

Diffuse Bier White Spots

Haitham Al-Qari, MD* Mersal Al-Arayedh, MD** Janel Gracey, MD***
Khalid Al-Ghaithi, MD**** Cohen Steven, MD***** Fatima Khamdan, MD*****

A seventeen-year-old Hispanic male presented with diffuse Bier white spots. The patient presented with asymptomatic diffuse hypopigmented anemic macules distributed mainly on the proximal extremities and torso for four months.

-
- * Consultant
 - ** Chief Resident
Department of Dermatology
Salmaniya Medical Complex
 - *** Senior Resident
 - **** Consultant
Department of Dermatology
Mount Sinai School of Medicine
New York
 - ***** Senior Professor
Department of Dermatology
Albert Einstein College of Medicine
New York, Bronx
 - ***** Senior Resident
Salmaniya Medical Complex
Department of Dermatology
Kingdom of Bahrain
Email: fatimakhamdan@gmail.com

Bier spots were first described in 1898, under experimental conditions when a protracted external compression of the brachial artery was performed which resulted in white spots on the forearm and hand¹. He considered these spots to be an exaggerated vasoconstrictive physiologic response caused by venous stasis-induced hypoxia. However, the Bier spots were replaced later by physiologic anemic macules which give a better reflection of the physiologic and clinical nature of these transient lesions. Gniadecki et al thought that Bier spots occurred because of the lack of vasoconstriction of ascending arterioles in response to venous filling². The prevalence of this skin disorder is uncertain, but not uncommon. This entity is rarely reported in the English literature. It is considered as a benign vascular anomaly in which vasoconstrictive response is induced by failure of the venoarteriolar reflex in dermal ascending arterioles in response to venous filling or secondary to venous stasis-associated hypoxia^{1,3}. This condition is characterized clinically by small hypopigmented anemic macules that are distributed on the upper and lower extremities and found to disappear on elevation of the extremity or when the normal venous return is restored. Bier spots might be associated with other systemic involvement such as scleroderma, cryoglobulinemia, renal crisis and aortic hypoplasia which makes it important to rule out any other systemic involvement⁴.

The aim of this presentation is to report a rare case of Bier spots in GCC region and stress its systemic involvement.

THE CASE

A seventeen-year-old Hispanic male was referred to the dermatology clinic in Salmaniya Medical Hospital in March 2005. He revealed a history of four-month of asymptomatic, white spots on his skin. He had been diagnosed with pulmonary TB and treated with isoniazid and rifampin and completed his treatment regimen two years ago. Currently, he is disease-free and on regular follow-up with the pulmonary clinic.

The patient stated that the rash becomes more obvious while standing and in cold temperature, and disappears while raising his arm above his head. The rash is characterized by diffuse ill-defined anemic hypopigmented macules lesions, 1 to 5 mm in diameter, distributed over the upper and lower extremities and torso, interspersed with blanchable erythematous macules giving rise to mottled appearance, see figure 1. The face, mucous membranes, palms and soles were spared. The rash completely disappears when the extremities were elevated and reappear upon dependency, see figure 2. There was no acrocyanosis.



Figure 1: Diffuse Bier White Spots Characterized by Diffuse Ill-Defined Anemic Hypopigmented Macules over the Upper Extremities



Figure 2: Diffuse Bier White Spots Appearance upon Dependency Posture

Systemic clinical examination was unremarkable. Laboratory investigations were normal; complete blood count, erythrocyte sedimentation rate, liver function tests, sugar, urea, creatinine, rheumatoid factor, coagulation profile, ANA, RNA, Anti-DNA, antiphospholipid autoantibodies, chest x-ray, C-reactive protein, hepatitis profile, serum proteins C and S and cryoglobulins were

negative or normal. Skin biopsy was not performed because of the benign course of this condition.

DISCUSSION

Bier spots are also known as physiologic anemic macules, nevus anemicus, speckled mottling of the limbs, exaggerated physiologic speckled mottling of the skin, and constitutive speckled vascular mottling of the skin and angiospastic macules⁴⁻⁷.

It is a benign vascular anomaly described by Bier in 1898. This rare entity is characterized by asymptomatic diffuse ill-defined anemic hypopigmented macules with adjacent erythematous skin that blanches with pressure, which leads to the disappearance of the hypopigmented macules. These spots could be elicited by putting the extremities in dependent posture. On the contrary, they disappear against gravity. The lesions gradually increase in number and definition.

Other pigmentary disorders could be confused with Bier spots such as vitiligo, pityriasis versicolor, achromic nevus and post inflammatory hypopigmentation^{3,5,8}. Speckled vascular mottling may resemble Bier spots which occur mainly in young age group, the mottling may occur spontaneously when venous stasis is produced by venous occlusion or dependency³. Laser Doppler studies showed a higher blood flux within the pale spots (vasoconstriction area) than in the surrounding skin³.

Bier concluded that the cause of the spots was due to micro vessels vasoconstriction induced by the lack of oxygen⁸⁻⁹. Vascular mottling is the result of venoconstriction that leads to pale, anemic macules while venodilation leads to erythematous areas. The other explanation for this exaggerated physiologic condition is due to venoarteriolar reflex failure on the dermal ascending arterioles in response to venous filling¹⁰.

Two associated conditions are reported in the literature, cryoglobulinemia and pregnancy. The first case is an elderly female patient with diffuse Bier white spots revealing the presence of mixed cryoglobulinemia. The observation emphasizes that hyperviscosity induced by cryoglobulinemia could play a pathogenic role in Bier spots¹. The second case of Bier spots started in the eighth month of a normal pregnancy and disappeared gradually after delivery. The condition is considered as exaggerated physiologic response of the micro vessels to venous hypertension in predisposed women¹¹. Other associated disorders include palmar hyperhidrosis, insomnia, tachycardia, scleroderma, renal crisis, aortic hypoplasia, varicosity, lichen planus, alopecia areata, and Peutz Jeghers syndrome.

Histopathological findings of the biopsy specimen were normal. There was no need to treat Bier spots as they are asymptomatic and possibly transient⁴.

CONCLUSION

Bier spot is a rare entity characterized by asymptomatic diffuse ill-defined anemic hypopigmented macules with adjacent erythematous skin that blanches with pressure; it is thought to be due to micro vessels vasoconstriction induced by lack of oxygen. It is a benign condition with normal histopathological findings and does not need treatment. However, it may be associated with other systemic and cutaneous disorders, which include scleroderma, cryoglobulinemia, renal crisis, aortic hypoplasia, varicosity, Peutz Jeghers

syndrome, lichen planus and alopecia areata. Therefore, it is crucial to be aware of systemic and cutaneous disease in patients with Bier spots.

Author Contribution: All authors share equal effort contribution towards (1) substantial contribution to conception and design, acquisition, analysis and interpretation of data; (2) drafting the article and revising it critically for important intellectual content; and (3) final approval of manuscript version to be published. Yes.

Potential Conflicts of Interest: None.

Competing Interest: None. **Sponsorship:** None.

Submission Date: 1 January 2015. **Acceptance Date:** 5 May 2015.

Ethical Approval: Approved by Dermatology Department, Salmaniya Medical Complex, Bahrain.

REFERENCES

1. Bessis D, Dereure O, Rivire S, et al. Diffuse Bier White Spots Revealing Cryoglobulinaemia. *Br J Dermatol* 2002; 146(5):921-2.
2. Gniadecki R, Gniadecka M. Constitutive Speckled Vascular Mottling of the Skin Resembling Bier White Spots: Lack of Venoarteriolar Reflex in Dermal Arterioles. *Bier White Spots: Lack of Venoarteriolar Reflex in Dermal Arterioles. Arch Dermatol* 2000; 136(5):674-5.
3. Peyrot I, Boulinguez S, Sparsa A, et al. Bier's White Spots Associated with Scleroderma Renal Crisis. *Clin Exp Dermatol* 2007; 32(2):165-7.
4. Khera P, English JC 3rd. Physiologic Anemic Macules. *Cutis* 2008; 81(6):477-8.
5. Cabanillas M, Suárez-Amor O, Loureiro M, et al. Bier's Spots in Association with Hypoplasia of the Aorta. *Dermatology* 2007; 215(2):166-7.
6. Graham RM, James MP. Exaggerated Physiologic Speckled Mottling of the Limbs. *Arch Dermatol* 1985; 121(3):415-7.
7. Baker C, Kelly R. Other Vascular Disorders. In: Bologna JL, Jorizzo JL, Rapini RP, et al, eds. *Dermatology*. 2nd ed. Spain: Elsevier Ltd; 2008:1624.
8. Wilkin JK, Martin H. Bier's Spots Reconsidered: A Tale of Two Spots, with Speculation on a Humerus Vein. *J Am Acad Dermatol* 1986; 14(3):411-9.
9. Collier JG, Dornhorst AC. Bier's Spots--Evidence that they are Mediated by an Intravascular Venoconstrictor Substance. *J Physiol* 1970; 209(1): Suppl:12P+.
10. Grosshans E. Multiple Anemic Macules or Bier's Spots? *Dermatology* 2001; 202(3):272.
11. Schoenlaub P, Dupré D, Redon JY, et al. Numerous and Large Bier's Spot Associated with Pregnancy. *Eur J Dermatol* 1999; 9(3):230-1.