births; the affected patients generally do not survive beyond infancy.[1] In those patients who reach adolescence, acne is an important cutaneous manifestation. Most cases are sporadic, although autosomal dominant inheritance as well as germinal mosaicism have been reported.[2] The syndrome is caused by nucleotide alterations resulting in amino acid substitutions, leading to a mutation in the fibroblast growth factor receptor-2 (FGFR-2) gene mapped to chromosome 10q26.[3] Premature fusion of the bones is responsible for bony abnormalities; premature fusion of coronal sutures lead to brachycephaly. Other craniofacial abnormalities include a prominent forehead with skin wrinkling, a broad cranium, and a flat occiput. The shortened bony orbit leads to hypertelorism, proptosis and strabismus. Additional features include a short broad nose with a bulbous tip, micrognathia, and a cleft palate. Intracranial anomalies include megalocephaly, progressive hydrocephalus, hypoplastic white matter, and agenesis of the corpus callosum and limbic structures, leading to cognitive impairment. Cardiac abnormalities including atrial and ventricular septal defects and renal anomalies such as hydronephrosis occur in about 10% of these patients.

The cutaneous abnormalities reported are acne, hyperhidrosis and oculocutaneous albinism. Acne is now known to represent the dermatological hallmark of Apert's syndrome; it usually appears between the age of nine and 12 years. The acneiform lesions involve not only the face and upper trunk, but also affect the forearms, buttocks, and thighs. Oily skin is striking by the time of adolescence but the etiology of these acneiform lesions remains controversial. Increases in circulatory androgens or the density of sebaceous gland androgen receptors have not been documented. End-organ androgen metabolism defects, structural malformation of pilosebaceous apparatus and a role of FGFR-2 in regulating androgen sensitivity of the pilosebaceous unit have been suggested hypotheses for the acneiform lesions.[4] The skin, eyes and hair may show pigmentary dilution. Other occasional cutaneous abnormalities include interrupted eyebrows, forehead wrinkling, paronychial infections, skin dimpling over the knuckles, shoulders and elbows, as well as lateral plantar hyperkeratosis.

There is no definitive treatment for this syndrome. The prognosis varies from child to child and a multidisciplinary approach is recommended.

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Hydroxychloroquine versus chloroquine in polymorphic light eruption

Sir,

We read the article titled "Comparative study of efficacy and safety of hydroxychloroquine and chloroquine in polymorphic light eruption: A randomized, double-blind, multicentric study" with great interest. However, we wish to point out the following incongruities observed in this study.

The conclusion drawn about superior efficacy and tolerability of hydroxychloroquine over chloroquine is not corroborated by the findings of the study, where only a marginal difference was observed in the efficacy of the two drugs, and in fact, chloroquine fared better in the side effect profile.[1]

Negative serological tests for light eruption (LE) are pertinent and desirable for the diagnosis of polymorphic light eruption (PLE) patients as PLE and LE may often coexist and PLE precedes LE in a subset of the patients.[2,3]

The lack of ocular toxicity observed in the hydroxychloroquine treatment group after a short course of therapy is understandable as the same is expected after long-term use only. The important risk factors for ocular toxicity associated with hydroxychloroquine therapy are: excessive daily dosage, increasing cumulative dosage, the duration of treatment and the patient's age as well as coexistent renal or liver disease and concomitant retinal disease.[4,5]

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Spider nevi: A presenting feature of chronic liver disease

Sir,

The vascular spider, arterial spider or spider angioma is the most classical vascular lesion that is sometimes a presenting sign of chronic liver disease. Spider telangietasia occur in up to 15% of normal individuals and may also be seen in pregnant women. The main vessel of the spider is an arteriole represented by a red point from which numerous, small, twisted vessels radiate. Application of pressure on the central arteriole with the head of a pin or a match stick causes blanching of the whole lesion. We report here a case with profusion of spider nevi predating the onset of liver disease.

A 36 year-old, non-alcoholic man presented to the Dermatology OPD for evaluation of multiple, eight months old, erythematous, asymptomatic macules distributed mainly over the front of the chest, with a few on the upper arm and back [Figure 1]. The rash was earlier diagnosed as an allergic reaction and was treated with antihistamines by a general physician. Six months after the patient presented to us, he developed jaundice and was investigated and found to have chronic liver disease.

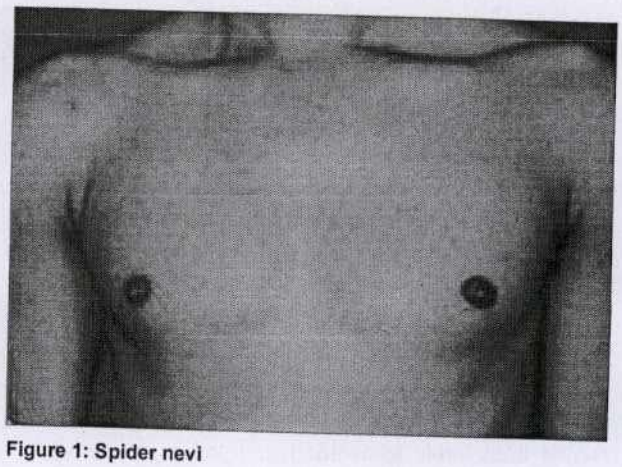

Figure 1: Spider nevi

Dermatological examination showed hundreds of erythematous macules 2–5 mm in size, mainly over the front of the chest, upper abdomen and a few on the upper arm and back [Figure 1]. The central body and the vessels radiating from it, could be seen clearly in a few lesions; mucous membranes were spared. There was no other dermatological evidence of chronic liver disease.

Hematological investigations revealed thrombocytopenia: 1,25,000 (Normal = 1,50,000–4,50,000); prothrombin time = 17 s (control: 13 s) and activated partial thromboplastin time = 36 s (control: 26 s) were prolonged. Viral markers for hepatitis were all absent. Total estrogen level was 85.7 pg/mL (normal < 56 pg/mL) and the total testosterone level was 218 mg/dL (normal: 245–1836 mg/dL). Tests for antinuclear antibody and alpha fetoprotein were also negative. Liver function tests showed elevated bilirubin (total 4.3 mg/dL) and elevated enzyme levels (ALT: 93 IU/L, ALP: 328 IU/L) (expand abbreviations). Blood sugar and renal parameters were normal. An ultrasound of the whole abdomen showed a shrunken liver with a coarse and nodular echo texture, suggestive of chronic liver disease, splenomegaly and ascites. A liver biopsy was not done as the patient was unwilling to have it done. Based on the above findings, a diagnosis of cryptogenic cirrhosis was made.

Spider angioma or nevus araenus is a dilatation of preexisting vessels under several circumstatnces.[1] Common causes of spider nevi are listed in Table 1.

Spider nevi are commonly distributed over the face, necklace area, forearms, hands and the upper part of chest, *i.e.,* mainly over the region drained by the superior vena cava.[2] Vascular spiders have been attributed to excessive levels of estrogen because estrogens cause blood vessels to enlarge and dilate.[3] Serum estradiol and total testosterone