Clinical Report
Open Access
CrossMark

"Deep Purple": a cutaneous presentation with a spectrum of diagnostic possibilities: what lies beneath the skin?

Volume 15 Issue 4 - 2023

Abdala Brian Marcelo, Ringer Ariana, Ruffino Juan Pablo, Abdala, Marcelo
Rheumatology Unit, Hospital Provincial del Centenario, Rosario, Santa Fe, Argentina

Correspondence: Abdala Brian Marcelo, Rheumatology Unit, Hospital Provincial del Centenario, Rosario, Santa Fe, Argentina, Email bmabdal@gmail.com

Received: July 25, 2023 | **Published:** August 11, 2023

Clinical report

Woman, 34 years old, with a history of aplastic anemia, no prior pregnancies, presents with a 7-day history of painful purpuric lesions on the cheeks, eyebrows, ears, trunk, and limbs. Denied illicit substance use. Physical examination reveals multiple palpable erythematous-violaceous plaques with a peripheral erythematous halo and central hemorrhagic bullae.

Laboratory findings show Hct 24%, reticulocytes 3.6%, leukocytes 5400/mm3, platelets 61,000/mm3, ESR 132 mm/h, CRP 48 mg/L, normal renal and hepatic function, normal coagulation times, LDH 1900 IU/L, hypoalbuminemia, and polyclonal increase of globulins. Urinary sediment is normal. Serology for HIV, HCV, HBV, and VDRL is negative. ANA, RF, ACL, anti-B2GP1, ANCA, and cryoglobulins are negative; C3 and C4 levels are normal. Infections were ruled out during the diagnostic process. Due to initial suspicion of autoimmune vasculitic and/or thrombotic pathology, the patient received pulses of intravenous methylprednisolone after a skin biopsy.

Skin biopsy

Tissue necrosis, fibrosis, vascular neoformation, and thrombosis in arterioles and venules were observed, but no evidence of vasculitis. Deposits of C3 and fibrinogen (++) were found in capillary walls of the papillary dermis. Flow cytometry in peripheral blood revealed clones of erythrocytes and leukocytes with absent expression of CD59 and FLAER/CD157, 22.4% and 61.4%, respectively. With this result, a diagnosis of paroxysmal nocturnal hemoglobinuria (PNH) was established, and anticoagulation with enoxaparin and specific treatment with eculizumab were initiated, resulting in subsequent resolution of clinical symptoms.

Paroxysmal nocturnal hemoglobinuria (PNH) is a rare acquired disorder characterized by abnormal destruction of red blood cells due to deficiency in glycosylphosphatidylinositol (GPI), leading to complement system activation. Clinically, it presents with episodes of intravascular hemolysis, particularly during the night, resulting in the presence of hemoglobinuria. The diagnosis of PNH relies on the detection of GPI-deficient blood cell clones, such as erythrocytes and leukocytes, using flow cytometry and other techniques.[1–5]

In addition to paroxysmal nocturnal hemoglobinuria (PNH), the clinical picture may be associated with various conditions that present similar features. These differential diagnoses include vasculitis, thrombotic microangiopathies, systemic lupus erythematosus, drug-induced skin reactions, paroxysmal cold hemoglobinuria, infections, cryoglobulinemia, and hematological malignancies. A thorough evaluation of the patient's medical history, laboratory tests, and imaging studies is crucial for an accurate diagnosis, and a multidisciplinary approach may be necessary for appropriate management (Figure 1).[6–12]

Cutaneous lesions pose diagnostic challenges in rheumatology. PNH presents with signs and symptoms that can be shared with other disease groups, such as vasculitis. Differential diagnoses in rheumatology are broad, and we should not limit ourselves solely to our area, requiring a high index of suspicion and critical thinking to address other clinical areas.

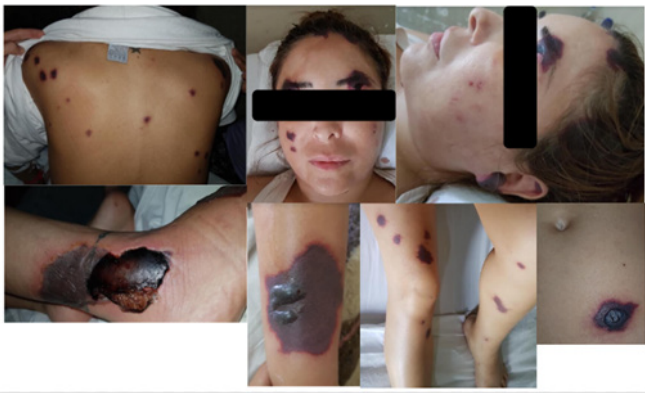

Figure 1 Skin lesions

Acknowledgments

None.

Conflicts of interest

The author declares no conflicts of interest.

References

1. Borowitz MJ, Craig FE, DiGiuseppe JA, et al. Guidelines for the diagnosis and monitoring of paroxysmal nocturnal hemoglobinuria and related disorders by flow cytometry. *Cytom Part B - Clin Cytom*. 2010;78(4):211–230.

2. Schrezenmeier H, Muus P, Socié G, et al. Baseline characteristics and disease burden in patients in the international paroxysmal nocturnal hemoglobinuria registry. *Haematologica*. 2014;99(5):922–929.
3. Höchsmann B, Murakami Y, Osato M, et al. Complement and inflammasome overactivation mediates paroxysmal nocturnal hemoglobinuria with autoinflammation. *J Clin Invest*. 2019;129(12):5123–5136.
4. Hill A, Kelly RJ, Hillmen P. Thrombosis in paroxysmal nocturnal hemoglobinuria. *Blood*. 2013;121(25):4985–4996.
5. Brodsky RA. Paroxysmal nocturnal hemoglobinuria. *Blood.* 2014;124(18):2804–2811.
6. Sobrino Grande C, Villalobos Sánchez L, Valero Expósito M, et al. Síndrome antifosfolípido. *Med*. 2017;12(27):1551–1559.
7. Moyo VM, Mukhina GL, Garrett ES, et al. Natural history of paroxysmal nocturnal haemoglobinuria using modern diagnostic assays. *Br J Haematol*. 2004;126(1):133–138.
8. Araten DJ, Thaler HT, Luzzatto L. High incidence of thrombosis in African-American and Latin-American patients with Paroxysmal Nocturnal Haemoglobinuria. *Thromb Haemost*. 2005;93(1):88–91.
9. Gallo D, Del Carmen M, Sánchez Luceros D, et al. Sociedad Argentina De Hematología Trombofilias Coordinadores. 2017.
10. Zhao H, Shattil S. Cutaneous thrombosis in PNH. *Blood*. 2013;122(19):3249.
11. Frumholtz L, Laurent-Roussel S, Lipsker D, et al. Cutaneous Vasculitis: Review on Diagnosis and Clinicopathologic Correlations. *Clin Rev Allergy Immunol*. 2021;61(2):181–193.
12. Brodsky RA. How I treat paroxysmal nocturnal hemoglobinuria. *Blood*. 2021;137(10):1304–1309.

Citation: Marcelo AB, Ariana R, Pablo RJ, et al. "Deep Purple": a cutaneous presentation with a spectrum of diagnostic possibilities: what lies beneath the skin?. *MOJ Orthop Rheumatol.* 2023;15(4):145–146. DOI: 10.15406/mojor.2023.15.00635