childhood. This contrasts with sporadic late-onset nemaline myopathy, which has an autoimmune aetiology and usually manifests after the third decade of life.

Serum muscle enzymes in nemaline myopathy tend to be in the normal range, though CK may be elevated. Electromyography usually shows myopathic changes, while MRI may show areas of hyperintensity [4]. Histopathology with the Gomori trichrome technique typically shows red-stained, rod-shaped, predominantly cytoplasmic nemaline bodies [3].

CNM presenting in adulthood is rare, and can mimic other more common rheumatological diseases. PM, which presents with a similar pattern of muscle weakness, can exhibit nemaline bodies in muscle biopsies [5]. Nemaline bodies may also be seen in HIV, SS, monoclonal gammopathy and primary hypothyroidism [5, 6]. As in the described case, immune dysfunction may also worsen symptoms, unmasking hitherto undiagnosed CNM [6].

Treatment for nemaline myopathy is supportive, with breathing assistance, mobility aids and nasogastric feeding as required. Research into L-tyrosine and gene therapy may provide the potential to alter the disease course in the future [3]. To our knowledge, this is the first description of a patient co-presenting with CNM and SLE, and in such patients with a relevant childhood history, myalgia and weakness, further neuromuscular investigations may be appropriate, even in the face of a normal CK and imaging.

Funding: No specific funding was received from any bodies in the public, commercial or not-for-profit sectors to carry out the work described in this manuscript.

Disclosure statement: The authors have declared no conflicts of interest.

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Revised version accepted 2 June 2017

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Rheumatology 2017;56:2035–2036
doi:10.1093/rheumatology/kex294
Advance Access publication 27 July 2017

Osteitis as a manifestation of familial Mediterranean fever

Rheumatology key message

- Unifocal osteitis with fever may be a new manifestation of FMF.

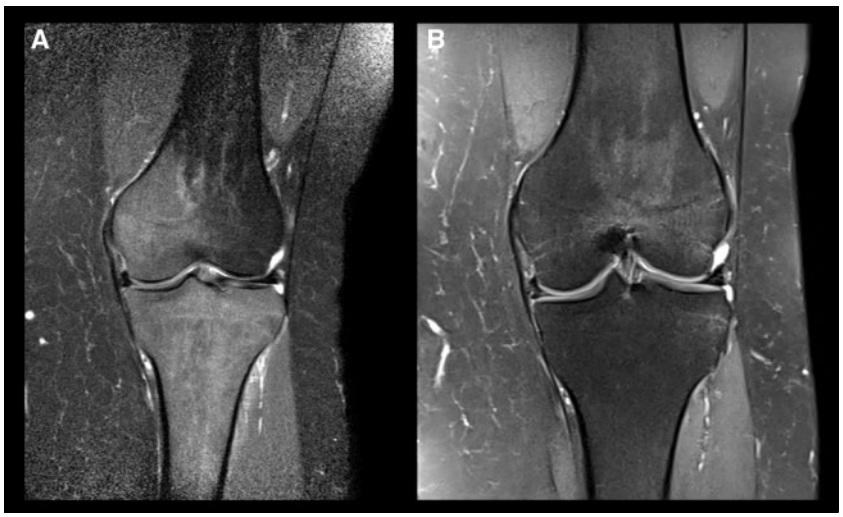
Sir, FMF is the most common autoinflammatory disease characterized by recurrent self-limited attacks of fever, sterile serositis, arthritis and erysipelas-like erythema [1]. Non-canonical manifestations of FMF are increasingly recognized in recent years, such as calf pain, vasculitides and neurological disease. Herein, we describe unifocal osteitis in an FMF patient who responded well to anakinra.

A 40-year-old Turkish female patient was previously diagnosed with FMF (V726A/–), with recurrent 1- to 3-day episodes of fever, peritonitis and intermittent arthritis. The maximal dose of colchicine was not effective enough to prevent the bouts of disease; therefore, anakinra 100 mg/day was added to colchicine treatment 18 months before. After this treatment, disease control was achieved, but the patient had borderline low leucocyte counts. Anakinra was withheld 3 weeks before the last presentation because of leukopenia. Shortly after, the patient complained of sharp pain around her left knee, accompanied by fever. Physical examination was unremarkable except for reproducible pain with compression and warmth on the affected site. The patient denied any trauma or overuse. MRI of the knee demonstrated significant bone marrow oedema of the femoral condyles and proximal tibia, with sparing of the joint space

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Fig. 1 MRI of the affected area, before and after treatment



(**A**) Coronal fat-saturated proton density image shows bone marrow oedema of femoral condyles and proximal tibia. (**B**) One month later, complete resolution of pathological signals and oedematous pattern.

(Fig. 1A). Her laboratory findings showed low white blood cell count (3.8×10^9/l) and increased acute phase responses; CRP 105 mg/l (normal, 0–5 mg/l) and ESR 66 mm/h. The procalcitonin concentration was normal. Blood and urine cultures were negative. A peripheral blood smear was compatible with her blood count, and there were no bizarre cells. Bone scintigraphy demonstrated uptake of the tracer in the affected area. As no evidence of any other disease was found and white blood cell counts improved, anakinra was reinstituted on the ninth day of complaints. The patient was relieved dramatically from pain and fever soon after the anakinra injection. Two weeks after anakinra treatment, her CRP and ESR values had returned to normal values and knee MRI showed complete resolution of pathological findings (Fig. 1B).

Osteitis is a feature in certain autoinflammatory diseases, such as chronic recurrent multifocal osteomyelitis, Synovitis, acne, pustulosis, hyperostosis syndrome (SAPHO) and Deficiency of the IL-1 receptor antagonist (DIRA). Recently, some patients with heterozygous *MEFV* mutations reported atypical clinical presentations, such as colchicine-responsive recurrent episodes of muscle pain and chronic recurrent multifocal osteomyelitis-like syndrome [2, 3]. Osteitis is also a feature of spondyloarthritis, and it is known that the frequency of spondyloarthritis increases in FMF patients irrespective of colchicine prophylaxis [4]. To the best of our knowledge, this case is the first FMF-associated unifocal osteitis with fever in the literature. This case brings to mind that some of the attacks considered as arthritis might, in fact, be osteitis. As a limitation, we did not perform gene analysis for other autoinflammatory diseases in this patient; however, the geographical region and clinical picture other than osteitis are typical of FMF.

Funding: No specific funding was received from any bodies in the public, commercial or not-for-profit sectors to carry out the work described in this manuscript.

Disclosure statement: The authors have declared no conflicts of interest.

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Revised version accepted 19 June 2017

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