CASES FOR DIAGNOSIS

Focal Hypopigmented Macules

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Clinical History

A 29-year-old man with a past history of appendectomy and morbid obesity, was seen for asymptomatic lesions that had appeared 7 to 8 years earlier on the hands and forearms.

Physical Examination

Multiple hypopigmented macules of 4 to 6 mm in diameter, with poorly defined borders and with surrounding erythema, were observed on the external aspect of both forearms and, more intensely, on the dorsum of the hands (Figures 1 and 2). The lesions disappeared on pressure under glass and on raising the limbs; they did not fluoresce under Wood light. Palmar hyperhidrosis was not observed.

Histopathology

The patient did not agree to a skin biopsy because of the asymptomatic nature of the lesions.

Additional Tests

Blood tests were normal, except for elevated aminotransferase, cholesterol, and triglyceride levels (aspartate aminotransferase, 85 U/L; alanine aminotransferase, 104 U/L; triglycerides, 511 mg/dL; total cholesterol, 260 mg/dL). Coagulation studies, antinuclear, extractable nuclear, and antiphospholipid antibodies, lupic anticoagulant, and complement levels were normal or negative.

What Was the Diagnosis?

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Diagnosis

Bier spots.

Clinical Course and Treatment

No treatment was given. The patient was informed of the benign and chronic nature of the condition.

Discussion

Bier spots were described by Bier in 1898 as alterations caused by an increase in the external pressure occurring in the upper limbs of some patients. The lesions disappear on normalization of the pressure. The spontaneous appearance of these lesions has been described as a response to venous stasis in a limb or to the application of local heat or cold; the term constitutive speckled vascular mottling of the skin is used in such cases. There are authors who favor differentiating between these 2 terms, as the latter cases show no pathologic associations.

There is no clear difference in the incidence and prevalence of this condition between men and women. The age at onset is variable, with cases reported between 17 and 75 years.

Clinically, the lesions present as irregular, hypopigmented macules, with a maximum diameter of less than 1 cm, and appear progressively. The majority of cases are revealed by application of a tourniquet around the arms, although their spontaneous appearance has also been reported. A key finding is that in many cases they disappear on raising the limb and on pressure under glass. Application of local heat or cold can exacerbate the lesions. The lesions do not fluoresce under Wood light, in contrast to other skin conditions.2

A number of pathophysiological mechanisms have been proposed to explain the pathogenesis of this disorder:

1. A physiologic increase in the resistance of the microcirculation, the release of vasoconstrictor substances in hypoxic tissue, or a failure of the arteriolar vasoconstrictor response.1-5
2. Increased sympathetic tone in the arterioles of the skin.
3. Increase blood viscosity in the microcirculation.

The histology of the Bier spots or anemic macules is unremarkable; there are no changes in the number or morphology of the blood vessels.

Early publications did not report any pathologic associations. However, since that time, there have been reports of cases associated with type II mixed cryoglobulinemia, coarctation of the aorta, and systemic scleroderma with renal involvement.

The differential diagnosis must include other disorders of pigmentation, particularly, nevus anemicus, vitiligo, pityriasis versicolor, achromic nevus, and achromic mycosis fungoides.

Few therapeutic options have been published in the literature; the most widely used agents have been the calcium channel blockers, which have shown no efficacy.

Conflicts of Interest

The authors declare no conflicts of interest.

References