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Letter to the editor

L5-S1 Charcot spine induced by diffuse idiopathic skeletal hyperostosis in chronic tetraplegia: 2 cases

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Dear Editor

Charcot spine, or spinal neuroarthropathy, is a rare and underestimated arthropathy occurring in the setting of neurological impairment [1] like spinal cord injury [2]. It is characterized by disc and vertebral degeneration followed by massive bone formation of the articulation that can mimic infective spondylitis or vertebral tumor [3]. It results in impaired joint innervation with loss of proprioception and sensitivity to pain, associated with repeated microtraumas due to instability [4]. It can cause mechanical low back pain, spinal deformation, audible noises, and altered neurological function including spasticity or autonomic dysfunction [5].

Diffuse idiopathic skeletal hyperostosis (DISH), also known as Forestier disease, is characterized by ankylosis of the spine due to non-inflammatory ossification of both ligaments and entheses [6,7]. It most commonly affects the thoraco-lumbar spine, but involvement is variable and it can affect the entire spine. DISH is more common in men than women and the incidence increases with age.

The exact cause of DISH is not known. Here, we report 2 cases of L5-S1 Charcot spine preceded by DISH after chronic tetraplegia and loss of proprioception.

The first case involved a 62-year-old man with complete C7 sensorimotor tetraplegia (Frankel A and American Spinal Injury Association [ASIA] motor 20/100, sensory 20/112) due to a C7 burst fracture and C6–C7 dislocation in 1980. The clinical evolution was characterized by the progressive appearance of spine stiffness in extension. When the patient was age 46 (in 2000), spine radiography revealed progressive signs of DISH with the development of spine osteophytis from the cervical to lumbar spine (Fig. 1), beginning up to C6. At 34 years after the initial diagnosis of tetraplegia, the patient presented sequela of autonomic dysreflexia, as evidenced by labile blood pressure and heart rate, associated with urinary symptoms. By searching for "irritative thorns", we found early-stage L5-S1 Charcot spine, which was confirmed by CT, with an aspect of horizontal hypertrophic pseudarthrosis. The Charcot spine was associated with vertical ossification of the anterior longitudinal ligament of the thoraco-lumbar spine and also heterotopic ossifications of hips (Fig. 2). The Charcot spine was in contact with the rectal ampulla and the

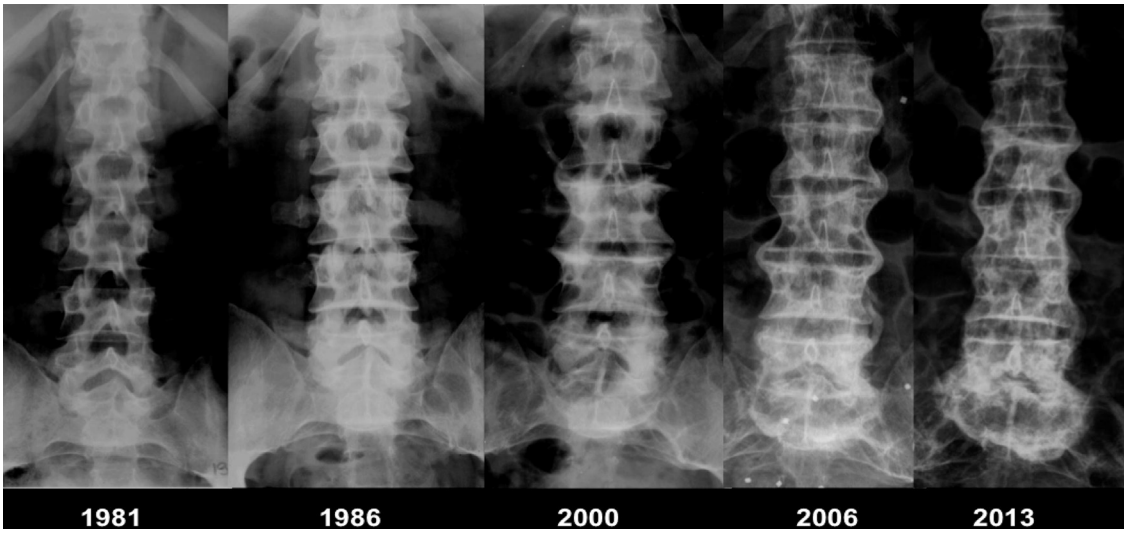

Fig. 1. Radiographic evolution of diffuse idiopathic skeletal hyperostosis (DISH) and L5-S1 Charcot spine in a C7 tetraplegic patient (case 1).

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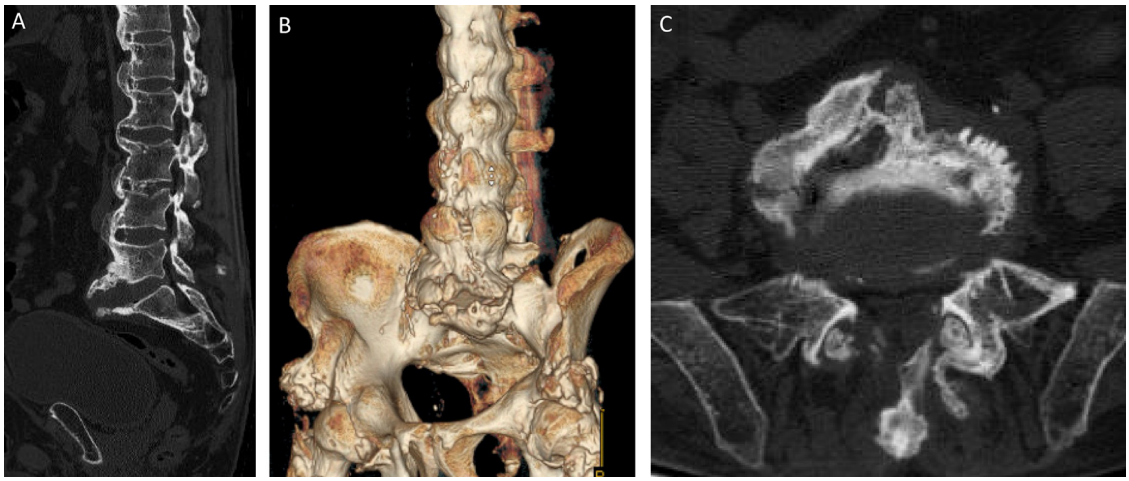

Fig. 2. L5-S1 Charcot spine in a C7 tetraplegic patient with DISH (case 1) shown by sagittal CT-scan (A) and 3D spine reconstruction CT-scan (B) with an aspect of hypertrophic pseudarthrosis on 3D reconstruction (B) and axial CT-scan (C).

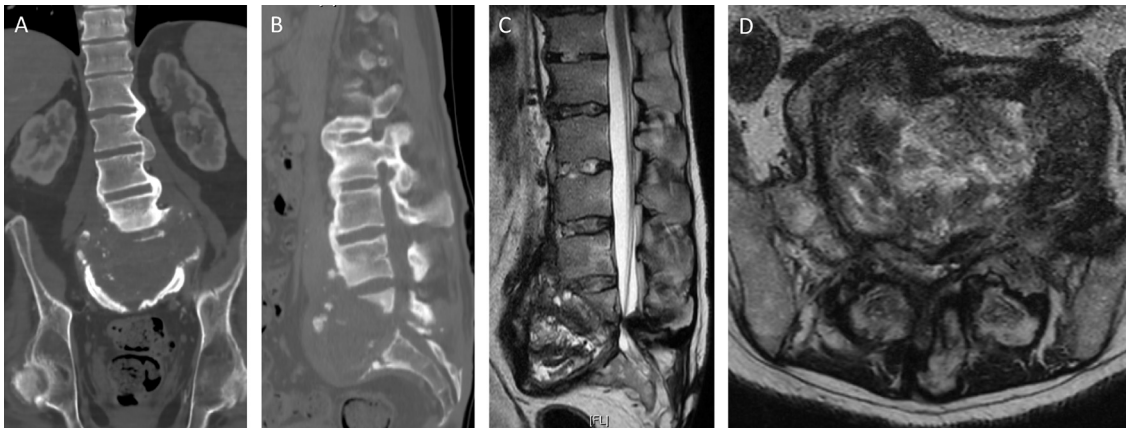

Fig. 3. Aspect of pseudotumoral L5-S1 Charcot spine in a C5 tetraplegic patient with DISH (case 2). Coronal (A) and sagittal (B) reconstructions from axial spiral CT scan on the left side, and sagittal (C) and axial (D) T2-weighted MRI images of the lumbar rachis on the right side.

distended bladder. Sequential spine radiographs showed no instability. The patient benefited from a corset to limit disease evolution. A urinary-incontinent prosthesis was required to reduce urinary symptoms. There were no clinical modifications after 2 years of follow-up.

The second case involved a 47-year-old man with complete C5 tetraplegia (Frankel C and ASIA motor 17/100, sensory 64/112) secondary to a C5 fracture with C5–C6 dislocation in 1987. At 26 years after the fracture, he presented low back pain, increased spasticity mainly during sitting, diaphoresis superior to the lesion, and unusual constipation. Lumbar spine radiography and CT (Fig. 3) revealed a significant lysis of the joint space between L5-S1 with notable reconstruction, evocative of lytic pseudotumoral L5-S1 Charcot spine. A follow-up MRI revealed no evidence of tumor growth (Fig. 3) and no spinal puncture was necessary. Spine radiography suggested a diagnosis of DISH, as evidenced by vertical osteophytis formation at several vertebral joints without joint destruction. Clinically, this arthropathy caused incorrect posture with axial tilt during the supine position and during sitting. Stabilization surgery has so far not been accepted because of the moderate functional impairment. For sitting, a rear support was fitted in the chair for a more stable position.

From these 2 observations and analysis of previous X-rays, we realized that the ossification of the anterior longitudinal ligament, characteristic of DISH, occurred before the onset of Charcot spine (Fig. 1). In the literature, there is not enough evidence to confirm this chronological link. Morita et al. [8] reported that in 9 patients with Charcot spine, 7 had ankylosing vertebral hyperostosis. This finding suggests the existence of an otherwise unnoted association between neuroarthropathy and vertebral ankylosing hyperostosis. The radiographs for the 2 case reports of Barrey et al. [4] also showed DISH above the Charcot spine. Our study is the first to show this chronological link, with a radiographic follow-up of 32 years.

With the current pathophysiological hypothesis [4], Charcot spine always occurs in the context of impaired joint innervation, with insensitivity to pain and loss of proprioceptive articular information. It leads to a loss of joint protection reflex. This neurological impairment is necessary but insufficient for the condition. Mechanical stress or excessive mobility must be added, leading to the joint destruction. Thus, Charcot spine is classically described for the thoraco-lumbar and lumbosacral spine, probably because these junctions are more stressed. Charcot spine also occurs in areas of surgery around an arthrodesis (overstress in proximal or distal junction segment) or even after laminectomy without arthrodesis, which increases the instability. Therefore, we can therefore hypothesize that DISH, via exuberant osteophytosis, leads to a loss of mobility, as a natural arthrodesis, with risk of

junction overstress. In other words, mobilization of the first non-ankylosed segment under DISH might play a role of mechanical stress that precedes the development of Charcot spine in patients with spinal cord lesions.

Furthermore, as in other cases in the literature [4,8], we found important osteophytis (compatible with DISH) in our patients after spinal cord injury. DISH is a frequent finding in the general population, but its prevalence is difficult to estimate mainly because the methods for diagnosis are heterogeneous (radiography, CT scan, autopsy) and multiple factors vary (e.g., age of screening, type of population [8.7% in Japanese patients [9], 17% in The Netherlands [10]]). This variation leads to an inconsistent measure of incidence. Nevertheless, Morita et al. [8] suggested that DISH could be more frequent in neurological patients (31% with mean age 55 years in their survey of 42 complete spinal cord injuries) than in general population at same mean age. Moreover, heterotopic ossifications (mainly in the hip and elbow) are also more frequent after spinal cord injury, as in our case 1. The pathophysiology of these neurogenic heterotopic ossifications is still debated [11], with different hypotheses: neurological, mechanical, biological, metabolic.

Our cases confirm the precedence of complete sensory deafferentation (by spinal cord injury) and abnormal mobility of the vertebral joint (by DISH) as a precursor to the development of Charcot spine. The importance of radiological monitoring in patients with traumatic spinal cord injury cannot be understated.

Disclosure of interest

The authors declare that they have no competing interest.

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