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Clinical Vignette

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Nerve pathology and cutaneous features of acute rheumatoid vasculitis

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A 60-year-old woman with a 12-year history of RA on prednisone 5 mg/day and hydroxychloroquine 200 mg/day presented with stepwise progression of pain and weakness of the left followed by right foot over 1 month. Physical examination was notable for livedo racemosa (Supplementary Figure S1, available at *Rheumatology Advances in Practice* online) and asymmetric distal lower extremity weakness (MRC grade 4+ on the right and 4– on the left), sensory loss and hyporeflexia. There were no signs of synovitis. Electrophysiologic studies showed an asymmetric axonal polyneuropathy. Rheumatoid factor (327 IU/ml; negative <15), anti-cyclic citrullinated peptide and inflammatory markers were elevated. Left sural nerve biopsy revealed acute occlusion of multiple large epineurial arterioles (Fig. 1A and B), inflammatory collections (Fig. 1A and B), fibrinoid necrosis (Fig. 1A–D), vessel-wall destruction (Fig. 1E) and fulminant axonal degeneration (Fig. 1F). These findings were diagnostic of acute necrotizing vasculitic neuropathy [1]. Treatment with high-dose steroids and rituximab led to ongoing clinical improvement. Rheumatoid vasculitis is a life-

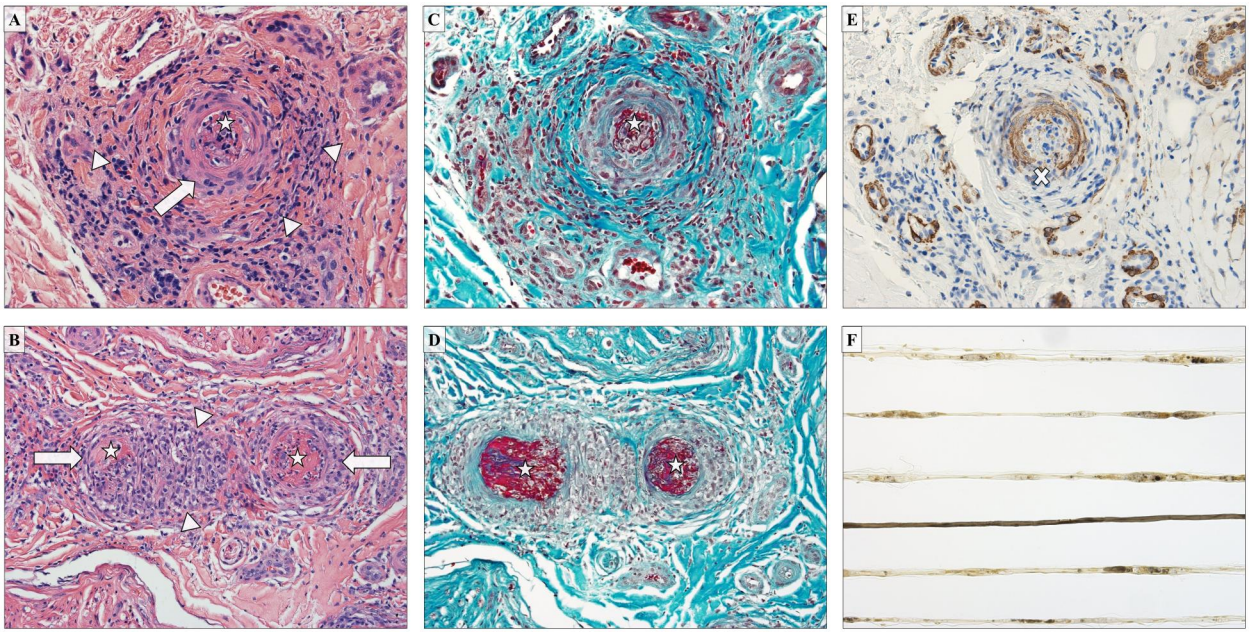

Figure 1. Sural nerve biopsy. Acute occlusion [arrows (panels A and B), haematoxylin and eosin] of large epineurial arterioles with perivascular inflammatory collections [arrowheads (panels A and B), haematoxylin and eosin] and fibrinoid necrosis [asterisks (panels A and B), haematoxylin and eosin; (panels C and D), Masson's trichrome], vessel wall destruction [X (panel E), smooth muscle actin] and fulminant axonal degeneration (panel F, teased fibres). Scale bar: 40× (panels A,C, and E) and 20x (panels B, D and F) magnification

threatening complication of RA, most commonly occurring in patients with long-standing disease (generally >10 years duration) with low (burned-out) articular disease activity [2]. Cutaneous and nerve vasculitis are the most common manifestations of rheumatoid vasculitis [2]. Nerve biopsy can help to distinguish neuropathy due to vasculitis from alternative aetiologies (e.g. toxic, nerve entrapment) and tailor appropriate treatment.

Supplementary material

Supplementary material is available at *Rheumatology Advances in Practice* online.

Data availability

Data are available upon reasonable request by any qualified researchers who engage in rigorous, independent scientific research and will be provided following review and approval of a research proposal and Statistical Analysis Plan and execution of a Data Sharing Agreement. All data relevant to the study are included in the article.

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