeen Ber Rahman

Symptomatic cutaneous leishmaniasis is diverse in its presentation and outcome in a tropical country like Pakistan where the disease is endemic. The study describes the clinical profile and atypical presentations in 41 cases among 718 patients of cutaneous leishmaniasis. Extremity was the most common site of involvement and lupoid cutaneous leishmaniasis was the most common atypical form observed. Authors suggest that clustering of atypical cases in a geographically restricted region could possibly be due to emergence of a new parasite strain.



23

Forehead plaque: A cutaneous marker of CNS involvement in tuberous sclerosis

G. Raghu Rama Rao, P. V. Krishna Rao, K. V. T. Gopal, Y. Hari Kishan Kumar, B. V. Ramachandra

In a retrospective study of 15 patients of tuberous sclerosis, eight patients had central nervous system involvement. Among these 8 cases, 7 cases had forehead plaque. This small study suggests that presence of forehead plaque is significantly associated with CNS involvement.

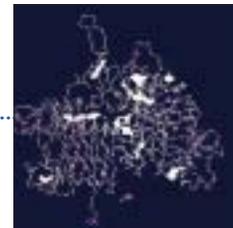


28

BRIEF REPORTS

Ligand-binding prediction for ErbB2, a key molecule in the pathogenesis of leprosy

Viroj Wiwanitkit.....



32

SCORTEN: Does it need modification?

Col. S. S. Vaishampayan, Col. A. L. Das, Col. R. Verma

35

CASE REPORTS

Universal acquired melanosis (Carbon baby)

P. K. Kaviarasan, P. V. S. Prasad, J. M. Joe, N. Nandana, P. Viswanathan



38

Adult onset, hypopigmented solitary mastocytoma: Report of two cases

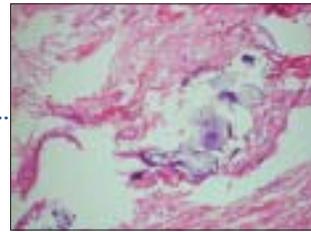
D. Pandhi, A. Singal, S. Aggarwal.....



41

Incidental finding of skin deposits of corticosteroids without associated granulomatous inflammation: Report of three cases

Rajiv Joshi



44

Erythromelanosus follicularis faciei et colli: Relationship with keratosis pilaris

M. Augustine, E. Jayaseelan



47

Naxos disease: A rare occurrence of cardiomyopathy with woolly hair and palmoplantar keratoderma

R. Rai, B. Ramachandran, V. S. Sundaram, G. Rajendren, C. R. Srinivas



50

Granular parakeratosis presenting with facial keratotic papules

R. Joshi, A. Taneja



53

Adult cutaneous myofibroma

V. Patel, V. Kharkar, U. Khopkar



56

LETTERS TO THE EDITOR

Extragenital lichen sclerosus of childhood presenting as erythematous patches

N. G. Stavrianeas, A. C. Katoulis, A. I. Kanelleas, E. Bozi, E. Toumbis-Ioannou



59

Leukocytoclastic vasculitis during pegylated interferon and ribavirin treatment of hepatitis C virus infection

Esra Adisen, Murat Dizbay, Kenan Hize, Nilsel İlter

60

Poland's syndrome

Saurabh Agarwal, Ajay Arya..... 62

Hereditary leiomyomatosis with renal cell carcinoma

Sachin S. Soni, Swarnalata Gowrishankar, Gopal Kishan Adikey,
Anuradha S. Raman 63

Infantile onset of Cockayne syndrome in two siblings

Prerna Batra, Abhijeet Saha, Ashok Kumar 65

Multiple xanthogranulomas in an adult

Surajit Nayak, Basanti Acharjya, Basanti Devi, Manoj Kumar Patra 67



Bullous pyoderma gangrenosum associated with ulcerative colitis

Naik Chandra Lal, Singh Gurcharan, Kumar Lekshman, Lokanatha K..... 68



Sporotrichoid pattern of malignant melanoma

Ranjan C. Rawal, Kanu Mangla..... 70



Acitretin for Papillon-Lefèvre syndrome in a five-year-old girl

Didem Didar Balci, Gamze Serarslan, Ozlem Sangun, Seydo Homan 71

Bilateral Becker's nevi

Ramesh Bansal, Rajeev Sen..... 73



RESIDENTS' PAGE

Madarosis: A dermatological marker

Silonie Sachdeva, Pawan Prasher 74

FOCUS

Botulinum toxin

Preeti Savardekar 77

E-IDVL

Net Studies

A study of oxidative stress in paucibacillary and multibacillary leprosy

P. Jyothi, Najeeba Riyaz, G. Nandakumar, M. P. Binitha 80

Clinical study of cutaneous drug eruptions in 200 patients

M. Patel Raksha, Y. S. Marfatia 80

Net case

Porokeratosis confined to the genital area: A report of three cases

Sujata Sengupta, Jayanta Kumar Das, Asok Gangopadhyay 80

Net Letters

Camisa disease: A rare variant of Vohwinkel's syndrome

T. S. Rajashekar, Gurcharan Singh, Chandra Naik, L. Rajendra Okade 81

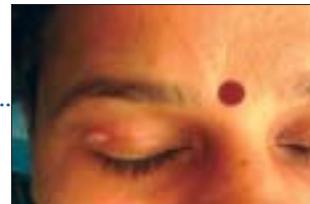
Cross reaction between two azoles used for different indications

Arika Bansal, Rashmi Kumari, M. Ramam 81

Net Quiz

Asymptomatic erythematous plaque on eyelid

Neeraj Srivastava, Lakhan Singh Solanki, Sanjay Singh 82



QUIZ

A bluish nodule on the arm

Ragunatha S., Arun C. Inamadar, Vamseedhar Annam, B. R. Yelikar 83



REFEREE INDEX-2007

INSTRUCTIONS FOR AUTHORS

The copies of the journal to members of the association are sent by ordinary post. The editorial board, association or publisher will not be responsible for non-receipt of copies. If any of the members wish to receive the copies by registered post or courier, kindly contact the journal's / publisher's office. If a copy returns due to incomplete, incorrect or changed address of a member on two consecutive occasions, the names of such members will be deleted from the mailing list of the journal. Providing complete, correct and up-to-date address is the responsibility of the members. Copies are sent to subscribers and members directly from the publisher's address; it is illegal to acquire copies from any other source. If a copy is received for personal use as a member of the association/society, one cannot resale or give-away the copy for commercial or library use.

Porokeratosis confined to the genital area: A report of three cases

Sujata Sengupta, Jayanta Kumar Das, Asok Gangopadhyay

Department of Dermatology, R.K.M. Seva Pratisthan and V.I.M.S, Kolkata, India

Address for correspondence: Dr. Sujata Sengupta, UV 24/3C, Udayan, 1050/1, Survey Park, Kolkata - 700 075, West Bengal, India.

E-mail: senguptasujata@yahoo.co.in

ABSTRACT

Genital involvement in porokeratosis (PK) is a rare occurrence even in disseminated forms. We encountered three patients who had porokeratosis affecting only the genital area. Two of them were male with involvement of the penis and scrotum and only the scrotum respectively. The lady with vulvar involvement is a hitherto unreported instance of porokeratosis confined to female genitalia. None of the cases were very easy to diagnose clinically but biopsies proved confirmatory. The male patients were advised light electrocautery under local anesthesia while the female patient underwent surgical excision. No malignant change has been reported in them till date.

Key Words: Female genitalia, Genital porokeratosis, Porokeratosis

Porokeratosis (PK) is a clonal disorder of keratinization showing one or multiple atrophic patches surrounded by a distinct ridge-like border. The common clinical variants include classical plaque-type porokeratosis of Mibelli (PM), disseminated superficial porokeratosis, linear porokeratosis, porokeratosis palmaris et plantaris disseminate and punctate porokeratosis.^[1] Genital porokeratosis (GP), though uncommon, can occur as a part of a more generalized involvement.^[2] But PK localized to the genital area is a rare entity. We report such an occurrence in one female patient and two male patients.

CASE REPORTS

Case 1

A 36-year-old housewife from a rural socioeconomic background presented with a dry, itchy vulvar lesion for the last 8 months. There was no history of sexual promiscuity or local irradiation and her family history was not significant. Topical steroids, antifungal and antibiotic creams had yielded no results. A hyperkeratotic verrucous plaque, 2 × 1.5 cm, was seen involving the vestibule, fourchette and adjacent perineal skin [Figure 1]. No similar skin lesion was present anywhere else in her body and systemic examination was normal. Histology from the edge of the skin lesion showed

a parakeratotic column (cornoid lamella) in the epidermal invagination and underlying hypogranulosis [Figure 2]. A mild perivascular infiltrate was seen in the dermis. She was diagnosed to have vulvar PK. She was advised surgical excision.

Case 2

A 35-year-old businessman presented with slowly spreading multiple asymptomatic brownish lesions on the scrotum and penis for the last 8 months. There was no history of extramarital sexual exposure, drug intake; and his family history was noncontributory. Topical and systemic steroids had produced no results. Several well-defined hyperpigmented annular plaques of varying sizes were seen on the penile shaft and scrotum [Figure 3]; each one had a mildly scaly atrophic center and a raised border [Figure 4]. The rest of the skin and systemic examination was normal. We initially considered the genital lesions to be annular lichen planus (LP), but a biopsy was advised. Histopathology was typical of PK with the cornoid lamella and epidermal invagination.

Case 3

A 34-year-old man reported with an asymptomatic plaque on the scrotal skin for the last 3 months. It had started as a small papule, which expanded slowly to its present size. His



Figure 1: Vulvar porokeratosis

family and sexual history was not significant. He had a 1.5 × 1 cm depigmented annular keratotic plaque on the scrotal skin that was surrounded by a raised border [Figure 5]. On close inspection, the border was found to be traversed by a groove. Skin biopsy was consistent with PK. The lesion healed, with a thin scar, after electrodesiccation under local anesthesia.

None of the three patients had inguinal adenopathy or any evidence of sexually transmitted disease (STD). Routine hematological and biochemical tests were normal and VDRL and ELISA for HIV were negative. Till date, malignant change has not been seen.

DISCUSSION

PK confined to the genital area is rarely seen. We came across



Figure 2: Cornoid lamella with underlying hypogranulosis (H and E stain, X400)



Figure 4: Porokeratosis on scrotum



Figure 3: Annular plaques of porokeratosis on penis and scrotum



Figure 5: Keratotic depigmented plaque on scrotum

about 20 such cases in English literature.^[3] Surprisingly, all the cases were males. PK is two to three times more common in males than in females, but GP in females seems even rarer. Robinson *et al.*^[4] gave the first report of vulvar PK in a 39-year-old lady who had disfiguring lesions in the perineal area, medial thigh and sole. But our report of the female patient is probably the first report of PK confined to female genitalia.

PK localized to the male genitalia has been reported most commonly in the scrotum, followed by penis, buttock, natal cleft, groins and adjacent thighs.^[3] Porter *et al.*^[5] came across a case involving the external urethral meatus that was treated with topical 5-fluorouracil. In our cases PK was localized to the penis and/or scrotum and involvement of skin adjacent to genitalia was not seen. Though the inheritance of PK is known to be autosomal dominant, none of our cases had a positive family history. Similarly, family history was noncontributory in all the 10 cases studied by Chen and his colleagues.^[3] In contrast to their study, none of our patients had diabetes or sexually transmitted diseases (STDs). Malignant transformation into squamous or basal cell carcinoma has been seen in almost all forms of PK.^[6] We have earlier reported multicentric squamous cell carcinoma (SCC) of the inguinal lesions of disseminated PK.^[7] But that patient did not have genital lesions. In fact, malignant change has not been found in GP till date.^[8]

Clinically, none of the cases were very easy to diagnose. We thought of condyloma acuminata in Case 1; but there was no sexual promiscuity and the response to imiquimod was unsatisfactory. This uncommon verrucous form of PK has been reported to occur over scrotum and buttocks.^[3,9] In Case 2, the annular plaques had raised borders but no typical ridge was seen. We considered lichen planus but extragenital or oral lesions were not seen. Annular syphilide was another possibility but VDRL was negative. A distinct keratotic ridge helped to diagnose Case 3 even though the center was depigmented.

The treatment options of GP are cryotherapy (liquid N₂), surgery, CO₂ laser, topical 5% 5-fluorouracil and imiquimod cream.^[2,5,10] We found that all our patients had been previously treated for GP with either antifungals or topical and systemic steroids. This highlights the possibility that PK confined to the genital area is an under-diagnosed entity and can be easily confused with STDs and other nonvenereal diseases affecting genitalia. A thorough clinical evaluation is mandatory and skin biopsy may be diagnostic in such cases. In addition, keeping in mind the potential for malignancy, all diagnosed cases should receive regular follow-up.

REFERENCES

1. Spencer LV. Porokeratosis. Available from: <http://www.emedicine.com/derm/topic343.htm>. [Last accessed on 2006 Oct 9].
2. Levell NJ, Bewly AP, Levene GM. Porokeratosis of Mibelli on the penis, scrotum and natal cleft. *Clin Exp Dermatol* 1994;19:77-8.
3. Chen TJ, Chow YC, Chen CH, Kuo TT, Hong HS. Genital porokeratosis: A series of 10 patients and review of the literature. *Br J Dermatol* 2006;155:325-9.
4. Robinson JB, Im DD, Jockle G, Roshenshein NB. Vulvar porokeratosis: Case report and review of literature. *Int J Gynecol Pathol* 1999;18:169-73.
5. Porter WM, Du P Menagé H, Philip G, Bunker CB. Porokeratosis of the penis. *Br J Dermatol* 2001;144:643-4.
6. Judge MR, McLean WH, Munro CS. Disorders of keratinization. *In: Burns T, Breathnach S, Cox N, Griffiths C, editors. Rook's Textbook Of Dermatology. 7th ed. Blackwell Science: Oxford; 2004. p. 34.1-34.111.*
7. Sengupta S, Das JK, Gangopadhyay A. Multicentric squamous cell carcinoma over lesions of porokeratosis palmaris et plantaris disseminata and giant porokeratosis. *Indian J Dermatol Venereol Leprol* 2005;71:414-6.
8. Neri I, Marzaduri S, Passarini B, Patrizi A. Genital porokeratosis of Mibelli. *Genitourin Med* 1995;71:410-1.
9. Wallner JS, Fitzpatrick JE, Brice SL. Verrucous porokeratosis of Mibelli on the buttocks mimicking psoriasis. *Cutis* 2003;72:391-3.
10. Agarwal S, Berth Jones J. Porokeratosis of Mibelli. *Br J Dermatol* 2002;146:338-9.