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A clinicoepidemiological study of polymorphic light eruption

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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.



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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

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.

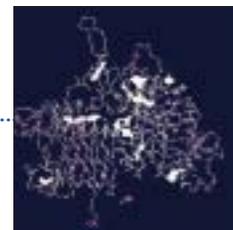


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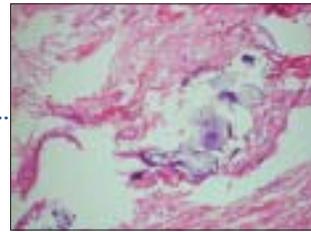
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Hereditary leiomyomatosis with renal cell carcinoma

Sir,

Hereditary leiomyomatosis with renal cell carcinoma (HLRCC) is a rare syndrome of cutaneous leiomyoma, uterine leiomyomatosis and renal cell carcinoma.^[1] It is an autosomal dominant disorder caused by mutations in the gene encoding the enzyme fumarate hydratase (FH). Launonen *et al.*,^[1] identified that the predisposition gene 'HLRCC' is mapped to 1q42-q44. It is often an unrecognized tumor syndrome. Its awareness is important because of its association with aggressive uterine fibroids and renal cell cancer. We report a case with multiple cutaneous leiomyomas and papillary renal cell cancer.

A 40-year-old male, a known case of hypertension controlled with one drug from two years presented with complaints of fullness in abdomen, anorexia, weakness, weight loss and intermittent fever since one month. Clinical examination revealed stable vitals and bilateral ballotable kidneys. He also had firm brown-colored non-painful, small skin nodules, six in number on the right arm, three on the left arm and four in the right thigh region. He was the eldest of the six siblings (two sons, four daughters). His father died of renal carcinoma at the age of 50 and his grandmother had similar skin nodules. The eldest of his four younger sisters had multiple uterine fibroids.

His investigations revealed hematocrit of 30 Vol%, serum creatinine-1.2 mg/dl, and serum calcium-10.4 mg/dl. Computerized tomography of abdomen showed bilateral enlarged kidneys with a mass lesion of 8 × 6 cm in each kidney [Figure 1]. Fine needle aspiration biopsy of a mass was suggestive of papillary renal carcinoma. Biopsies of skin nodules from the right arm and the right thigh showed cutaneous leiomyomatosis [Figure 2]. Assessment for the spread of malignancy was positive

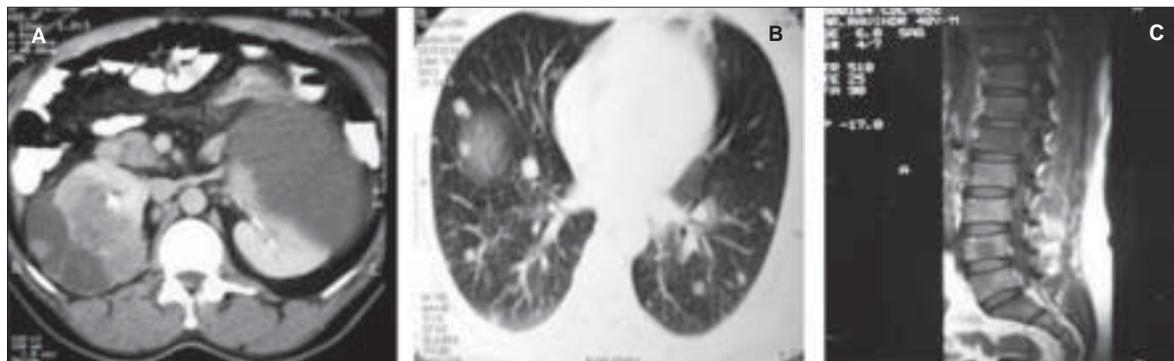


Figure 1: A. Computed tomography (CT) scan of abdomen showing bilateral renal masses B. CT scan of chest showing multiple pulmonary metastases C. Magnetic resonance imaging (MRI) scan showing metastasis to L4 vertebra

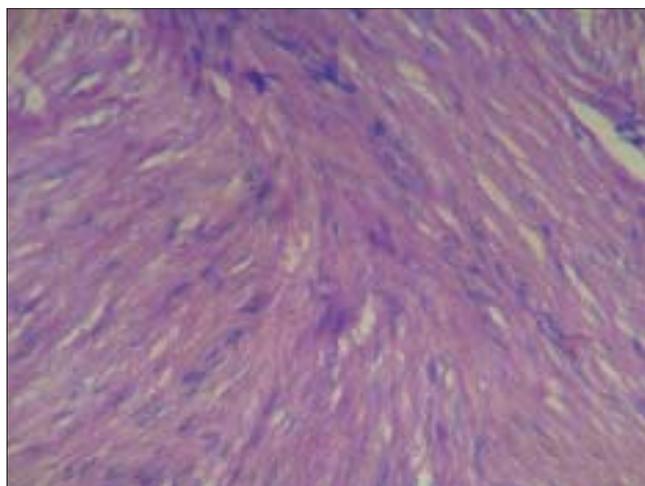


Figure 2: Proliferation of smooth muscle cells in fascicles suggestive of leiomyoma (H and E stain, X400)

with multiple metastases. Bilateral papillary renal malignancy at a young age, distant metastases at diagnosis, positive family history and cutaneous leiomyomatosis make it a case of hereditary leiomyomatosis with renal cell carcinoma.

Tomlinson *et al.*^[2] studied individuals with germline mutations in the FH gene. The activity of this enzyme of the TCA (tricarboxylic acid) cycle was reduced in lymphoblastoid cells from cases with leiomyomatosis. Chan *et al.*^[3] suggested that the 'HLRCC' is likely to be a tumor suppressor gene. Kiuru *et al.*^[4] concluded that this is a two-hit condition.

The majority (76%) of individuals with HLRCC present with a single or multiple cutaneous leiomyomas. Forty per cent of individuals with HLRCC have mild cutaneous manifestations with five or fewer lesions.^[5] Histopathologically, proliferation of interlacing bundles of smooth muscle fibers with centrally located long blunt-ended nuclei is observed. Classical

histological findings, family history of renal cell carcinoma and leiomyoma (both cutaneous and uterine) make the diagnosis of HLRCC likely in this case. Molecular diagnosis has not been made due to lack of facilities. This is the first case of the syndrome being reported from India. In view of the recent identification of this syndrome and lack of adequate clinical data, prognosis of these patients is not well defined although it appears to depend upon the staging of the tumor.

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