Clinical Medicine Insights: Dermatology

CASE REPORT

Does Lymphocytic Thrombophilic Arteritis Have a Wider Histopathological Spectrum? A Case Displaying Clinical Features of Macular Arteritis with Histolopathological Features of Lymphocytic Thrombophilic Vasculitis

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Abstract: Macular arteritis is a benign condition characterised clinically by livedo racemosa and histopathologically by lymphocytic vasculitis involving the medium sized arterioles. We report a case displaying clinical features of macular arteritis with a lymphocytic vasculitis involving the vessels of the superficial and mid dermis histopathologically.

Keywords: macular arteritis, lymphocytic thrombophilic vasculitis, superficial and mid dermal vessels

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A 37 year old woman was seen with a 20 year history of recurrent episodes of livedo racemosa involving her lower limbs (Fig. 1). She was initially diagnosed with Henoch Schonlein purpura however this diagnosis was later rejected by two haematologists and a dermatologist but an alternate diagnosis could not be established.

Past medical history was venocuff surgery 13 yrs previously for bilateral varicose veins. This procedure involves the application of a small silicon coated Dacron belt around sites of venous incompetence which restores normal valvular function. There was no previous history of thrombo-embolic phenomenon. Systemically the patient was well.

Clinical examination (Fig. 1) revealed livedo patterning and macular hyper pigmentation involving the lower limbs only. There was no history of palpable purpura or ulceration. There was no evidence of atrophie blanche.

Negative investigations included full blood count, electrolytes, urea, creatinine and liver function tests, anti-nuclear antibody, extractable anti-nuclear antigen, double-stranded DNA, rheumatoid factor, protein electrophoresis, anti-neutrophil cytoplasmic antibody, complement studies, hepatitis B, hepatitis C, HIV, urine MCS, protein C, S, anti-thrombin 111, factor V Leiden, homocysteine and cryoglobulins. Anti-cardiolipin IgM antibodies were persistently elevated in low titre but anti-cardiolipin IgG and lupus anticoagulant were negative.

A provisional diagnosis of lymphocytic thrombophilic arteritis was made and biopsy taken.

Histopathology (Figs. 2 and 3) showed a lymphocytic vasculitis involving vessels of the superficial and mid dermis with mural and luminal fibrin. The findings differ from the described histopathology in showing involvement of superficial and mid dermal vasculature. Medium sized arterioles of the deep dermis and upper sub cutis were not evident on biopsy. Livedoid patterning supports their involvement but is not diagnostic as livedoid patterning is also seen with venous disorders. The long duration of this condition would be consistent with obliteration of many these vessels and a permanent reduction in their number so it was not surprising a vessel was not found. It was not felt clinically appropriate to repeat the biopsy.

Macular arteritis was first reported by Fein, Sheth and Mutasim.[1] They reported three patients with a progressive macular hyperpigmentation associated with a lymphoplasmocytic infiltrate centred on the arterioles of the deep reticular dermis and subcutaneous fat. A benign prognosis was reported. A further 2 Japanese patients were reported by Sadahira C, Yoshida T, Matsuoka Y, Takai I et al[2] Identical histology was reported.

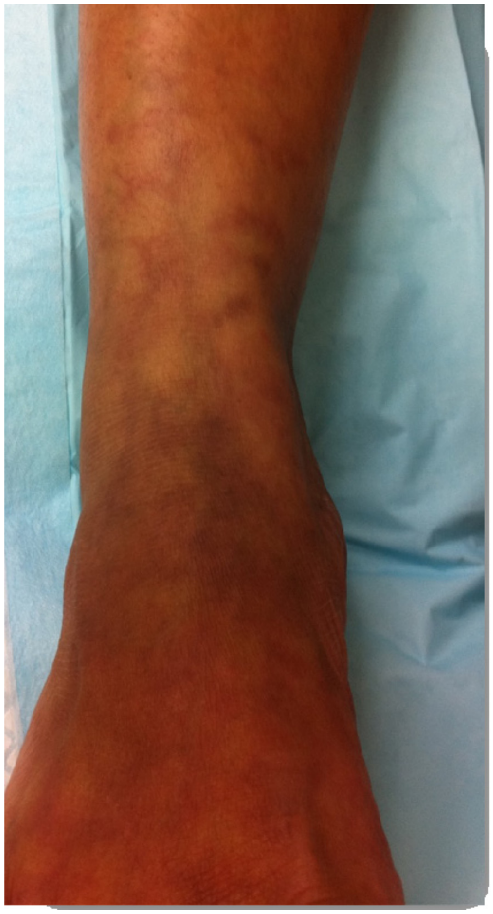

Figure 1. Lower limb showing macular hyper pigmentation and livedo racemosa.

The term lymphocytic thrombophilic arteritis was introduced by Siong-See Lee, Kossard and McGrath[3] to describe this condition. They reported five patients with patchy reticular hyperpigmentation and livedo racemosa associated with identical histopathological findings. Four of the five patients in Siong et al's series[3] had anticardiolipin, antiphospholipid, lupus anticoagulant or antiglycoprotein antibodies. None the less the authors postulated this was an epiphenomenon related to endothelial damage. Although this patient had low titre anti-cardiolipin IgM antibodies she failed to meet the current criteria for antiphospholipid syndrome.[4]

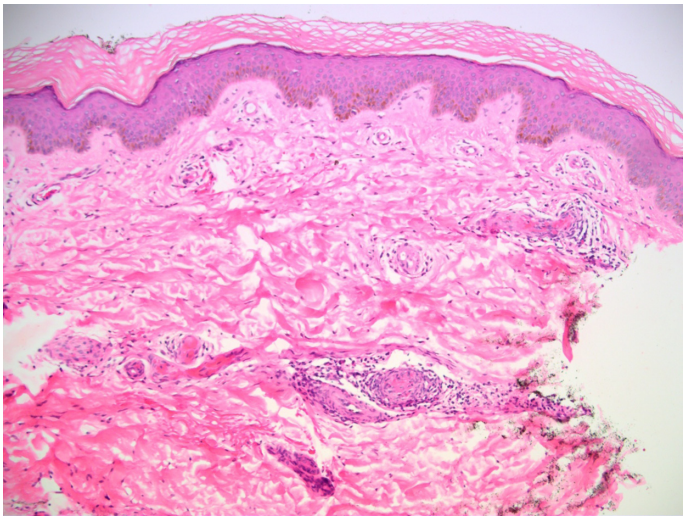

Figure 2. Low power view showing a moderately intense lymphocytic infiltrate surrounding and involving the wall of the superficial and mid dermal vessels.

It is presumed these findings are part of the spectrum of lymphocytic thrombophilic arteritis although the possibility they represent a separate disorder cannot be discounted. However the clinical findings, benign history and negative laboratory results are consistent with the entity reported as macular arteritis. Livedoid vasculopathy secondary to venous hypertension leads to fibrin deposition within the vessels walls of the superficial to mid dermis but significant extravasation of red blood cells and a less intense lymphocytic infiltrate is usually evident. In addition, despite the previous history of venocuff surgery this patient displayed no clinical features of venous hypertension.

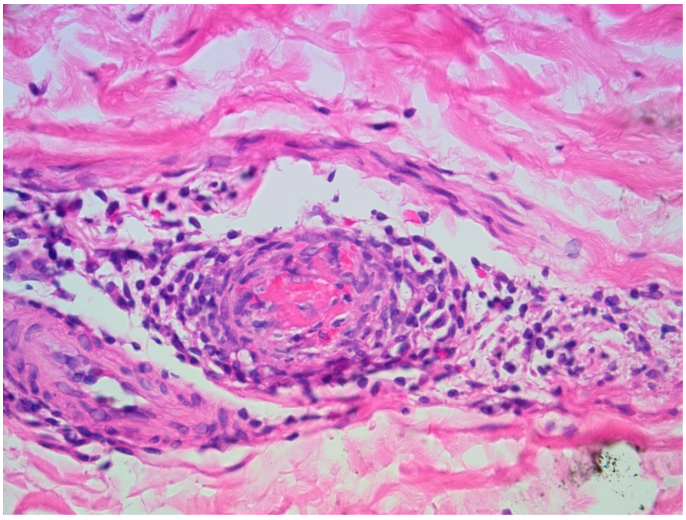

Figure 3. High power view showing luminal and mural fibrin deposition.

We thus report a clinical case of macular arteritis with atypical histopathological features. It was not possible to make a definitive diagnosis of lymphocytic thrombophilic arteritis as a medium sized arteriole could not be identified on the biopsy specimen. None the less the clinical and laboratory features are consistent with the entity described as lymphocytic thrombophilic arteritis. Although livedoid vasculopathy results in fibrin deposition within the superficial and mid dermal vessels there were no clinical features to support this diagnosis and the histopathology was not typical.

Author Contributions

Wrote the first draft of the manuscript: RN. Contributed to the writing of the manuscript: RN, KW. Agree with manuscript results and conclusions: RN, KW. Jointly developed the structure and arguments for the paper: RN, KW. Made critical revisions and approved final version: RN, KW. All authors reviewed and approved of the final manuscript.

Disclosures and Ethics

As a requirement of publication author(s) have provided to the publisher signed confirmation of compliance with legal and ethical obligations including but not limited to the following: authorship and contributorship, conflicts of interest, privacy and confidentiality and (where applicable) protection of human and animal research subjects. The authors have read and confirmed their agreement with the ICMJE authorship and conflict of interest criteria. The authors have also confirmed that this article is unique and not under consideration or published in any other publication, and that they have permission from rights holders to reproduce any copyrighted material. Any disclosures are made in this section. The external blind peer reviewers report no conflicts of interest.

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