Necrosis of the gastrocnemius muscle in microscopic polyangiitis

Takahiko Akagi,[1] Hirotake Nishimura,[2] Tomoyuki Mukai,[1] Yoshitaka Morita[1]

[1]Department of Rheumatology, Kawasaki Medical School, Kurashiki, Japan
[2]Department of Pathology, Kawasaki Medical School, Kurashiki, Japan

Correspondence to
Dr Yoshitaka Morita,
morita@med.kawasaki-m.ac.jp

Accepted 17 January 2018

DESCRIPTION

A 59-year-old man presented with a 1-month history of bilateral lower leg pain. The pain was more severe in the left leg, with associated swelling of the lower leg. He did not have any eye, nose, paranasal sinus or respiratory symptoms. Physical examination of the chest and abdomen was unremarkable. Neurological examination was normal. He had a body temperature of 36.5°C, a blood pressure of 97/71 mm Hg, and a pulse of 77 beats/min. Blood tests revealed a C-reactive protein level of 155 mg/L, leucocyte count of 10.3×10^9/L (neutrophil count 8.4×10^9/L), haemoglobin level of 14.2 g/dL and platelet count of 328×10^9/L. Serum creatine kinase and lactate dehydrogenase levels were slightly elevated at 237 U/L (normal 41–153) and 231 U/L (124–222), respectively. Serum creatinine was 0.51 mg/dL. Serum complement (C3 and C4) concentrations were normal. Antinuclear antibody and myeloperoxidase-specific antineutrophil cytoplasmic antibody (ANCA) were negative. Proteinase 3-specific ANCA was positive with a titre of 241 U/mL (normal <3.5). Urinalysis showed 2+ haematuria and 1+ proteinuria with a few granular casts, red blood cell casts and white blood cell casts. Plain X-ray and high-resolution CT of the chest showed no pulmonary abnormality. MRI revealed diffuse oedema in the left gastrocnemius muscle.

Biopsy of the left gastrocnemius muscle revealed a small-vessel vasculitis composed of fibrinoid necrosis and infiltration of inflammatory cells in the arterial walls (figure 1A). The muscle tissues were clearly divided into necrotic lesions and normal muscle fibres, and necrotic vasculitis was found near the infarct-like muscle lesions (figure 1B). A

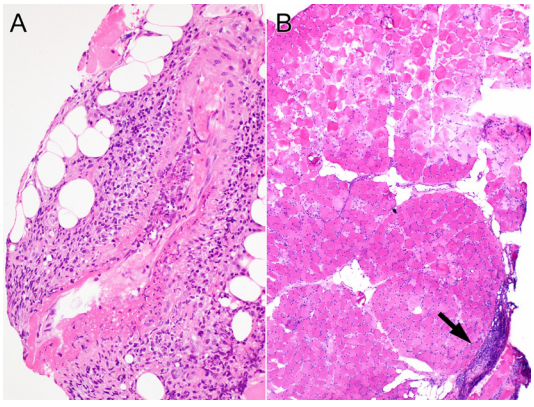

Figure 1 A gastrocnemius muscle biopsy specimen. H&E staining showed (A) small-vessel vasculitis and (B) ischaemic infarct-like necrotic changes. Necrotic vasculitis was seen in the same biopsy specimen (*arrow*).

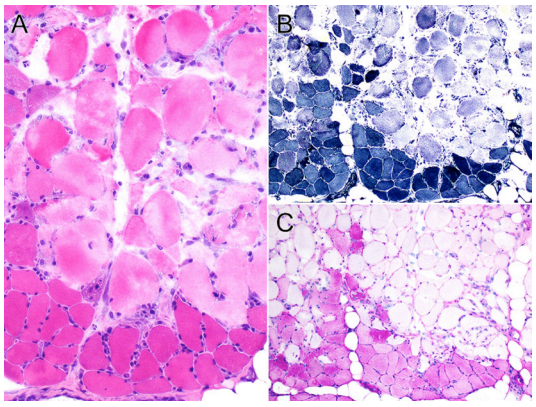

Figure 2 Ischaemic necrotic muscle tissues stained with (A) H&E, (B) nicotinamide adenine dinucleotide tetrazolium reductase and (C) periodic acid-Schiff.

diagnosis of muscle necrosis associated with small-vessel vasculitis was considered. The necrotic muscle tissue did not react with nicotinamide adenine dinucleotide tetrazolium reductase or with glycogen on periodic acid-Schiff staining, which also supported muscle ischaemia (figure 2). No granuloma was observed. A diagnosis of ANCA-positive microscopic polyangiitis was made. The patient was successfully treated with prednisolone 50 mg/day and intravenous cyclophosphamide, followed by methotrexate.

Small to medium-sized vessel vasculitis can develop in the skeletal muscle of the lower limb. Our case is unusual in that a muscle biopsy revealed ischaemic infarct-like necrosis. A clinicopathological review of 40 cases of skeletal muscle vasculitis identified neurogenic muscle atrophy in the majority (95%) of biopsy specimens but did not describe muscle necrosis.[1] A literature search revealed only one report of muscle necrosis in ANCA-negative microscopic polyangiitis.[2] The reason why muscle necrosis is uncommon in skeletal muscle vasculitis has yet to be identified. More reports on skeletal muscle vasculitis and investigations in experimental

Learning points

- Small to medium-sized vessel vasculitis can develop in the skeletal muscle of the lower limbs.
- Neurogenic muscle atrophy is observed in the majority of biopsy specimens from patients with skeletal muscle vasculitis, but muscle necrosis is uncommon.

To cite: Akagi T, Nishimura H, Mukai T, *et al*. *BMJ Case Rep* Published Online First: [*please include* Day Month Year]. doi:10.1136/bcr-2017-223585

animal models are needed to better understand the histopathological findings in this disease.

Contributors TA, HN and YM are involved in conception or design of the work. TA and HN are responsible for acquisition of data. TA, HN, TM and YM are responsible for analysis and interpretation of data. TA, TM and YM drafted or revised the manuscript.

Funding This research received no specific grant from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests TA, TM and YM received scholarship donations from AbbVie, Actelion, Astellas, Bristol-Myers, Chugai, Daiichi-Sankyo, Eisai, Eli Lilly, Japan Blood Products Organization, Mitsubishi-Tanabe, MSD, Pfizer, Shionogi, Takeda, Teijin and UCB.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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