Our Dermatology Online

Case Report

From macular lymphocytic arteritis to cutaneous polyarteritis nodosa: A spectrum of the same disease? Two cases of the same clinical presentation yet different histopathological findings

Judith Monserrat Corona-Herrera[1], Luis Asdruval Zepeda-Gutiérrez[2], Orlando Emmanuel Falcon-Antonio[3], Alejandro Barrera-Godínez[1], Silvia Méndez Flores[1], Judith Guadalupe Domínguez-Cherit[1]

[1]Department of Dermatology, National Institute of Medical Sciences and Nutrition Salvador Zubirán, Ciudad de Mexico, México, [2]Department of Internal Medicine, National Institute of Medical Sciences and Nutrition Salvador Zubirán, Ciudad de Mexico, México, [3]Department of Pathology, National Institute of Medical Sciences and Nutrition Salvador Zubirán, Ciudad de Mexico, México

Corresponding author: Judith Monserrat Corona-Herrera, MD, E-mail: jch.fesi.mc@gmail.com

ABSTRACT

Macular lymphocytic arteritis (MLA) is a type of indolent cutaneous vasculitis that affects small to medium-sized blood vessels. Histopathologically, it is characterized by a predominantly lymphocytic leukocytic infiltrate (unlike polyarteritis nodosa, in which neutrophils predominate) in the blood vessels of the deep dermis and superficial hypodermis. The disease mainly affects females (17:3), with a mean age of forty years. In general, this entity is considered benign with a variable clinical presentation. The importance of knowing this entity lies in the fact that the pathological findings are discordant with the clinic and an erroneous diagnosis may lead to unnecessary treatments. This vasculitis usually has a benign course and no progression to systemic disease. Herein, we present two cases of this rare vasculitis.

Key words: Macular lymphocytic arteritis, Cutaneous polyarteritis nodosa, Vasculitis, Systemic lupus erythematosus

INTRODUCTION

Macular lymphocytic arteritis (MLA) is a type of indolent, cutaneous vasculitis that affects small to medium-sized blood vessels. Histopathologically, it is characterized by a predominantly lymphocytic leukocytic infiltrate (unlike polyarteritis nodosa, in which neutrophils predominate) in the blood vessels of the deep dermis and superficial hypodermis [1]. The disease mainly affects females (17:3) with a mean age of forty years [2,3]. In general, this entity is considered benign with a variable clinical presentation. It has been reported from an asymptomatic presentation (66%) to the appearance of oval macules of erythematous to brown coloration, especially on the lower limbs, or the appearance of leveloid patches (65% of symptomatic cases). Other symptoms that may accompany skin lesions are pruritus (10%), pain in the lesions (7.5%), weakness in the extremities (5%), and Raynaud's phenomenon (2.5 %) [3,4]. The importance of knowing this entity lies in the fact that the pathological findings are discordant with the clinic, and an erroneous diagnosis may lead to unnecessary treatments. This vasculitis usually has a benign course and no progression to systemic disease [1]. Herein, we present two cases of this rare vasculitis.

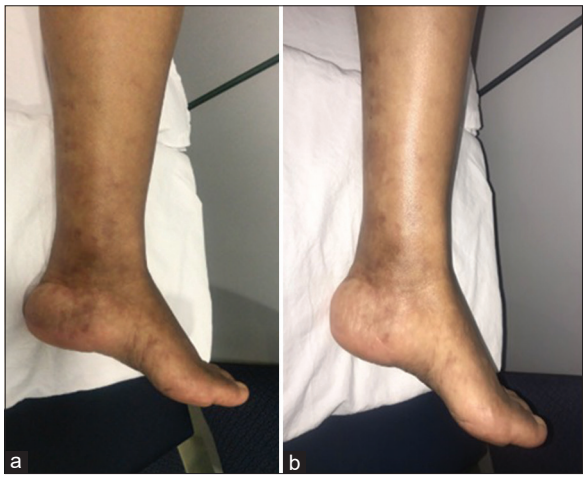
Case 1

The first case was a 52-year-old female with a history of systemic lupus erythematosus (SLE) with positive

How to cite this article: Corona-Herrera JM, Zepeda-Gutiérrez LA, Falcon-Antonio OE, Barrera-Godínez A, Méndez Flores S, Domínguez-Cherit JG. From macular lymphocytic arteritis to cutaneous polyarteritis nodosa: A spectrum of the same disease? Two cases of the same clinical presentation yet different histopathological findings. Our Dermatol Online.2024;15(1):64-66.

Submission: 08.06.2023; **Acceptance:** 03.08.2023
DOI: 10.7241/ourd.20241.15

antiphospholipid antibodies (anticardiolipin). She presented a localized dermatosis (Figs. 1a and 1b) on the lower extremities with a tendency to symmetry, which affected the legs on their anterior, internal, and external lateral aspects, as well as on the dorsum of the feet, characterized by erythematous, hyperpigmented spots and brown with irregular and poorly defined borders, some circular, others oval converging and giving the appearance of livedo racemosa of one week of evolution and no other associated symptoms. A punch biopsy was performed (Figs. 2a – 2d), which reported panarteritis nodosa (PAN). Subsequently, renal activity was documented and three boluses of methylprednisolone plus cyclophosphamide were administered. During follow-up, a clinical improvement was observed in all lesions, and no other systemic symptoms were noted.



Figures 1: (a and b) Erythematous, brown, hyperpigmented spots with irregular and poorly defined borders, some circular, others oval converging and giving the appearance of livedo racemosa.

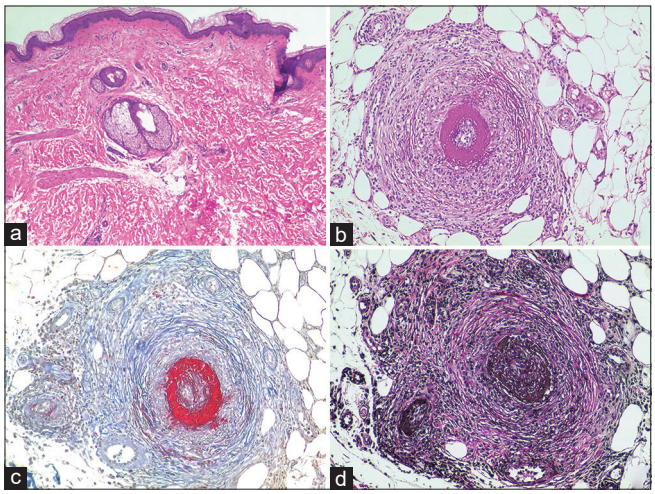
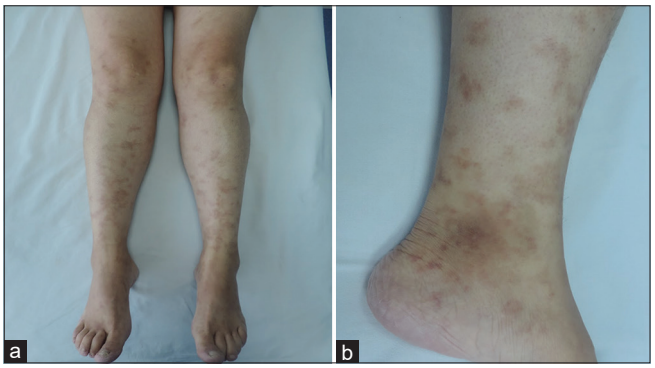


Figure 2: (a) The biopsy showing epidermal maturation and no affection in the superficial and medium dermis. (b) The damage observed in medium and small-sized arteries within subcutaneous cell tissue, necrotizing vasculitis involving all layers of the vascular wall accompanied by fibrinoid degeneration; neutrophils, eosinophils, and lymphocytes present. (c) Histochemical stains (Masson's trichrome) marking fibrinoid necrosis. (d) Elastic fiber stain showing the inflammatory infiltrate dissecting through the wall of the vessel.

Case 2

The second case was a 37-year-old female with a history of SLE. He presented a localized dermatosis (Figs. 3a and 3b) in the lower extremities with a tendency to symmetry, which affected the anterior and inner thighs, all sides of the legs, the malleolus, and the back of the feet, characterized by discretely erythematous, irregular, poorly defined, hyperpigmented spots, some with a reticulated appearance. A punch biopsy was performed, which reported panarteritis nodosa (Figs. 4a – 4d). One month later, renal activity was documented due to the presence of proteinuria, and a short course of prednisone and mycophenolate mofetil was initiated. During follow-up, a clinical improvement



Figures 3: (a and b) Hyperpigmented spots with a reticulated appearance.

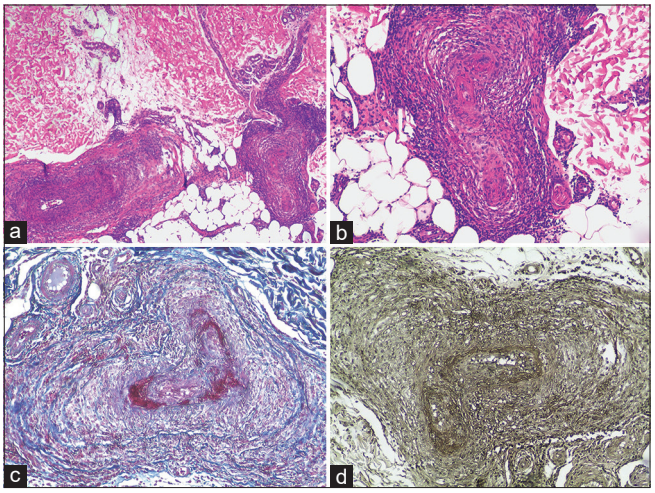


Figure 4: (a) The lesion observed in the deep planes, also involving medium-sized vessels in the deep dermis and subcutis. (b) Early lesions showing fibrinoid necrosis with thickening and infiltration of the vessel wall and leukocytoclastic also observed. (c and d) Vessel partial obliteration occurring secondary to intimal and mural fibrosis.

was observed in all lesions, and no further treatment was needed.

DISCUSSION

These two patients with a history of SLE presented with a similar clinical course. The biopsies revealed findings suggestive of PAN, yet the first with neutrophil, eosinophil, and lymphocyte infiltrate and the second just with lymphocyte infiltrate. However, during the course of both conditions, the skin lesions were self-limiting and were not accompanied by other systemic symptoms characteristic of polyarteritis nodosa. To date, there still has been a debate regarding whether this entity (AML) is misdiagnosed as cutaneous PAN (cPAN) or is a spectrum in and of itself [3.4]. Clinically, cPAN is characterized by painful, erythematous nodules on the lower extremities associated with livedo racemosa, followed by ulcers, and rarely gangrene and digital necrosis. Histopathological findings include septal panniculitis with necrotizing vasculitis of small vessels in the deep dermis and superficial hypodermis. In a longer series, it was impossible to demonstrate the development of a systemic disease in the PAN variety after a follow-up of seven years [5]. cPAN, due to its benign course and medium vessel vasculitis, may resolve with reticulated hyperpigmentation (post-inflammatory hyperpigmentation), yet to our knowledge, hyperpigmented macules never occur as primary lesions of vasculitis, and based on overlapping clinicopathological characteristics, macular arteritis may represent a latent, not nodule-forming chronic variant of cutaneous polyarteritis nodosa [6,7]. Regarding histopathological findings, it is important to make the differential diagnosis of other entities and make clinical pathological correlation, since cases of histopathological findings have been reported, mimicking thrombophlebitis. Although the differential diagnosis is sometimes difficult, thrombophlebitis should be considered when diagnosing cPAN and AML to avoid misdiagnosis and excessive treatment [8]. Our patients presented skin lesions characterized by hyperpigmented and erythematous spots, consistent with what has been reported in the literature (same lesions in up to 65% of cases) [3,4]. Regarding the histopathological findings, both biopsies highlighted the presence of fibrinoid necrosis and predominantly lymphocytic infiltrate in small and medium caliber vessels in the deep dermis and hypodermis, consistently with an AML case series [2].

CONCLUSION

Up to now, there have been no reports of an association of AML with LEG activity. However, in these two cases, it was highlighted that, simultaneously or in a short period of time after the dermatosis appeared, activity by the LEG was documented on other levels. During the follow-up of both cases, they never met the criteria for polyarteritis nodosa.

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients gave their consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

REFERENCES

1. Zampella JG, Vakili S, Doig S, Girardi N, Kwatra SG, Philip S, et al. Macular lymphocytic arteritis: Clinical-pathologic correlation of a rare vasculitis. JAAD Case Rep. 2017;3:116-20.
2. Morita TCAB, Trés GFS, Criado PR. Is macular lymphocytic arteritis limited to the skin? Long-term follow-up of seven patients. An Bras Dermatol. 2020;95:32-9.
3. Vakili S, Zampella JG, Kwatra SG, Blanck J, Loss M. Lymphocytic thrombophilic arteritis: A review. J Clin Rheumatol. 2019;25:147-52.
4. Valverde R, Garrido C, Leis V, Ruiz-Bravo E. Macular arteritis: A pole of the spectrum of cutaneous polyarteritis nodosa? Actas Dermosifiliogr. 2013;104:263-5.
5. Martinez Braga G, Di Martino Ortiz B. Septal panniculitis: Clinico-pathological review of the literature and case presentation. Our Dermatol Online. 2014;5:74-82.
6. Al-Daraji W, Gregory AN, Carlson JA. "Macular arteritis": A latent form of cutaneous polyarteritis nodosa? Am J Dermatopathol. 2008;30:145-9.
7. Fein H, Sheth AP, Mutasim DF. Cutaneous arteritis presenting with hyperpigmented macules: Macular arteritis. J Am Acad Dermatol. 2003;49:519-22.
8. Hiraiwa T, Yamamoto T. Superficial thrombophlebitis mimicking cutaneous polyarteritis nodosa as an early and sole cutaneous manifestation of Behçet's disease. Our Dermatol Online. 2015;6:367-8.

Source of Support: This article has no funding source.

Conflict of Interest: The authors have no conflict of interest to declare.