

BRIEF REPORT

Bier anaemic spots, cyanosis with urticaria-like eruption (BASCULE) syndrome on trunk and upper limbs

Abstract

Bier anaemic spots, cyanosis with urticaria-like eruption syndrome is a recently described benign vasomotor dermatosis that mainly affects the lower limbs. Histopathology shows multiple dilated dermal vessels with scattered eosinophils. We report a case on the trunk and upper limbs.

1 | CASE REPORT

An 8-year-old boy with no relevant medical history presented with asymptomatic red spots on the trunk and upper limbs. The lesions had appeared 3 years earlier. Physical examination showed multiple,

asymptomatic, urticaria-like, red papules on anemic macules with a surrounding erythrocyanotic background (Figure 1). Cutaneous lesions observed symmetrically on the trunk and upper limbs became more evident in the orthostatic position. The skin lesions relieved slightly within minutes of lying down or walking around. A pediatric cardiologist evaluated the patient. His resting heart rate was 90 beats per minute. Orthostatic postural tachycardia syndrome was excluded. Echocardiography; abdominal and upper limb arterial and venous ultrasonography; and laboratory tests including complete blood count and coagulation tests were normal. Antibody tests for

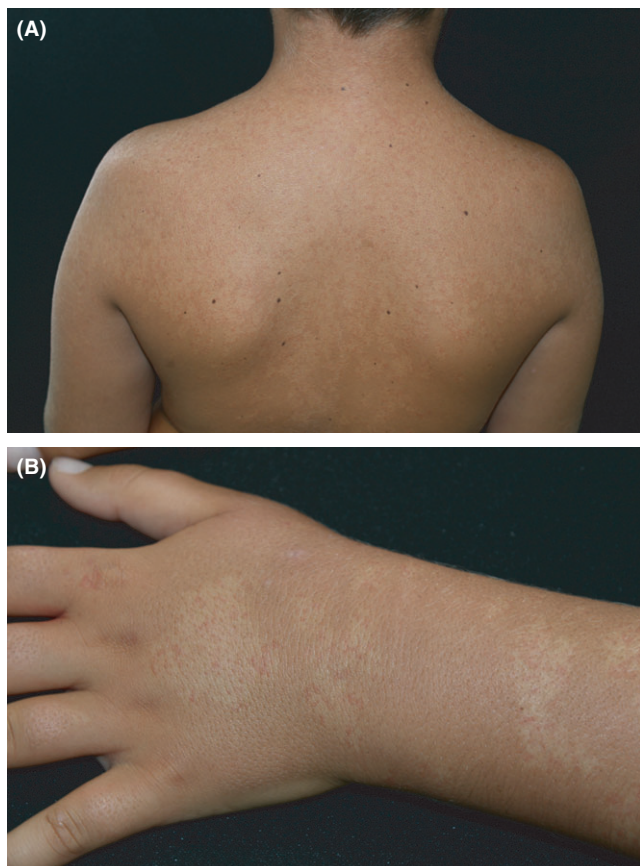


FIGURE 1 (A, B) Clinical images of Bier anaemic spots, cyanosis with urticaria-like eruption syndrome. Multiple urticaria-like red papules are super-imposed on anemic white macules with surrounding erythrocyanotic background

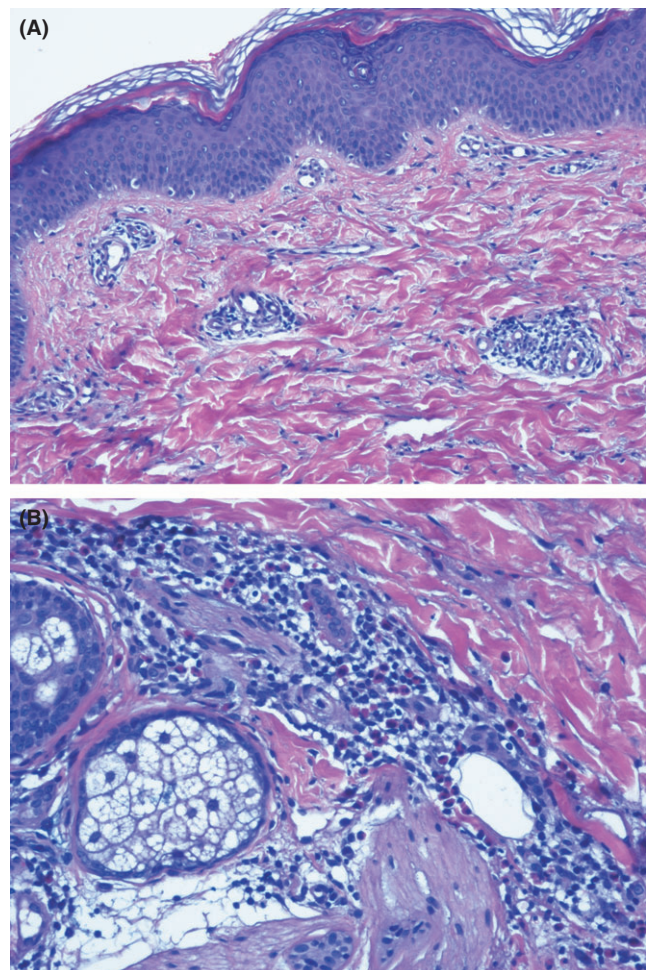


FIGURE 2 Histologic examination of Bier anaemic spots, cyanosis with urticaria-like eruption syndrome from a urticaria-like papule. Multiple dilated dermal vessels with a perivascular lymphocytic infiltrate and scattered eosinophils [hematoxylin and eosin, original magnifications: (A) $\times 100$ (B) $\times 200$]

TABLE 1 Reported cases of Bier anaemic spots, cyanosis with urticaria-like eruption syndrome

References	Sex	Age	Location	Age at onset	Symptoms	Outcome	Associated conditions	Laboratory test results
Bessis et al. ^{1,3}	Female	14 y	Lower limbs	2 y	Pruritus	Lesions remained unchanged	None	Normal
	Female	15 y	Lower limbs	10 mo	Pruritus and ankle edema	Lesions remained unchanged	None	Normal
	Male	19 y	Lower limbs	18 mo	Asymptomatic	Spontaneous resolution (2 y later)	None	Normal
	Male	3 mo	Upper and lower limbs	Recent	Tenderness and edema of left leg	Lesions remained unchanged	None	Normal
Martín et al. ²	Female	13 y	Lower limbs	2 y	Pruritus	Lesions remained unchanged	Exercise-related syncopal episodes	Normal
Current report	Male	8 y	Trunk and upper limbs	3 y	Asymptomatic	Lesions remained unchanged	None	Normal

autoimmune disorders were negative. Skin biopsy from an urticaria-like red papule showed multiple dilated dermal vessels with a perivascular lymphocytic infiltrate and scattered eosinophils (Figure 2). No vasculitis was observed. A diagnosis of Bier anaemic spots, cyanosis with urticaria-like eruption (BASCULE) syndrome was made based on clinical and histologic findings. Antihistamines resulted in no changes to the cutaneous lesions.

2 | DISCUSSION

Bier anaemic spots, cyanosis with urticaria-like eruption syndrome is a new dermatosis that Bessis and colleagues described in 2016 (Table 1)^{1,2} that is characterized by multiple urticaria-like red papules on anemic white macules with a surrounding cyanotic background. These variably pruritic lesions are mainly located on the lower limbs.^{1,3} BASCULE syndrome differs from Bier spots in that it also has urticaria-like papules on the anemic macules. Histopathology from Bier spots reveals multiple dilated capillaries but no significant inflammatory cell infiltrate.⁴ In contrast, histopathologic examination of BASCULE syndrome from an urticaria-like papule reveals multiple dilated dermal vessels with scattered eosinophils. No damage to the vessel walls of the upper dermis has been described.^{1,3} These histologic findings that Bessis and colleagues described were the same as those observed in our patient. The exaggerated vasoconstrictive response of arterioles to hypoxia induced by venous stasis appears to be the main pathologic mechanism of BASCULE syndrome,^{1,3} although venous stasis does not solely explain the involvement of the trunk and upper limbs. The presence of eosinophils is the main histologic difference between BASCULE syndrome and Bier spots. It is unknown whether eosinophils are the cause or outcome of this syndrome. We hypothesized that the eosinophilic infiltrate develops urticaria-like papules. Hypoxia has been shown to regulate eosinophil viability.^{5,6} It may be that the extension or intensity of the anemic macule-associated hypoxia triggers this eosinophilic activation. We highlight the observation of urticaria-like red papules and tissue eosinophils to distinguish it from "classic" Bier spots.

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Keywords

anaemic spots, Bier spots, genetic disease, genodermatoses, mechanisms, urticaria

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