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CASE REPORT

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Behçet's disease with severe destructive arthritis

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Abstract Behçet's disease is a systemic disease characterized by oral aphta, genital ulcers, and ocular lesions, and arthritic manifestations also appear to be common. However, this disease rarely produces loss of function or deformity in arthritic joints. We report the case of a 52-year-old woman with Behçet's disease who had a history of recurrent oral aphta, genital ulcerations, and intestinal lesions for almost 30 years. When she was about 30 years old, she began to notice significant morning stiffness and polyarthritis, and progressive destructive arthritic changes in the bilateral fingers, wrists, and left ankle. Behçet's disease with severe destructive arthritic changes is rare, and the underlying mechanism is still unknown.

Key words Arthrodesis · Arthropathy · Behçet's disease · Rheumatoid arthritis (RA)

Introduction

Behçet's disease, originally described as a triad of ocular inflammation, and oral and genital ulcerations, is better understood as a multisystem disease involving the skin, joints, central nervous system, large bowel, and peripheral veins. Arthritic manifestations appear common in Behçet's disease. Arthropathy, which occurs in at least half of all reported patients, is usually monoarticular or oligoarticular. It mainly affects the joints of the lower extremities, recurs occasionally, and is rarely chronic.[1] The arthritis is not deforming, and destructive joint lesions are rarely observed. In rare cases, loss of articular cartilage and pannus formation with erosive damage has been reported, but the underlying mechanism is still unknown.[2] In this paper we describe a case of Behçet's disease with severe destructive arthritic changes.

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Case report

The patient, a 52-year-old woman, had experienced recurrent oral aphta and genital ulcerations since the age of 22 years. She was operated on for hemorrhage from intestinal lesions at another hospital when she was 30 years old. Subsequently, cutaneous erythema and soft-tissue swelling often developed on both legs. When she was 41 years old and had hemorrhaged from ulcerations in small intestinal lesions, she was diagnosed as having Behçet's disease based on the International Study Group Criteria.[3] She was then started on prednisolone (15 mg/day). The dose of prednisolone was gradually reduced to a maintenance dose of 7.5 mg/day, and except for polyarthritis, her symptoms were under control. Ocular and neurological examination showed no signs of eye lesions or nervous system involvement at that time. However, when she was around 30 years old, she began to notice significant morning stiffness and polyarthritis. She then complained of progressively worsening pain, especially in the bilateral wrists and left ankle. Salazosulfapyridine was added to her treatment after the diagnosis of rheumatoid arthritis (RA) was established. However, she still complained of pain, especially in her left ankle joint, despite medical treatment, and gradually became unable to walk because of the severe pain.

In December 1997, when she was 51 years old, she was referred to us for treatment of the ankle joint. Pain in her left ankle joint did not disappear despite medication, rehabilitation, and an ankle brace. She was admitted to this hospital for surgery on the left ankle joint on July 6, 1998. The range of motion in the left ankle joint in dorsiflexion was 0°, and in plantar flexion it was 45°. A radiograph of the left ankle showed destructive arthritic changes and the disappearance of the joint space (Fig. 1a). A radiograph of the

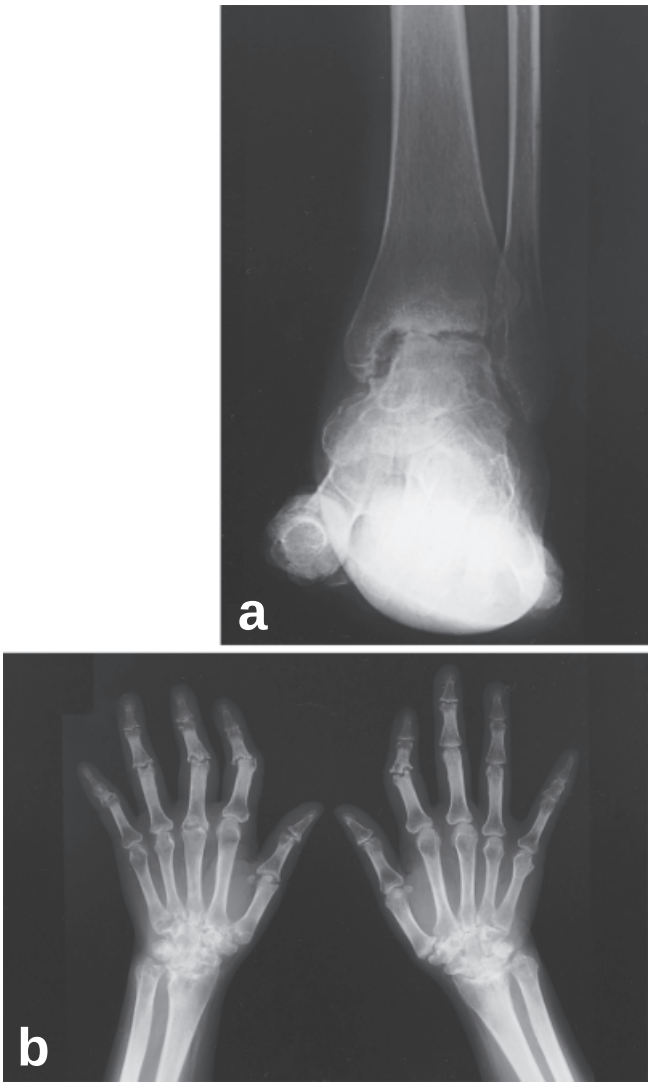

Fig. 1. **a** Radiograph showing destructive arthritic changes in the left ankle. The partial lesions in the subchondral bone were sclerotic. **b** Radiograph of bilateral hands and wrists showing destructive changes, especially in the bilateral carpal bones and some proximal interphalangeal joints

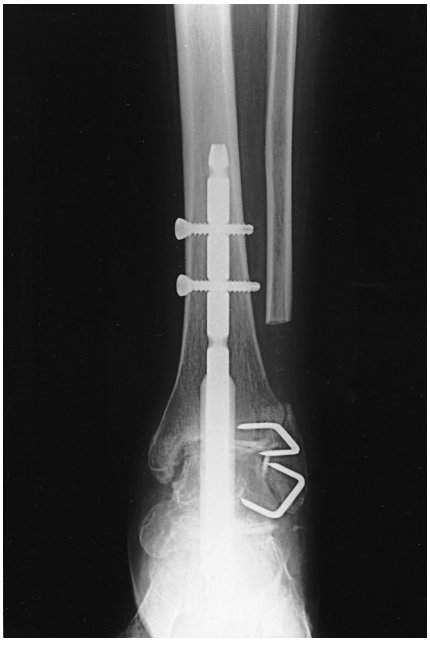

Fig. 2. Ankle arthrodesis surgery was performed following the lateral approach to the ankle and using internal fixation with an intramedullary nail with four distal fins from the bottom of the foot

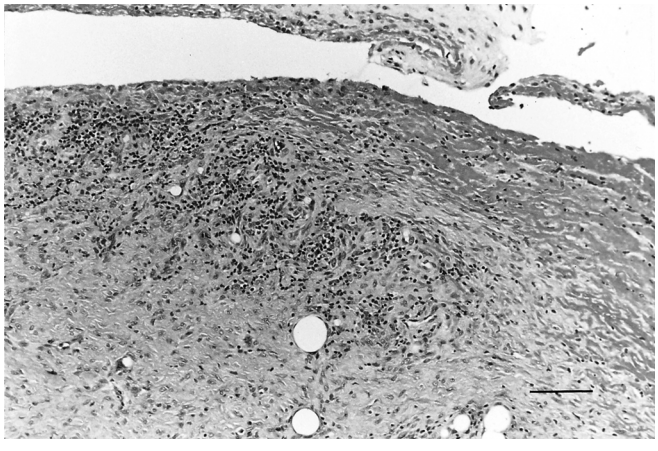

Fig. 3. Hematoxylin–eosin-stained section of synovia obtained from the left ankle, showing the inflammatory infiltration of lymphocytes and sedimentation of fibrin, and the disappearance of articular cartilage. The surface is covered by reparative fibrocartilage. *Bar* 100 μ

bilateral hands and wrists also revealed destructive changes, especially in the carpal bones and proximal interphalangeal joints (Fig. 1b). Laboratory studies showed a white blood cell (WBC) count of 6300/mm^3, Hb 11.6 g/dl, erythrocyte sedimentation rate (ESR) 23.6 mm/h, C-reactive protein (CRP) 0.3 mg/dl, rheumatoid factor latex (−), RAPA $<$ $\times$40, and antinuclear antibodies $\times$20. Other investigations, such as platelet count, blood urea nitrogen (BUN), creatinine, alkaline phosphatase, and transaminase levels were within the normal range. Her human leukocyte antigen (HLA) phenotype was HLA-A2, A-24(9), B-7, B-46, CW-1, CW-7, DR-1, DR-8, and DQ-1. A chest radiograph and electrocardiogram were normal.

Ankle arthrodesis surgery was performed following the lateral approach to the ankle and using internal fixation with an intramedullary nail with four distal fins from the bottom of the foot (Fig. 2). Macroscopically, our impression was that the tissues in the joint had been destroyed. There was no articular cartilage or synovial fluid in the left ankle joint. A specimen of synovia taken from that joint showed the inflammatory infiltration of lymphocytes and sedimentation of fibrin, which indicated a nonspecific inflammatory reaction, and the disappearance of articular cartilage, with the surface being covered by reparative fibrocartilage (Fig. 3). The patient used a non-weight-bearing short leg cast for 2 weeks after the operation. She then began to walk with crutches, and her cast was removed. The patient could walk without any support 4 weeks after the operation. The

severe pain in her left ankle diminished, and she was discharged on August 1, 1998. To date, she feels no pain in her left ankle and can walk normally.

Discussion

In most studies, arthritis is considered to be a manifestation of Behçet's disease. The prevalence varies among the populations studied, and ranges from 40% to 60%.[4] Park[5] reported that joint manifestations appeared in 97% of patients, while Benamour et al.[6] reported that joint manifestations were present in more than half the patients in a series that included 601. However, this disease rarely produces a loss of function or deformity of the articular joints. Radiological findings are usually unremarkable except for erosive changes that are limited to a minority of patients.[2,7] In most cases (76.4%), the articular symptoms are short-lived, and chronic arthritis is rare.[5] Mason and Barnes[8] reported that a mean of 5.5 joints were affected per patient, whereas Yurdakul et al.[2] reported that most patients had monoarticular arthritis. Kim et al.[9] reported that most patients had 2–4 joints affected during an episode of arthropathy, and of these, 13% of patients reported monoarticular attacks as well. Monoarthritis and oligoarthritis were seen in 16.2% and 11.8% of the cases, respectively, and polyarthritis involving the large limb joints and the small joints of the hands and feet was found in 17.1% of cases.[6] Monoarticular involvement is more common, particularly in the knee (50%), wrist (23%), ankle (11%), and elbow (5%) joints.[10] Moral et al.[10] also observed that the pattern of involvement was oligoarticular in 75%, monoarticular in 23%, and polyarticular in 2% of patients.

Destructive joint lesions are not usually observed. However, loss of cartilage and pannus formation with erosive damage have been reported in rare cases.[2,4,8,11]

Benamour et al.[6] described eight unusual patients who had polyarthritis with deformities and/or destruction, including two patients who also met the criteria for RA, among 340 cases with joint manifestations identified among 601 cases of Behçet's disease seen over a 15-year period.

In Behçet's disease, the latex rheumatoid factor test and an antinuclear antibody estimate were reported to be negative,[2,9,12] and the laboratory parameters of ESR and CRP were reported to be elevated during the arthritis.[4,9,13] Our patient also met the American Rheumatism Association 1987 revised criteria for the classification of RA. However, laboratory investigations showed that she was negative for rheumatoid factor, and neither ESR nor CRP was elevated at the time of the operation. In this case, severe destructive arthritic changes were observed not only in the hands and wrists, but also in the left ankle, and the radiological findings were similar to those for RA. Therefore, we must consider whether this patient should have been diagnosed with Behçet's disease associated with RA.

Usually, synovial fluid analysis of the arthritic joint in Behçet's disease is reported to show a cloudy fluid with high viscosity, and the WBC ranges from 300 to 36200/mm^3 with polymorphonuclear predominance,[2] or from 1100 to 17300/mm^3.[14] The protein and glucose contents are normal or lower than in blood. Histological examination shows a superficial inflammatory reaction on the synovial membrane, and increased vascularity with perivascular lymphocyte infiltration. It is very difficult to establish a diagnosis from the histology alone.[15] We could not obtain synovial fluid from the left ankle of this patient, but we observed chronic nonspecific and hemorrhagic changes in the synovium and complete disappearance of the articular cartilage, with the surface being covered by reparative fibrocartilage.

In conclusion, although the pathological condition that caused destructive polyarthritis in this patient remains unclear, we diagnosed Behçet's disease with severe destructive arthritic changes. The disease pattern of Behçet's disease, which has mainly gastrointestinal symptoms, is slightly different from the patterns of other systemic manifestations of this disease. We will follow this patient over the long term.

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