Charcot arthropathy in relation to autonomic dysreflexia in spinal cord injury

Case report and review of the literature

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Charcot spinal arthropathy has been described as a late complication of spinal cord injury. In patients with these injuries in whom the spine below the level of injury is insensate, joint trauma can progress until spinal instability ensues. The authors describe the case of a 50-year-old man with complete C-8 tetraplegia who experienced a 4-month history of episodic severe headaches, profuse sweating over his face and arms, and episodic severe hypertension in addition to a “grinding” sensation in the lower back. Charcot arthropathy at the T11–12 levels with pathological mobility was demonstrated on neuroimaging. Intraoperatively, a complete spinal cord transection was identified. Anterior and posterior thoracolumbar fusion across the mobile segment resulted in complete amelioration of signs and symptoms of autonomic dysreflexia. This entity, a common condition in the setting of spinal cord injury, has many triggers. Definitive treatment is targeted at the removal of the underlying cause. As demonstrated here, Charcot spinal arthropathy can act as a powerful trigger for induction of autonomic dysreflexia. Treatment of the associated spinal instability resulted in eradication of all signs and symptoms of the dysreflexia.

KEY WORDS • neuropathic arthropathy • autonomic dysreflexia • spinal cord injury • spinal instability

EUROPATHIC spinal arthropathy represents a mechanical joint degeneration that occurs as a result of impaired deep sensation to facet joints. Although initially described by Mitchell, it is commonly referred to as a Charcot joint, after Charcot described its association with tabes dorsalis. In 1884, Kronig was the first to describe its manifestation in the spinal column. Today, neuropathic spinal arthropathy is more commonly encountered as one of the sequelae of spinal cord injury. Rare reports describe its occurrence in patients with diabetes, patients with tumors, and those with congenital insensitivity to pain. These manifestations indicate an injury to the afferent pathways of pain and proprioception, which can occur in fibers ranging from peripheral nerves to the spinal cord. These deficits, in conjunction with ongoing joint trauma, ultimately result in bone destruction.

The most common presenting symptoms in patients with Charcot spine are progressive kyphosis and back pain. In patients with spinal cord injury, in whom a quadriplegia or paraplegia renders the spine below the cord lesion partially or completely insensitive to pain, a Charcot joint can progress unrecognized until joint destruction leads to production of a hypermobile segment and spinal instability.

Autonomic dysreflexia is an acute syndrome of imbalanced reflex sympathetic discharge occurring in patients with spinal cord injury above the splanchic sympathetic outflow (T-6 level). This condition was originally recognized in 1890 when a young man with a spinal cord injury experienced profuse sweating during bladder catheterization. The syndrome was later described in its more severe form as a sudden increase in blood pressure with compensatory bradycardia, headache, and profuse sweating. Because of its hemodynamic sequelae, this condition represents a medical emergency. This, along with its high incidence after upper spinal cord injury (~48–70% with injury above T-6), makes its early recognition and swift treatment essential.

In this report we describe a patient who presented with autonomic dysreflexia and who after a thorough workup was found to have a grossly unstable thoracolumbar junction with near-complete dissociation as a result of Charcot arthropathy. He underwent a posterolateral fusion across the hypermobile joint, which resulted in dramatic cessation of all his symptoms.
Charcot arthropathy and autonomic dysreflexia in spinal cord injury

Case Report

History. This 50-year-old man with complete C-8 tetraplegia sustained in a motorcycle accident 20 years earlier presented with a 4-month history of episodic profuse sweating over his face and arms, along with severe headaches. His blood pressure during one of these episodes was recorded as 220/115 mm Hg. He also reported a “grinding” noise in his back when transferring from his wheelchair to his bed. The patient reported no association between urinary self-catheterization and the onset of his symptoms. He did report that extension of his torso was the most consistent stimulus for producing his symptoms. Initially, the episodes of diaphoresis and headaches were occurring once a week, but during the month prior to presentation the patient was experiencing symptoms two or three times per day.

Examination. Results of his physical examination were remarkable for the absence of all voluntary motor activity and sensation below the level of his original injury. In addition, a stepoff and focal kyphosis were identified at the thoracolumbar junction. Laboratory analysis revealed no signs of discitis/osteomyelitis or urinary tract infection.

Neuroimaging Studies. Plain x-ray films of the thoracolumbar junction showed extensive osteophyte formation throughout the lower thoracic spine. More important, irregular endplate destruction and sclerosis at the T11–12 level with a resultant focal kyphosis of 34° was apparent. In addition, there was a large paraspinous soft-tissue mass at this level. Flexion–extension views revealed extensive anterior subluxation (1.8 cm) of T-11 on T-12 with flexion (Fig. 1). Extension views obtained after the patient was placed in a thoracolumbar spinal orthotic brace revealed a 4-cm widening at the anterior margin of T11–12 endplates. An MR image (Fig. 2) and a noncontrast-enhanced CT scan (Fig. 3) obtained through this region revealed T11–12 disc space destruction and distraction with formation of a cystic central portion between the two osseous endplates. Interestingly, radiographs of the thoracolumbar spine obtained at the time of his original injury had revealed no evidence of a fracture or dislocation in the thoracic or lumbar spine, as mentioned in radiology reports. The actual images, however, were not available for review.

Operation. Given this degree of instability, the patient underwent an anterior–posterior fusion across the hypermobile segment (Fig. 4). Intraoperatively, retrolisthesis of T-11 on T-12 was identified. After we performed laminectomies at T-11 and T-12, it was apparent that the dural tube and the spinal cord were completely transected at this level. Fibrous scarring of the two ends of the dural tube was apparent and no evidence of cerebrospinal fluid leakage was noted. The endplates of T-11 and T-12, which were composed of mostly sclerotic bone, were debried. Because the spinal cord was transected, an expandable titanium cage was fitted between the endplates of T-11 and T-12 via the same surgical approach. A posterolateral fusion from T-8 to L-3 was performed along with pedicle fixation. Histopathological findings in the resected bone and soft tissues were consistent with Charcot arthropathy.

Postoperative Course. The patient had an unremarkable postoperative course, with no episodes of diaphoresis, headaches, or episodic hypertension. He was fitted with a thoracolumbar spinal orthotic jacket and was mobilized in his wheelchair 4 days after surgery. Follow-up imaging, including upright x-ray films obtained at 3, 6, and 18 months, revealed a stable construct and evidence of bone fusion between the T-11 and T-12 VBs. The patient has remained symptom free during the 18 months since his thoracolumbar fusion.

Discussion

Sensory deprivation and repetitive trauma are the only requirements for the development of Charcot joints. With the loss of protective sensation, ligamentous and capsular structures become insensitive to stretching and fail to stimulate protective muscle contractions to stabilize the spine. Repetitive trauma results in the destruction of articular cartilage and joint capsules. Sclerosis occurs once bone is denuded of cartilage. Periarticular and periosteal bone formation ensues, producing large osteophytes that bridge the VBs. With continued trauma, retrolisthesis or complete dislocation can occur, as was seen in our patient. In patients with spinal cord injuries, early diagnosis of this condition is difficult because of the absence of pain sensation below the spinal cord injury.
Fig. 2. Sagittal and axial T₂-weighted MR images demonstrating destruction of the endplates of T-11 and T-12 and a fluid-filled cavity in between. Letters on the sagittal MR image correspond to the axial images in the right panels.  
  A: Axial MR image obtained through the inferior endplate of T-11.  
  B: Axial MR image obtained through the fluid-filled cavity.  
  C: Axial MR image obtained through the endplate of T-12.

Fig. 3. Sagittal CT reconstruction of the thoracolumbar spine from T-10 to L-2 and axial images of T11–12 demonstrating the bone destruction of the T11–12 endplates and the complete dislocation of the spine at this level.  
  A: Axial bone window CT obtained through the VB of T-11.  
  B: Axial bone window CT obtained through the fluid-filled cavity.  
  C: Axial bone window CT obtained through the VB of T-12.
Loss of sitting balance appears to be the most common presentation of Charcot arthropathy in the setting of spinal cord injury (Table 1).

Understanding the pathogenic mechanisms of autonomic dysreflexia can shed light on the nature of its association with Charcot arthropathy. A variety of triggers for induction of this entity in the setting of spinal cord injury, including bladder distension, urinary tract infection, epididymitis, bowel distension, and detrusor–sphincter dyssynergia have been described. Individuals with a complete spinal cord injury above the major splanchnic outflow (T-6) are at greatest risk of suffering autonomic dysreflexia. Below the injury, intact peripheral sensory nerves transmit noxious impulses that ascend in the spinal cord. As a result of the cord injury, the ascent of these impulses through normal pain pathways is inhibited. It is postulated that the sympathetic neurons in the intermediolateral gray matter are stimulated by these impulses, resulting in severe arterial vasoconstriction. Intact baroreceptor function results in vasodilation above the level of injury. Patients commonly report a headache caused by vasodilation of pain-sensitive intracranial vessels. Vaso-motor brainstem reflexes attempt to lower blood pressure by increasing parasympathetic stimulation to the heart.

### TABLE 1

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of Patients</th>
<th>Presenting Symptoms</th>
<th>Injury</th>
<th>Treatment</th>
<th>Resolution of Symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>Slabaugh &amp; Smith, 1978</td>
<td>1</td>
<td>loss of sitting balance</td>
<td>complete</td>
<td>AP fusion</td>
<td>yes</td>
</tr>
<tr>
<td>Sobel, et al., 1985</td>
<td>5</td>
<td>loss of sitting balance</td>
<td>complete</td>
<td>2 of 5 w/ fusion</td>
<td>1 died/fusion, 1 planned fusion, 3 no</td>
</tr>
<tr>
<td>Kalen, et al., 1987</td>
<td>5</td>
<td>incidental finding</td>
<td>complete</td>
<td>1 pst fusion, 1 nonsurgical, 1 ext orthosis</td>
<td>1 yes, 2 no</td>
</tr>
<tr>
<td>Kapila &amp; Lines, 1987</td>
<td>1</td>
<td>loss of sitting balance</td>
<td>complete</td>
<td>pst fusion</td>
<td>not mentioned</td>
</tr>
<tr>
<td>Crim, et al., 1988</td>
<td>4</td>
<td>audible noise, loss of sitting balance</td>
<td>3 complete, 1 incomplete</td>
<td>3 pst fusion</td>
<td>yes</td>
</tr>
<tr>
<td>Mikawa, et al., 1989</td>
<td>1</td>
<td>loss of sitting balance</td>
<td>complete</td>
<td>AP fusion</td>
<td>yes</td>
</tr>
<tr>
<td>Hoppnfeld, et al., 1990</td>
<td>1</td>
<td>back pain</td>
<td>incomplete</td>
<td>ext orthosis</td>
<td>yes</td>
</tr>
<tr>
<td>McBride &amp; Greenberg, 1991</td>
<td>4</td>
<td>progressive kyphosis</td>
<td>complete</td>
<td>AP fusion</td>
<td>yes</td>
</tr>
<tr>
<td>Brown, et al., 1992</td>
<td>15</td>
<td>back pain, loss of sitting balance</td>
<td>complete</td>
<td>8 AP fusion, 7 nonsurgical</td>
<td>8 yes, 2 asymptomatic, 2 refused op, 3 planned fusion</td>
</tr>
<tr>
<td>Glennon, et al., 1992</td>
<td>3</td>
<td>new deficit, loss of sitting balance</td>
<td>complete</td>
<td>2 pst fusion, 1 brace</td>
<td>yes</td>
</tr>
<tr>
<td>Standaert, et al., 1997</td>
<td>5</td>
<td>back pain, loss of sitting balance</td>
<td>complete</td>
<td>AP fusion</td>
<td>yes</td>
</tr>
<tr>
<td>Thumbikat, et al., 2001</td>
<td>1</td>
<td>autonomic dysreflexia</td>
<td>complete</td>
<td>pst fusion</td>
<td>yes</td>
</tr>
<tr>
<td>Selmi, et al., 2002</td>
<td>2</td>
<td>autonomic dysreflexia</td>
<td>complete</td>
<td>bedrest</td>
<td>yes</td>
</tr>
</tbody>
</table>

* Ext = external; pst = posterior.
through the vagus nerve, which causes compensatory bradycardia.

Considering the progressive nature of Charcot spinal arthropathy, immobilization of the affected joints appears to be of paramount importance. Most authors advocate conservative therapy in the absence of disabling pain, neurological deficit, or obvious spinal instability. In cases of spinal instability such as that seen in the patient described here, surgical stabilization is warranted. The most successful outcomes have been reported for combined anterior and posterior approaches.

Long-term outcomes of both surgical and nonsurgical management are not well reported. Brown, et al., described 15 patients with Charcot spinal arthropathy following traumatic paraplegia, eight of whom were treated surgically with anterior and posterior fusion. A postoperative follow-up duration ranging from 1 to 8 years revealed successful fusion in all eight patients. In our patient, a posterior approach was used to provide anterior column support by the use of an expandable titanium cage as well as posterolateral segmental fusion. At the 6-month follow-up visit a solid construct, evidence of bone fusion, and cessation of autonomic dysreflexia were noted. It is therefore likely that the gross spinal instability that occurred as a result of the Charcot arthropathy was the underlying trigger for initiation of autonomic dysreflexia.

Although two previous case reports have detailed the association of autonomic dysreflexia and Charcot arthropathy in patients with spinal cord injury, we believe that the case described here is unique because of the severity of the pathological features and because we have outlined a successful and enduring treatment strategy supported by long-term postoperative follow up. Selmi, et al., have described prolonged bed rest as a treatment for autonomic dysreflexia in two patients with spinal cord injury in whom Charcot spinal arthropathy developed. It is unlikely that such a strategy would be successful in our patient in light of his severe pathological condition and gross spinal instability. The lack of patient compliance with complete and prolonged bed rest was also a factor in our case. Successful treatment of autonomic dysreflexia with spinal fusion in Charcot spinal arthropathy has also been described in a patient with paraplegia at the T-5 level. In this case, a freely mobile spondylolisthesis at L4–5 (the site of a previous laminectomy for insertion of a spinal cord stimulator) was identified. Although spinal fusion resulted in amelioration of the patient’s symptoms postoperatively, long-term follow-up data for this patient were not provided. In our case, we have demonstrated the absence of any signs and symptoms of autonomic dysreflexia during a period of 18 months after spinal fixation.

References

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