

A case of Bier spots with bilateral nevoid telangiectasia

Sir,

A 65-year-old woman complained of multiple hypochromatic macules on the lower limbs for four years. She was asymptomatic and denied a history of rheumatic, cardiovascular, hematologic or hepatic diseases, or any previous surgery. She neither had any family history of similar lesions. Her physical examination revealed numerous suborbicular white macules against a mild erythematous background on the extensor surface of the lower limbs [Figure 1a]. The macules were comparatively clear under Wood's light [Figure 1b] and were prominent in the vertical position of the limbs, while disappeared in the horizontal position [Figure 2a]. Punctate telangiectasias were seen at the center of white macules. These remained unchanged in the

horizontal position [Figure 2a] but faded significantly after the compression. The laboratory tests, including blood cell count, c-reactive protein, liver and renal function tests, antibodies to hepatitis B and C, testosterone, dehydroepiandrosterone, estradiol, luteinizing hormone, follicle-stimulating hormone, prolactin, sex hormone-binding globulin were normal. The ultrasonography of the arteries and veins of the lower limbs was also normal. Dermoscopy (Dermlite Foto II Pro 3Gen, contact, non-polarized, $\times 20$) revealed significant decrease or disappearance of vessels in the white macules while the surrounding skin revealed numerous reticular vessels [Figure 2b]. "Spider nevi"-like telangiectasias were also seen in the middle of white macules, which faded significantly after the compression [Figures 2b and c]. Based

on the typical clinical manifestations and dermoscopic features, a diagnosis of Bier spots with bilateral nevoid telangiectasia was established.

Bier spots are benign, physiologic, vascular anomaly, with less than 100 cases reported in the English literature. It was first described by August Bier in 1898. Bier spots are usually found in people aged 20–40 years. The clinical manifestations include asymptomatic, multiple, small, irregular, anemic macules on the extensor surface of limbs, predominantly on the upper extremities and rarely on the lower limbs and trunk. The incidence is almost equal in men and women. These are similar to other white macules, such as vitiligo, pityriasis versicolor, nevus anemicus, achromic nevus and post-inflammatory hypopigmentation. The pathogenesis of this condition remains obscure. Bier spots have been reported in association with various conditions such as scleroderma renal crisis, pregnancy, cryoglobulinemia, coarctation of the aorta, hypoplasia of the aorta, palmar hyperhidrosis, tachycardia, varicosity, lichen planus, alopecia areata, Peutz-Jeghers syndrome and heredity. Some scholars considered that the disease was caused by abnormal vasoconstriction of the ascending arterioles, leading to venoconstriction in pale areas and venodilation in erythematous areas.

Nevoid telangiectasia is characterized by superficial dermal telangiectasias. It is usually confined to the unilateral face, neck and upper chest. Nevoid telangiectasia is also rare and

about 100 cases have been reported till date. However, only 11 cases were bilateral, and all cases were acquired. Two types of nevoid telangiectasia have been described: punctate type and patchy type. Punctate type is the most common. The exact pathogenesis of nevoid telangiectasia is unknown. Some authors considered it to be connected with hyperestrogenic



Figure 1a: Punctate telangiectasias with a surrounding white halo against a mild erythema background on the lower limbs in the vertical position

Table 1: Bier spots with nevoid telangiectasia reported in the literature

Case	Year	Author	Sex	Age	Course (year)	Location	Laboratory and instrument examinations	Symptom	Underlying disease	Treatment
1	2011	Tan <i>et al.</i>	Female	23	6	Right hand	Liver function tests; antibodies to hepatitis A, B and C; and levels of estrogen, progesterone, testosterone and dehydroepiandrosterone were all normal	None	None	None
2	2015	Pinho <i>et al.</i>	Female	12	Since birth	Left hand and arm	Blood count, coagulation studies, cryoglobulins, kidney and liver function tests, proteinogram, immunoglobulins, sedimentation rate, autoantibodies, thyroid function, sex hormones, HIV and hepatitis and Doppler ultrasound of the upper limbs were normal	None	Not mentioned	None
3	2017	Lang <i>et al.</i>	Male	16	4	Left side of the chest, arm, and shoulder	Blood cell count; liver and renal function tests; coagulation parameters; testing for hepatitis A, B, and C; testosterone, dihydrotestosterone; androstenedione; dehydroepiandrosterone; dehydroepiandrosterone sulfate; sex hormone-binding globulin; free androgen index; progesterone; estradiol; prolactin; luteinizing hormone; follicle-stimulating hormone; cortisol; thyroid-stimulating hormone; and ultrasound of the liver, biliary tract and pancreas were normal	None	None	None

HIV: Human immunodeficiency virus

conditions, estrogen-sensitivity or increased estrogen/progesterone receptors. Bilateral nevoid telangiectasia is analogous to the unilateral form, but it is distributed on both sides of the body and follows the characteristic dermatomal distribution. In 2004, Tang *et al.* reported the first case of bilateral nevoid telangiectasia in a pregnant lady, who also had gastrointestinal involvement and which regressed completely after delivery.¹ Turan *et al.* also described the condition in a man, associated with gastrointestinal involvement but without any known etiology.² Jee *et al.* reported nine patients (none in puberty or pregnant), of which two had liver disease which may have altered the serum estrogen levels, and two were postmenopausal.³ The patient in the present case reported onset of lesions more than 10 years after menopause. Hence, estrogen might not be a trigger for bilateral nevoid telangiectasia or bier spots. Consequently, the pathogenesis remains to be further studied.

Bier spots accompanied by nevoid telangiectasia are extremely rare. Only 3 cases have been reported to date. In 2011, Tan and Zhu reported the first case of unilateral nevoid telangiectasia superimposed on bier spots and considered that this dichotomy of vascular phenomena was a new type

of vascular twin spotting.⁴ Subsequently, Pinho *et al.* and Lang *et al.* reported similar cases separately.^{5,6} None of these patients had hepatitis, sex hormone abnormality or other diseases [Table 1]. The cause of this condition was unknown. Bier spots with bilateral nevoid telangiectasia have not been reported yet. These two opposite kinds of vascular lesions occur in the same region simultaneously, as a special type of vascular twin spotting. This phenomenon occurs due to the local vasomotor dysfunction, but the intrinsic pathogenesis and triggering factors are still to be clarified.

The patient in the present case had no underlying disease or symptoms of discomfort, and she refused any treatment.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal the identity, but anonymity cannot be guaranteed.



Figure 1b: Punctate telangiectasias and white macules were more clear under the Wood's light (black arrow)

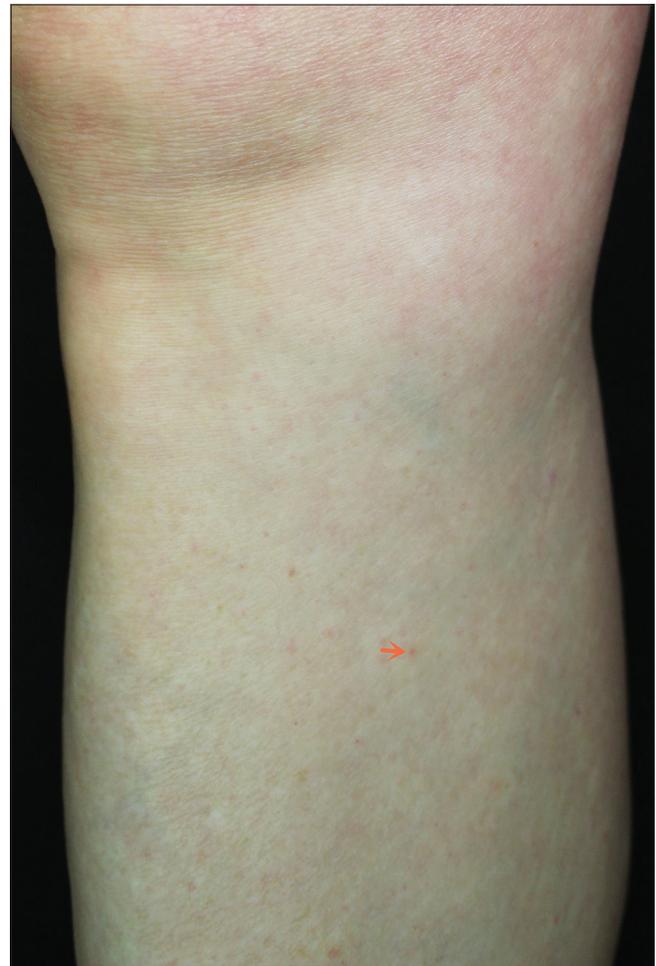


Figure 2a: White macules disappeared but the telangiectasias remained in the horizontal position

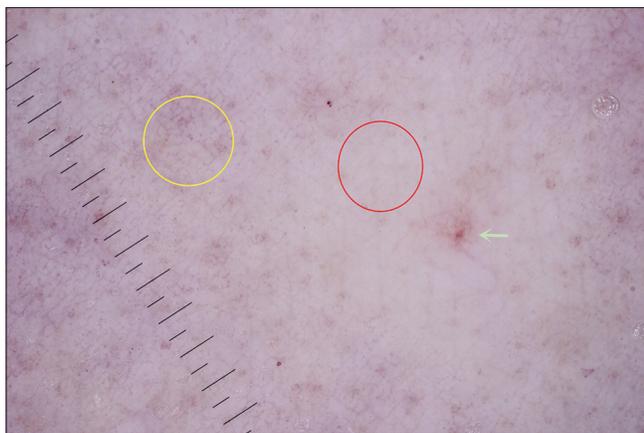


Figure 2b: Dermoscopy (Dermlite Foto II Pro 3Gen, nonpolarized, ×20) revealed that the vessels in the white macules significantly decreased or disappeared (red circle) compared with the numerous reticular vessels in the surrounding skin (yellow circle). “Spider nevi”-like telangiectasias were also seen at the center of the white macules (green arrow)

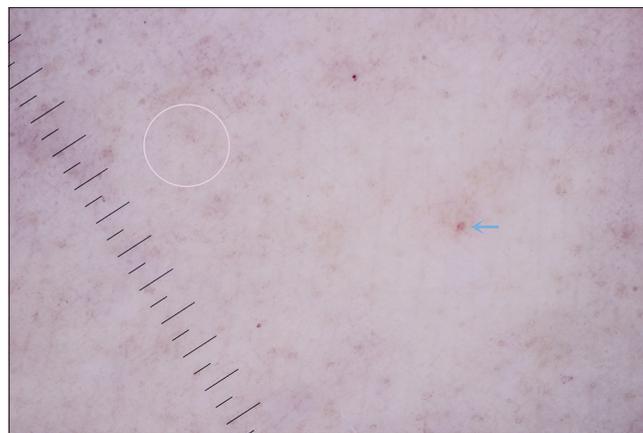


Figure 2c: Dermoscopy (Dermlite Foto II Pro 3Gen, nonpolarized, ×20) revealed that the “spider nevi”-like telangiectasias (blue arrow) and reticular vessels in the surrounding skin (pink circle) faded significantly after the compression

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Conflicts of interest

There are no conflicts of interest.

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