Annals of Clinical Case Reports

Case Study
Published: 30 Mar, 2020

Purpura Pigmentosa Linearis Unilateralis Chronic - A Case Report with a Review of the Literature

*Vessel Kantardjiev[1], Kristina Petkova[1] and Valentina Broshtilova[1,2]**

[1]Department of Dermatology and Venereology, Military Medical Academy, Bulgaria

[2]Department of Infectious diseases, Parasitology and Dermatovenereology, Medical University "Prof. Dr Paraskev Stoyanov", Bulgaria

Abstract

Purpura pigmentosa linearis unilateralis chronica is a rare subtype of pigmented purpuric dermatoses, characterized by linear or segmental distribution of purpuric macules, predominantly on the lower extremities. It is usually not associated with underlying systemic disease and follows a benign course with spontaneous regression. No specific treatment modalities are needed. Herein, an anecdotal case of purpura pigmentosa linearis unilateralis chronica is presented in the highlights of a scientific literature update.

Keywords: Purpura pigmentosa; Segmental distribution; Spontaneous regression

Introduction

Purpura Pigmentosa Linearis Unilateralis Chronica (PPLUC) is a rare capillaritis [1], a subtype of Pigmented Purpuric Dermatoses (PPD). It is characterized clinically by linear or segmental distribution of pigmented purpuric macules located predominantly on the lower extremities. First, in 1992 Riordan et al. [2] described four cases of distinctive type of purpuric eruption, occurring in a linear and pseudo-dermatomal distribution in young males.

Case Presentation

A 23-year-old male was admitted to our department with a eight-month history of an asymptomatic rash, starting from the posterior left lower leg and ascending towards his left buttock. The patient denied any accompanying disease. The clinical presentation revealed well-defined purpuric patches, partly confluent into circumscribed ecchymoses (Figure 1), following the lines of Blaschko in a linear S1/S2 distribution. Dermoscopy showed vascular lacunae, brown dots, and linear vessels (Figure 2). A punch biopsy from the left calf revealed mild perivascular lympho-plasmocytic inflammatory infiltration with erythrocyte extravasation in the papillary dermis (Figure 3a,3b). The routine blood count and coagulation tests were normal, thus excluding thrombocytopenic purpura.

Diagnosis of pigmented linear unilateral purpura was concluded based on clinical presentation and histological findings. The patient was advised to sustain low activity lifestyle. Vasoprotective regimen was additionally prescribed. At the second month follow-up visit no objective changes were registered.

Discussion

Purpura pigmentosa chronica is a chronic inflammation of the capillaries, usually affecting areas of hypostasis. Several clinical subtypes have been commonly described: Purpura annularis telangiectodes or Majocchi's disease, pigmented purpuric lichenoid dermatosis of Gougerot and Blum, eczematid-like purpura of Doucas and Kapetanakis, Schamberg's disease or purpura progressive, and lichen aureus [3,4]. Rare variants such as purpura granulomatosis, transitory pigmented purpuric eruption, purpura pigmentosa linearis unilateralis chronica have also been reported. All subtypes are histologically identical, manifesting capillaritis [5,6].

Unilateral Linear Capillaritis (ULC) is a benign condition with a tendency to regress spontaneously over several months to a few years [7]. In 2017 Elshimy et al. [8] critically reviewed the fourteen ULC cases reported up to that time worldwide and found no sex predominance (male:female = 8:6), mid-adulthood average onset age (21 years), predominant location of the lower extremities, and spontaneous regression between 3 to 36 months. As patients were largely asymptomatic, no active treatment was required.

OPEN ACCESS

Correspondence:
Valentina Broshtilova, Department of Dermatology and Venereology, Military Medical Academy, Sofia, Bulgaria, E-mail: broshtolova@mail.bg

Received Date: *09 Mar 2020*
Accepted Date: *26 Mar 2020*
Published Date: *30 Mar 2020*

Citation:
Kantardjiev V, Petkova K, Broshtilova V. Purpura Pigmentosa Linearis Unilateralis Chronic - A Case Report with a Review of the Literature. Ann Clin Case Rep. 2020; 5: 1816.

ISSN: 2474-1655

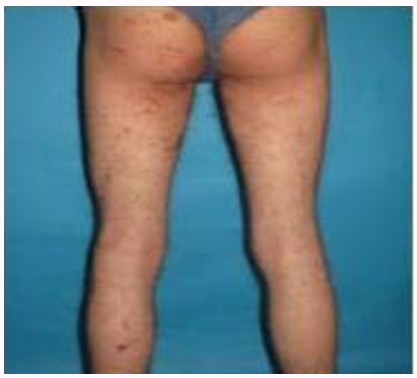

Figure 1: Purpuric macules linearly distributed on S1/S2 dermatome.

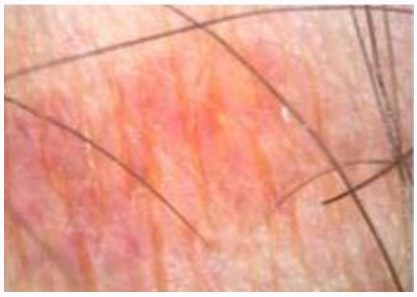

Figure 2: Vascular lacunae, brown dots, and linear vessels on dermoscopy.

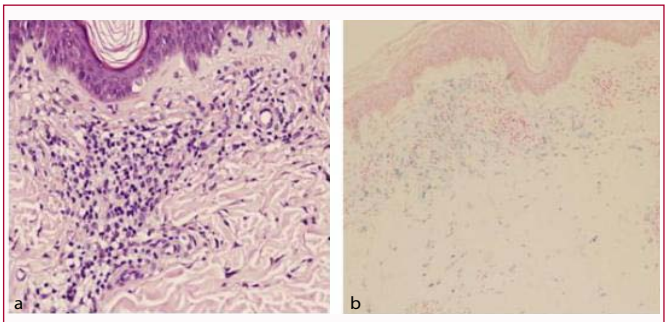

Figure 3: a) Acanthosis, focal interface, intense perivascular lympho-plasmocytic inflammatory infiltration in the papillary dermis (H&E, x200); b) Erythrocyte extravasation in the papillary dermis (Fe stain, x100).

Suggested causative factors include abnormalities in the cellular immune system, bacterial infections, capillary fragility, drug induction, and phlebostasis [8]. The unilateral or linear distribution of ULC along Blaschko lines might also be explained by an underlying genetic abnormality, such as mosaicism [10].

Three different pathogenetic mechanisms have been proposed: enhanced delayed hypersensitivity immune response causing dermal infiltrate of lymphocytes, macrophages and Langerhans cells; disturbed humoral immunity; and increased capillary fragility [11,12].

The main differential diagnosis includes thrombocytopenic purpura - a rare blood disorder, characterized by purpuric patches, fatigue, anemia, and neurologic disturbances. Often severe bleeding, fever and dark urine from hemoglobinuria can also be seen [13]. The disease is easily recognized by typical blood count findings.

The rather inconspicuous ULC course, lack of subjective symptoms, and the tendency of spontaneous regression evokes no active treatment [14]. However, few therapeutic modalities have been tried. Pentoxifylline proved beneficial, especially in patients with more extensive lesions [15], psoralen-PUVA treatment has been reported to improve the condition [5]. We preferred vasoprotective drugs, which unfortunately showed no effective impact on the 20th month follow-up visit.

Conclusion

ULC is a very peculiar clinical form of chronic pigmented purpura, usually not associated with underlying systemic condition. It follows a benign course with spontaneous regression, occurring up to 36 months upon onset. Hence, different treatment modalities should be accurately selected since they may expose patients to greater risk than benefit.

References

1. Pock L, Capkova S. Segmental Pigmented Purpura. Paediatr Dermatol. 2002;19(6):517-9.
2. Riordan CA, Darley C, Markey AC, Murphy G, Wilkinson JD. Unilateral linear capillaritis. Clin Exp Dermatol. 1992;17(3):182-5.
3. Zalaudek I, Ferrara G, Brongo S, Giorgio CM, Argenziano G. [Atypical clinical presentation of pigmented purpuric dermatosis]. J Disch Dermatol Ges. 2006;4(2):138-40.
4. Wong WR, Kuo TT, Chen MJ, Chan HL. Granulomatous variant of chronic pigmented purpuric dermatosis: Report of two cases. Br J Dermatol. 2001;145(1):162-4.
5. Sardana K, Sarkar R, Sehgal VN. Pigmented purpuric dermatoses: An overview. Int J Dermatol. 2004;43(7):482-8.
6. Ma HJ, Zhao G, Liu W, Dang YP, Li DG. Unilateral linear capillaritis: Two unusual Chinese cases. Eur J Dermatol. 2007;17(2):160-3.
7. Hersh CS, Shwayder TA. Unilateral progressive pigmentary purpura (Schamberg's disease) in a 15-year-old boy. J Am Acad Dermatol. 1991;24(4):651.
8. Elshimy N, Walker B. Unilateral Linear Capillaritis: A case and summary of the literature 25 years on from its original description. Clin Case Rep. 2017;3(4):1-3.
9. Mar A, Fergin P, Hogan P. Unilateral pigmented purpuric eruption. Australas J Dermatol. 1999;40(4):211-4.
10. Higgins EM, Cox NH. A case of quadrantic capillaropathy. Dermatologica. 1990;180(2):93-5.
11. Abdullah L, Abbas O. Answer: Can you identify this condition? Canad Famil Physic. 2011;57:564.
12. Taketuchi Y, Chinen T, Ichikawa Y, Ito M. Two cases of unilateral pigmented purpuric dermatosis. J Dermatol. 2001;28(9):493-8.
13. Ozkaya DB, Emiroglu N, Su O, Cengiz FP. Dermatoscopic findings of pigmented purpuric dermatosis. An Bras Dermatol. 2016;91(5):584-7.
14. Zainal A, Salama A, Alweis R. Immune Thrombocytopenic Purpura. J Commun Hosp Intern Med Prospect. 2019;9(1):59-61.
15. Mun JH, Jwa MS, Song M, Kim HS, Ko HC, Kim MB. Extensive Pigmented Purpuric Dermatosis Successfully Treated with Pentoxifylline. Annal Dermatol. 2012;24(3):363-5.