

Case report

Ankylosing spondylitis with predominant involvement of the cervical spine in a woman with Behçet's syndrome

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SUMMARY The case of a 45 year old woman affected by both Behçet's syndrome (BS) and ankylosing spondylitis (AS) is reported. Cervical spine involvement was observed long before the appearance of the sacroiliac and lumbar changes.

Key words: sacroiliitis, seronegative spondarthritides, computed tomography.

Roentgenographic evidence of sacroiliitis is an indispensable condition for the diagnosis of ankylosing spondylitis (AS).¹ Nevertheless, cases with spinal features typical of AS and normal sacroiliac joints have also been reported.²⁻⁴

Cervical changes are not infrequent in patients with AS.^{5,6} These tend to occur later than thoracic and lumbar features and are more prevalent in women.^{5,6} Abnormalities in the cervical spine and sacroiliac joints, with sparing of the thoracic and lumbar segments, are also relatively more prevalent in women.⁶

The inclusion of Behçet's syndrome (BS) among the seronegative spondarthritides is under consideration.⁷⁻¹¹ Some investigators have reported an increased prevalence of sacroiliitis and AS in patients with BS,⁸ but others have found a prevalence that does not differ from that of the general population.⁹⁻¹¹

The case described here illustrates the unusual course of AS, involving predominantly the cervical spine, in a woman with BS. Only recently have lumbar spine and sacroiliac joint features become evident.

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Case report

A 45 year old woman was referred to the rheumatology unit in June 1984 after suffering for two years from neck pain and decreasing range of movement of the cervical spine. The pain was most prominent in the early morning and was relieved by exercise.

Her medical history included episodic inflammatory low back pain and stiffness in the previous 20 years, four episodes of disseminated painful oral ulceration, one episode of erythema nodosum, and numerous episodes of disseminated papulopustular skin lesions in the previous 10 years. She denied any history of psoriasis, urethritis, conjunctivitis, peripheral arthritis, gastrointestinal, neurological, or cardiovascular symptoms.

Her family history was negative for seronegative spondarthritides, BS, or idiopathic recurrent aphthous stomatitis.

Physical examination showed a severe restriction of movement of the cervical spine in all directions. Extension, flexion (modified Shober test 19.5 cm), and lateral bending of the lumbar spine were slightly limited. Chest expansion was 4 cm. The skin prick test (pathergy test) was negative.

The only aspect of laboratory evaluation worthy

of note was the erythrocyte sedimentation rate of 30 mm/1st h (Westergren). HLA typing was positive for A2, A3, Bw51, and B12 antigens.

Cervical spine radiography showed anterior syndesmophytic bridging extending from C4 to C6, reduction in height of the C6-7 intervertebral disc, and narrowing, irregularity, and sclerosis of the apophyseal joints, all characteristic features of AS (Fig. 1). Radiographs of the sacroiliac joints and the thoracic and lumbar spine were normal.

During the last three years the patient had recurrent episodes of disseminated papulopustular skin lesions on the trunk and upper and lower limbs, and recurrent episodes of painful genital and buccal aphthous ulceration. The pathergy test was persistently negative.

During the last 18 months her inflammatory low back pain was persistent and unresponsive to non-steroidal anti-inflammatory drugs, including



Fig. 1 Lateral radiograph of the cervical spine, taken in June 1984, showing syndesmophytes extending from C4 to C6, apophyseal joint sclerosis, irregularity, and narrowing, together with a reduction in height of the C6-7 intervertebral disc.

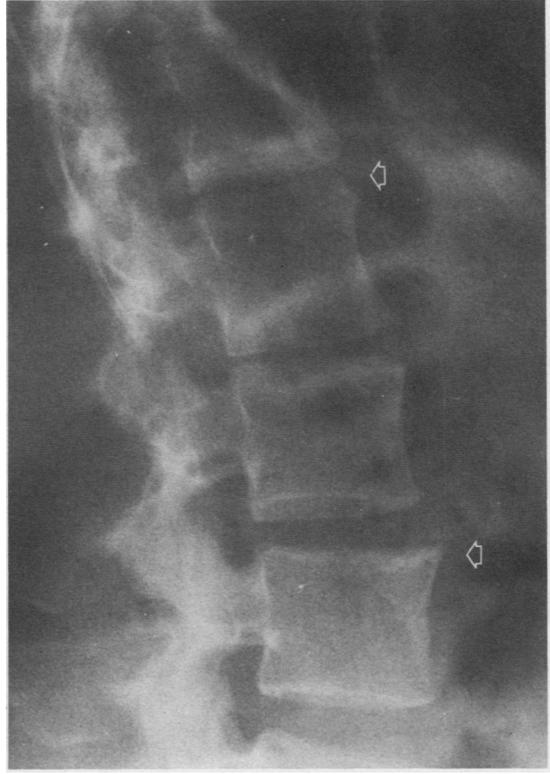


Fig. 2 Lateral radiograph of the lumbar spine, taken in November 1986, showing erosion surrounded by sclerosis ('Romanus lesion') of the upper corner of the second lumbar vertebra and proliferative spur of new bone of the fourth.

phenylbutazone. Movement of the lumbar spine decreased despite physical treatment. In June 1987 the modified Shober test gave a result of 18.5 cm. Chest expansion had decreased to 2.5 cm.

Radiological examination in November 1986 showed subchondral osteitis of the upper corner of the second and fourth lumbar vertebrae (Fig. 2), typical of AS, as well as an evolution of the features in the cervical spine (Fig. 3). The sacroiliac joints showed a mild sclerosis (Fig. 4a). Computed tomography, performed at that time, showed sclerosis and a slight irregularity in the left sacroiliac joint and severe sclerosis and erosions in the right joint (Fig. 4b).

Discussion

Our patient meets the diagnostic criteria for BS as suggested by Mason and Barnes,¹⁰ O'Duffy,¹² and

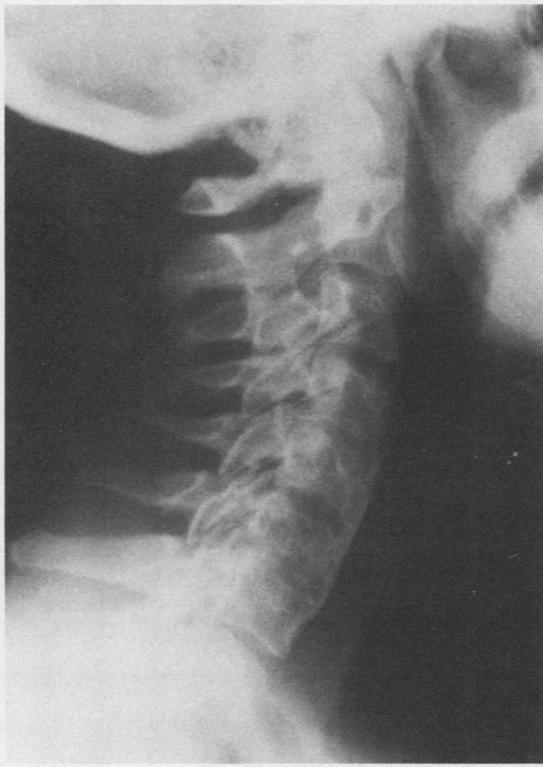


Fig. 3 Lateral view of the cervical spine, obtained in November 1986, illustrating the evolution of the features shown in Fig. 1.

the Japanese Committee on BS.¹³ She is positive for the HLA-B5 antigen (Bw51 split), which has been found to be associated with BS in Japanese,¹⁴ Turkish,¹⁵ and other Middle Eastern and Mediterranean populations, including Italians.¹⁶ No significant association has been observed in English¹⁷ or North American Caucasoid patients.¹⁸ According to Lehner and Batchelor only eye involvement is associated with the HLA-B5 antigen in English patients, whereas mucocutaneous involvement is associated with HLA-B12.¹⁷ Our patient has a 'mucocutaneous type' of BS and is positive for HLA-B12.

Our patient also meets the New York criteria for AS.¹ When we first saw the patient in 1984 she had radiological features typical of AS only in her cervical spine. The lumbar and the sacroiliac joints were normal. Although it was possible to exclude the other conditions that can cause cervical spine ossification and hyperostosis,¹⁹ we were reluctant to diagnose seronegative spondyloarthropathy. Radiological follow up has shown the appearance of typical features of AS in the lumbar spine and equivocal findings in the sacroiliac joints. Sacroiliac changes were better shown by computed tomography, which has been found to be useful in the early diagnosis of sacroiliitis.²⁰

The inclusion of BS among the seronegative spondyloarthritides is still being debated.⁷⁻¹¹ Dilşen *et al* have recently re-evaluated 331 Turkish patients with BS for sacroiliitis and AS.⁸ Thirty three patients (10%) were found to have AS and 112

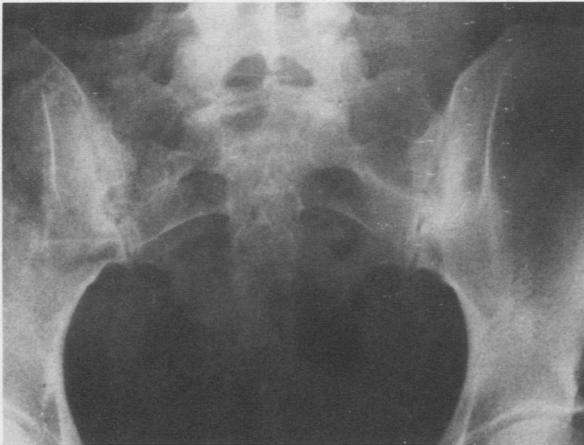


Fig. 4a



Fig. 4b

Fig. 4 (a) Conventional radiograph of the sacroiliac joints, taken in November 1986, showing sclerosis in both joints. (b) Computed tomography through the synovial portion of the sacroiliac joints, obtained at the same time, showing severe sclerosis, and narrowing and erosions of the right sacroiliac joint. The left joint shows a milder sclerosis and irregularity of the joint space.

(34%) sacroiliitis. None of the 92 controls had AS and only 5 (5%) had sacroiliitis. Patients with both AS and BS had a significantly higher incidence of both the HLA-B5 and HLA-B27 antigens than controls. Other authors have reported that there is no association between BS and AS.⁹⁻¹¹ Surprisingly, Yazici *et al* did not find any increase in the prevalence of sacroiliitis, in comparison with controls, in their Turkish series of BS.⁹ Only one of their 184 patients with BS presented definite AS. They have recently suggested that 'high observer variation' in interpreting the conventional antero-posterior view of the sacroiliac joints may account for the reported association between BS and sacroiliitis.²¹

A single case, of course, does not permit us to take sides in the controversy. We wish to emphasise, however, that, if BS and AS are really associated diseases, then secondary AS can begin in the spinal column rather than in the sacroiliac joints and may affect the cervical segment earlier and more severely than the lumbar one, as sometimes happens in primary AS, especially in women.

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