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Case Report

Cervical spinal cord injury in sapho syndrome

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Cervical spinal fracture and pseudarthrosis are previously described causes of spinal cord injury (SCI) in patients with spondylarthropathy. SAPHO (Synovitis Acne Pustulosis Hyperostosis Osteitis) syndrome is a recently recognized rheumatic condition characterized by hyperostosis and arthro-osteitis of the upper anterior chest wall, spinal involvement similar to spondylarthropathies and skin manifestations including palmoplantar pustulosis and pustular psoriasis. We report the first case of SAPHO syndrome disclosed by SCI related to cervical spine ankylosis.

Keywords: spinal cord injury; SAPHO syndrome; arthro-osteitis; sterno-clavicular hyperostosis; spondylarthropathy; psoriasis

Introduction

In 1987, Chamot and Kahn suggested the acronym SAPHO (Synovitis Acne Pustulosis Hyperostosis Osteitis) to describe a number of peculiar bone, joint and skin lesion reports published over the last 25 years.[1] The disease is characterized by hyperostosis and artho-osteitis of the sterno-clavicular and costo-sternal joints, oligoarticular peripheral synovitis, spinal and sacroiliac changes similar to spondylarthropathies and skin manifestations including palmoplantar pustulosis, pustular psoriasis or severe acne. We describe a case of SAPHO syndrome with spinal involvement disclosed by a SCI.

Case illustration

A 74-year-old man was admitted into our institution with cervical pain and a progressive limb weakness following a road accident at low speed. In his past medical history, the patient reported psoriasis of the calf and a painless sterno-clavicular hyperostosis of unknown aetiology. Physical examination revealed a C6 incomplete (ASIA scale C) sensory-motor tetraplegia. Cranial nerves were normal.

Laboratory investigation revealed an inflammatory syndrome: erythrocyte sedimentation rate was 32 mm/h in the first hour (<10 mm/h), CRP 5.5 mg/dl (<0.5 mg/dl) and fibrinogen 521 mg/dl (180–400 mg/dl). Leucocyte count was normal. Alkaline phosphatase was 177 UI/L (90–265 UI/L). Antinuclear antibodies, rheumatoid factor and HLAB27 test were negative.

Cervical plain radiographs disclosed an ankylosed spine from the C4-C5 to the C7-D1 level and a C6-C7 transdiscal fracture extending posteriorly to the ankylosed facets (Figure 1). T1 and T2-weighted sagittal magnetic resonance images showed a fracture involving the C6-C7 disc and the adjacent vertebral endplates, a spinal cord compression and an epidural hematoma (Figures 2 and 3). Radiographs and CT-scan (Figure 4) also revealed an oblique fracture of the sternum, an arthro-osteitis of the sterno-clavicular joints, an hyperostosis of the costo-sternal joints and anterior syndesmophyte formation of the dorso-lombar spine.

The association of cervical spine lesions similar to those met in seronegative spondylarthropathies, arthro-osteitis of the sterno-clavicular joints, hyperostosis of the costo-sternal joints, anterior syndesmophytes of the dorso-lombar spine and psoriasis led to the diagnosis of SAPHO syndrome complicated by C6-C7 cervical fracture and SCI. The treatment consisted of orthosis, cervical immobilization and rehabilitation. Clinical improvement was progressive with slow recovery of the muscle strength and of vesical function. Four months after the onset, the patient was able to walk with crutches and to use hands for daily living activities. MRI control at 8

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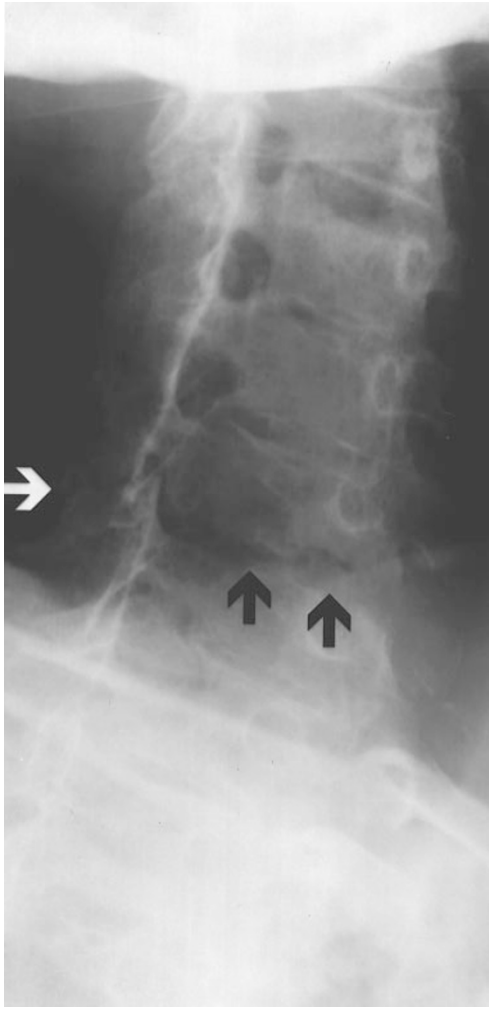

Figure 1 Right anterior oblique radiograph of the cervical spine shows a transverse fracture of the C6-C7 disc (black arrows) extending to the ankylosed posterior facets (white arrow)

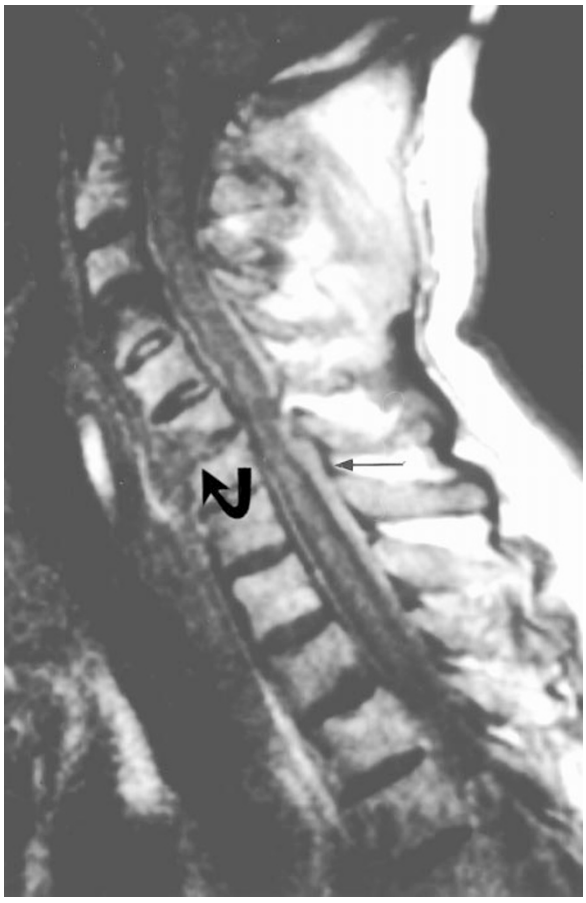

Figure 2 Sagittal T1-weighted MR image of the cervical spine: the ankylosed discs present a high signal intensity whereas the C6-C7 disc shows an intermediate signal intensity (large black curved arrow). A non-compressive epidural hematoma (straight black arrow) is also present

months showed the appearance of a progressive kyphosis of the cervical spine at the C6-C7 level and myelomalacia (Figure 5). With conservative treatment, the MRI control at 1 year showed a stabilization of the cervical kyphosis at 38°. This result was persistent 2 years after the accident.

Discussion

The fundamental component of SAPHO syndrome is an inflammatory, pseudo-infectious, usually sterile osteitis.[2] The upper anterior chest wall involvement is the most frequent manifestation occurring in 66% to 85% of the cases including sterno-clavicular and sterno-costal joints hyperostosis and arthro-osteitis.[3,4] Spinal involvement, peripheral bones osteomyelitis, peripheral arthritis, sacroliitis and enthesitis were also reported.[2,4] Skin manifestations include severe acne, palmoplantar pustulosis and pustular psoriasis. A long interval may be observed between bone involvement and skin lesion. So, in the presence of typical anterior chest wall involvement, skin manifestations are not necessary for the diagnosis.[5]

The aetiology of SAPHO syndrome is unknown. Proprionibacterium was isolated in surgically obtained bone specimens but, in the majority of cases, culture is negative. The bone lesions may be due to a pathogen characterized by low infectivity according to a mechanism similar to that of so called reactive arthritis, in which tiny amounts of inciting organisms have been revealed in synovium specimens.[2] A genetic background is suggested by the frequency of this condition in Japan and Scandinavian countries in contrast with its extreme rarity in the UK and France.[5]

Spinal involvement is described in 50% of the cases. The spinal radiological findings in SAPHO (syndesmophytosis, spondylitis, interspinal ossification and spinal posterior joint arthritis and ossification) are similar to those met in spondylarthropathy. In the study of Maugars, one third of the SAPHO cases met

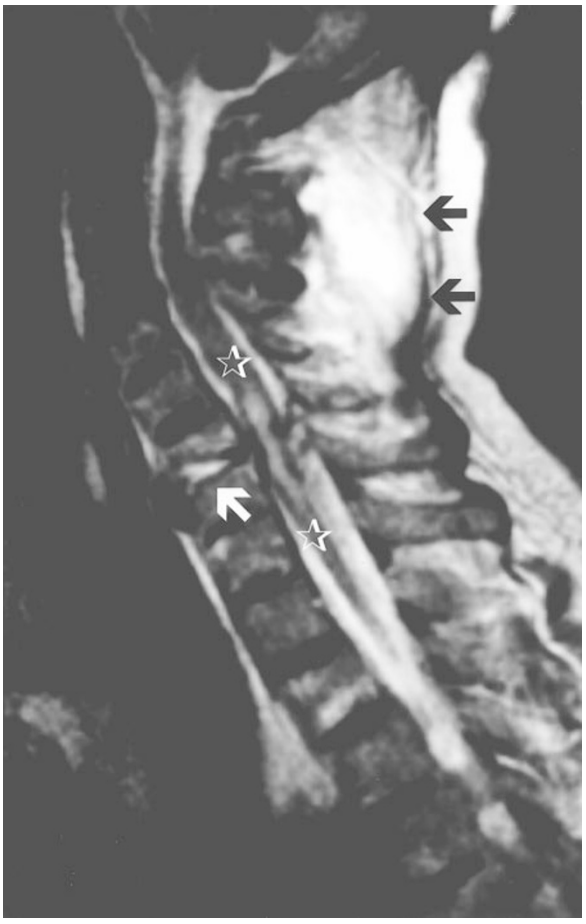

Figure 3 Sagittal T2-weighted MR image of the cervical spine: the C6-C7 fractured disc presents a high signal intensity suggestive of fluid (white arrow). The small prevertebral hematoma, the contusion of the posterior muscles (black arrows) and oedema of the spinal cord (white asterisks) also show a high signal intensity

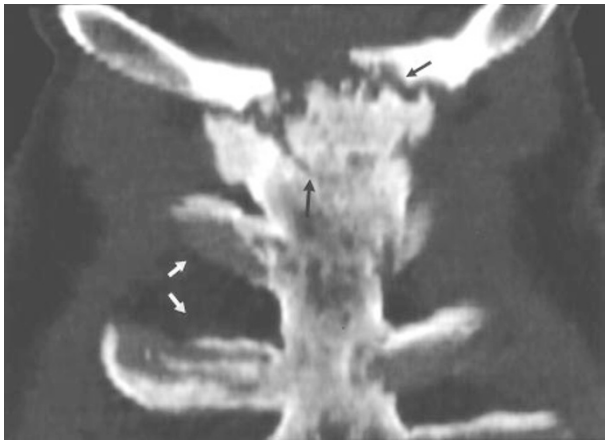

Figure 4 Coronal reformatted computed tomography of the sterno-costo-clavicular joints showing severe arthro-osteitis of the sterno-clavicular joints (small black arrow), hyperostosis of the costo-sternal joints (white arrows) and an oblique fracture through the manubrium (large black arrow)

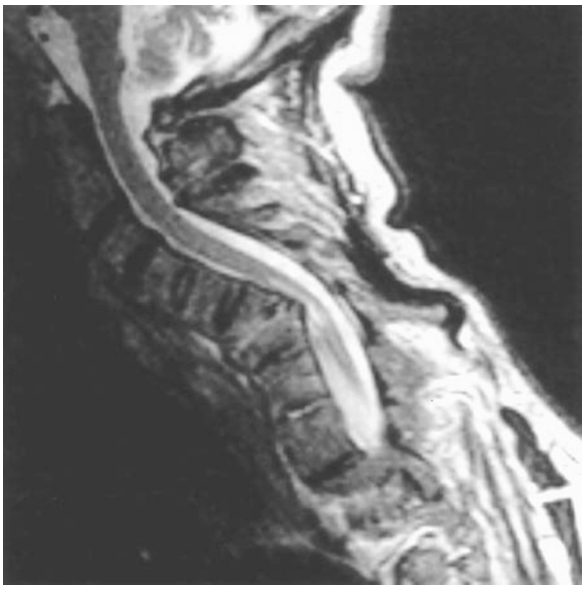

Figure 5 Sagittal T2-weighted MR image of the cervical spine performed 8 months later shows progressive cervical kyphosis of the C6-C7 level and myelomalacia

the criteria for diagnosis of spondylarthropathy.[4] Toussirot found sterile spondylodiscitis in 32% of the SAPHO syndrome but no neurological complications.[6]

Similar spinal radiological findings, frequent sacroiliitis and enthesitis suggest that a large proportion of the patients with SAPHO syndrome should be classified among patients with spondylarthropathy.[2] On the other hand, the higher incidence of anterior chest wall involvement and the lack of HLA B27 antigen, uveitis and urinary tract inflammation in the SAPHO syndrome argue for differentiation between the two entities.[4] The link between SAPHO and spondylarthropathy is still a matter of debate. As suggested by Maugars and Gladman, psoriatic arthritis is perhaps the 'missing link' between the SAPHO syndrome and spondylarthropathy.[4,7]

The aetiology of medullary compression in spondylarthropathies is atlantoaxial subluxation (AAS), ossification of the posterior longitudinal ligament (OPLL) and post-traumatic fracture.[8] Ramos-Remus found AAS in 21% of the cases with ankylosing spondylitis, OPLL in 2%, and clinical myelopathy in 18%.[9] Bohlman and Foo estimated the incidence of AS in SCI between 1.5 and 3%.[10,11] Ossified ligaments, loss of elasticity and osteopenia result in a rigid, brittle structure that is much more prone to fracture resulting from minor trauma.[12] Such fractures present a high rate of severe neurological deficit and a major risk of epidural hematoma and pseudarthrosis.[13]

Conservative treatment is associated with a high rate of fracture union and low rate of complications. Graham proposed a surgical intervention in evolving

neurologic lesions and in the management of the fractures that cannot be stabilized by nonoperative means.[14]

References

1 Chamot AM *et al*. Le syndrome Acne Pustulose Hyperostose Ostéite (SAPHO). Résultat d'une enquête nationale. 85 observations. *Rev Rhum Mal Osteoartic* 1987; **54:** 187–196.
2 Kahn MF. Why the SAPHO syndrome? *J Rheumatol* 1995; **22:** 2017–2019.
3 Goupille Ph, Valat JP. SAPHO syndrome and spondylarthropathy. *J Rheumatol* 1996; **23:** 1667–1668.
4 Maugars Y, Berthelot JM, Ducloux JM, Prost A. SAPHO syndrome: a followup study of 19 cases with special emphasis on enthesis involvement. *J Rheumatol* 1995; **22:** 2135–2141.
5 Kahn MF *et al*. Sternoclavicular pustulotic osteitis (SAPHO). 20-Year interval between skin and bone lesions. *J Rheumatol* 1991; **18:** 1104–1108.
6 Toussirot E, Dupond JL, Wendling D. Spondylodiscitis in SAPHO syndrome. A serie of eight cases. *Ann Rheum Dis* 1997; **56:** 52–58.
7 Gladman DD, Espinoza LR. International symposium on psoriatic arthritis. *J Rheumatol* 1992; **19:** 2–3.
8 Huang T *et al*. A rare neurological presentation due to ossification of the posterior longitudinal ligament of the thoracic spine and ankylosing sponylitis: case report. *Spinal Cord* 1997; **35:** 550–553.
9 Ramos-Remus C *et al*. Frequency of atlantoaxial subluxation and neurologic involvement in patients with ankylosing spondylitis. *J Rheumatol* 1995; **22:** 2120–2125.
10 Bohlman HH. Acute fractures and dislocations of the cervical spine. *J Bone Joint Surg* 1979; **61A:** 1119–1142.
11 Foo D, Sarkarati M, Marcelino V. Cervical spinal cord injury complicating ankylosing spondylitis. *Paraplegia* 1985; **23:** 358–363.
12 Broom MJ, Raycroft JF. Complications of fractures of the cervical spine in ankylosing spondylitis. *Spine* 1988; **13:** 763–766.
13 Deburge A, Guigui P, Ouahes M, Barre E. Cervical pseudarthrosis in ankylosing spondylitis. A case report. *Spine* 1996; **21:** 2801–2805.
14 Graham B, Van Peteghem PK. Fractures of the spine in ankylosing sponylitis. Diagnosis, treatment and complications. *Spine* 1989; **14:** 803–807.