A Case of Macular Lymphocytic Arteritis in a Caucasian Male

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ABSTRACT

Macular lymphocytic arteritis (MLA), also known as lymphocytic thrombophilic arteritis is a rare, medium vessel vasculitis that has previously been reported in literature as cases predominantly presenting in females,[1] and almost exclusively in individuals of African descent.[2] There have been no published cases of MLA presenting in a Caucasian male at this time; however, reports of such cases would help elude to any possible genetic predisposition for MLA as well as broadening differentials in lower extremity rashes in the Caucasian male population. We present a case of a 27-year-old Caucasian male with violaceous macules on his bilateral lower extremities. He denied joint pain, photosensitivity, mouth ulcers, or genitalia lesions. On physical exam were violaceous macules bilaterally on his feet and lower legs. A 4mm punch biopsies from the right lateral foot showed histological features consistent with MLA. The patient was treated with crisaborole, and halobetasol propionate lotion. He returned 17 days later for suture removal and reported improvement with the treatment and no new lesions. Our Caucasian male patient in his late twenties represents a unique case of rare pathology, which previously was only reported in either females or males of Asian or African descent.[3] We aim to contribute to understanding any possible genetic predisposition for MLA as wells as bringing awareness for the inclusion of a disease that is not traditionally present in this subset of the population.

INTRODUCTION

Macular lymphocytic arteritis (MLA):

- Rare medium vessel vasculitis.
- Presents as multiple asymptomatic hyperpigmented macules on the lower extremities.[2]
- Lesions have been described as macules, papules, livedoid patches, and ulcers.[4]
- Histology will show lymphocytic infiltration of arteries in the deep dermis and subcutaneous fat.[5]
- Current literature of cases presents predominantly in females, and almost exclusively in individuals of African descent. (3,2)

CASE REPORT

27-year-old Caucasian male with no contributing past medical history:

- Presents with violaceous macules on bilateral lower extremities that arose over the preceding two months.
- Denied joint pain, photosensitivity, mouth ulcers, or genitalia lesions.
- Lesions were treated with clobetasol propionate 0.05% cream with no significant improvements.
- Physical examination showed violaceous macules bilaterally on his feet and lower legs, many of which were turning brown or lightening in color (**Figure 1**).

WORK-UP

Two 4mm punch biopsies were performed

- Direct immunofluorescence (DIF) and hematoxylin and eosin (H&E) staining to evaluate for vasculitis.
- CBC, renal panel, liver function panel, ESR, anti-neutrophil cytoplasmic antibody, antinuclear antibody, antistreptolysin-o titer, cardiolipin Ab IgG, cardiolipin Ab IgM, hepatitis B, and C screening, and stool guaiac tests, were ordered and found to be negative.

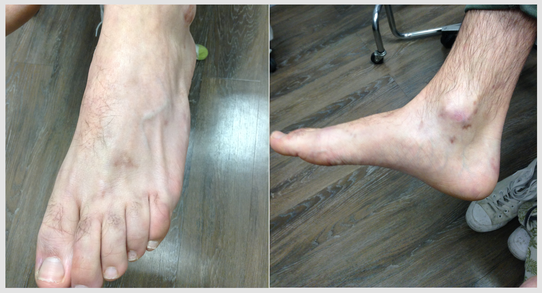

Figure 1. Violaceous macules bilaterally on feet and lower legs

HISTOLOGY

- Biopsy from the right lateral foot showed an unremarkable epidermis and superficial dermis.
- At the junction of the deep dermis and subcutis, there was a lymphocytic infiltrate that surrounds a muscular artery (**Figure 2**).
- Focal hyalinization and fibrin were noted in the wall and lumen without significant numbers of neutrophils or leukoclasia present (**Figure 3**).

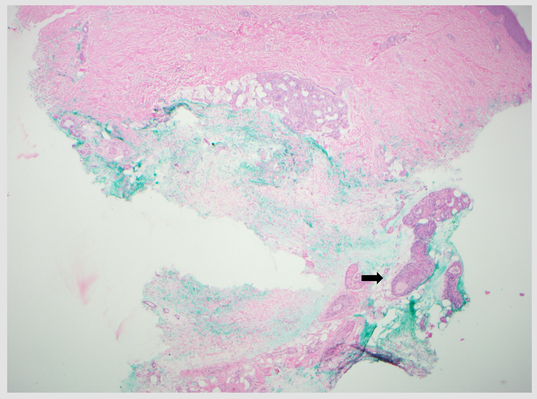

Figure 2. Lymphocytic infiltrate surrounding a muscular artery with focal hyalinization and fibrin in the wall and lumen at the junction of the deep dermis and subcutis.

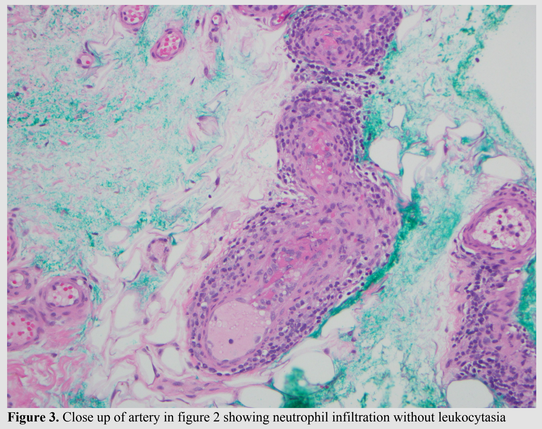

Figure 3. Close up of artery in figure 2 showing neutrophil infiltration without leukocytasia

MANAGEMENT

He was treated with crisaborole topically, twice daily, and halobetasol propionate lotion and returned 17 days later for suture removal and follow up.

- Reported improvement with the treatment and no new lesions.
- Violaceous lesions in his lower extremities appeared to be turning brown, resembling post-inflammatory hyperpigmentation.
- The patient was subsequently evaluated by a rheumatologist who felt the patient had cutaneous vasculitis.
- Serologies, clinical exams, and diagnostics did not exhibit any features of systemic vasculitis.
- No systemic therapy was recommended at that time.

DISCUSSION

Cases of macular lymphocytic arteritis (MLA) appear throughout literature presenting clinically with macules of hyper- or hypo-pigmentation[1] accompanied by histological findings similar to those of cutaneous polyarteritis nodosa (c-PAN) with lymphocytic infiltration of arteries in the deep dermis and subcutaneous fat.

- MLA differs from c-PAN in that it lacks neutrophilic infiltrate on histology.[4]
- The disease is typically indolent and limited to the skin, with one case being reported with progressive systemic involvement.(7,8)

The present case is a 27-year-old Caucasian male who was, otherwise, in good health, presenting with violaceous macules on his lower extremities bilaterally.

- Laboratory, clinical presentation and pathological findings support the diagnosis of MLA in this patient.
- Our patient is a unique case of rare pathology, which previously was only reported in either females or males of Asian or African descent.
- There have been no reported cases presenting in Caucasian males in literature at this time.
- Reports of such cases would be helpful in understanding possible genetic predisposition for MLA, and broadening differentials in lower extremity rashes in the Caucasian male population.

CONCLUSION

We have presented a case of macular lymphocytic arteritis, a rare form of medium vessel vasculitis, in a Caucasian male. No other cases of Caucasian males have been reported in the literature thus far. This case will contribute to enhancing a clinician's differential for lower extremity hyperpigmented macules, assist in determining possible genetic predispositions for this disease, and provide additional evidence for future treatments of macular lymphocytic arteritis.

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