Case Report

Ossification of the Posterior Longitudinal Ligament of the Cervical Spine and SAPHO Syndrome

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ABSTRACT. We describe a case of cervical cord compression due to ossification of the posterior longitudinal ligament of the spine (OPLLS) in a 43-year-old Vietnamese patient with SAPHO syndrome (synovitis, acne, pustulosis, hyperostosis, and osteitis). Idiopathic OPLLS is mainly reported in 50- to 60-year-old men, particularly in Japanese, with a prevalence of 2%. Cervical myelopathy may occur. In addition to OPLLS in patients of Asian origin, the condition has also been described in association with ossifying diseases, including ankylosing spondylitis (AS) and diffuse idiopathic skeletal hyperostosis (DISH) but not previously, to our knowledge, with SAPHO syndrome. (J Rheumatol 2005; 32:1361–4)

Key Indexing Terms:
OSSIFICATION
SAPHO SYNDROME
POSTERIOR LONGITUDINAL LIGAMENT
ANKYLOSING SPONDYLITIS

Myelopathy is a frequent complication of ossification of the posterior longitudinal ligament (ligamentum longitudinale posterius) of the spine (OPLLS)[1,2]. It has been reported in association with ossifying diseases such as ankylosing spondylitis (AS)[3] and diffuse idiopathic skeletal hyperostosis (DISH)[4], which are known to constitute predisposing factors for neurological complications[4,5].

SAPHO syndrome (synovitis, acne, pustulosis, hyperostosis, and osteitis) has been identified in patients presenting with dermatological features (severe acne or palmoplantar pustulosis) and osteoarticular involvement (i.e., anterior thoracic, vertebral or peripheral osteitis, or hyperostosis) similar to AS and other spondyloarthropathies[6]. We describe a patient of Vietnamese origin with cervical myelopathy due to OPLLS associated with SAPHO syndrome.

CASE REPORT

A 43-year-old male patient of Vietnamese origin was admitted in August 1992 for intense cervicalgia associated with severe acne of the thorax and plantar pustulosis. He had a 10-year history of dorsolumbar pain and had never received retinoids for his severe acne. Clinical examination revealed multidirectional cervical stiffness, an occiput wall distance of 6 cm, reduction of thoracic expansion to 2 cm, and painful stiffness of both hips. There was overall indefinite dysesthesia of both lower limbs and reduced osteotendinous responses. Radiographs showed mixed-type OPLLS of the cervical spine (Figure 1), syndesmophytes along the lumbar spine (Figure 2), ossification of the acetabula of both hips, and sparing of the joint space width (data not shown). Radiographs of the sacroiliac joints were normal. Technetium-99 bone scan showed increasing uptake of the left sternoclavicular, manubrial, and sternocostal joints (Figure 3). Erythrocyte sedimentation rate, C-reactive protein, blood cell count, and creatinine, phosphorus, and calcium levels were normal. There was untreated glucose intolerance. HLA–B27 antigen was absent. The pathologists were unable to distinguish between pustular psoriasis and isolated pustulosis on analysis of plantar cutaneous biopsy. SAPHO syndrome was diagnosed.

The patient was admitted in April 1993 for intermittent claudication. Myelography showed a narrowed lumbar canal at L3-L4 and L4-L5 with hypertrophy of zygapophyseal joint compromising the left L5 root. Laminectomy was performed and provided satisfactory post-operative improvement.

The patient was readmitted to the Department of Neurosurgery in April 1996 suffering from tetrapyramidal syndrome with paresis of the upper right limb. Computed tomographic myelography revealed complete extradural blocking of the contrast medium at C5 (Figure 4) and anterior compression of the dura mater due to OPLLS. Cervical laminectomy from C2 to T1 was performed. The dura mater was tight and had become very thin, and numerous distended perimedullary vessels covered the spinal cord. There was a partial post-operative improvement in the paresis of the right upper limb. The patient's neurological state stabilized in July 1996 and he was able to move about unaided using a wheelchair.

DISCUSSION

Although OPLLS is well defined by clinicians, its pathogenesis remains obscure. Some authors have speculated on the role of calcium and vitamin D metabolism[7], whereas others have hypothesized regarding the intervention of growth factors in the pathogenesis of the ossification[8-11]. Moreover, vitamin A[12] and retinoids[13] have also been implicated in the production of hyperostosis. Familial cases[14] and a higher frequency of certain HLA predisposing genes[15] support influence of genetic factors.

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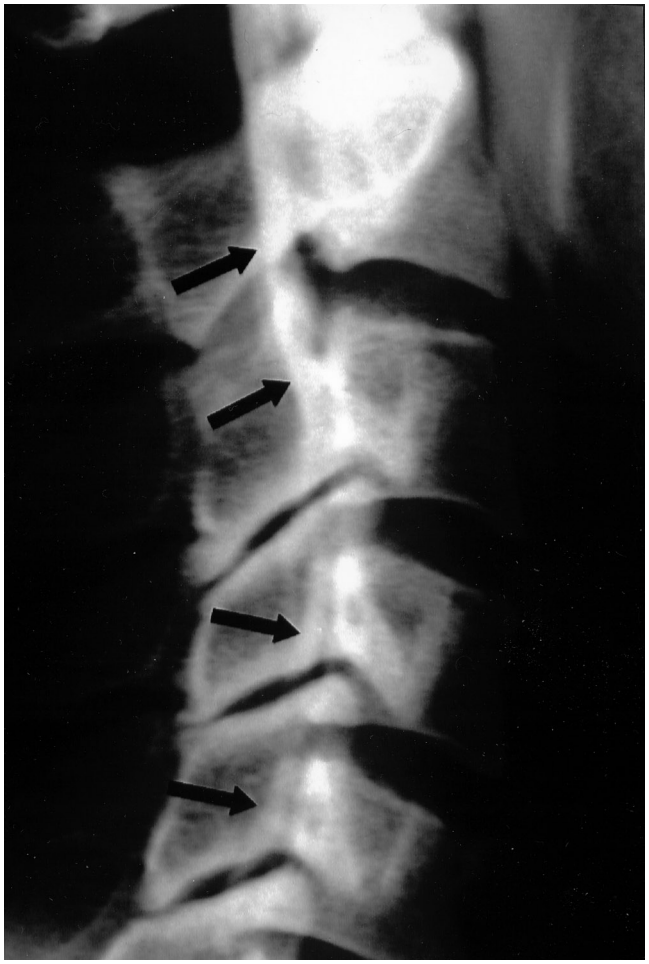

Figure 1. Radiographs of the cervical spine: ossification of the posterior longitudinal ligament (arrows).

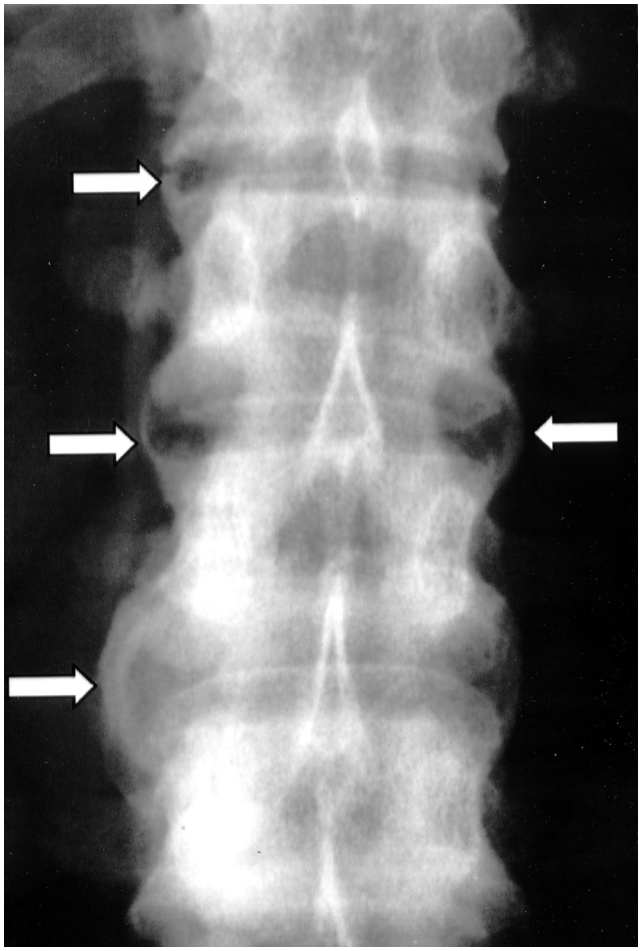

Figure 2. Spine radiographs: lumbar syndesmophytes (arrows).

The incidence of OPLLS is exceptionally high in Japan compared with non-Japanese Asians and Westerners. The incidence varies depending on the diagnostic approach and study design: the incidence of radiographically proven ossification is probably about 2% in Japan, but OPLLS has been found in up to 20% of Japanese over 60 years of age in a necropsy study[1].

The predominance of OPLLS in the cervical spine (92% of cases) is apparent compared with thoracic (4%) and lumbar (4 to 5%) levels[1,16]. Ossification of the ligamentum flavum may also be observed in OPLLS (7.4% of cases)[16].

According to Matsunaga, *et al* neurological signs may occur in 16% of asymptomatic patients after a 10-year followup[2]. The onset of signs of myelopathy in our patient occurred 3 years and 8 months after diagnosis of OPLLS. Occurrence of cervical myelopathy is probably multifactorial. Microtrauma may be encountered in association with segmental OPLLS although stiffening, continuous ossification tends to provide protection[2]. In addition, the posterior progression of OPLLS may reduce the diameter of the spinal canal as in our case. Vascular risk factors have also been reported to be significantly related to the risk of myelopathy, probably by enhancing ischemia of the spinal cord[2,5].

OPLLS has been considered as distinct from other hyperostotic conditions, i.e., DISH and AS. The main radiological characteristic of OPLLS is the presence of a radio-transparent area between the vertebral body and the ossification, whereas ossification is contiguous to the vertebra in AS.

Interestingly, overlap with DISH has been reported in 23.9 to 50% of cases[1,4] and more occasionally with AS (2% of cases)[1,3,5]. The diagnosis of DISH could be considered in our case but the radiographs of the lumbar spine are more suggestive of spondyloarthropathy; moreover, the association with severe acne, plantar pustulosis, and increased uptake by chest wall joints on bone scan allowed us to confirm the diagnosis of SAPHO according to criteria proposed by Chamot, *et al*[6]; finally, cases of coexistence of DISH and spondyloarthropathy have been reported[17]. No clear association between AS and OPLLS had been shown[18] before

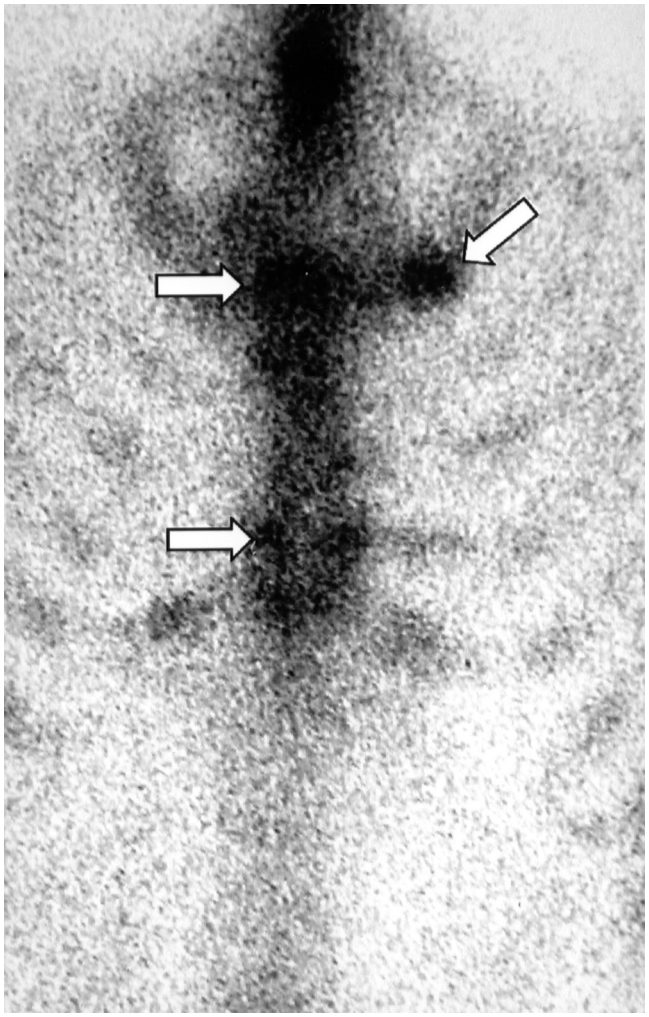

Figure 3. Technetium-99 bone scan. Uptake of the left sterno-clavicular, manubrial, and sternocostal joints (arrows).

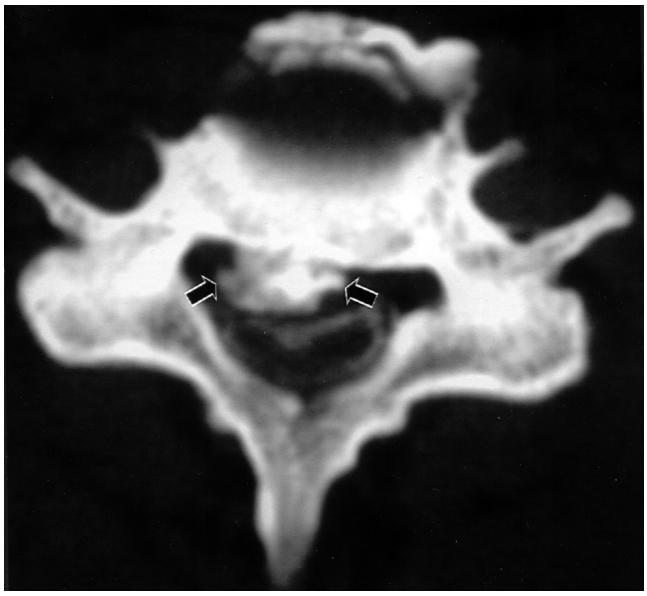

Figure 4. Computed tomographic myelography: epidural blocking of contrast medium reduced to a narrow strip at C5, due to OPLLS (arrows).

1998 when Ramos-Remus, *et al*[19] presented results of a cross sectional study. These authors attempted to assess the prevalence of OPLLS in 3 groups of patients with AS in Mexico and Canada and surprisingly found a frequency of 15.5%. Moreover, they also observed among 193 patients a few cases of psoriatic arthritis (3 patients), inflammatory bowel disease (2 patients), and reactive arthritis (one patient), but no SAPHO syndrome.

Although fairly rare, OPLLS should be investigated in cases of ossifying disease, particularly in patients of Asian origin. OPLLS overlapping with SAPHO syndrome has not been reported to our knowledge. As a significant association between OPLLS and AS has recently been reported, our case is probably not coincidental.

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