

characterized by large, fixed, geographic and symmetrical fine scaly erythematous plaques over the knees, elbows, shoulder girdle, hands and feet. Onset of PSEK is usually in early childhood. The condition progresses over the next few years and then becomes stable with the morphology, colour and sites remaining constant over time. A positive family history may be elicited in only about 50 percent patients, rest of cases are due to spontaneous mutation of the loricrin gene. Loricrin is a major structural component of the cornified cell envelope, formed beneath the plasma membrane of stratified squamous epithelial cells during terminal differentiation. Association of PSEK with palmoplantar keratoderma, ataxia and syndactyly has been reported.¹

PSEK needs to be differentiated from erythrokeratoderma variabilis. Unlike in PSEK, the lesions in erythrokeratoderma variabilis fluctuate in their extent and configuration, involve the abdomen and thorax in addition to the extremities and show seasonal variation. Biopsy findings essentially are a psoriasiform hyperplasia with focal parakeratosis and well preserved granular layer. There is no suprapapillary thinning of the epidermis or Munro's microabscesses as seen in psoriasis. The other differentials that need to be considered are psoriasis and pityriasis rubra pilaris which can aptly be ruled out on the basis of histopathology.^{2,3}

Therapeutic options for PSEK include emollients, topical and oral retinoids. Our patient was treated with tazarotene and emollients with which her condition improved remarkably.

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An interesting case of idiopathic patterned hypomelanosis – a new entity?

Sir, the practice of dermatology sometimes presents us with riddles, implication of which goes beyond the boundaries of hitherto published wisdom. Here, we present a case of idiopathic patterned hypomelanotic condition which showed unique histopathological features.

A 20-year-old male student presented to our hospital with asymptomatic, non-scaly, hypomelanotic macules over chest, abdomen and back for six months. The lesions were insidious in onset, with no history of pre-existing dermatosis. Morphologically three different types of lesions were present: oval, arcuate and annular (**Figure1**). All lesions had well-defined borders with no sensory change, anhidrosis or alopecia. Cutaneous and cranial

nerve examination depicted no abnormality. Sensory and motor functions were absolutely normal. Patient did not have any history of kala-azar or previous nodular or ulcerated lesions. Careful family history could not elicit



Figure 1 Three different morphological patterns of macular hypomelanosis: oval, arcuate and annular.

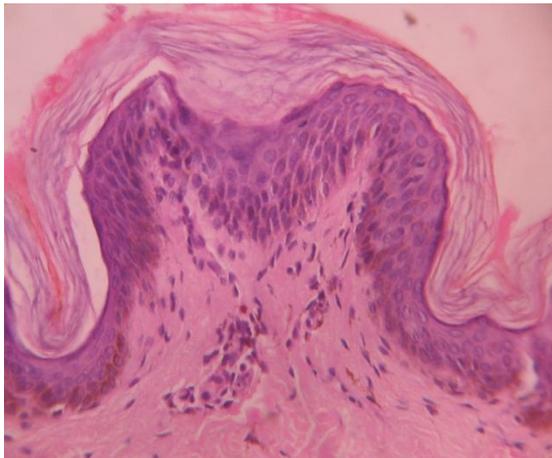


Figure 2 H&E staining showing hyperkeratosis, follicular plugging, irregular acanthosis, focal basal cell degeneration, pigment incontinence and patchy lymphocytic infiltrate in upper dermis (40x).

history of similar lesions in any family member or history of consanguineous marriages. Patients did not have any systemic features or atopy.

Routine investigations including blood counts and chest X rays were normal. VDRL was nonreactive and HIV ELISA was negative. Slit-skin smear failed to detect any acid-fast bacilli and skin scraping on KOH mount was negative for fungus. Biopsy for histopathology revealed hyperkeratosis, follicular plugging, irregular acanthosis, focal basal cell degeneration, pigment incontinence and patchy lymphocytic infiltrate in upper dermis (**Figure2**). Fite-Faracco staining was negative for acid fast bacilli.

Clinically we considered many differential diagnoses including Hansen disease, post-inflammatory hypopigmentation, post kala-azar dermal leishmaniasis, secondary syphilis, hypomelanosis of Ito, seborrheic dermatitis, tinea versicolor and early vitiligo. Absence of neurological changes clinically and absence of granuloma or acid-fast bacilli on histopathology ruled out a diagnosis of Hansen disease. A diagnosis of tinea versicolor or seborrheic dermatitis was not considered as the lesions were non-scaly and of unusual morphological pattern. Post-inflammatory hypopigmentation was unlikely due to absence of preexisting dermatosis or exposure to any chemical substance. Even when the patient was in our close follow up, he was developing *de novo* lesions. No history of sexual exposure, absent genital lesion and a non-reactive VDRL ruled out a possibility of secondary syphilis. Hypomelanosis of Ito has an earlier onset, a whorled pattern of pigmentation along Blaschko's line and characteristic absence of basal layer damage.¹⁻³ Vitiligo could easily be excluded by an absence of depigmentation, unusual distribution and morphology and an unsupportive histopathology. Idiopathic guttate hypomelanosis has smaller macular lesions in the photo-exposed area which shows decreased

basal melanin on histopathology.⁴ Cutaneous T cell lymphoma was not a possibility as both clinical picture and histopathology did not collaborate.

After extensive Pubmed search we could not find out any entity which closely resembled our case. Symmetric progressive leukopathy which is seen in young adults in Brazil and Japan is different from our case as in those cases there are punctuate leukoderma in bilaterally symmetrical distribution in arms and shins.⁵ Our case is unique because of its distinguishing arcuate and annular morphology, absence of symptoms and association with any preexisting condition. Moreover an unusual histopathological finding of basal cell degeneration along with very prominent pigment incontinence could not correlate with the clinically hypopigmented lesions. To the best of our knowledge we are reporting the first case of this unusual entity.

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Widespread gangrene: dermatological complication in cirrhosis

Cirrhosis is a serious disorder of liver and leads to wide range of complications. Cutaneous complications are uncommonly reported though these can be serious in nature. We report one such case of cirrhosis of liver who developed widespread gangrene of skin spontaneously. This complication was managed but the patient succumbed to his primary disease.

A 36-year-old male patient suffering from Wilson's disease with cirrhosis of liver and generalized anasarca developed painful, erythematous lesions over lower anterior abdominal wall and both thighs. The lesions deteriorated and within 4 days turned into wide gangrenous patches (**Figure 1** and **2**). There were no systemic features of toxemia like fever. Debridement was done and the raw areas were closed primarily and partly reconstructed with split thickness skin grafts. The excised specimens showed features of thrombosis of cutaneous vessels with infiltration of vessel wall with inflammatory cells. Postoperatively, the healing process was delayed but after 6 weeks all the wounds healed. The patient, however, succumbed to his cirrhotic liver disease after