Original article

Do all Charcot Spine require surgery?

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ABSTRACT

Introduction: Spinal neuroarthropathy (SNA), also called “Charcot spine”, is very uncommon disease of unknown etiology. Kronig first reported this pathology in 1884 on a patient with Tabes dorsalis (also known as syphilitic myelopathy). As syphilis tends to disappear in developed countries, spinal cord lesion is the most frequent etiology of SNA.

Objectives: To describe clinical and radiographic results in 12 patients suffering from spinal neuroarthropathy (SNA).

Methods: Twelve patients diagnosed with SNA were included in the study. All patients were wheelchair users. The average delay between the neurological disease and the diagnosis of SNA was 18 years. All patients were initially treated conservatively. Surgery was only indicated in persistent symptomatic or instable cases, and for infected SNA. Surgery was a circumferential arthrodesis.

Results: From 12 patients, with a median follow-up of 4 years, five patients were operated on and 7 patients were still conservatively treated. Two patients with back pain and evolutive destruction were declined for surgery. One suffered of bilateral hip ankylosis and extensive spinal surgery would have confined him to bed, and one due to an evolutive bedsores. One patient improved with a complete regression of back pain.

Conclusion: Nowadays, surgical treatment is recommended with an extensive and circumferential fusion, in order to prevent relapses. Good radiographic outcome is reported but functional results have not been studied. Natural evolution of SNA remains unknown but can be less disabling than surgery. This pathologic mobility can contribute to patient’s autonomy and can therefore be considered as opportune. Conservative therapy can be considered for SNA.

Level of evidence: Level IV.

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1. Introduction

Neuroarthropathy is a disease of unknown etiology. A failure of proprioceptive perception is supposed to be mandatory for its development. Peripheral involvement, such as “Charcot foot”, is the most common form. Spinal neuroarthropathy (SNA), also called "Charcot spine", is very uncommon. Kronig first reported this pathology in 1884 on a patient with Tabes dorsalis (also known as syphilitic myelopathy). As syphilis tends to disappear in developed countries, spinal cord lesion is the most frequent etiology of SNA. Due to its common asymptomatic presentation, prevalence of SNA remains unknown. Some authors report 6 to 21% of spinal involvement in patients suffering from peripheral neuroarthropathy [1–3]. Because of prevailing asymptomatic evolution, diagnosis is usually made at an advanced stage of bone destroying lesions.

For some authors, surgery is necessary for all SNA cases to prevent death, neurological complications or infections [4–6], but surgical management is demanding with high rates of complications and failures. Non-unions, infections and relapses are the most commonly reported complications with a reoperation rate as high as 40% [7]. Also surgery leads loss in mobility and autonomy. Conservative treatment has been formerly reported, but seldom studied.

In our experience, conservative treatment is the reference. Surgery was only indicated in persistent symptomatic or instable cases, and for infected SNA. The aim of this study is to describe results from a mainly conservatively treatment, in order to discuss the advisability of non-surgical management of this pathology.

2. Material and methods

This retrospective case study was conducted in a hospital specialized in surgery and rehabilitation for neurologically disabled...
patients. All patient diagnosed for SNA between January 1997 and December 2007 were considered.

Patients were diagnosed with SNA when they were suffering from a neurological deficiency and when clinical and radiological presentations were suggestive [8]. Clinical presentation is non-specific: back pain, audible sounds, loss in height, spinal progressive deformity and modification of neurological status. For radiological analysis, each patient underwent standard and dynamics X-rays, computed tomography and resonance magnetic imaging. Evocative imaging usually associates bone forming and bone destroying lesions. Lumbar or lower thoracic spine are the most common localizations of SNA. Both anterior and posterior parts of the spine are generally affected. Vacuum disks, facets involvement, vertebral dislocation or instability are important diagnosis criterions. Destructing lesions begin in disk space and posterior joints. They can be explosive with instability and dislocation of the spine. Bone forming lesions can manifest with the formation of osteophytes and endplate sclerosis. Imaging is not specific and care must be taken to exclude inflammatory, tumoral or infectious etiologies.

A total of twelve patients (8 men, 4 women) were reviewed in our institution (Table 1). A spinal surgeon and a rehabilitation practitioner specialized in cerebral palsy followed all patients in this study. Before the diagnosis of SNA, the average interval with spinal cord lesion was 17 years (5 to 35 years). Seven of them had post-traumatic neurologic deficiency, one had a myelitis paraplegia, one had a post aortic dissection paraplegia, one suffered from Friedreich ataxia, one had a chronic inflammatory demyelinating polyneuropathy (chronic counterpart of Guillain-Barré syndrome) and one had spinal cord tumor. Two had evolution etiology.

All patients were confined to wheelchairs at the time of diagnosis. Eight patients were ranked ASIA A and two ASIA B. Patient suffering from Friedreich ataxia lost the ability to walk 6 years before the diagnosis of the SNA and patient with chronic inflammatory demyelinating polyneuropathy lost the ability to walk 4 years before the diagnosis of the SNA. Nine patients had a previous spinal surgery.

Lumbar area was more frequently (11 patients) affected, involving both anterior and posterior columns. None of the patients had a history of peripheral neuroarthropathy. Six patients had serious comorbidities at the time of diagnosis: myocardiopathy, sacral bed sore and bilateral hip heterotopic ossifications for two patients, one complicated with sepsis.

2.1. Symptoms

Ten patients had predominant mechanical complaints such as loss of stature while sitting, back pain, instability or dislocation of the spine with audible cracking thuds. Transit or respiratory difficulties occurred, resulting from the collapse of the trunk. Four patients experienced also a modification of the neurological status with sciatalgia, dysautonomia with blood pressure lability, spasticity and lost of vesical reflex. One patient was almost asymptomatic with only slight back pain.

2.2. Imaging

At first evaluation, 3 patients had a dislocated spine, reducible in traction, and one patient had an unstable level with a mobility of about 25°. One patient had a spontaneous fusion of the involved levels. The three remaining patients had mobile lesions with a stable spine.

According to the balance of bone forming and bone destroying lesions, 2 patients had constructive predominant patterns while in the others it was mainly destructive.

Initial care was well codified. A multidisciplinary team with a spine surgeon, a rehabilitation practitioner and a pain management, carried out conservative treatment. For back pain, a custom-made thoracolumbar brace, molded on the iliac crests and supports clavicles was the main part of the conservative treatment. The hips were free in order not to decrease the mobility. Brace could be not tolerated; bed rest was an alternative proposal. Classic and neuropathic painkillers, and physiotherapy were used for back pain. The dysautonomic syndrome was treated by the stabilization of the lesion, physical adaptation in the wheelchair, bed rest and physiotherapy. Bladder instability required anticholinergic drugs. The surgical treatment was performed in case of resistant pain, neurologic evolution, progressive destruction and for infected SNA. The surgical treatment was an extended and circumferential arthrodesis in one or two steps. An evolutional sacral bed sore was considered as a contraindication for surgery.

A resistant pain was defined after 6 months of a completed conservative treatment. Unstable spine was defined as a dislocated spine or a mobility of more than 10° on the dynamic X-rays. Lesions were considered as progressive if major constructive or destructive phenomenon occurred during follow-up.

Therefore, all patients were routinely screened for infection by clinical, biological and imaging confrontation. Whenever it was necessary, biopsies were done with bacteriological and anatopathology analysis.

3. Results

The average time of follow-up was 4 years. At the last follow-up, surgery was performed for five patients. The median delay between diagnosis and surgery was 17 months. The same surgeon treated all of these 5 patients. A thoracolumbar brace was prescribed for at least three months for 3 of them. Strictly bed rest was needed for the two other patients for three months in order to prevent non-union.

For the five patients operated, fusion was acquired in all cases. One surgical site infection occurred during hospitalization with good evolution after reoperation and antibiotic therapy during three months. One patient had a pseudo-arthrosis and needed a new surgery. The same patient developed a junctional kyphosis above the arthrosis and needed a cervical extension.

In follow-up, three patients complain about difficulty for manual exonation and intermittent urologic catheterization. No neurologic complication occurred, excepted exacerbation of vesical dysautonomy in two cases.

From the seven patients still treated conservatively at the last follow-up (2 women and 5 men), symptoms of five patients were controlled. Even, one patient improved with a complete regression of back pain due to spontaneous fusion (Fig. 1).

One patient worsened with an increase of back pain, spasticity and bladder dysfunctions. He was declined for surgery due to sacral bed sore.

One patient showed evolutional radiological destruction with an important instability and neurologic risk. However, due to a bilateral hip ankylosis, surgery was declined. A spinal arthrodesis could lead to incapacity to stay in the sitting position (Figs. 2 and 3).

No secondary localization of spinal or peripheral neuroarthropathy developed. At follow-up, no modification of treatment was considered, neither by the medical team nor by the patients who previously refused surgery.

3.1. Discussion

From a case series of twelve Charcot Spine, at a median of four years follow-up, with seven patients treated conservatively and five patients operated, conservative treatment showed good efficacy. When surgery was needed, fusion was obtained in all cases, but with complain about difficulty for manual exonation.
Table 1
Patients: initial evaluation and outcome.

<table>
<thead>
<tr>
<th>Patient</th>
<th>Initial neurologic disease</th>
<th>Neurologic status</th>
<th>Previous treatment</th>
<th>Delay between neurological lesion and first symptoms (years)</th>
<th>Level of SNA</th>
<th>Initial symptomatology</th>
<th>Surgery</th>
<th>Symptomatology at follow-up</th>
<th>Radiographic evolution at follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>T10 fracture</td>
<td>ASIA A</td>
<td>T7L1 posterior fusion</td>
<td>20</td>
<td>L3L4</td>
<td>Back pain, lost of height</td>
<td>None</td>
<td>Unchanged</td>
<td>No evolution</td>
</tr>
<tr>
<td>2</td>
<td>T10 fracture</td>
<td>ASIA A</td>
<td>T8L1 posterior fusion</td>
<td>31</td>
<td>L4L5</td>
<td>Back pain, lost of height, respiratory and digestive discomfort, dysautonomia</td>
<td>None</td>
<td>Unchanged</td>
<td>No major evolution, constructive spur</td>
</tr>
<tr>
<td>3</td>
<td>T9–10 fracture</td>
<td>ASIA A</td>
<td>Orthoses</td>
<td>35</td>
<td>L1L2</td>
<td>Moderate back pain, lost spasticity and vesical reflex</td>
<td>None</td>
<td>Unchanged</td>
<td>No evolution</td>
</tr>
<tr>
<td>4</td>
<td>Aortic dissection</td>
<td>ASIA A</td>
<td>Aortic surgery</td>
<td>15</td>
<td>L4L5</td>
<td>Moderate back pain</td>
<td>None</td>
<td>Unchanged</td>
<td>No evolution</td>
</tr>
<tr>
<td>5</td>
<td>Transverse myelitis</td>
<td>ASIA A</td>
<td>None</td>
<td>7</td>
<td>L2L3</td>
<td>Back pain</td>
<td>None</td>
<td>Unchanged</td>
<td>No evolution</td>
</tr>
<tr>
<td>6</td>
<td>Friedreich ataxia</td>
<td>Evolutive status</td>
<td>None</td>
<td>19</td>
<td>L4L5</td>
<td>Back pain, sciatalgia</td>
<td>None</td>
<td>Asymptomatic</td>
<td>No evolution</td>
</tr>
<tr>
<td>7</td>
<td>T8–T9 fracture</td>
<td>ASIA B</td>
<td>T8-T10 posterior fusion</td>
<td>12</td>
<td>L5S1</td>
<td>Back pain, lumbosacral kyphosis, elevated spasticity</td>
<td>None</td>
<td>Partial lost of sensibility, bladder and anal dysfunction</td>
<td>No major evolution</td>
</tr>
<tr>
<td>8</td>
<td>T9 fracture</td>
<td>ASIA B</td>
<td>T7-L3 posterior fusion</td>
<td>30</td>
<td>L5S1</td>
<td>Back pain, audible cracking thuds, modification of neurological status (spasticity)</td>
<td>Extension sacrum, iliac crest (01/2006), Surgical site infection</td>
<td>Decrease of back pain, kyphosis (12/2011)</td>
<td>Fusion</td>
</tr>
<tr>
<td>9</td>
<td>C6-C7 dislocation</td>
<td>ASIA A</td>
<td>Arthrodesis T12S1, pseudoarthrodese T12-L1, surgery T9-T2</td>
<td>25</td>
<td>T10-T11</td>
<td>Moderate back pain, kyphosis, modification of neurological status (dysautonomia)</td>
<td>Extension arthrodose T4-L4</td>
<td>No back pain, stabilization of neurologic aggravation, BUT loss in mobility and autonomy</td>
<td>Fusion</td>
</tr>
<tr>
<td>10</td>
<td>Spinal cord tumor</td>
<td>ASIA A</td>
<td>T2S1 posterior arthrodese, instrumentation removal L5S1</td>
<td>15</td>
<td>L5S1</td>
<td>Back pain, lost of height, kyphosis</td>
<td>Circumferential arthrodose, extension sacrum, iliac bone (10/2011)</td>
<td>No back pain</td>
<td>Fusion</td>
</tr>
<tr>
<td>11</td>
<td>Chronic inflammatory</td>
<td>Evolutive status</td>
<td>None</td>
<td>5</td>
<td>L4L5</td>
<td>Modification of neurological status (vesical and anal dysfunction)</td>
<td>Posterior arthrodose, anterior arthrodose T10-S1 (03/2004), pseudarthrodisis L5S1: circumferential Arthrodose (05/2006), junctional kyphosis: extension C7</td>
<td>No back pain, stabilization of neurologic symptom</td>
<td>Fusion</td>
</tr>
<tr>
<td>12</td>
<td>T8–T9 dislocation</td>
<td>ASIA A</td>
<td>T5–L1 arthrodese</td>
<td>11</td>
<td>L4-L5</td>
<td>Modification of neurological status (spasticity)</td>
<td>Circumferential arthrodose, extension sacrum—and iliac crest</td>
<td>Decrease of spasticity, but loss in autonomy</td>
<td>Fusion</td>
</tr>
</tbody>
</table>

SNA: spinal neuroarthropathy.
and intermittent urologic catheterization or exacerbation of vesical dysautonomy.

SNA is a very uncommon pathology. Mitchel, who also suggested an association with spinal disorders, reported the first case of neuroarthropathy in 1831 [9]. A French neurologist named Jean-Martin Charcot diffused knowledge of peripheral neuroarthropathy in 1868, now known as Charcot disease [10]. The first case of spinal involvement is attributed to Kronig in 1884, in a tabetic patient [11]. With the eradication of neurosyphilis in modern countries, spinal cord pathologies are the most common causes of SNA [5,7,12].

Traumatic spinal cord injury was first reported as a cause of SNA by Slabaugh in 1978 and tends to be nowadays one of the most frequent etiologies. The delay between the spinal cord injury and the onset of neuroarthropathy is usually long, ranging from 2 to more than 30 years. The incidence of SNA in post-traumatic paraplegic patients is not known. No predisposing factor is recognized, even if laminectomy as been suggested [13–16].

Peripheral neuroarthropathy is usually painless due to the loss of sensitivity. Similarities exist with spinal involvement and the disease is often diagnosed at an advanced stage of destruction, after a clandestine and asymptomatic evolution [14,15,17–19].

Back pain is common in paraplegic wheelchair users who often over-use their lumbar spine. The appearance of a painless progressive deformity is more suggestive. It could worsen because of increasing mechanical strains when instability, reducible dislocation or cracking thuds develop. Modification of the neurological state such as motor deficit, dysautonomia, loss or increase of spasticity can also be part of the evolution [4].

Before the advent of spinal instrumentation, management of SNA was essentially approached by employing conservative means. By 1945, Key suggested not to operate on SNA since any attempt to fuse was supposed to lead to failures and thus recommended bed rest, orthosis and braces [14]. Radiotherapy has once been used by Feldman, without any success [19]. No particular therapeutic scheme has been evaluated in the literature and conservative treatment must be adapted to each patient.

Some authors report good clinical outcomes after 4 to 6 months of treatment without any radiographic change [4–6,13,20,21].

Some others report failures of conservative management with evolution of radiographic findings [3,15]. Only radiographic supervision was found to give evidence of SNA activity.

The natural evolution of SNA is unknown but considering peripheral manifestations, a 3-stage path can be suggested with fragmentation, coalescence and reconstruction [22,23].

During the active phase, the objective of therapy should be the immobilization of concerned joints to prevent fractures and deformities. Satisfactory results can be achieved in peripheral joints by casts or splints but paraplegic patients do usually not tolerate spinal orthoses and braces. Progression of active Charcot spine may not be stopped by conservative measures [3].

During the residual phase, whilst orthoses cannot reduce dislocated spine, they can have a positive effect on instability and back pain.

No secondary localization has been described with conservative treatments. Spontaneously infected Charcot spine has been seldom reported [24–26] whereas infectious complications of
surgical management are common. Therefore, surgical procedure may not be done in order to prevent sepsis.

Due to late diagnosis, destructive lesions are frequently explosive and challenge surgical reconstruction [19]. Slaubaugh reported the first case of SNA treated by instrumented fusion in 1978. Surgery took root with the advent of segmental spinal devices. Arnold considered that surgery was a better alternative than non-surgical management and that it would become the norm with modern instrumentation [27]. For most contemporary authors, conservative treatment is the usual recourse for patients who refuse surgery or who manifest contraindications [4–6]. Nowadays, circumferential instrumented fusion is recommended but nevertheless, the rate of complication remains high with mainly non-unions, metal failures, relapses or infections [13,16,18,28–30]. Brown and Devlin reported an overall 50% complication rate with a reoperation rate up to 40% [5,7]. Most paraplegic patients would like an efficient single surgery to relieve their symptoms and they can be reluctant to engage a procedure with such a rate of overall complications.

The optimal extent of fusion is not known. In order to prevent relapse, most authors recommend the fusion of all segments below a previous instrumented area or below the levels of laminectomy [7,12]. Brown observed a rate of relapse as high as 25% when the lumbosacral junction was not fused [5]. However, he underlined the fact that a fusion to the pelvis would dramatically reduce the autonomy of wheelchair patients.

In our study, one patient out of seven showed Evolutional lesions. Bilateral hip ankylosis was caused by heterotopic ossification but patient strictly refused hip surgery. Siting position and wheelchair use were permit by SNA mobility and spinal fusion would have confined him to bed. As long as he refuses hip surgery, no spinal arthrodesis can be considered.

The goal of surgery is to obtain a solid fusion and most studies reports satisfactory radiographic outcomes. Less is known about functional outcomes and to date, no study has addressed the relationship between radiographic fusion and functional evaluation.

From our surgical experience, an increase of the handicap can be induced by extensive arthrodesis. This pathologic mobility is sometimes considered as providential as it permits patients to rotate and bend while seated.

Charcot spine is an uncommon pathology. All in all, only 12 patients were diagnosed with SNA in our institution during this ten years period and from our knowledge, only five authors reported case series of more than seven patients [2,5,7,12,19], with generally mixed surgical and conservative management.

Clinical and radiological presentations at first assessment were not similar and therefore conservative measures could not be standardized. Some patients don’t require any specific treatment while some others need bed rest andorthoses in order to improve pain.

Thus, an evaluation of conservative therapy could not have been conducted and its contribution in the improvement of the outcome remains unknown.

For ambulatory and incomplete paraplegic patients, neurological preservation necessitates surgical therapeutic in order to stabilize and protect the spinal cord. Fusion should be circumferential at the level of SNA whereas posterior instrumentation should extent from a sensitive level down to the pelvis, in order to prevent relapse.

For wheelchair and paraplegic patients, the loss of autonomy must be considered. We recommend that a brace test should be done before surgery, to simulate fusion and estimate the functional outcome. Triggering and stopping factors of active SNA remain undiscovered. Natural stabilization of SNA lesions is possible and does not always lead to complications. Patients dependent on this mobility for their daily lives should be informed on the anticipated consequences of spinal fusion on their autonomy.

In our study, even patients with dislocated spine and back pain refused surgery, because of the lack of autonomy or the risk of relapse.

For paraplegic patients, insofar as we cannot propose a simple and efficient procedure that responds to their functional needs, conservative measures should be considered as part of first line therapeutic options.

Patients with spontaneously fused or stable spine rarely benefit from surgery. Suggested surgical indications are Evolutional deformity, infection or neurological deterioration, even if the question of the indicated span of fusion still remains unanswered [18,24].

4. Conclusion

Charcot spine is a challenging disease. This pathologic mobility can contribute to patients ‘ autonomy while seated and can therefore be considered as opportunite. Surgical intervention requires circumferential and extended fusion, often including the pelvis, and can increase the handicap with an expected loss of autonomy.

Its natural evolution remains unknown but is not always unfavorable and can be less disabling than extensive surgery. The functional state of patients with SNA tends to remain stable after the active phase. Unless spine infection or neurological deterioration occurs, conservative therapy can be considered for complete or non-ambulatory paraplegic patients. To our belief, patients with SNA should not systematically undergo surgery.

Disclosure of interest

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