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CASE REPORT

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

Rowland Noakes¹ and Kevin Whitehead²

¹Terrace Dermatology, Morris Towers, Wickham Terrace, Brisbane. ²Histopathologist, Histopathology, Sullivan Nicolaides Pathology. Corresponding author email: ky_n_urenine@hotmail.com; kevin_whitehead@snp.com.au

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 Schönlein 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 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 al2 Identical histology was reported.

The term lymphocytic thrombophilic arteritis was introduced by Siong-See Lee, Kossard and McGrath3 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 series3 had anticardiolipin, antiphospholipid, lupus anticoagulant or antilygycoprotein 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
A case of macular arteritis with involvement of the superficial and mid dermal vessels

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**
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