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CASE REPORT

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Behçet's disease and sacroiliitis in a child

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Abstract We describe the case of a Japanese girl who suffered from both Behçet's disease (BD) and bilateral sacroiliitis. This patient was diagnosed with BD at 8 years of age, and began to complain of buttock pain when she was 10 years old. X-rays of her pelvis showed bilateral grade 3 sacroiliitis, and bone scintigraphy showed an increased uptake in the sacroiliac joints. Movements of the spine and chest expansion were not restricted. HLA typing was positive for B44 and DR4, but negative for B27. The patient was not diagnosed as having seronegative spondyloarthropathy. This case indicates that erosive sacroiliitis is an intrinsic condition of BD.

Key words Behçet's disease (BD) · Child · Sacroiliitis · Spondyloarthropathy

Introduction

Behçet's disease (BD) is known as a multisystem illness involving the skin, joints, gastrointestinal tract, blood vessels, and central nervous system, as well as other sites. This disease is rare, but it is most frequently encountered in Eastern Mediterranean countries along the Silk Road, and in the Far East, especially Japan.[1] BD is very uncommon in children, and is often unrecognized because of the prolonged interval between the onset of the first symptoms and a definitive diagnosis. X-ray findings in BD are usually unremarkable, although in some adult BD patients with back pain, erosive sacroiliitis has been reported.[2,3] This report concerns a child suffering from both BD and sacroiliitis, which to our knowledge has never been reported before.

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Case report

An 8-year-old Japanese girl was first referred to our outpatient clinic in August 1996 with low-grade fever, oral aphthae, genital ulcers, perianal ulcers, and arthritis of the left ankle and right knee joints. The patient had had oral aphthae from 2 or 3 years of age. There were no abnormal ocular findings. Plain radiograms showed no abnormalities in the ankle or knee joints. Hematological test results were: hemaglobin (Hb) 11.0%; hematocrit (Hct) 34.3 g/dl; WBC 10 800/mm^3; Plt 318 000/mm^3; C-reactive protein (CRP) 7.1 mg/dl; erythrocyte sedimentation rate (ESR) 107 mm/h. Biological tests showed: IgG 2938 mg/dl; IgA 495 mg/dl; IgM 430 mg/dl; IgD 14 mg/dl; C_3 122 mg/dl; C_4 65 mg/dl. Rheumatoid factor was negative. Urinalysis and coagulation tests were normal. No microorganisms could be found in the urine, and a pathergy test was negative. HLA-A,B,C typing tests were positive for A33 and B44. The phenotype of the HLA-DR locus was DR4. A diagnosis of BD was made according to the International Study Group criteria for BD.[4] Initially, the patient received prednisolone at 20 mg/day for a short period, and her clinical symptoms and laboratory abnormalities began to improve. Therefore, the dosage of prednisolone was tapered of and then ceased.

In 1998, the patient began to complain of buttock pain. A physical examination showed no restriction of movement of the lumber spine or chest expansion. A plain radiogram of the pelvis showed unequivocal erosion, especially in the iliac edge, and a widening of the right sacroiliac joint. These findings corresponded to grade 3 sacroiliitis according to the New York criteria (Fig. 1).[5] Bone scintigraphy revealed an uptake of radioactivity in the right sacroiliac joint (Fig. 2), while laboratory tests showed inflammatory changes (ESR 25 mm/h, CRP 1.2 mg/dl). The patient continued to complain of pain in the left buttock. Radiological examinations 8 months later showed bilateral sacroiliitis (Fig. 3). Her

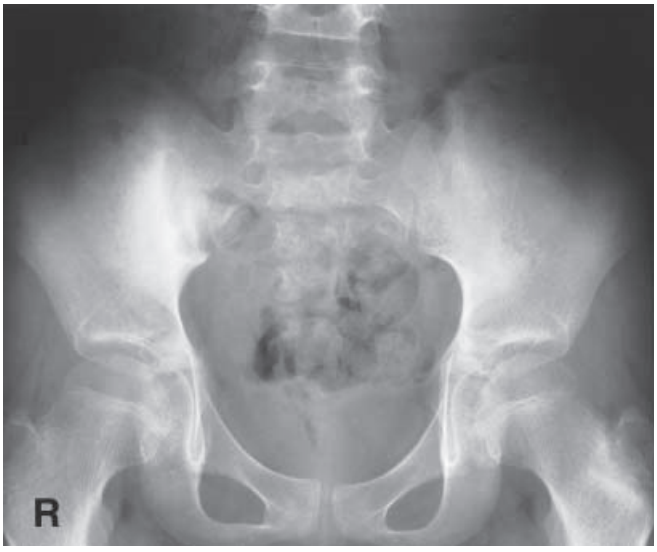

Fig. 1. Radiograph of the pelvis, taken in 1998 (10 years of age), showing obvious erosion in the right (*R*) sacroiliac joint

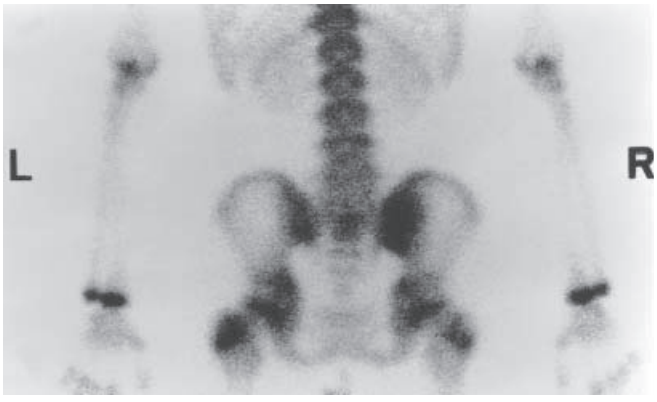

Fig. 2. Bone scintigraphy, taken in 1998, revealed increasing uptake in the right sacroiliac joint. *L*, left

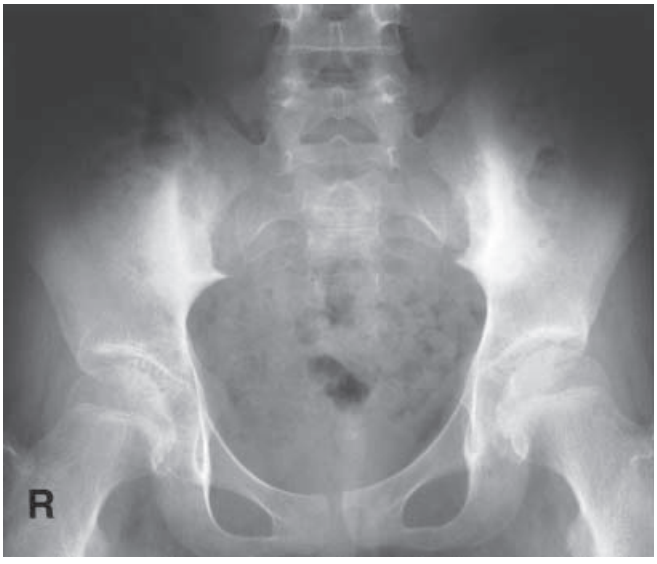

Fig. 3. Radiograph taken in 1999 (11 years of age), showing bilateral grade 3 sacroiliitis

family history was negative for BD, rheumatoid arthritis, and spondyloarthropathy. The patient did not meet the New York criteria for ankylosing spondylitis (AS),[5] or the European Spondylarthropathy Study Group criteria for definite forms of seronegative spondyloarthropathy.[6]

The patient gradually improved with nonsteroidal anti-inflammatory drugs, and acute-phase reactants decreased to within the normal range.

Discussion

Patients with BD frequently suffer from recurrent oligo- or polyarthritis.[2,3] Knee, wrist, and ankle joints tend to be affected, although the arthritis is usually nondestructive. Pivetti-Pezzi et al.[7] reported that arthritis was equally distributed in children and adults (62.5% vs. 74.6%). Another study reported that articular involvements were observed in 55% of BD patients, and the affected sites were mainly large joints in both children and adults.[8] Some reports have indicated an increased prevalence of erosive sacroiliitis in patients with BD,[2,9] and Caporn et al.[2] reported that in 14 patients with BD, 7 (50%) had mild erosive sacroiliitis. In contrast, other reports have indicated that the prevalence of sacroiliitis in BD does not differ from that in the general population.[10,11] There is no known correlation between BD and HLA-B27. There has been some debate about whether BD can be included among the seronegative spondyloarthropathies, as several adult cases of coexisting BD and ankylosing spondylitis or seronegative spondyloarthropathy have been reported.[12–18] Characteristically, the sacroiliitis was radiologically mild to moderate, and no case showed severe destruction or ankylosis, while 70% had phenotype B27. BD is very rare in children, and our case is the first report of sacroiliitis in a child with BD.

Yazici et al.[19] suggested that there were intra- and interobserver variations in the diagnosis of sacroiliitis. Further, the prevalence of osteoarthrosis in sacroiliac joints is reported to be relatively high, and findings such as erosion, sclerosis, narrowing, osteophytes, and subchondral cyst have been observed in young adults.[20,21] Our patient had erosive bilateral sacroiliitis, which was confirmed by both plain radiography and bone scintigraphy. The degenerative changes were not great and the HLA-B27 antigen was negative. Therefore, we suspect that erosive sacroiliitis is an intrinsic feature in BD patients.

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