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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.

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The Journal is printed on acid free paper.
A clinicoepidemiological study of polymorphic light eruption
Lata Sharma, A. Basnet

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 housewives 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

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.

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.

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

Viroj Wiwanitkit

SCORTEN: Does it need modification?

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

Universal acquired melanosis (Carbon baby)

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

Adult onset, hypopigmented solitary mastocytoma: Report of two cases

D. Pandhi, A. Singal, S. Aggarwal
Incidental finding of skin deposits of corticosteroids without associated granulomatous inflammation: Report of three cases
Rajiv Joshi ..................................................................................................................................................................... 44

Erythromelanosis 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 İltuer ........................................................................................................... 60
Poland's syndrome
Saurabh Agarwal, Ajay Arya

Hereditary leiomyomatosis with renal cell carcinoma
Sachin S. Soni, Swarnalata Gourishankar, Gopal Kishan Adikey, Anuradha S. Raman

Infantile onset of Cockayne syndrome in two siblings
Prerna Batra, Abhijeet Saha, Ashok Kumar

Multiple xanthogranulomas in an adult
Surajit Nayak, Basanti Acharjya, Basanti Devi, Manoj Kumar Patra

Bullous pyoderma gangrenosum associated with ulcerative colitis
Naik Chandra Lal, Singh Gurcharan, Kumar Lekshman, Lokanatha K

Sporotrichoid pattern of malignant melanoma
Ranjan C. Rawal, Kanu Mangla

Acitretin for Papillon-Lefèvre syndrome in a five-year-old girl
Didem Didar Balci, Gamze Serarslan, Ozlem Sangun, Seydo Homan

Bilateral Becker's nevi
Ramesh Bansal, Rajeev Sen

Madarosis: A dermatological marker
Silonie Sachdeva, Pawan Prasher
CONTENTS (Contd.)

FOCUS

Botulinum toxin
Preeti Savardekar .......................................................... 77

E-IJDL

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

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INTRODUCTION

Cutaneous leishmaniasis (CL) is a major world health problem that is growing epidemically in Pakistan and Afghanistan. It is a parasitic disease caused by Leishmania and transmitted by the bite of some species of sandflies and it affects various age groups. Phlebotomus papatasi, Phlebotomus sergenti and Phlebotomus salehi are the sandfly species reported from Pakistan. Depending on the infecting Leishmania species and host immunocompetence, there are cutaneous, mucocutaneous and visceral forms of the disease. CL typically presents with a skin ulcer over exposed regions of the body after a sandfly bite and generally heals spontaneously within 3-6 months. In addition to classical clinical picture, several unusual and atypical clinical features of the disease have been reported in the literature.

Lesions may appear at unusual sites or the disease may present with atypical morphologies. We recorded all the atypical and unusual morphological variants of the disease that we encountered during the last 5 years. Our cases have actually extended the spectrum of clinical presentations of CL in the Old World.

METHODS

Patients of all ages and both sexes, military personnel as well as civilian population, reporting to dermatology departments of Military Hospital, Rawalpindi (January 2002 to December 2002); Pakistan Air Force Hospital, Sargodha (January 2003 to December 2005); and Combined Military Hospital, Muzaffarabad (January 2006 to December 2006), were included in the study. They either belonged to an
enigmatic area of the disease or had traveled to some endemic region during the last 6 months. Apparently infected patients who showed clearance of their lesions after short courses of antibiotics were excluded from the study. All patients having clinically suggestive lesions (one or more discrete nodules, plaques, non-healing ulcers on exposed sites or any atypical lesion anywhere on the body not responding to repeated courses of antibiotics and anti tuberculosis drugs) were subjected to slit skin smear examination for Leishman Donovan (LD) bodies. Negative skin smears were followed by skin biopsy for histopathological diagnosis. They all had clinical and parasitological diagnosis of cutaneous leishmaniasis, but some of them exhibited unusual clinical pattern or atypical morphology of the disease. Patients initially diagnosed differently but later proving to be leishmaniasis on slit skin smears and histopathology were also included. Number of the lesions, type of the lesions, site of involvement and gross morphology of the lesions were noted before starting treatment. Any change in morphology of the lesions and new developments during progression of the disease were also recorded. Out of all these, patients with unusual lesions (other than typical papulo-nodulo-ulcerative lesions and crusted plaques) were isolated and categorized accordingly using simple descriptive statistics. All unusual lesions were also photographed. Majority of the patients were treated indoors with once-daily intramuscular injection of meglumine antimonate for 2-3 weeks. Some with isolated lesions over non-articular and nonfacial sites were given intralésional injection of meglumine antimonate twice weekly for a total of six to eight injections in the outpatient clinic. In a few patients having chronic, large, heaped-up lesions not adequately responding to systemic treatment, cryotherapy twice weekly was used as an adjunctive treatment modality.

RESULTS

The study included 500 (69.6%) males and 137 (19.1%) females and 81 (11.3%) children. There age ranged from 2 to 68 years (mean age 25.7 years). Majority of the patients had lesions over exposed parts of the body. The duration of the disease varied between 1 and 18 months. The sites commonly affected were upper limbs (40%), lower limbs (32%), face (21%) and trunk (6%). The clinical profile that emerged was basically of the following three types: dry type (slowly evolving nodule or plaque with late ulceration, urban type) in 529 (73.7%) patients, wet type (rapidly evolving papule, nodule or plaque with early ulceration, rural type) in 156 (21.7%) patients and miscellaneous type (lupoid and other unusual morphologies) in 33 (4.6%) patients. Almost all patients who contracted the disease in Punjab had dry type lesions and patients with wet type disease were either from Kashmir or were those who had been in some endemic regions of Baluchistan or interior Sindh in the recent past or at least had visited the same areas during the last 1 year. Out of the total 718 patients, 41 (5.7%) had unusual presentations. Most common was lupoid leishmaniasis 14 (34.1%) [Figure 1], followed by sporotrichoid 5 (12.1%) [Figure 2] and paronychial 3 (7.3%) [Figure 3], erysipeloid, lid leishmaniasis [Figure 4], psoriasiform, mycetomatous and chancriform lesions were seen in 2 (4.9%) patients each. Scar leishmaniasis, zosteriform, whitlow, palmar/planter [Figure 5], DLE-like, squamous cell carcinoma, eczematous and verrucous morphology [Figure 6] were seen in 1 (2.4%) patient each. Mucocutaneous lesions were seen in 1 (2.4%) child. Chronic cutaneous disease (recidivans cutaneous leishmaniasis) was not seen in any of the patients; however, one patient presented with persistent boggy swelling over the dorsum of foot for the last 18 months, which mimicked squamous cell carcinoma. Geographical distribution of these unusual presentations is given in Table 1. Atypical presentations of cutaneous leishmaniasis are detailed in Figure 7.

DISCUSSION

Symptomatic CL is diverse in its presentation and outcome. This clinical diversity is basically governed by parasite and host factors and immuno-inflammatory responses.[3] Clinically the wet (rural) type of the disease is more common in Baluchistan (southern province which borders with Iran and Afghanistan) and interior regions of Sindh province, while dry (urban) type is more prevalent in Punjab province, suggesting the presence of the two main strains of parasite (Leishmania major and leishmania tropica) in the country.[5,9] Vast majority of our patients had dry (urban) type lesions, as the causative strain of parasite in these geographical regions was Leishmania tropica. Wet (rural type) lesions were only seen in patients who had a history of stay in or visit to, Baluchistan or Sindh province, where the prevalent strain was Leishmania major. We encountered a number of atypical variants of localized cutaneous disease. The proportion of these unusual morphologies was significantly higher in our study and the spectrum we observed was also greater than that seen in any of the previous studies.[5-8] Unusual variants were seen in 5.7% of our patients. This was twice to what has been reported earlier by Raja et al,[5] Shams et al[7] and Calvopena et al.[8] We had 17 different atypical presentations. Some of these atypical variants were already described by Raja et al,[5] (paronychial, chancriform, annular, palmoplantar,
Bari, et al.: Cutaneous leishmaniasis

zosteriform and erysipeloid) and more recently by Shams et al.,[7] (paronychial, annular, eyelid, chancriform, zosteriform, palmoplantar, DLE-like and eczematous CL). Both of these studies were done in Baluchistan, while all of our patients belonged to central and northern Punjab and the northern
areas of the country from where such atypical cases were not reported previously.

The commonest atypical pattern (34%) was lupoid leishmaniasis and surprisingly these were not very chronic cases of CL or the leishmaniasis recidivans as described in the past. They had duration of less than 1 year (mostly 4-8 months) and 12 out of the 14 cases were from the Kashmir region. In these patients, the lesion started as a small painless plaque on the nose or a side of the face, which progressively enlarged to involve most of the face in 2-3 months' time. Ten of the 14 affected patients were females. This geographical restriction points towards the possibility of some different strains of parasite in the region or the altered host-immune response and predominance in females could be due to social or cultural factors (as most Kashmiri women work outdoors in fields and the face is the only uncovered part of the body apart from hands and feet).

Sporotrichoid pattern was restricted to wet type and was seen in patients who visited the endemic areas of Baluchistan or Sindh province. These nodules are thought to represent an immune reaction from direct lymphatic extension of leishmania organisms or antigens. Relative rarity of this pattern was due to the fact that a vast majority of our patients had dry type of lesion due to *Leishmania tropica*, where sporotrichoid spread is not commonly seen.

Most of the other atypical presentations, like paronychial, whitlow, lid, scar, palmoplantar and chancriform, were probably related to the normal host response to the bite of the sandfly at these atypical sites. Verrucous, psoriasiform, erysipeloid, zosteriform, mycetomatous, DLE-like, squamous cell carcinoma-like and eczematous morphologies were likely to be due to some altered or over-expressed immune host responses. One case of mucocutaneous disease in a child was probably due to local extension of the lesions over the lip and nose to the mucosal surfaces and was not due to different strain of the parasite. We did not see any patient with chronic recidivans lesions or scalp lesions. Recidivans cases were not encountered probably due to variable immune host response or different causative strain of parasite in the region. It may be a chance finding as recidivans cases had been reported earlier from Baluchistan. Similarly, no case of post-kala-azar dermal leishmaniasis, diffuse cutaneous leishmaniasis and cutaneous disease in immunocompromised patient was encountered, possibly because of low prevalence of visceral leishmaniasis and AIDS.

<table>
<thead>
<tr>
<th>Regions</th>
<th>Males</th>
<th>Females</th>
<th>Children</th>
<th>Total no</th>
<th>Typical</th>
<th>Atypical</th>
<th>Atypical presentations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Central Punjab</td>
<td>134</td>
<td>26</td>
<td>20</td>
<td>180</td>
<td>172</td>
<td>8 (4.4%)</td>
<td>Sporotrichoid, eczematous, scar, mucocutaneous, chancriform</td>
</tr>
<tr>
<td>North Punjab</td>
<td>256</td>
<td>45</td>
<td>34</td>
<td>335</td>
<td>321</td>
<td>14 (4.2%)</td>
<td>Sporotrichoid, zosteriform, DLE like, mycetomatous, paronychial, erysipelas, palmar, whitlow, lupoid</td>
</tr>
<tr>
<td>Northern Areas</td>
<td>8</td>
<td>2</td>
<td>2</td>
<td>12</td>
<td>11</td>
<td>1 (9.1%)</td>
<td>SCC like</td>
</tr>
<tr>
<td>Kashmir</td>
<td>102</td>
<td>64</td>
<td>25</td>
<td>191</td>
<td>173</td>
<td>18 (9.5%)</td>
<td>Lupoid, lid, chancriform, paronychial, psoriasiform, verrucous</td>
</tr>
<tr>
<td>Total</td>
<td>500</td>
<td>137</td>
<td>81</td>
<td>718</td>
<td>677 (94.3%)</td>
<td>41 (5.7%)</td>
<td></td>
</tr>
</tbody>
</table>

Figure 7: Pattern of atypical clinical presentations
Many of these atypical variants have already been described in literature, though very infrequently; but such a large spectrum with 17 different types of unusual morphologies was never described earlier. Moreover, the clinical spectrum was quite different compared to two other studies from the country. Most of the atypical morphologies were correctly diagnosed, keeping a high index of suspicion regarding the endemicity of the disease in the regions; but in a few cases, there was substantial delay, as in cases of chancriform, psoriasiform, zosteriform, erysipeloid, mycetoma and SCC-like lesions of CL. These were initially diagnosed as syphilitic chancre, psoriasis, herpes zoster, erysipelas, mycetoma and SCC; but it was after therapeutic unresponsiveness and/or further investigations that an ultimate diagnosis of atypical CL could be made. Response to treatment in these atypical cases encountered in the study was quite satisfactory.

Atypical clinical presentations of cutaneous leishmaniasis are increasingly seen in Pakistan and clustering of atypical cases in a geographically restricted region could possibly be due to a new parasite strain. To confirm this, further studies should be focused for species characterization in patients presenting with unusual morphology. It is further recommended that CL should be included in the differential diagnosis of common dermatological diseases like erysipelas, chronic eczema, herpes zoster, paronychia; and uncommon disorders like lupus vulgaris, squamous cell carcinoma, sporotrichosis, mycetoma and other deep mycoses.

REFERENCES