Massive Charcot spinal disease deformity in a patient presenting with increasing abdominal girth and discomfort

Case report

FRANK S. BISHOP, M.D., ANDREW T. DALEY, M.D., AND MEIC H. SCHMIDT, M.D.

Department of Neurosurgery, Clinical Neurosciences and Spine Center, University of Utah, Salt Lake City, Utah

Charcot spinal disease is a destructive degenerative process involving the vertebrae and surrounding discs, resulting from repetitive microtrauma in patients who have decreased joint protective mechanisms due to loss of deep pain and proprioceptive sensation. The typical presentation of the disease is back pain and progressive spinal instability and deformity. The authors report an unusual case of massive Charcot spinal disease deformity in a patient presenting with increasing abdominal girth and discomfort. (DOI: 10.3171/2009.12.FOCUS09277)

KEY WORDS • Charcot spinal disease • vertebral destruction • spinal neuropathic arthropathy • spinal neurogenic arthropathy

CHARCOT disease of the spine, also known as spinal neuropathic or neurogenic arthropathy, is a destructive degenerative process involving the vertebral bodies and surrounding discs. This condition results from repetitive microtrauma in patients who have decreased joint protective mechanisms from loss of deep pain and proprioceptive sensation, typically because of spinal cord injury or sensory neuropathies. The patient typically presents with back pain and progressive spinal instability and deformity. We report a unique case of massive Charcot spinal disease and deformity in a patient presenting with increasing abdominal girth and discomfort.

Case Report

History and Examination. This 33-year-old man suffered a complete spinal cord injury after a 25-foot fall from a parking garage. He was unable to move or feel his legs at the scene, and upon arrival to the emergency department was found to have a complete spinal cord injury with loss of sensation at the T-10 level. A CT scan of the thoracic spine demonstrated a severe T-11 fracture dislocation with complete obliteration of the spinal canal and a moderate local kyphotic deformity (Fig. 1A). Multiple mild compression and burst fractures from T-4 to T-7, in addition to multiple transverse process fractures, were also noted.

Initial Operation. On hospital Day 2, the patient underwent a T8–12 posterior spinal fusion, with pedicle screw instrumentation at T-9, T-10, T-12, and L-1, and posterolateral onlay fusion. This surgery was followed by a second-stage procedure on hospital Day 9 for anterior column reconstruction. A T-11 corpectomy via a left-sided thoracotomy was performed, with insertion of a Synex expandable titanium cage (Synthes, Inc.) and bone allograft (Fig. 1B). A chest tube was placed during the operation, which was removed on postoperative Day 2. The patient recovered appropriately from these procedures and was discharged to inpatient rehabilitation on hospital Day 12. Unfortunately, the patient did not recover any neurological function and remained completely paraplegic. The patient was monitored for 2 years using serial radiographs, which demonstrated mild scoliosis without evidence of hardware failure (Fig. 1C). He was asymptomatic and elected to continue with conservative treatment.

Charcot Spinal Disease. Five years after undergoing reconstruction and fusion, the patient presented with increasing abdominal girth and discomfort. He had no other significant complaints, including no symptoms of back pain. A CT scan of his abdomen demonstrated a large, abdominal, cystic paraspinal mass with hyperemia of the surrounding soft tissue and musculature (Fig. 2D). Thoracolumbar CT scans further demonstrated osseous...
destruction and complete destabilization of the spine below the level of fusion from T-12 to L-3, consistent with Charcot spinal disease (Fig. 2A–C). Infection was considered in the differential diagnosis but was believed to be unlikely because of the patient’s near-normal inflammatory markers (white blood cell count 10.5 × 10³/mm³, erythrocyte sedimentation rate 9 mm/hour, C-reactive protein 1.1 mg/dl).

Second Operation. A 3-stage procedure was planned for decompression and resection, realignment, and circumferential stabilization and reconstruction. The first surgery involved a posterior approach for open debride-ment and exploration. Free-floating bone fragments were identified and removed, and the visible cyst was aspirated. Laboratory studies on the cyst fluid were negative for infection. Severe spinal instability was apparent at the levels involved with spinal arthropathy. The second procedure was performed on postoperative Day 5 and involved a posterolateral spinal fusion from T-3 to the ileum, with pedicle screw fixation (Fig. 3A). The L1–3 levels showed severe osseous destruction and were omitted from the fusion construct. After recovering for 11 days, the patient underwent the third staged procedure, which involved a retroperitoneal approach for L1–3 corpectomies, anterior reconstruction using a Synex II expandable titanium cage (Synthes, Inc.), and anterolateral instrumentation using an MACS TL plate and vertebral body screws (Aesculap; Fig. 3B).

Postoperative Course. The patient experienced resolution of his abdominal symptoms postoperatively. Bone union was successfully achieved as demonstrated on follow-up radiographs. Although the Synex II cage has been recalled since it was implanted in this patient, the fusion construct and cage have remained solid. At his 1-year follow-up appointment, the patient demonstrated continued spinal stability and appropriate alignment without progressive deformity or pseudarthrosis.

Discussion
Charcot joint disease was initially described in the 19th century in patients inflicted with tabes dorsalis from tertiary syphilis and is a progressive destructive disease of the peripheral joints and the spine. Also known as neuropathic or neurogenic arthropathy, this condition is observed in patients with decreased joint protective mechanisms due to loss of deep pain and proprioceptive sensation from various causes including spinal cord injury (secondary to trauma, tumor, or infection), diabetic and other peripheral neuropathies, myelomeningocele, syrin-
Charcot spinal disease presenting with abdominal discomfort

The patient with this disease typically presents with symptoms of worsening back pain and audible noises during postural changes. Other presentations include sitting imbalance from progressive spinal instability and deformity, a decrease in lower-limb spasticity or sensation, cutaneous fistulas to paraspinal cysts, development of autonomic dysreflexia, and even deep vein thrombosis from prevertebral compression.5,7,11 This patient presented with unique complaints of abdominal discomfort and increasing girth from massive bone destruction and cyst formation in the retroperitoneal space, causing compression of the abdominal organs, without other typical presenting complaints. The patient experienced resolution of his abdominal symptoms after surgical decompression of the cyst, resection of disorganized bone, correction of the deformity, and reconstruction and stabilization, which is the current treatment for Charcot spinal disease.5,11

Disclosure

Dr. Schmidt has served as a consultant for Aesculap.

Author contributions to the study and manuscript preparation include the following. Conception and design: FS Bishop. Drafting the article: FS Bishop. Critically revising the article: FS Bishop, AT Dailey, MH Schmidt. Reviewed final version of the manuscript and approved it for submission: MH Schmidt.

References


Accepted December 16, 2009.
Address correspondence to: Meic H. Schmidt, M.D., Department of Neurosurgery, University of Utah, 175 North Medical Drive East, Salt Lake City, Utah 84132. email: meic.schmidt@hsc.utah.edu.