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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.

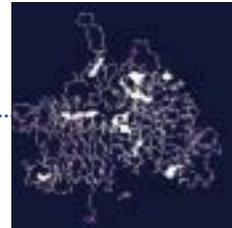


28

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

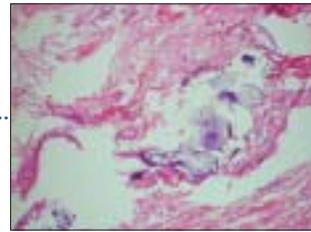
D. Pandhi, A. Singal, S. Aggarwal.....



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. Yelikor 83



REFEREE INDEX-2007

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Camisa disease: A rare variant of Vohwinkel's syndrome

Sir,

Vohwinkel's syndrome is a rare, dominantly inherited keratoderma of palms and soles with a characteristic honeycomb appearance, linear and/or starfish keratoses on the extensor surfaces of the elbows, knees, knuckles and hands with flexion contractures and constricting bands (pseudoainhum) of digits resulting in autoamputation.^[1,2] We report a case of Camisa disease, a rare variant of Vohwinkel's syndrome.

A 3-year-old female child born of 2nd degree consanguineous marriage presented with hyperkeratosis of the palms and soles, linear hyperkeratotic plaques over dorsa of hands with loss of left great toe, of 1-year duration [Figure 1]. Constricting fibrous bands were seen over the right great toe and right 5th toe [Figure 2]. Generalized ichthyosis was present, being more prominent over extremities [Figures 1-2]. Ridging and onychodystrophy was seen in toe nails though hair growth was normal.

Her audiogram, eyes and dental examination were normal. There was no delay in developmental milestones. Hematological and biochemical investigations were within normal limits and peripheral smear showed microcytic hypochromic anemia with mild eosinophilia. Blood serological investigations (HIV and VDRL) were normal. Abdominal scan showed hepatomegaly. Skin biopsy revealed hyperkeratosis, focal parakeratosis, acanthosis, elongation of rete ridges and sparse dermal lymphocytic infiltrate with normal appendages.

Camisa disease is a rare variant of Vohwinkel's syndrome associated with generalized ichthyosis and without deafness.^[1,2] On the basis of recent molecular studies, it is now clear that Vohwinkel's syndrome associated with ichthyosis is caused by mutations in *loricrin* gene.^[3-5] However, a variant of Vohwinkel's syndrome which had all the classical clinical features of Vohwinkel's syndrome but lacking atypical associations like ichthyosis and sensorineural deafness with negative gene mapping for *loricrin* mutation has been reported recently.^[6]



Figure 1: Linear keratosis over dorsum of hands associated with ichthyosis



Figure 2: Constricting fibrous bands over right great toe and fifth toe with loss of left great toe

Along with the features of Vohwinkel's syndrome, our patient had generalized ichthyosis, which is similar to the previous case reports of Camisa variant of Vohwinkel's syndrome.^[1,4] The other clinical variant of Vohwinkel's syndrome is associated with deafness but no ichthyosis.^[3] However, audiogram

revealed no hearing loss in our patient. Histologically the skin lesion showed hyperkeratosis, focal parakeratosis, acanthosis, elongation of rete ridges and sparse dermal lymphocytic infiltrate with normal appendages, which were consistent to earlier case report of an ichthyotic (or Camisa) variant of Vohwinkel's syndrome.^[4] Thus our case represents a rare variant of Vohwinkel's syndrome, termed as Camisa disease.

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