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SHORT REPORT

Incomplete attack and protracted sacroiliitis: an unusual manifestation of FMF in a child

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We describe an unusual presentation of a patient with familial Mediterranean fever (FMF) in whom the protracted sacroiliitis was the dominant clinical feature with incomplete abdominal attacks.

An 11-year-old Turkish girl was referred for evaluation of recurrent abdominal and low back pain. Recurrent abdominal pain started while she was 6 years old without fever lasting for 1–2 days and localising at the right lower quadrant with full remission after the attack. She had a history of appendicectomy at the age of 7 years which was not productive. The complaints recurred every 1–2 months. As for her low back pain, complaints started when she was 2 years of age with exacerbation on exercise. Low back pain recurred every 2–3 months and persisted for more than a month. Throughout the period of 6–11 years of age, she had been prescribed nonsteroidal anti-inflammatory drugs (NSAID) which proved to be non-beneficial.

She had no history of recurrent upper respiratory infection, psoriasis, chronic diarrhoea, oral aphthae or genital ulcers. There is no family history of periodic fever, FMF, spondyloarthropathy, rheumatoid arthritis, psoriasis or inflammatory bowel disease or Behçet's disease.

Physical examination revealed tenderness on pressure of the pelvis in the lateral position and no restriction on movement of the lumbar spine or on chest expansion. There was no abnormal ocular finding; neurological and other joint examinations were normal.

Laboratory investigations revealed that HLA-B27 antigen, rheumatoid factor and antinuclear antibody were negative. During the attacks, C-reactive protein was 11.6 mg/dl and fibrinogen was 524 mg/dl while complete blood count, erythrocyte sedimentation rate, blood biochemistry and urinary analyses were normal. An anterior-posterior plain radiogram of the pelvis showed negative findings consistent with her complaints. Pelvic magnetic resonance imaging (MRI) revealed a chronic bilateral sacroiliitis grade 2 (Fig. 1). Molecular analysis showed a homozygous M694V mutation in the Mediterranean fever (MEFV) gene.

For a period of 2 years under regular colchicine therapy, the patient experienced neither FMF attacks nor low back pain with no positive physical findings of sacroiliitis.

Discussion

Sacroiliitis due to FMF is very rare in paediatrics and is mostly HLA-B27 negative. Chronic arthritis of FMF, defined as arthritis with a duration of more than 30 days, occurs in 5% of FMF patients particularly affecting one joint, continues for several months and remits without destruction [6].

The patient's diagnosis depended on the clinical course, gene mutation analysis and favourable response to colchi-

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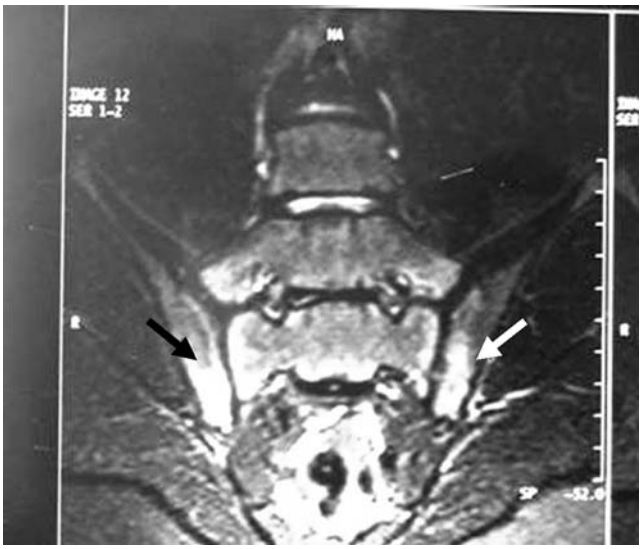

Fig. 1 Pelvic MRI, revealing marked sclerosis and erosion changes obscuring one-third of the sacroiliac joint (*arrows*), which is accepted as grade 2

cine treatment. Diagnostic criteria of FMF are recurrent fever, serositis, ethnicity, family history and response to colchicine therapy [4]. Also the MEFV gene has been shown to be a useful diagnostic tool implicated in atypical clinical presentations, including patients who do not satisfy clinical criteria [3, 5]. Patients usually have short abdominal attacks without fever with full remission after the attack, which is a feature of an incomplete attack. The simplified criteria set by Livneh et al., who used an incomplete abdominal attack as a major criterion, solved our diagnostic dilemma [4].

There are very few reports of sacroiliitis associated with childhood FMF in the literature. Sacroiliitis of an FMF case without any findings on pelvic X-ray was reported for the paediatric age group and was diagnosed by computed tomography (CT) [1]. Contrast pelvic MRI was performed since our patient had inflammatory back pain despite NSAID treatment and negative X-ray findings. Pelvic MRI has been shown to be superior to conventional radiography, scintigraphy and CT [2].

This patient went undiagnosed for almost a decade. This treatable disease should not be left undiagnosed, as missing the diagnosis can have severe consequences. Special efforts have to be made to diagnose FMF, particularly when the ethnic background is appropriate.

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