Spinal neuroarthropathy associated with Guillain–Barré syndrome

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Abstract Since the rarefaction of neurosyphilis, axial neuroarthropathy is mostly secondary to spinal cord pathologies. Peripheral manifestations of neuroarthropathy resulting from Guillain–Barré syndrome have already been reported but to our knowledge, this is the first case of a patient diagnosed with axial involvement. After the acute phase, a 47-year-old woman suffered of paraparesia with a partial loss of sensibility under the tenth thoracic vertebra. As a consequence, she developed first of all neuroarthropathy of both knees and few years later, a spinal involvement was discovered. Multiple surgeries of both knees and of the spine were required, due to non-union, relapse, and infection. Natural evolution of Charcot spine remains unknown. Surgical treatment is recommended but even with circumferential fusion, failures do frequently occur. This observation allows us to report an original case and to discuss the etiology of axial neuroarthropathy, the classical radiographic findings, and the inherent difficulties of its treatment.

Keywords Guillain–Barré syndrome · Charcot spine · Axial neuroarthropathy · Spinal neuroarthropathy

Introduction

The first description of spinal neuroarthropathy is generally attributed to Jean Martin Charcot in 1868 [4]. However, in

1703, Musgrave described a case of arthritis during the evolution of a neurosyphilis [25] and in 1831 Mitchell mentioned two cases of peripheral joint destruction associated with spinal cord lesions [14].

The spine was not identified as a location of axial arthropathy until Kronig reported a case in 1884 in a patient suffering from tabes dorsalis [13].

From then on, a prevalence of 6–21% of spinal involvement has been reported in patients with peripheral Charcot joints [2, 29].

The precise pathophysiology remains unknown but a sensorial neurological deficit seems to be *sine qua non* for the diagnosis of this lesion. Moreover, with the eradication of tabes in developed countries, spinal cord lesions have become the dominating cause of Charcot spine.

We report a unique case of a patient presenting with axial and peripheral Charcot arthropathy after having been diagnosed with Guillain–Barré syndrome. This case allows us to discuss the therapeutic options of spinal neuroarthropathy and to underline the inherent difficulties of its treatment.

Case

A healthy 47-year-old woman had a past history of otitis and flu-like symptoms a month before the onset of the first neurological symptoms.

She rapidly developed paresthesias, ataxia, and weakness of the four limbs, with an acme of neurological symptoms after 1 month of evolution. Sensorial symptoms were predominant but no cranial nerves, autonomic nervous system, respiratory involvement, or sphincteric dysfunction occurred. Tendon and abdominal reflexes were absent.

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Repeated lumbar punctures showed an albumino-cytological dissociation (elevated cerebrospinal protein rate at 0.6 g/l, normal cell count and glucose level). Cultures remained sterile.

Electroneuromyograms were performed showing conduction slowing and blocks, with prolonged distal latencies. A large biologic panel of serodiagnostic tests and antibodies dosages showed to be normal. Cervical spine magnetic resonance imaging ruled out spinal cord compression or myelitis.

The patient was thus diagnosed as an acute Guillain-Barré syndrome. No specific treatment was introduced, due to a delayed diagnosis.

Fig. 1 X-ray of the right knee showing peripheral neuroarthropathy initiating the Charcot disease

After 5 months of evolution, the patient's health improved to a state where she could again walk, albeit with the use of a walking aid. She had, however, a persistent paraparesia associated with a severe sensory deficit of the four limbs and the trunk, predominating below the tenth thoracic vertebrae.

The suspicion of Charcot arthropathy started with bilateral knee pain, which evolved rapidly toward a complete epiphyseal destruction of both tibias (Fig. 1).

Bilateral total knee arthroplasties were performed in another medical center. With the evolution of neuroarthropathy, ensuing prosthetic dislocation occurred.

After 4 years, the patient was referred to our service because of the emergence of a painless and progressive deformity of the spine. X-rays revealed a kyphoscoliosis secondary to the destruction of L2–L3 and L4–L5 discs and posterior articular joints. After having ruled out infectious causes with biologic investigations, as imaging was considered as typical, the diagnosis of Charcot spine was established (Fig. 2).

To counter the rapid evolution of the spinal deformation, a posterolateral correction and fusion were performed from level T10 to S1, in order to bypass the thoracolumbar junction. Cultures of surgical biopsies were negative. A simultaneous nerve biopsy from the musculocutaneous nerve of right leg showed no abnormality.

This procedure was firstly complicated by a sepsis necessitating the removal of a sacral screw and 3 months of antibiotics. Secondly, a lumbosacral non-union required an additional surgery in order to ensure consolidation. A posterior approach with screw placement in iliac wings was performed, followed by a retroperitoneal approach with graft and a fixation of the L5/S1 disc.

Four years later, when the patient presented with a kyphosis associated with painless cracking sounds, a relapse of the spinal neuroarthropathy above the previous instrumentation was diagnosed. The spinal fusion had to be extended using a posterior approach and pedicular instrumentation (Fig. 3). T10 screws of the previous construct

Fig. 2 Lumbar spine roentgenograms showing axial neuroarthropathy (L2/L3, L4/L5) before surgery (a, b) and after first posterior surgery with lumbosacral iterative circumferential fusion (c, d)

Fig. 3 Extension of the posterior arthrodesis up to the T3 vertebra for T9–T10 relapse of spinal neuroarthropathy with vertebral numbering

were removed and rod connectors were used, in order to instrument and fuse all the thoracic kyphosis up to T3. Cultures of surgical biopsies were negatives.

Two months later, when exercising a simple cervical flexion, patient's T3 vertebra fractured (Fig. 4). It was complicated by an incomplete paraplegia ranked ASIA C (American Spinal Injury Association) [1], necessitating an upper thoracic laminectomy with an extension of the previous spinal instrumentation up to the seventh vertebra (Fig. 5). Anatomopathologic analysis of biopsies from T3 vertebra showed vertebral necrosis, which remained of unknown etiology after biologic investigation.

One year later, the patient showed a partial neurological recuperation with an almost normal strength on the upper limbs but persistent and incapacitating paresia of both legs. Sensory deficit remained with epicritic and proprioceptive troubles. She recovered full sphincter and bladder control.

Simultaneously, removal of both knee arthroplasties had to be performed because of secondary infection, further to a lateral malleolar wound. Prosthetic reimplantation succeeded for the left knee but iterative infection of the right knee occurred, necessitating an arthrodesis.

At the last follow-up, she was able to walk few steps with the aid of technical apparatus but was most of the time confined to wheelchair.

Fig. 4 MRI T2-weighted sagittal image showing a T3 fracture associated with spinal cord compression

Discussion

Charcot arthropathy corresponds to a destructive arthropathy of unknown causes. Initially, it was attributed to vasomotor phenomena [4, 12] but nowadays, the majority of the authors believe that the causative lesion of this pathology is a loss of proprioceptive sensitivity [7, 15, 21, 29]. This deficit can be linked to a lesion of the spinal cord or of the peripheral nervous system, of either congenital or acquired origin. The loss of the proprioceptive sensitivity would lead to the failure of protective mechanisms in joints, after excessive and repetitive use [27]. Micro fractures in joints are thought to generate the accumulation of intrasynovial osteo-cartilagineous debris [9]. Secondly, an inflammatory mechanism would lead to articular destruction [10, 24].

However, patients with congenital insensitivity to pain may develop this type of vertebral lesions without having proprioceptive troubles [20, 26].

The prevalence of axial neuroarthropathy in patients who have central or peripheral neurological lesions is unknown. According to Piazza, 6–21% of patients with tabetic peripheral Charcot disease have spinal implications

Fig. 5 Full spine roentgenograms with posterior instrumentation and fusion from T3 to iliac wings, at last follow-up

[5, 15, 16]. This rate is probably underestimated because of the frequent asymptomatic nature of this complication [9].

In the literature, lumbar spinal implication is more frequently reported than affection of the lower thoracic area [19, 23, 24]. To our knowledge, the upper thoracic and cervical spine are exceptionally concerned by this pathology [6].

Two types of radiographic presentations have been described in the literature: bone destroying and bone-forming lesions [7]. Both of them usually affect anterior and posterior elements of the spine, which can help to differentiate from infection.

At the outset, disc space narrowing and endplate sclerosis can be the earliest radiographic findings.

Vacuum discs, debris, bone fragmentation, joint disorganization, and facet involvement suggest the diagnosis of Charcot spine. When major vertebral destructions are observed, a dislocation of the spine can occur.

Besides this, bone-forming lesions can sometimes result in a natural stabilization of the spine.

Guillain–Barré syndrome is an acute inflammatory demyelinating polyneuropathy of autoimmune origin. This demyelination most commonly involves both sensory and motor aspects. The fine tactile and proprioceptive elements are usually the most affected sensitivity aspects. Neurological after-effects are observed in up to 50% of the patients [8, 17]. Guillain–Barré has already been associated in one case with a peripheral localization of Charcot disease [18] but to our knowledge, it has never been reported as the cause of axial neuroarthropathy.

The treatment of spinal neuroarthropathy is based on the immobilization of the lesion either surgically or conservatively.

The natural evolution of this disease with conservative treatment was briefly touched upon and studied by few authors. Selmi reported paraplegic patients suffering from dysautonomical disorders, secondary to axial neuroarthropathy, that were efficiently treated by prolonged immobilization or bed rest [19]. Even though Hoppenfeld also reported good outcomes of conservative treatments, most

authors advocate surgery and believe that conservative management leads to permanent neurological deficit, infection or death [11, 22, 23].

For ambulatory patients with incomplete paraplegia or peripheral nerve disease, the functional risk of axial neuroarthropathy is higher than for complete paraplegic patients. Surgical management is usually mandatory before definitive spinal cord lesions occur.

Given the nature of this arthropathy and its circumferential involvement of the spine, anterior and posterior fusion of the spine is recommended. Patients treated with elective anterior or posterior arthrodesis are highly exposed to complications such as non-union or metal failure [3, 20, 26–28].

Despite this, complications such as non-union, failure, or infection are still common. In a study conducted in 10 patients, Devlin observed a 50% rate of complications and 40% of reoperation.

In a paraplegic patient, the presence of a sacral bedsore or its past history is an important infectious risk factor for spinal surgery.

The majority of the authors strongly recommend performing a circumferential fusion but little is known on where to limit the arthrodesis. Adjacent level relapse is well-documented as a complication in the literature [20, 21, 26]. The necessity of performing extended fusion in order not to leave any mobile segment below an existing fused area or in the lumbosacral area is recognized [7, 23, 24].

In our case, an associated anterior fusion of the lumbar spine could have been performed, as recommended by most authors, but the lumbosacral non-union occurred at a previously intact level, which raised the question of the extent of the interbody fusion.

The relapse of the neuropathic vertebral arthropathy at the level T9–T10 was most probably due to elevated mechanical strains above the fused area, in a zone of sensory deficient zone. A hypolordosis may have participated in the increase of junctional strains. Retrospectively, fusion should have at least included the thoracic kyphosis above the last thoracic sensory level. No etiology of the T3 vertebra necrosis and fracture was brought to light after exploration.

Conclusion

To our knowledge, this particular case of acute axial neuroarthropathy is the first to be presented as secondary to a Guillain-Barré syndrome.

If a conservative treatment can be debated for paraplegic patients, surgical intervention is recommended as a course of treatment in ambulatory patients.

Often proposed with advanced destructions, surgical intervention usually requires a circumferential spinal fusion. In order to prevent relapse of the arthropathy, posterior fusion should extend from the first sacral vertebrae to at least the first level in a sensitive area.

Failure to consolidate, relapse in adjacent levels and infections constitute the majority of the complications, thus exposing the patient to a high rate of reoperation.

Conflict of interest statement None of the authors has any potential conflict of interest.

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