SACROILIITIS IN FAMILIAL MEDITERRANEAN FEVER*
An Unusual Presentation in Childhood

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Familial Mediterranean fever (FMF) is an autosomal recessively transmitted disease characterized by attacks of fever and serositis. The course of arthritis, which is a common manifestation of FMF, is generally benign. Sacroiliitis due to FMF has been reported by several authors, but all the patients described so far had roentgenographic abnormalities, and most of them were adult cases. Here we report the youngest FMF patient with sacroiliitis without any abnormality on sacroiliac x-ray. She is also the first FMF patient in whom sacroiliac involvement was diagnosed by computed tomography (CT) in childhood. It is concluded that CT is a useful technique for the early diagnosis of destructive arthritis in FMF patients even in early childhood.
Key words: familial Mediterranean fever, sacroiliitis, childhood, computed tomography.

Familial Mediterranean fever (FMF) is an autosomal recessive disease of unknown origin characterized by recurrent and self-limited episodes of fever accompanied by peritonitis, pleuritis and arthritis[1,2]. FMF attacks are usually short in duration, typically lasting 12-72 hours, but protracted episodes of arthritis lasting approximately one year, mainly affecting the large joints of the lower extremities, are also seen[3]. The course of arthritis is typically benign without any destruction or incapacity, but cases with destructive arthritis have also been reported in the literature[4,5].

In 25 percent of FMF patients, arthritis is the initial symptom. HLA-B27-negative sacroiliitis has been reported in the literature[6-8], but the diagnosis of destructive sacroiliitis has been based on roentgenographic findings in all the patients previously reported[4,6,7,9]. Destructive sacroiliitis presents a considerable problem, sometimes requiring surgical treatment[5]. The diagnosis may be difficult and, in cases presenting with arthritis only, this may cause a delay in the specific therapy of FMF.

We report an FMF patient with HLA-B27-negative destructive sacroiliitis without any abnormality on conventional x-ray studies.

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Case Report

A $5^1/_2$ year-old Turkish girl was admitted to Hacettepe Children's Hospital in January 1994 for evaluation of hip pain and intermittent fever with attacks of abdominal, chest and lower extremity joint pain At $1^1/_2$ years of age, her left ankle became swollen and painful together with systemic fever lasting two days. Subsequently, she suffered from intermittent attacks of fever with severe abdominal and chest pain, sometimes with arthritis that ceased spontaneously in two or three dyas. Duration of symptom-free intervals were between three weeks and six months. The patient was also complaining of continuous right hip pain for approximately three years, which was aggravated during the attack periods. Family history revealed similar complaints in a female cousin on her father's side, and her father's uncle was diagnosed as end stage renal failure due to amyloidosis at the age of 44.

On examination, she was at the 25^{th} percentile for weight and 50^{th} percentile for height; her blood pressure was 100/65 mmHg. Minimal atrophy of the right leg was observed; otherwise, physical examination including ophthalmological evaluation was normal.

Complete blood cell count and the results of urinalysis, electrolytes, blood urea nitrogen, creatinine concentrations, erythrocyte sedimentation rate and fibrinogen were all normal. Antinuclear antibodies, C-reactive protein and rheumatoid factor tests were negative. Repeated values of erythrocyte sedimentation rate, fibrinogen (normal: 200-400 mg/dl) and C-reactive protein (normal: < 2 mg/dl) levels during an attack were: 80 mm/hr, 518 mg/dl and 8.3 mg/dl, respectively. Although there was no abnormality on her sacroiliac joint roentgenogram (Fig. 1), a computed

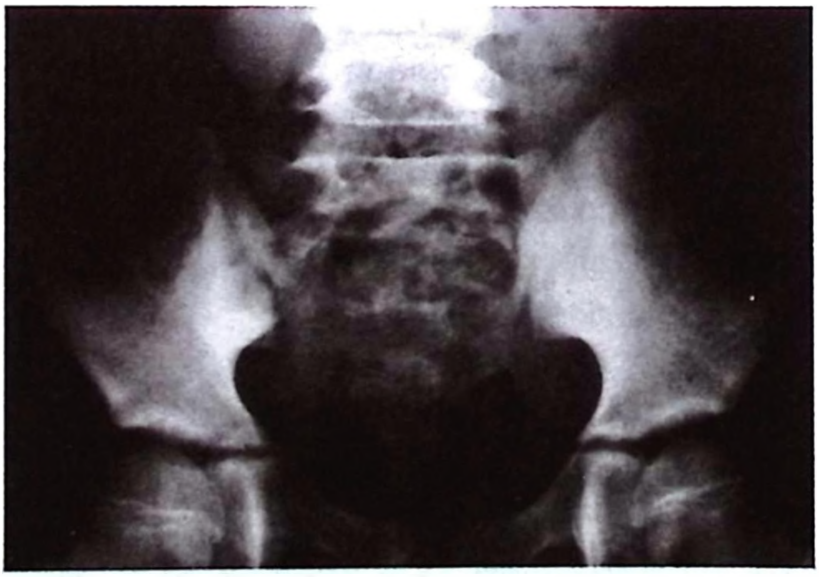

Fig. 1: Anteroposterior radiogram of pelvis is unremarkable except for the incidental spina bifida noted at the L-5 vertebrae.

tomography (CT) of the sacroiliac joints was performed since the patient had severe hip pain. Irregularity in the iliac aspects of the sacroiliac joints, more pronounced in the right, was demonstrated, indicating bilateral sacroiliitis (Fig. 2). HLA-B27 was not present. Colchicine therapy (1 mg/day orally) was started, and responded well to the therapy. The patient has been free of symptoms, including continuous hip pain, since July 1994.

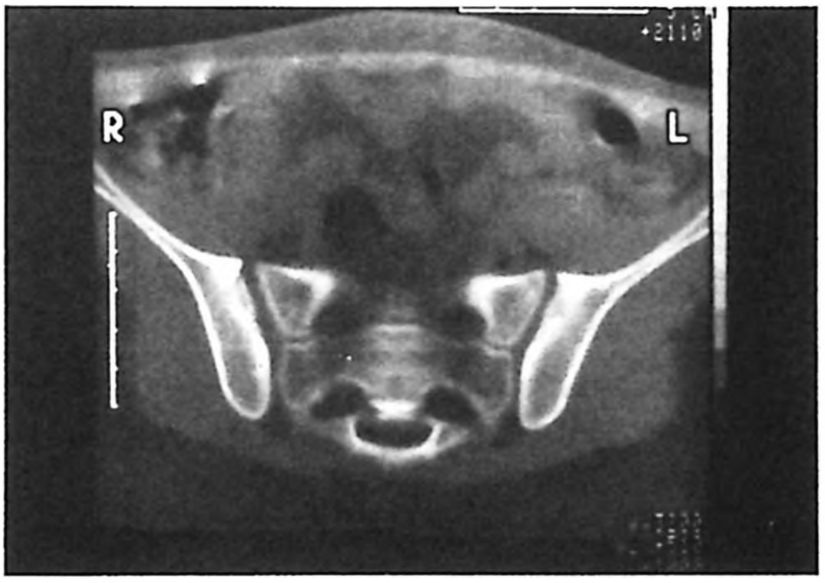

Fig. 2: Axial tomography section through the sacroiliac joints demonstrates the widening of the joint space on the right. Irregularity in the iliac aspect of the sacroiliac joints, more pronounced on the right, indicates destruction of the articular surfaces.

Discussion

Familial Mediterranean fever is a lifelong disease, usually appearing after the first two years of life. The diagnosis is mainly based on clinical findings together with a positive family history and exclusion of any other disorder that may cause similar attacks[1,8]. Arthritis of FMF manifests itself as an episodic monoarthritis or oligoarthritis of large joints that appears in childhood or adolescence, and may precede other manifestations of the disease. Although the episodes are short and self-limited (days to weeks) in nature, five percent may occasionally last for several months and even up to one year[3,4]. Permanent joint damage is a rare finding, but transient osteoporotic periarticular changes are reported[3,9]. Sacroiliac joint involvement due to FMF has been reported, especially in adult patients, with the diagnosis based on roentgenographic abnormalities[4,10]. There are only two reports of sacroiliitis associated with childhood FMF in the literature, with the youngest case reported so far being ten years old[6,7].

Sacroiliitis is commonly seen in Reiter's syndrome, ankylosing spondylitis, Crohn's disease, ulcerative colitis, Whipple's disease and psoriatic arthritis. In these disorders, HLA-B27 positivity is higher than in the normal population[11]. However,

HLA-B27 is not present in FMF patients with sacroiliitis, as was the case with our patient. She complained of a continuous hip pain aggravated during the periods of fever and abdominal pain. The roentgenographic appearance of the sacroiliac joints was unremarkable. A CT examination was performed in order to explain the atypical complaints of the patient, and destructive changes were observed which confirmed the diagnosis of sacroiliitis. The patient was diagnosed as FMF with sacroiliac joint involvement based on the clinical course of her disease: recurrent fever accompanied by abdominal and joint pain history with elevated acute phase reactants during the attacks, together with a positive family history and her ethnic origin. All complaints, including continuous hip pain, resolved after colchicine treatment.

This is the youngest FMF patient with sacroiliitis reported so far. All previously reported cases were diagnosed by x-rays. Our patient is the first case diagnosed by CT as sacroiliitis due to FMF in the absence of pathologic findings on x-ray. We conclude that CT is a useful technique for the early diagnosis of destructive arthritis in FMF patients with long-standing joint involvement.

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