Case Report

Late Onset Peripheral Seronegative Spondyloarthropathy: Report of Two Additional Cases

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Abstract. Two more cases of late onset peripheral seronegative spondyloarthropathy (SpA) are reported. Like the patients reported by Dubost and Sauvezie, they had extensive pitting edema of the lower limbs, constitutional symptoms, elevated erythrocyte sedimentation rate and minimal involvement of the axial skeleton, constitutional symptoms and an elevated erythrocyte sedimentation rate (ESR)1.

We describe 2 more cases of this syndrome.

CASE REPORTS

Case 1. A 64-year-old man was admitted to the 5th Internal Medicine Department of Pisa Hospital on May 31, 1991. Ten days before, he had developed a temperature together with painful swelling of his right lower limb, suggesting acute venous thrombosis.

Examination showed synovitis of the right ankle together with an extensive pitting edema of his right lower limb and dactylitis of his left great toe. There was no limitation of chest expansion or spine movement. He was obese and hypertensive.

His past medical history revealed short, mild episodes of inflammatory low back pain in the last 15 years. He denied any history of peripheral arthritis, peripheral enthesitis, conjunctivitis, uveitis, diarrhea, urethritis, psoriasis or cardiac symptoms. His family history revealed that his father had had ankylosing spondylitis (AS).

Laboratory evaluation showed an erythrocyte sedimentation rate (ESR) (Westergren) 112 mm/h, C-reactive protein (CRP) 49 mg/l (normal < 5), alpha-1-acid glycoprotein 1.64 g/l (normal 0.55-1.40), alpha-2-globulin 12.2 % (71 g/l total protein), fibrinogen 7.4 g/l (normal 1.5-4.5), triglycerides 229 mg/dl (normal < 190), gamma glutamyl transferase (GGT) 243 U/l (normal < 50), aspartate aminotransferase (AST) 54 U/l (normal < 45), HLA typing showed A1, A24, B8, and B27 antigens.

Spine radiographs revealed findings typical of diffuse idiopathic skeletal hyperostosis (DISH) in his dorsal spine: a continuous flowing ossification along the right anterolateral aspect of the vertebrae and disc spaces, together with radiolucencies between the posterior aspect of the new bone and the subjacent anterior aspect of the vertebral body (Figure 1). Mild changes of DISH were present also in his cervical and lumbar spine. Features typical of AS were not seen in any part of the spine. An anteroposterior view of his pelvis showed grade 3 bilateral sacroiliitis. Computed tomography revealed an anterior capsular bridging typical of DISH, in addition to erosion, joint space narrowing, ankylosis and sclerosis typical of AS (Figure 2). Radiographs of his knees and feet revealed mild new bone formation with no erosion at the sites of attachment of the quadriceps tendon, plantar fascia and Achilles tendon.

A few days after admission, he developed pain and swelling with no obvious effusion at his right knee, and pain at the insertion of the left plantar fascia.

He was given diclofenac at a dose of 150 mg/day. His arthritis improved and he was discharged on June 15. Laboratory evaluation at the end of July, 1991 revealed normal acute phase reactants. Examination was negative. He has remained asymptomatic.

Case 2. A 66-year-old man was referred to the rheumatic disease unit in March, 1988 with an 8-month history of inflammatory spinal pain and synovitis of some joints of his upper and lower limbs.

His medical and family history were negative for SpA and other B27 associated syndromes.

Examination disclosed synovitis of the left elbow, the right wrist and the 5th proximal interphalangeal (PIP) joint of his left hand. His left foot showed extensive pitting edema extending to the distal part of his leg and pain mostly at the tarsus. Cervical and lumbar spine movement were moderately restricted, while chest expansion was normal.

Laboratory evaluation showed an ESR 38 mm/h, CRP 22 mg/l (normal < 5), serum IgA level 5.7 g/l (normal 0.5-3.5), glucose 142 mg/dl (normal 70-110). HLA typing was positive for B27 antigen.

Radiographs of the peripheral joints involved revealed only soft tissue swelling. Spurs were seen at the insertions of both Achilles tendons. Cer-
Fig. 1. Patient 1. Lateral view of the dorsal spine showing flowing ossification and radiolucent lines (arrow).

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vical (Figure 3) and dorsal spine radiographs showed a continuous flowing ossification along the anterior aspect of the vertebrae and disc spaces. Radiolucencies could be seen between the new bone and the underlying anterior vertebral body border. In the dorsal spine, the flowing bony outgrowths were most marked on the right lateral aspect of the vertebral bodies. In the lumbar spine, only mild signs of DISH were seen. No features of AS were seen at any spinal segment. Sacroiliac joint radiographs were normal, except for caudal incomplete bridging, typical of DISH.

Our patient was given naproxen at a dose of 1 g/day. Steroid injections were given in the right wrist and the left 5th PIP joint. His symptoms subsided in December, 1988 and have not reappeared. Lumbar spine movement became normal while cervical spine flexion remained restricted. ESR returned to normal in November, 1989 while CRP remained high till April, 1991. New spine and sacroiliac joint radiographs, obtained in March, 1992, showed an evolution of spinal findings of DISH with respect to those observed 4 years before. No signs of AS were seen in either spine or sacroiliac joints.

DISCUSSION

The clinical spectrum of SpA has been broadened in the last few years. Two sets of diagnostic criteria have been proposed for the classification and diagnosis of all forms of SpA, including those considered unclassified or undifferentiated. Our 2 patients meet both sets of criteria. Their form of SpA is similar to that described by Dubost and Sauvezie. Both patients were more than 50-years-old at onset and had constitutional symptoms together with an elevated ESR. Both showed peripheral oligoarthritis, together with extensive pitting edema of the lower limbs and a little or no axial skeleton involvement. Patient 1 also showed dactylitis and peripheral enthesopathy. Like the 10 patients studied by Dubost and Sauvezie, Patient 2 had symptoms for more than 12 months. Symptoms remitted in a few months in Patient 1.

According to Dubost and Sauvezie, late onset peripheral SpA differs from the remitting seronegative symmetrical synovitis with pitting edema (RS,PE) syndrome reported by McCarty, et al. Patients with RS,PE syndrome showed swelling and synovitis of the hands (rare in late onset SpA), no constitutional symptoms, high frequency of the B7 antigen and a good response to salicylates. But the distinction may not be so clearcut, since 2 of the 4 patients reported by Chouat and Le Parc with arthritis and pitting edema resembling the RS,PE syndrome were B27 positive. The 4 patients were not investigated for other features of SpA.

Another important point suggested by our 2 patients is that care must be taken in patients with late onset peripheral SpA not to mistake spinal findings of DISH for AS. DISH and SpA are completely different diseases having in common the involvement of the axial skeleton and extraspinal entheses. DISH affects middle aged and elderly patients and is often asymptomatic or associated with mild dorsolumbar pain and/or slight restriction of spine movement; AS affects young adults and produces inflammatory spinal pain and stiffness, gradual limitation of spine movement, and characteristic posture abnormality. The radiological findings of spine involvement of SpA and DISH are so different that in patients with coexisting DISH and AS, it is possible to distinguish between the changes due to the 2 diseases. The first of our 2 patients showed only findings of DISH in his spine and findings of both DISH and SpA in his sacroiliac joints, as shown by computerized tomogram. The second patient showed findings of DISH in both spine and sacroiliac joints. During the period of SpA activity 4 years ago, he suffered from inflammatory spinal pain and showed restricted cervical and lumbar spine movement, mostly due to pain. After the resolution of the active phase, he showed only restricted movement due to DISH. Findings of AS have not appeared during followup, suggesting that the spinal pain present in the active phase was due to enthesitis.


