Dermatology Practical & Conceptual

Familial BASCULE Syndrome in Two Siblings

Belén Romero-Jiménez[1], Catalina Axpe-Gil[1], Jorge Román-Sainz[1], Fernando Gruber-Velasco[1], Marcela Martínez-Pérez[1], Adrián Imbernón-Moya[1]

1 Department of Dermatology, Hospital Universitario Severo Ochoa, Leganés, Madrid, Spain

Citation: Romero-Jiménez B, Axpe-Gil C, Romàn-Sainz J, Gruber-Velasco F, Martìnez-Pérez M, Imbernón-Moya A. Familial BASCULE Syndrome In Two Siblings. *Dermatol Pract Concept*. 2023;13(4):e2023224. DOI: https://doi.org/10.5826/dpc.1304a224

Accepted: April 25, 2023; **Published:** October 2023

Funding: None.

Competing Interests: None.

Authorship: All authors have contributed significantly to this publication.

Corresponding Author: Belén Romero-Jiménez, Department of Dermatology, Hospital Universitario Severo Ochoa, Leganés, Madrid, Spain. ORCID https://orcid.org/0000-0002-1557-9284 E-Mail: belenromjim96@yahoo.es

Case Presentation

We present two siblings, a girl and a boy of 12 and 15 years respectively, who present asymptomatic multiple symmetrical hypopigmented macules with central telangiectasias surrounded by a background of erythrocyanosis on the dorsum of their hands since childhood (Figure 1, A and B). The lesions appeared after 1-2 minutes of keeping down their arms and disappeared when they were elevated. The girl also had similar lesions on her lower extremities which appeared after a few minutes in standing position and disappeared when sitting or lifting her legs. They had no history of cardiovascular disorders.

Laboratory tests with TSH and autoimmunity were performed without anomalies. Cardiological abnormalities were ruled out in both siblings. The diagnosis was familial BASCULE syndrome. None of the parents presented similar lesions in childhood or adolescence.

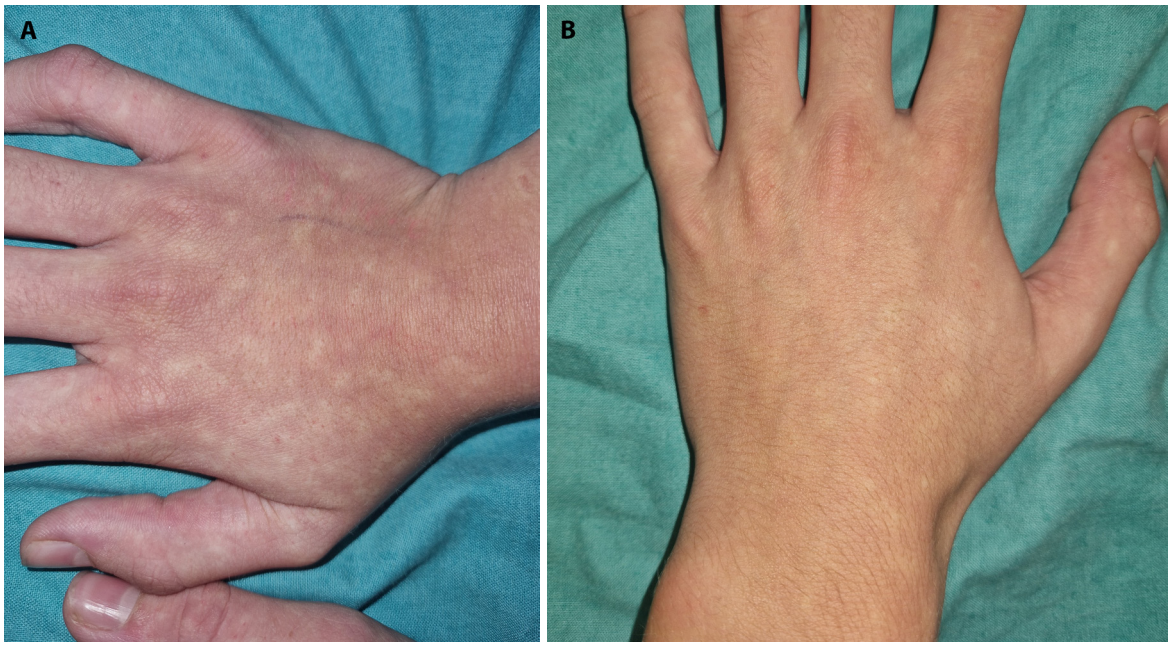

Figure 1. (**A**) Clinical image of a 12-year-old girl with hypopigmented macules with central telangiectasias in a background of erythrocyanosis on the dorsum of her hands. (B) Clinical image of a 15-year-old boy with hypopigmented macules on the dorsum of his hands.

Teaching Point

BASCULE syndrome (Bier anemic spots, cyanosis and urticaria-like eruption) is a benign vasomotor dermatosis that typically affects the lower extremities of children and adolescents [1,2]. No familial cases have been reported. The diagnosis is mostly clinical and biopsy is usually not necessary [2]. It is recommended to perform a blood test with TSH and autoimmunity and refer to cardiology to rule out underlying heart disease. Treatment with antihistamines is not effective. The prognosis is favorable with resolution of the lesions with pubertal growth [1,2].

Herein, we present a BASCULE syndrome in two members of the same family, which may suggest the possibility of having a hereditary component.

References

1. El Nemnom P, Lauwerys B, Marot L, Tennstedt D, Dekeuleneer V. Bier anemic spots, cyanosis, and urticaria-like eruption (BASCULE) syndrome: Report of two new cases and literature review. *Pediatr Dermatol*. 2020;37(5):864-867. DOI: 10.1111/pde.14213. PMID: 32648305.
2. Barbé J, Escobar G, Bursztejn AC. Bier anemic spots, cyanosis with urticaria-like eruption (BASCULE) syndrome: a simple benign pathology, but probably underdiagnosed. *Int J Dermatol*. 2021;60(9):1163-1164. DOI: 10.1111/ijd.15452. PMID: 33611787.