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Case Report

Management of autonomic dysreflexia associated with Charcot spinal arthropathy in a patient with complete spinal cord injury: Case report and review of the literature

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Abstract

Background: Charcot spinal arthropathy (CSA) clearly represents a challenge in long-term spinal cord injury patients, one that can have extremely uncomfortable and potentially lethal outcomes if not managed properly.

Case Description: A 66-year-old man with a history of complete C7 quadriplegia presented with new-onset autonomic dysreflexia that resulted from Charcot spinal arthropathy (CSA). Pathologic instability, in the atypical site of the mid-thoracic spine, spanning from the T8–T9 vertebral levels was appreciated on physical exam as an audible, palpable, and visible dynamic kyphosis; kyphosis was later confirmed on neuroimaging. Based on the CSA severity and sequelae, the patient underwent bilateral decompression laminectomy with lateral extracavitary arthrodesis and posterior instrumentation. Symptoms dramatically improved and at 1-year follow-up, dynamic thoracic kyphosis and most symptoms of autonomic dysreflexia had resolved.

Conclusions: Based on our case and published reports, vigilant imaging and thorough physical examination in long-standing spinal cord injury could help early diagnosis and treatment of CSA, theoretically preventing development of cord atrophy and subsequent long-term sequelae. Surgical correction rather than bracing may be recommended in patients who have complete injury at or above T6 in patients with symptoms of autonomic dysreflexia associated with CSA confirmed on neuroimaging.

Key Words: Charcot spine, complications, long-term spinal cord injury, neuropathic spinal arthropathy, spinal neuroarthropathy

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INTRODUCTION

Charcot spinal arthropathy (CSA), also called Charcot spine, spinal neuroarthropathy, or neuropathic spinal arthropathy, is rare mechanical destructive process that affects the intervertebral disc and adjacent vertebral bodies in patients who have loss of joint protective mechanisms.[3,7] Patients with CSA commonly present with progressive kyphosis, back pain, and sitting imbalance,[26,34] whereas CSA symptoms in patients with

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http://surgicalneurologyint.com/Management-of-autonomic-dysreflexia-associated-with-Charcot-spinal-arthropathy-in-a-patient-with-complete-spinal-cord-injury:-Case-report-and-review-of-the-literature/

spinal cord injury (SCI) can progress with no perceived symptoms until joint destruction occurs. At this point, the hypermobile region of the spinal column can become unstable and impinge the cord. Patients often report a "grinding" noise during transfers, and, rarely, symptoms of autonomic dysreflexia. Typical physical exam findings include a step-off and focal, often dynamic, kyphosis at the site of the Charcot joint.

CSA should be included in the differential diagnosis of patients with SCI who develop autonomic dysreflexia symptoms with transfers or torso movement.[11,20,22,23,27,29] Autonomic dysreflexia typically presents as a sudden increase in blood pressure with associated headaches, flushing, and profuse sweating above the level of the cord injury.[12] It can occur in any SCI patient whose initial injury was at or above the T6 level because this region contains the sympathetic connections between the brain and splanchnic vascular bed. Most importantly, autonomic dysreflexia can represent a medical emergency when hypertension reaches malignant, life-threatening levels that warrant urgent intervention.[9]

Our case highlights a new-onset autonomic dysreflexia that resulted from CSA in a patient with a long history of complete C7 quadriplegia. Following the course from initial presentation to 1-year postoperative examination, we discuss recommendations for treatment based on our case and review of the literature. Vigilant care is needed for patients living with long-standing SCIs, where one can face extremely uncomfortable or potentially lethal outcomes if not managed properly.

CASE REPORT

History

Our patient initially sustained a C5/6 fracture dislocation while playing rugby in 1974 that resulted in complete C7 tetraplegia (as defined by American Spinal Injury Association as ASIA A). Decades later, he came to our clinic with a 3-month history of gradual onset blood pressure fluctuations (up to 240/135) associated with change in position. Concerning to our 66-year-old patient were episodes of other symptoms, including severe headaches, blurry vision, and hyperhidrosis that affected his left arm and face. Neither the blood pressure lability nor symptoms improved with medications (i.e. clonidine, hydralazine, chlorothiazide) or other interventions (i.e. excision of hemorrhoids, colostomy, abdominal exploration with colon resection). Bladder distention, issues with his straight catheterization regimen, anal fissure, and skin ulcers were also all ruled out as potential causes of the autonomic dysreflexia.

Examination

Physical examination noted no voluntary function below the C7 level, good biceps and deltoid power, normal reflexes for C6 and above, and complete sensory level beginning a few inches above the nipple line. An audible, palpable, and visible dynamic kyphosis was seen in the thoracic spine. CT-guided joint aspiration, along with negative findings for gram stain, bacterial culture, acid-fast bacilli testing, and fungal culture, proved negative for indications of discitis or osteomyelitis but were consistent with CSA. In addition, laboratory analysis of blood and urine revealed no signs of sepsis or urinary tract infection.

Imaging

Plain X-ray films showed evidence of mild spondylosis and well-healed intervertebral body fusion of C5-6; mild lumbar spondylosis; and degenerative changes with anterior osteophytes of multiple thoracic levels with no major deformities, spondylolisthesis, or acute compression fractures. CT showed no evidence of an acute or chronic pathologic process in the brain and marked fluid collection was seen in the T8–T9 interspace with destruction of the intervertebral disc space and degradation of the vertebral bodies [Figure 1]. Magnetic resonance imaging showed a nonenhancing expansive spinal cord lesion that extended from C4-5 to mid-body of T2, which was consistent with syrinx [Figure 2]; its intensity characteristics, which were similar to cerebrospinal fluid (CSF), had worsened compared with an earlier image. Marked kyphotic deformity and high-grade compression of the T8-9 spinal cord were characteristic of Charcot joint features, specifically, destruction of the disc space, formation of a central cystic component in the joint between osseous endplates, and chronic fracture of the T8 and T9 vertebral bodies.

Treatment

We believed that the cause of the syrinx was secondary to the caudal compression that occurred in the thoracic cord because of the Charcot joint. Given this assessment and the CSA severity and sequelae, the patient underwent surgical decompression of the spinal cord with instrumented arthrodesis. With exposure of the posterior elements from T6 through T11, a hypertrophic reactive mass was identified at the articulation of T8–T9 with benign characteristics on visual inspection. A bilateral decompressive laminectomy of T8 and T9 was then performed, along with discectomies of T7–T8, T8–T9, and T9–T10. The epidural mass associated with the Charcot joint was excised, and both T8 and T9 nerve roots were ligated. Pedicle screws were placed into the T6, T7, T10, and T11 vertebrae, then rods connected and reduced. After a complete lateral extracavitary vertebrectomy of T8 and T9, a titanium vertebral body replacement cage placed into the interspace helped to restore anterior column support and achieve interbody arthrodesis from T7 to T10. Finally, a posterolateral arthrodesis was performed from T6 to T10 using a combination of allograft and autograft bone. Biopsy of

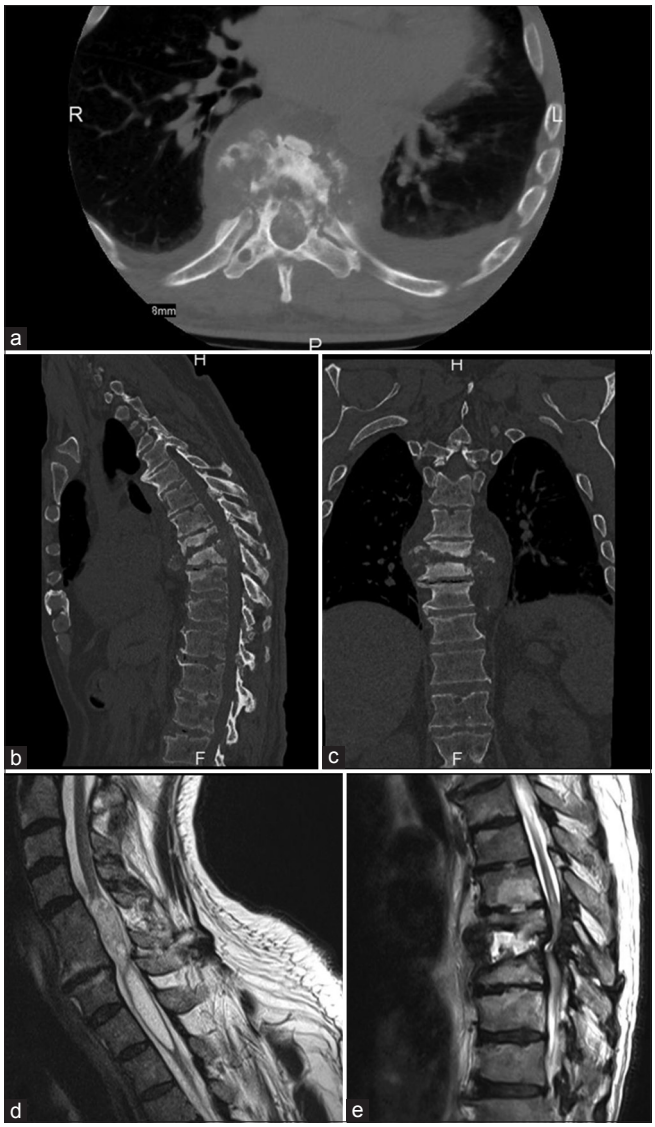

Figure 1: Preoperative imaging in patient with new-onset autonomic dysreflexia and long history of complete C7 quadriplegia. CT imaging in axial (a), sagittal (b), and coronal (c) views showing sclerosis, joint erosion and effusion, paraspinal osseous fluid collection, T8–9 vertebral body destruction with air spaces, and intervertebral disc destruction at the T8–9 interspace. T2-weighted noncontrast MRIs in sagittal view (d and e) shows debris and disorganized fluid within the disc space of T8–9, significant vertebral body erosion of T8–9, complete loss of intervertebral disc integrity at the T8–9 interspace, inflammation and erosion of posterior elements, and proximal syrinx formation from C4–5 to mid-body T2. formation from C4–5 to mid-body T2

the soft tissue and bone at T8–T9 sent for pathologic evaluation was consistent with histological characteristics of CSA.

Postoperative course

The procedure was well tolerated, the postoperative course uncomplicated, and adequate decompression and proper location of all instrumentation confirmed on postoperative imaging [Figure 2]. The patient noted immediate resolution of the hyperhidrosis and

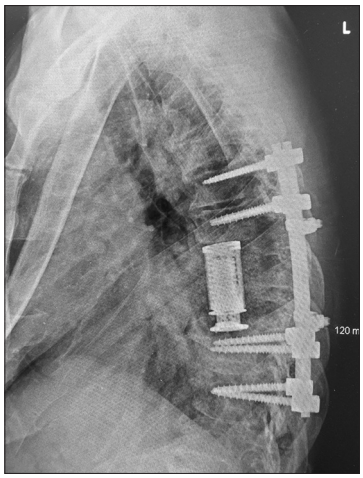

Figure 2: Postoperative plain thoracic X-ray (lateral view) showing vertebral body cage placement and posterior construct with fixation at T6–T7 and T10–T11

flushing; increased stability at the joint was seen with no visible dynamic or static kyphosis. At 1-year examination, MRI and upright X-ray films demonstrated a stable construct, good cord decompression, and resolution of the syrinx [Figure 3]. However, marked atresia of the cord was noted at the site of previous compression. Despite resolution of his most concerning symptoms (hyperhidrosis and flushing), blood pressure lability issues have persisted, likely due to the atretic cord segment visualized on imaging and during the operation.

DISCUSSION

Our patient's urgent symptoms of autonomic dysreflexia were associated with a relatively rare CSA owing to the effects of a long-standing spinal cord injury. Most reported cases of CSA have been found at the thoracolumbar and lumbosacral junctions[4] or at levels immediately adjacent to laminectomies or arthrodesis[34] because of the increased flexibility, weight bearing, and subsequent forces at those locations. Our patient's Charcot joint occurred atypically in the mid-thoracic region and had classic symptoms (i.e., dynamic kyphosis on physical exam, subjective feeling of instability, sterile culture of joint aspirate), characteristic neuroimaging, and histopathologic findings. As supporting care and treatments evolve for patients with spinal cord injuries, our case advocates for routine imaging and thorough physical examination, allowing early diagnosis and treatment of CSA and theoretically preventing further cord atrophy and subsequent long-term sequelae.

Charcot joint, a disease progression

After its initial description by Charcot, the term Charcot joint emerged based on the mid-19th century work that demonstrated an association with tabes dorsalis.[7,32] Following Kronig's 1884 report first describing its manifestation in the spinal column,[15]

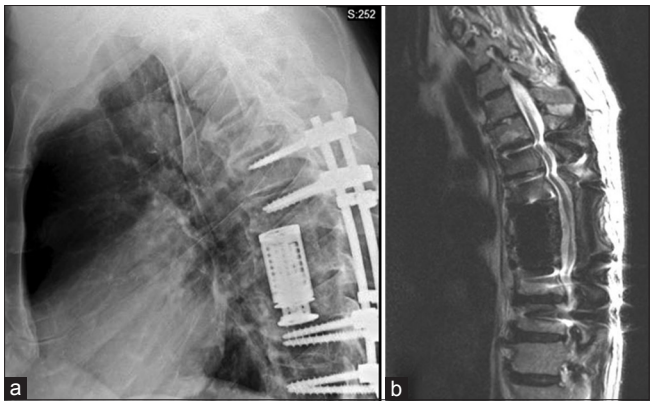

Figure 3:At 1-year follow-up, imaging of thoracic spine. Plain lateral X-ray (a) and (b) sagittal T2-weighted non-contrast MRI (b) showing stable construct, cord decompression, restoration of lordosis, and syrinx resolution

Charcot spinal arthropathy has since been associated with other neurological disorders including syringomyelia, peripheral neuropathies, severe degenerative spinal diseases, hemiplegia, and diabetic neuropathy.[14,30,33,37] Charcot joint develops after spinal cord injury because of repeated microtrauma to joints that no longer have intact protective proprioceptive mechanisms.[36] The early signs of paraspinal bony masses and effusions, progressive malalignment, and signs of stretching ligaments reflect processes are consequences of chronic hyperemia from loss of sympathetic innervation and pro-inflammatory cytokine-mediated osteolysis.[2,17,19] Over time, the intervertebral disc and facet joints are destroyed, resulting in the clinical syndrome of CSA.[18]

The time between onset of neurological impairment and diagnosis of CSA averages 17.3 years.[13] Loss of seated balance is the most common presentation of CSA in SCI; most patients report some kind of back pain or instability.[18]

Charcot spinal arthropathy, considerations in long-term care of spinal cord injury

Our CSA patient's urgent symptoms of autonomic dysreflexia highlight one complexity for SCI patients aiming to live actively long-term. Although the most common cause of autonomic dysreflexia is genitourinary affecting the bladder, large bowel, rectum, or anal canal, other causes include infection, muscle spasms, trauma, and syringomyelia.[10,13,20,21,24,25] In each of these cases, AD symptoms are relieved by sitting up. However, for the AD patient who has symptoms with movement, including exacerbation rather than relief on sitting up, the potential sources of noxious stimuli to consider should be the spine, hips, and sacrum.[4,31] Such noxious stimuli (e.g., caused by cord compression secondary to CSA) can cause a reflex peripheral sympathetic surge via thoracolumbar sympathetic nerves and subsequent subdiaphragmatic vasoconstriction below the level of SCI occurs, mainly in the splanchnic vascular beds. Via carotid baroreceptors, the brain perceives the resulting peripheral hypertension and attempts to correct it via two mechanisms: (1) sending descending inhibitory impulses via the sympathetic tract and (2) slowing the heart rate via the vagus nerve (parasympathetic).[1,5,6] When the cord injury above T6 blocks sympathetic signals from reaching below, vasodilation only occurs above the level of injury. This manifests as hyperhidrosis and flushing above the level of the injury, bradycardia, and extreme blood pressure lability.

Typical, though nonspecific, imaging characteristics of CSA include the "six Ds:" distension (soft-tissue mass), density (sclerosis with preserved bone density), debris (osseous fragments), disorganization (articular contour distortion with intervertebral joint abnormalities), dislocation (spondylolisthesis), and destruction (of endplates and facets).[35] Plain film radiograph findings are often not present until late in disease progression, including decreased fine osseous detail, severe juxta-articular bone destruction, fluffy marginal osteophytes, and dense new bone formation.[8,28] Gas within the disc space, debris, and disorganization have the highest specificity for CSA.[35] Early CT findings of joint space narrowing appear similar to disc and facet joint degeneration.[16] Later in the disease course, soft tissue and bone debris can replace the normal disc space, large paraspinal osseous fluid collections can be present, and both anterior and posterior intervertebral joint elements can be involved. MRI provides the best soft-tissue resolution; compared with CT, its higher sensitivity to inflammatory changes make it best for evaluation of early- and end-stage disease.[16] Typical MRI findings include bone marrow edema and enhancement of vertebral bodies, compression deformities, malalignment; facet joint edema, bone destruction, and disorganization; and paravertebral soft-tissue masses with osseous elements. Characteristics that discriminate CSA from another pathology on MRI are disorganization of the entire intervertebral joint, endplate and facet destruction, listhesis, peripheral disk enhancement, and homogeneous T2 signal hyperintensity with enhancement of the vertebral bodies.[18,35] Important differential diagnoses with similar imaging characteristics to consider include spinal infection, degenerative disc disease, hemodialysis-related spondyloarthropathy, pseudarthrosis, tumor, Andersson lesions, and SAPHO (synovitis, acne, pustulosis, hyperostosis, and osteitis) syndrome.[18]

CONCLUSIONS

Vigilant, routine imaging, and thorough physical examination in patients with long-standing SCI can aid early diagnosis and treatment, thus theoretically preventing the development of cord atrophy and

subsequent long-term sequelae. We believe that symptomatic CSA is best managed with surgical stabilization of the affected joint. In our patient and several other reports, the cervical syrinx resolved after cord decompression, which may contribute to the observed positive outcomes. In asymptomatic CSA, either conservative or surgical treatments can be appropriate, largely based on patient preferences and comorbidities. CSA clearly represents a challenge in long-term spinal cord injury patients, one that can have extremely uncomfortable and potentially lethal outcomes if not managed properly.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initial will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Arnold JM, Feng QP, Delaney GA, Teasell RW. Autonomic dysreflexia in tetraplegic patients: Evidence for alpha-adrenoceptor hyper-responsiveness. Clin Auton Res 1995;5:267-70.
2. Baker N, Green A, Krishnan S, Rayman G. Microvascular and C-fiber function in diabetic charcot neuroarthropathy and diabetic peripheral neuropathy. Diabetes Care 2007;30:3077-9.
3. Barrey C, Massourides H, Cotton F, Perrin G, Rode G. Charcot spine: Two new case reports and a systematic review of 109 clinical cases from the literature. Ann Phys Rehabil Med 2010:53:200-20.
4. Beard JP, Wade WH, Barber DB. Sacral insufficiency stress fracture as etiology of positional autonomic dysreflexia: Case report. Paraplegia 1996:34:173-5.
5. Bloch R. Autonomic dysfunction in management of spinal cord. In: Bloch RF, Basbaum M, editors. Management of Spinal Cord Injuries**.** Baltimore, MD: Williams and Wilkins; 1986. pp 149-63.
6. Brown R, Burton A, Macefield VG. Input-output relationships of a somatosympathetic reflex in human spinal injury. Clin Auton Res 2009;19:213-20.
7. Charcot J. Sur quelques arthropathies qui paraissent dépendre d'une lésion du cerveau ou de la moelle épiniére. Arch Physiol Norm Pathol 1868;1:161-78.
8. Crim JR, Bassett LW, Gold RH, Mirra JM, Mikulics M, Dawson EG, *et al*. Spinal neuroarthropathy after traumatic paraplegia. Am J Neuroradiol 1988;9:359-62.
9. Curt A, Nitsche B, Rodic B, Schurch B, Dietz V. Assessment of autonomic dysreflexia in patients with spinal cord injury. J Neurol Neurosurg Psych 1997;62:473-7.
10. Devlin VJ, Ogilvie JW, Transfeldt EE, Boachie-Adjei O, Bradford DS. Surgical treatment of neuropathic spinal arthropathy. J Spinal Disord 1991;4:319-28.
11. Glennon TP, Madewell JE, Donovan WH, Bontke CF, Spjut HJ. Neuropathic spinal arthropathy after spinal cord injury. A report of three cases. Spine (Phila Pa 1976) 1992;17:964-71.
12. Guttman L, Whitteridge D. Effects of bladder distension on autonomic mechanism after spinal cord injuries. Brain 1947;70:361-404.
13. Karlsson AK. Autonomic dysreflexia. Spinal Cord 1999;37:383-91.
14. Key J. Clinical observations of tabetic arthropathies (Charcot joints). Am J Syph 1932;16:429.
15. Kronig G. Spondylolisthese bei einem Tabiker. Zeit Klin Med 1884;7:5-6.
16. Lacout A, Lebreton C, Mompoint D, Mokhtari S, Vallee CA, Carlier RY. CT and MRI of spinal neuroarthropathy. AJR Am J Roentgenol 2009;193:W505-514.
17. Larson SA and Burns PR. The pathogenesis of Charcot neuroarthropathy: Current concepts. Diabet Foot Ankle 2012;3.
18. Ledbetter LN, Salzman KL, Sanders RK, Shah LM. Spinal neuroarthropathy: Pathophysiology, clinical and imaging features, and differential diagnosis. Radiographics 2016;36:783-99.
19. Mascarenhas JV, Jude EB. Pathogenesis and medical management of diabetic Charcot neuroarthropathy. Med Clin North Am 2013;97:857-72.
20. Mathias C and Frankel H. Autonomic disturbances in spinal cord lesions. In: Mathias C, Bannister R, editors. Autonomic failure: A textbook of clinical disorders of the autonomic nervous system, 4th ed. Oxford: Oxford University Press; 1999. pp 494-513.
21. Mathias C, Frankel H, Cole J. Management of cardiovascular abnormalities caused by autonomic dysfunction in spinal cord injury. In: Illis L, editor. Spinal Cord Dysfunction: Intervention and Treatment. Oxford: Oxford University Press; 1992. pp 101-20.
22. McNeel DP, Ehni G. Charcot joint of the lumbar spine. J Neurosurg 1969;30:55-61.
23. Milligan J, Lee J, McMillan C, Klassen H. Autonomic dysreflexia: Recognizing a common serious condition in patients with spinal cord injury. Can Fam Physician 2012;58:831-5.
24. Mohit AA, Mirza S, James J, Goodkin R. Charcot arthropathy in relation to autonomic dysreflexia in spinal cord injury: Case report and review of the literature. J Neurosurg Spine 2005;2:476-80.
25. Montgomery TJ, McGuire RA Jr. Traumatic neuropathic arthropathy of the spine. Orthop Rev 1993;22:1153-7.
26. Morita M, Miyauchi A, Okuda S, Oda T, Yamamoto T, Iwasaki M. Charcot spinal disease after spinal cord injury. J Neurosurg Spine 2008;9:419-26.
27. Morita M, Iwasaki M, Okuda S, Oda T, Miyauchi A. Autonomic dysreflexia associated with Charcot spine following spinal cord injury: A case report and literature review. Eur Spine J. 2010;19 (Suppl 2):S179-82.
28. Park YH, Taylor JA, Szollar SM, Resnick D. Imaging findings in spinal neuroarthropathy. Spine (Phila Pa 1976) 1994;19:1499-504.
29. Phillips S, Williams AL, Peters JR. Neuropathic arthropathy of the spine in diabetes. Diabetes Care 1995;18:867-69.
30. Race MC, Keppler JP, Grant AE. Diabetic Charcot spine as cauda equina syndrome: An unusual presentation. Arch Phys Med Rehabil 1985;66:463-5.
31. Selmi F, Frankel HL, Kumaraguru AP, Apostopoulos V. Charcot joint of the spine, a cause of autonomic dysreflexia in spinal cord injured patients. Spinal Cord 2002;40:481-3.
32. Slabaugh PB, Smith TK. Neuropathic spine after spinal cord injury. A case report. J Bone Joint Surg Am 1978;60:1005-6.
33. Sobel JW, Bohlman HH, Freehafer AA. Charcot's arthropathy of the spine following spinal cord injury. A report of five cases. J Bone Joint Surg Am 1985;67:771-6.
34. Standaert C, Cardenas DD, Anderson P. Charcot spine as a late complication of traumatic spinal cord injury. Arch Phys Med Rehabil 1997;78:221-5.
35. Wagner SC, Schweitzer ME, Morrison WB, Przybylski GJ, Parker L. Can imaging findings help differentiate spinal neuropathic arthropathy from disk space infection? Initial experience. Radiology 2000;214:693-9.
36. Young MJ, Marshall A, Adams JE, Selby PL, Boulton AJ. Osteopenia, neurological dysfunction, and the development of Charcot neuroarthropathy. Diabetes Care 1995;18:34-8.
37. Zucker G, Marder MJ. Charcot spine due to diabetic neuropathy. Am J Med 1952;12:118-24.