Scholars Journal of Medical Case Reports **ISSN 2347-6559 (Online)**
Sch J Med Case Rep 2015; 3(2):92-94 **ISSN 2347-9507 (Print)**

(An International Publisher for Academic and Scientific Resources)
www.saspublishers.com DOI: 10.36347/sjmcr.2015.v03i02.007

Familial Bier's Spots: A Father And Two Sons

Betul Tas[1], Saadet Pilten[2], Ramazan Albayrak[3]
[1]Department of Dermatology, Bagcilar Research and Training Hospital, Istanbul/Turkey
[2]Department of Biochemistry, Bagcilar Research and Training Hospital, Istanbul/Turkey
[3]Department of Radiodiagnostic, Bagcilar Research and Training Hospital, Istanbul/Turkey

***Corresponding Author:**
Name: Betul Tas
Email: betulavc@yahoo.com

Abstract: Bier spots are small, hypopigmented macules that are usually found on the arms and legs of young adults. They are usually considered benign vascular anomaly and usually occur sporadically. No familial case has been reported so far. Herein, we report of three cases with Bier's spots in the same family consisting of a father and his 2 sons. To the best of our knowledge our cases are the first examples of familial Bier's spots in literature.
Keywords: Vascular skin diseases, physiological phenomenon, familial

INTRODUCTION

Bier's spots are exaggerated benign vascular speckled lesions characterized by transient, multiple irregular, whitish or hypopigmented macules with surrounding blanching erythema of the skin, and are also called as angiospastic or anemic macules which were originally described by Bier in 1898 [1,2]. They usually appear on the arms and legs of young adults [2] with female predominance [1]. The lesions usually occur sporadically and sometimes they can involve wider areas [1, 2].

CASE REPORT

A 54-year-old man (patient 1) and his 23 and 18-year-old-sons (patient 2 and 3) were admitted to our dermatology clinic for their asymptomatic white macules on their forearms and hands. The lesions of the father had been present for 30 years. Patient 2 and 3 were aware of their lesions for 7 and 3 years, respectively. Similar lesions were not seen in anyone else in the family (mother and two doughters) or previous generation. The father was a teacher and his sons were students, and they did not describe any precipitating factors such as cold, heat or exercise, and did not indicate that their lesions were related to any reason. They did not have any other diseases or take medicine. According to their histories, the spots were noticeable only when their arms were in down, and went back to normal when the arms were upright position in a few minutes. On dermatological examinations of each patient, there were multiple, irregular but well-defined white macules measuring between 3-7 mm in diameter on the forearms and back of the hands of patient 1, 2 and 3 when they lowered their arms below the heart level. (Fig. 1a, b, c, respectively). When the arms were upright and above the heart level, the spots disappeared in 7 to 10 minutes (Fig. 1d, e, f). Furthermore, in all patients, even though the arms were above the heart level, the white macular views were induced by inflating a sphygmomanometer cuff on their upper arm to a pressure exceeding systolic blood pressure for 3-5 minutes, and they disappeared by loosening the tournique. There were no lesions on their faces, bodies or lower extremities. No changes such as scaling, erythema, scarring, induration or atrophy were seen in the lesional areas. No temperature differences were detected between the lesional and non lesional areas. Other dermatological examinations of the patients, including the nails or mucosa were normal. The remaining physical examinations did not reveal signs of regional and systemic lymphadenopathy or hepatosplenomegaly, or venous and arterial hypertension. Neurological or cardiological examinations were normal. Reynaud's phenomenon was not detected in the patients. Laboratory examinations of the patients including a red blood cell count, hemoglobin, hematocrit, erythrocyte sedimentation rate, C reactive protein, the rates of peripheral blood venous and arterial gas markers (Ph, pO_2 and pCO_2), serum biochemistry and urinalysis were within the normal limits. No other pathology was detected in the vitamin B12, iron, ferritin, TSH, T4, LDH, anti-extractable nuclear antigen antibodies (anti-ds DNA, anti nRNP/sm complex, anti SS-A(Ro), anti SS-B(La), anti-Scl-70, anti Jo-1, anti Sm, anti histon) , protein S and C, and the cryoglobulin tests. Requested bilateral upper extremity arterial and venous Doppler ultrasonography (Dopp.USG), and jugular and carotis Dopp.USG did not reveal vascular abnormalities in the patients. Based on these typical histories, clinical and

laboratory findings, the lesions were diagnosed as " spontaneous Bier's spots".

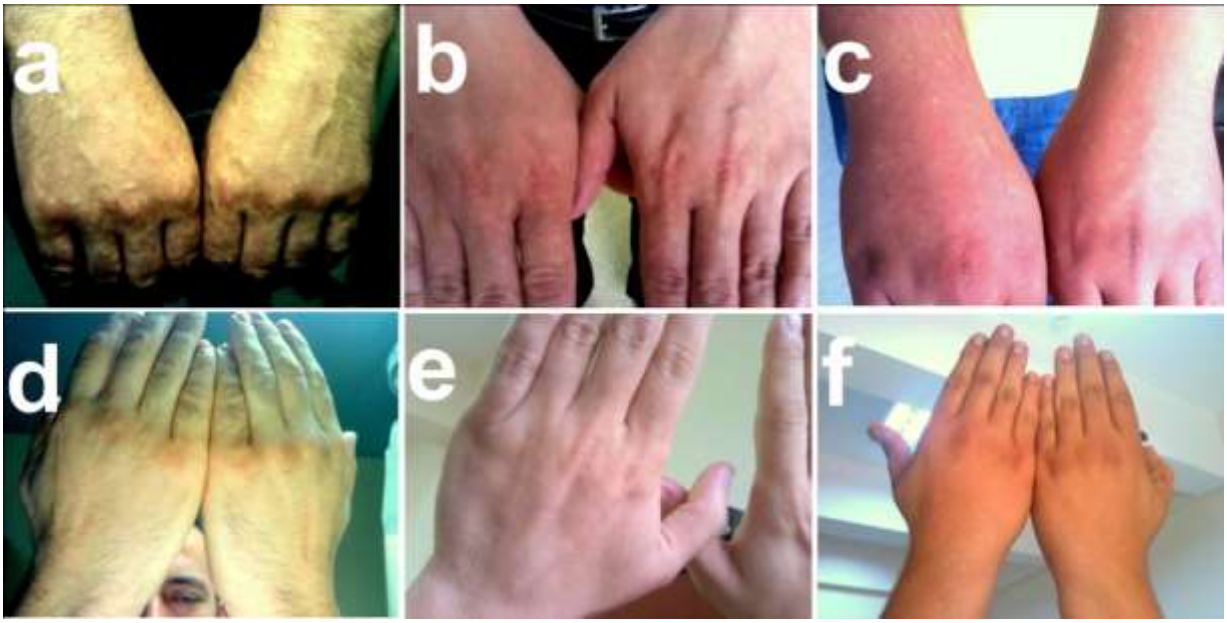

Fig. 1: Scattered pale, whitish and macular spots on the wrists and back of the hands of the patients when arms below the heart level (a, b, c,). The spots disappeared after the patients raised their arms (d, e, f,) respectively.

DISCUSSION

Bier's spots or angiospastic macules can be diagnosed easily with the clinical features. It has been stated that while the erythematous areas show venodilation, the pale areas show venoconstriction [1, 2]. They are created by extending down an affected limb under the heart level with mild mild venous stasis, and completely disappear when the extremity is raised with the venous pressure normalized [2]. Although they have usually been regarded as physiological but abnormal vasoconstrictive reactions of small vessels, they can be seen in a variety of diverse conditions such as pregnancy, cryoglobulinemia, venous stasis [2], scleroderma, renal crisis or lymphoma [1]. Because the lesions can appear during pregnancy and be resolved in postpartum period, these cases have been evaluated as exaggerated physiological responses of small vessels to venous hypertension [3]. Bessis *et al*. reported a case accompanying cryoglobulinemia, and they speculated that hyperviscosity related with cryoglobulinemi could play a role in that condition [4]. Cabanillas *et al*. stated that Bier's spots can be associated with aortic malformations, and an increased sympathetic tone in cutaneous arterioles may be responsible for the pathogenesis [5]. For our patients, due to the lack of any accompanying autoimmune, vascular, occlusive or thrombophilic pathologies in the laboratory tests, the patients were diagnosed with spontaneous Bier's spots. Additionally, due to the provocation of the lesions with the tourniquet test when the arms were upright position, we think that a reflex sympathetic activation caused by a hypoxic vasoconstriction in small arterioles is more likely to be responsible for the pathogenesis rather than a venous stasis or thrombotic condition. In differential diagnoses, other disorders in which similar white macules are seen such as vitiligo, postinflammatory hypopigmentation, pityriasis versicolour, pityriasis alba, nevus anemicus [1] and constitutive speckled vascular mottling should be considered [2, 6]. This last condition can be induced by cold temperatures [6]. Some previous attempts with nifedipine, cimetidine and loratadine have failed in treatment, and is unnecessary [1, 2]. However, even though Bier's spots are commonly regarded as benign conditions, and only lead to cosmetic or psychological complaints, sometimes they can accompany some vascular or thrombotic processes. Therefore, recognizing of this condition is important to avoid ineffective or redundant treatments in especially spontaneous ones. Only 17 spontaneous cases including ours have been reported so far. Previous patients in literature have been described as sporadic. To the best of our knowledge, our patients are the first familial and spontaneous cases in literature. Additionally, because the presented lesions were found only in the father and his two sons, but not in the mother and two doughters, we think that an X-linked recessive inheritance can be responsible for at least one part of pathogenesis for the patients 2 and 3.

REFERENCES

1. Liaw FY, Chiang CP; Bier spots. CMAJ, 2013; 185(7): E304.
2. Heller M; Diffuse Bier spots. Dermatol Online J., 2005; 11(4): 2.
3. Schoenlaub P1, Dupré D, Redon JY, Plantin P; Numerous and large Bier's spot associated with

pregnancy. Eur J Dermatol., 1999; 9(3): 230-231.

4. Bessis D, Dereure O, Rivire S, Ravi N, Le Quellec A, Guilhou JJ; Diffuse Bier white spots revealing cryoglobulinaemia. Br J Dermatol., 2002; 146(5): 921-922.
5. Cabanillas M, Suárez-Amor O, Loureiro M, Ginarte M, Toribio J; Bier's spots in association with hypoplasia of the aorta. Dermatology, 2007; 215(2): 166-167.
6. Gniadecki R, Gniadecka M; Constitutive speckled vascular mottling of the skin resembling Bier white spots: lack of venoarteriolar reflex in dermal arterioles. Arch Dermatol., 2000; 136(5): 674-675.