

## Bier spots: An uncommon cause of mottled skin

Sir,

Bier spots represent a very uncommon pattern of vascular mottling often confused with pigmentary changes leading to unnecessary psychological distress. Only few cases have been reported since its original description by Bier in 1898. This 28-year-old male of Tibetan origin presented with numerous, light coloured spots over his forearms and hands in a mottled pattern [Figure 1]. As the spots were asymptomatic, he became aware of these only two years ago. He was treated for pityriasis versicolor previously at a peripheral center without any benefit. His family and medical history was unremarkable. He was a nonsmoker, and did not consume alcohol. There was no drug history. Examination revealed hypopigmented macular lesions having irregular borders with a background blanching erythema spread over the forearms and hands. Lesions were more marked over palms and became prominent when his arms were in a dependent position [Figure 2], and disappeared on limb elevation [Figure 3]. Clinical examination of the abdomen, cardiovascular, central nervous and pulmonary systems, and skin over the trunk and lower extremities was normal. Laboratory investigations including blood counts, erythrocyte sedimentation rate, serum biochemistry, antinuclear antibody, rheumatoid factor, coagulogram, and urinalysis revealed no abnormality. Histology of a forearm lesion showed no significant pathology except for dilated capillaries in the upper dermis [Figure 4].

Bier spots (syn: Angiospastic macules, physiologic anemic macules, exaggerated physiologic speckled mottling of skin) most commonly occur in young adults aged 20-40 years affecting females more often than males.<sup>[1,2]</sup> Although no racial or genetic preponderance is suggested, interestingly most reported cases have been of Chinese origin.<sup>[2]</sup> The marbled mottling pattern of skin consists of irregular shaped white macular areas against a blanching erythema or occasionally blue cyanotic background. The lesions involve upper extremities, the legs and the trunk in order of frequency. They are generally induced by venous congestion and can be elicited or made prominent when the affected limb is put in a dependent position or by placing a tourniquet around the limb. The lesions become imperceptible on diascopy (akin to nevus anemicus), with limb elevation or after the tourniquet is removed. Bier spots are idiopathic, self-limiting and except for counseling, require no treatment in most instances.

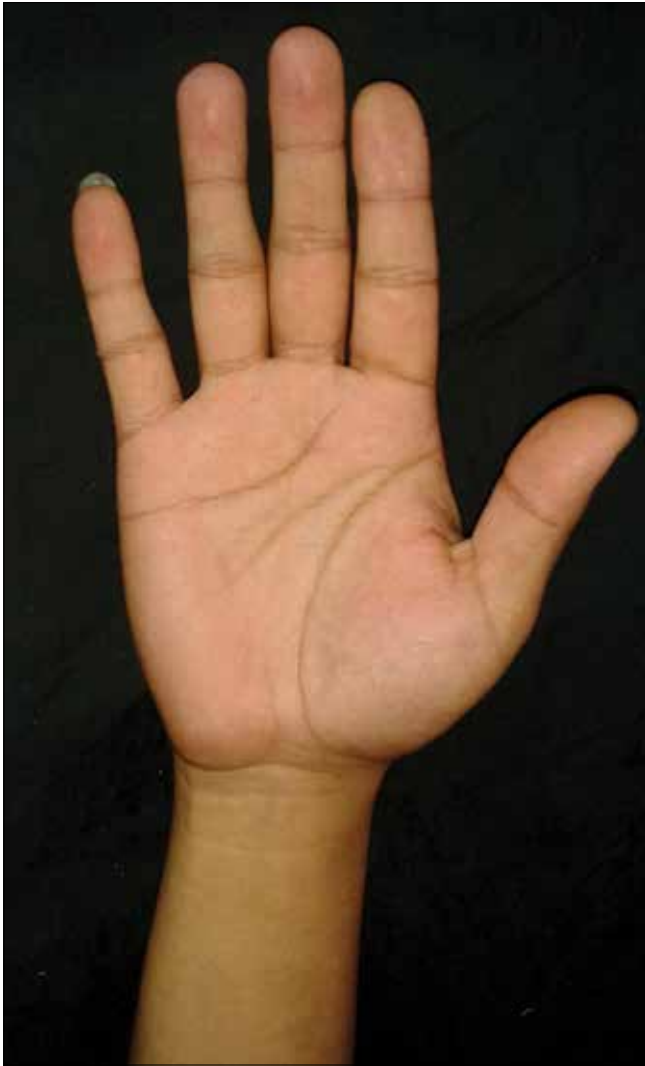


**Figure 1:** Innumerable hypopigmented macular lesions with faint, irregular borders against blanching erythema in a mottled pattern over forearms and palms. Mottling is more marked over the palms



**Figure 2:** The lesions over palms became prominent after the limb was in a prolonged dependent position

Nevertheless, it is imperative to differentiate the condition from hypopigmented macules of vitiligo, pityriasis alba, postinflammatory hypopigmentation, pityriasis versicolor and nevus anemicus/hypochromicus to avoid unnecessary treatment/distress. Their association with scleroderma renal crisis, mixed cryoglobulinemia, lymphoma, and pregnancy remains anecdotal.<sup>[2-5]</sup> Bier spots presenting together with insomnia and tachycardia in a rare case has been termed as Marshall-White syndrome.<sup>[6]</sup> The pathogenesis of this uncommon condition remains obscure. Peyrot *et al.*<sup>[3]</sup> considered these to be from a benign physiological response of small cutaneous vessels to venous hypertension or perhaps tissue hypoxia as a result of vasoconstriction of small vessels giving it a characteristic vascular pattern. However, laser Doppler velocimetry studies show that these red spots are caused by relative vasodilation with higher blood flow and vasoconstriction in the pale areas.<sup>[7]</sup> Although dilated capillaries seen on histology in our patient appear to corroborate these features, the overall histopathological patterns remain unmapped due to paucity of cases.



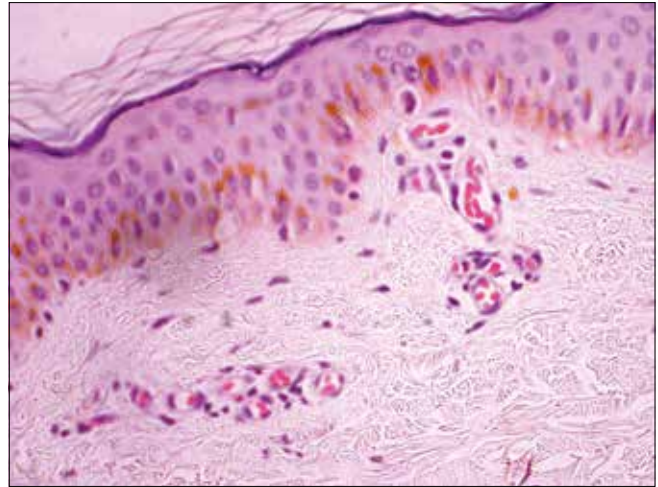
**Figure 3:** All lesions disappeared on limb elevation imparting a normal appearance to the skin

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**Figure 4:** Histopathology showing stratum corneum having characteristic normal basket weave appearance, normal epidermis and basal cell layer. Upper dermis shows multiple dilated capillaries with normal vessel walls and no vasculopathy/vasculitis, melanin incontinence or significant inflammatory cell infiltrate (H and E,  $\times 40$ )

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