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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.

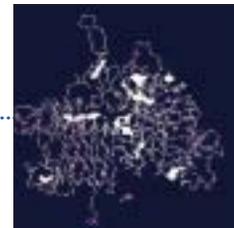


28

**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**

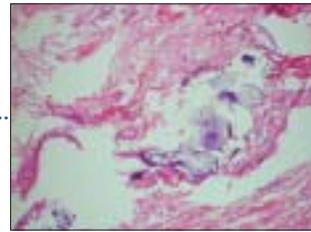
D. Pandhi, A. Singal, S. Aggarwal.....



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

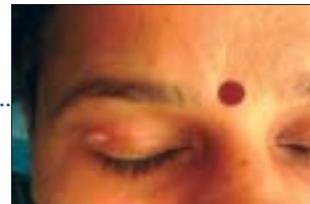
**Cross reaction between two azoles used for different indications**

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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## 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

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### ABSTRACT

**Background:** Tuberous sclerosis complex (TSC) is a neurocutaneous genodermatosis characterized by hamartoma formation in multiple organs. There are no definite cutaneous markers suggestive of central nervous system (CNS) involvement in TSC. **Aims:** To study association of forehead plaque seen in tuberous sclerosis patients and CNS involvement in TSC. **Methods:** This is a retrospective study of 15 cases of tuberous sclerosis in varying age groups - from 1.5 to 50 years. All the cases were thoroughly evaluated with detailed history; clinical examination; and relevant investigations like X-rays of chest, skull, hands and feet; ultrasound abdomen and computed tomography of brain. **Results:** Out of the 15 cases, CNS involvement was seen in 8 cases. Seizures were present in 8 cases (53.33%) and mental retardation was seen in 6 cases (40%). Computerized tomography of brain revealed subependymal nodules (SENs) in eight cases (53.33%). In addition to SENs, subependymal giant cell astrocytomas and cortical tubers were seen in 2 cases each. Out of these 8 cases having CNS involvement, in 7 cases forehead plaque was observed. In 1 case, no forehead plaque was observed ( $X^2 = 1.07$ ,  $P < 0.05$ ). **Conclusion:** This study shows that there is a statistically significant relationship between the presence of a forehead plaque and CNS involvement in TSC. Therefore, forehead plaque may be considered as a novel cutaneous marker to know the CNS involvement in TSC at an early stage.

**Key Words:** Central nervous system manifestations, Fibrotic forehead plaque, Tuberous sclerosis complex

### INTRODUCTION

Tuberous sclerosis complex (TSC) is an autosomal dominant neurocutaneous genodermatosis characterized by hamartoma formation in multiple organs - like skin, brain, kidney, lung, heart and eyes.<sup>[1-3]</sup> The incidence of TSC is about 1 in 10,000, with half of the TSC families linked to chromosome 9q34 and the other half to 16p13. Tuberin (TSC1) and hamartin (TSC2), proteins having tumor-suppressor activity, located on chromosomes 9 and 16 respectively are the known defective proteins in TSC.<sup>[4,5]</sup> Approximately 60% of cases occur as apparent sporadic cases without any family history, due to germline mosaicism.<sup>[6]</sup> The definitive diagnosis of TSC is made by the presence of either one primary feature like facial angiofibromas, subungual fibromas, cortical tubers, etc.; or two secondary

features or one secondary plus two tertiary features.<sup>[2,7]</sup>

CNS manifestations like seizures occur in 86%, mental retardation in 49% and cutaneous manifestations are seen in almost 96% patients of TSC.<sup>[5,8]</sup> Cutaneous manifestations of TSC include facial angiofibromas, subungual fibromas, hypomelanotic macules, forehead fibrous plaques and shagreen patches.<sup>[2,7,9]</sup> In 1961, Nickel and Reed observed fibromatous forehead plaques in patients with advanced mental retardation. They opined that presence of fibrotic forehead plaque was a poor prognostic sign in tuberous sclerosis.<sup>[10]</sup> Till now, there are no specific studies to observe the relationship between forehead plaque and CNS manifestations. The objective of the present study is to examine the relationship between the presence of forehead plaque and CNS involvement in TSC.

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## METHODS

This retrospective study was conducted in the Department of Dermatology, King George Hospital and Andhra Medical College, Visakhapatnam, between May 2003 and October 2004. The study group included 15 cases of tuberous sclerosis. Diagnosis of tuberous sclerosis was made on the basis of the presence of at least one primary feature, which included facial angiofibromas, multiple subungual fibromas, cortical tubers, subependymal nodules or giant cell astrocytomas and multiple retinal astrocytomas.<sup>[2,7]</sup>

In all patients, a detailed clinical history was taken with reference to age at onset of various cutaneous lesions, infantile spasms, seizures or mental retardation. Family history was taken in all patients, including details of any affected first-degree relative, consanguinity and genetic pedigree. In all patients, thorough dermatological and CNS examination was carried out. Complete ophthalmologic examination was also done in all patients with direct and indirect ophthalmoscopy and fundoscopy to detect any retinal hamartomas. In all 15 cases, computed tomography of brain was performed to find out any CNS lesions.

Relevant investigations like routine hematological and biochemical tests; X-rays of the chest, skull, hands and feet; and ultrasound abdomen were performed. Elliptical biopsy of the forehead plaque was done in seven patients to study the histopathological features.

## RESULTS

Out of the 15 TSC patients, 8 were males and 7 were females.

The age of the patients varied from 1.5 years to 50 years. Mean age was 15.9 years. Seven of the 15 patients gave a family history of TSC, with at least one affected first-degree relative. Consanguinity of parents was found in 3 cases.

The various clinical features of our cases are given in Table 1. Forehead plaque was observed in 7 of the 15 cases (47%). In 4 cases, a single forehead plaque was present since birth [Figure 1]. In the 3 other cases, two or more forehead plaques were present, which developed at the age of 2, 3 and 4 years respectively [Figure 2]. Histopathological examination of the forehead plaques revealed features suggestive of connective tissue hamartoma consisting of vascular, fibrous and dermal tissues.

Specific CNS manifestations and their relationship with forehead plaque are shown in Table 2. Out of the 15 cases, CNS involvement was seen in 8 cases. History of seizures was present in 8 of the 15 cases (53.33%). Out of these 8 cases, 3 cases had infantile spasms; and in 6 cases, mental retardation was observed. CT scan of brain revealed subependymal nodules (SENs) in 8 of the 15 cases (53.33%) [Figure 3] In addition to SENs, subependymal giant cell astrocytomas and cortical tubers were seen in 2 cases each and retinal phakomas were seen in 1 case [Figures 4]. Out of these 8 cases having CNS involvement, in 7 cases forehead plaque was observed ( $\chi^2 = 1.07, P < 0.05$ ). In 1 case, no forehead plaque was observed. In the remaining 7 cases, neither CNS involvement nor fibrotic forehead plaque was seen.

Routine hematological, biochemical investigations and X-ray studies were within normal limits in all patients. Ultrasound scanning of the abdomen revealed renal angiomyolipomas

**Table 1: Clinical features (N = 15)**

Case no.	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	Total
Facial angiofibromas	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	14
Shagreen patches	+	+	+	+	+	+	+	+	+	+	+	-	+	-	-	12
Hypomelanotic macules	-	+	+	+	-	+	+	+	+	+	+	-	+	+	+	12
Subungual fibromas	-	-	+	-	-	-	-	-	-	+	-	-	-	-	-	2
Forehead plaque	+	-	+	+	-	-	+	-	-	-	-	-	+	+	+	7
CNS Involvement	+		+	+	-	-	+	-	-	-	-	+	+	+	+	8
Renal changes	-	-	-	-	-	-	+	-	-	-	-	-	-	-	-	1
Eye changes	+	-	-	-	-	-	-	-	-	-	-	-	-	-	-	1

**Table 2: Forehead plaque and CNS involvement (N = 8)**

Case no.	1	3	4	7	12	13	14	15
Forehead plaque	+	+	+	+	-	+	+	+
Convulsions	+	+	+	+	+	+	+	+
Age at onset of seizures	2 yrs	3 yrs	10 m	2 yrs	8 yrs	4 m	12 yrs	5 m
CT findings	SENS, SEGA, RP	SENS	SENS	SENS	SENS, CT	SENS	SENS, CT	SENS, SEGA

SENS - Subependymal nodules, SEGA - Subependymal giant cell astrocytoma, CT - Cortical tubers, RP - Retinal phakomas



Figure 1: Single forehead plaque

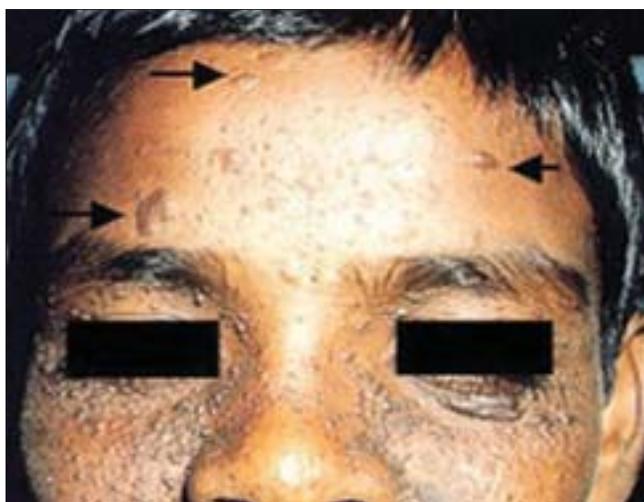


Figure 2: Multiple forehead plaques

and distal aortic aneurysm in Case 7.

## DISCUSSION

The systemic nature of tuberous sclerosis was first described by Vogt in the clinical triad of seizures, mental retardation and adenoma sebaceum, all of which are not consistently present in all cases.<sup>[11]</sup> TSC is usually classified as one of the phakomatoses or neurocutaneous syndromes, a group which includes more than 50 entities.<sup>[11,12]</sup> It is differentiated from the other members of the group by its involvement of nearly all organ systems and tissues.<sup>[1,2]</sup> Pathologically, it is a disorder of cellular migration, proliferation and differentiation.<sup>[13]</sup>

The cutaneous manifestations of TSC are hypomelanotic macules, confetti skin lesions, facial angiofibromas, unguinal

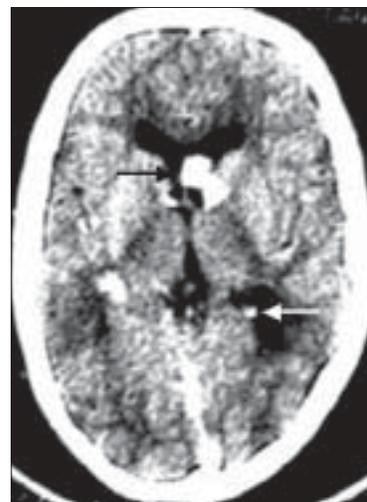


Figure 3: Subependymal nodule (white arrow) and subependymal giant cell astrocytoma (black arrow)

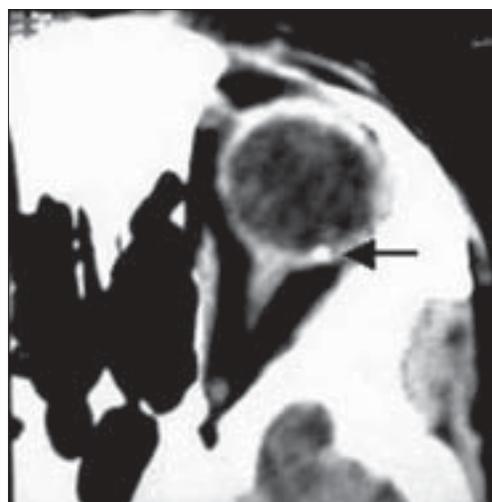


Figure 4: Retinal phakoma

fibromas, shagreen patches and forehead plaque. The last four of these provide strong support for a diagnosis of TSC.<sup>[2,5,7]</sup> According to Jozwiak S *et al.*, hypopigmented macules were the most frequent finding (97.2%) and facial angiofibromas were the next common cutaneous lesion.<sup>[14]</sup> Forehead plaques were observed in 20 of 103 cases (18.9%). Webb *et al.*, reported forehead plaque in 36% of cases.<sup>[15]</sup> In our study, in 47% of cases of TSC, forehead plaque was observed. Fibrotic forehead plaques are large connective tissue hamartomas that are fibromatous, soft, compressible, doughy-to-firm tumorous or plaque-like lesions commonly present on the forehead, eyelids, upper cheeks and scalp. These are present in up to 25% of patients with TSC, are often multiple, and are commonly seen on the forehead.<sup>[10,16]</sup> Recently, it was found that forehead plaques are more frequently seen in TSC2 patients than in TSC1 patients.<sup>[17]</sup> The individual lesions tend to be much larger than the

angiofibromas on the face, are often unilateral and may be present at birth.

Nickel and Reed in 1961 had observed that in tuberous sclerosis, fibromatous forehead plaque was not observed in normal patients but was common in hospitalized patients with advanced mental retardation. They suggested that the presence of forehead plaque was a poor prognostic sign.<sup>[10]</sup> Since then, no attempts have been made to establish the association between forehead plaque and CNS involvement. In various previous studies, though forehead plaque was observed, no correlation between these lesions and CNS involvement was made.<sup>[8,9,14,15,18-20]</sup> In one study, forehead plaque along with retinal phakomas and multiple intracranial periventricular calcifications was reported.<sup>[20]</sup> In our study, various CNS manifestations like infantile spasms, persistent seizures, mental retardation, subependymal nodules, subependymal giant cell astrocytomas, cortical tubers and retinal phakomas were seen in 8 of the 15 cases (53.33%); and in 7 of these cases, forehead plaque was observed ( $\chi^2 = 1.07$ ,  $P < 0.05$ ). In the remaining 7 cases, though other cutaneous manifestations like angiofibromas, shagreen patches and subungual fibromas were seen, no clinical or radiological evidence of CNS involvement was seen. Our findings show that there is a significant relationship between the presence of forehead plaque and CNS involvement. Therefore, whenever fibrotic forehead plaques are seen in TSC patients, a thorough radiological search may be carried out to rule out the involvement of other organ systems, especially CNS, even in the absence of clinical manifestations.

We suggest that forehead plaque can be considered to be a novel cutaneous marker of CNS involvement at an early stage so that proper and timely prophylactic measures can be undertaken to prevent seizures, mental retardation and permanent CNS damage. However, larger clinical studies are warranted to establish forehead plaque as one of the important prognostic markers.

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