Eur Spine J (2010) 19 (Suppl 2):S179–S182
DOI 10.1007/s00586-010-1296-y

CASE REPORT

Autonomic dysreflexia associated with Charcot spine following spinal cord injury: a case report and literature review

Masahiro Morita · Motoki Iwasaki · Shinya Okuda · Takenori Oda · Akira Miyauchi

Received: 16 June 2009 / Revised: 16 November 2009 / Accepted: 15 January 2010 / Published online: 4 February 2010

Abstract We report the case of a 50-year-old man presenting symptoms of autonomic dysreflexia associated with Charcot spine following complete C8 spinal cord injury. After posterior lumbar interbody fusion of L2/3 with simultaneous posterior instrumentation from L1 to L5, the patient recovered from the symptoms of autonomic dysreflexia. Although the patient began to faint when he sat up and transferred after surgery, it began to be resolved by continuous urinary catheterization, setting a limit to activity and prescription of alpha-, beta-stimulants. Within a few weeks after performing these treatment strategies, he could return to active wheelchair life, and no recurrence of any symptoms was noted at the 6-year follow-up. Although there are only a small number of cases with Charcot spine presenting autonomic dysreflexia, surgical stabilization of the affected lesion for patients with this condition should be recommended.

Keywords Charcot spine · Autonomic dysreflexia · Spinal fusion surgery

M. Morita · S. Okuda · T. Oda · A. Miyauchi
Department of Orthopaedic Surgery, Osaka Rosai Hospital, 1179-3 Nagasone, Kitaku, Sakai, Osaka 591-8025, Japan

M. Iwasaki
Department of Orthopaedic Surgery, Osaka University Graduate School of Medicine, 2-2 Yamadaoka, Suita, Osaka 565-0871, Japan

M. Morita (✉)
Department of Orthopaedic Surgery, Izumi Municipal Hospital, 4-10-10 Fuchu, Izumi, Osaka 594-0071, Japan
e-mail: m1908130@msic.med.osaka-cu.ac.jp

Introduction

The pathological mechanisms underlying Charcot joints have been thought to involve repetitive minor trauma to joints that have lost their protective mechanisms due to destruction of afferent proprioceptive fibers by primary disease such as diabetic neuropathy, tertiary syphilis, anesthetic leprosy, syringomyelia, congenital absence of pain syndrome and spinal cord injury [3]. Destruction of all three columns of the spine can also generate Charcot spine (neuropathic spinal arthropathy).

Spinal cord injury can be responsible for Charcot spine; at the same time, spinal cord injury affecting the cord above the sixth thoracic level is sometimes complicated by autonomic dysreflexia (AD) [7].

Although reports of Charcot spine following spinal cord injury have often been described, Charcot spine presenting AD following spinal cord injury is extremely rare. A case of AD associated with Charcot spine in a 50-year-old man with a 30-year history of complete C8 spinal cord injury is presented.

Case report

This 50-year-old man had a 30-year history of complete C8 spinal cord injury. He had been injured in a fall and sustained a fracture of the cervicothoracic junction. Neurological findings revealed anesthesia in the trunk and lower extremities, along with complete bowel and bladder dysfunction. He had never complained of any symptoms involving the upper extremities, and neurological findings revealed only weakness of abduction and adduction of the fingers. Anterior spinal fusion of the lesion (from C6 to T1) had been performed at the time of spinal injury, and the

postoperative period, during which he was confined to a wheelchair, was uneventful. The lower extremities initially displayed spastic paralysis, but the spasticity disappeared and gradually became flaccid. Thirty years later, he was referred to our hospital with headache and sweating of the head associated with trunk movements, such as sitting and turning over in bed, and the movement to perform self-urinary catheterization caused forced trunk flexion and led to hyperhidrosis and hypertension. His blood pressure during such an episode was recorded as 250/150 mmHg, though the pressure recorded in the decubitus position was within the normal range. He also complained of a crunching sound in his back with postural changes. Radiography of the lumbar spine showed irregular endplate destruction and sclerosis at L2/3. A lateral radiograph in sitting/extension in the decubitus position demonstrated gross instability of the segment, and the range of motion at the lesion site was 24° (sitting: +34°/extension in decubitus: +10°) (Fig. 1a, b). Magnetic resonance imaging showed destruction of the endplates and a fluid-filled cavity at the lesion (Fig. 1c). There were no other abnormal features that suggested malignancy or infection. No other conditions except for trunk movement induced the AD reaction; therefore, the abnormal spinal instability was thought to be the cause of the symptoms. At first, we hypothesized that the spinal instability exerted a stretching force on the renal artery, which activated the renin–angiotensin system and resulted in hypertension and headaches. However, Doppler examination of the renal artery and measurements of the renin–angiotensin system in the sitting and decubitus position were negative. Although we could not determine why postural change caused the AD reaction, a posterior lumbar interbody fusion of L2/3 with simultaneous posterior instrumentation from L1 to L5 was performed. Intraoperatively, manual motion of the lesion reproduced the hypertension, and intraoperative cultures gave negative results for infection. The AD symptoms completely resolved soon after the surgery, and no major complications associated with surgery were encountered, such as instrumentation failure or postoperative infection. However, the patient began to complain of heat and hypohidrosis of the head and sometimes he became unconscious when he sat up and transferred about a week after surgery. Although these symptoms continued for about 2 months, postural hypotension began to be resolved by continuous urinary catheterization, setting a limit to activity and prescription of alpha-, beta-stimulants. Within a few weeks after performing these treatment strategies, he could return to active wheelchair life without pharmacological management of blood pressure, and no recurrence of any symptoms was noted at the 6-year follow-up. Radiographs showed solid arthrodesis at the lesion and no obvious degeneration at the adjacent segments (Fig. 1d).

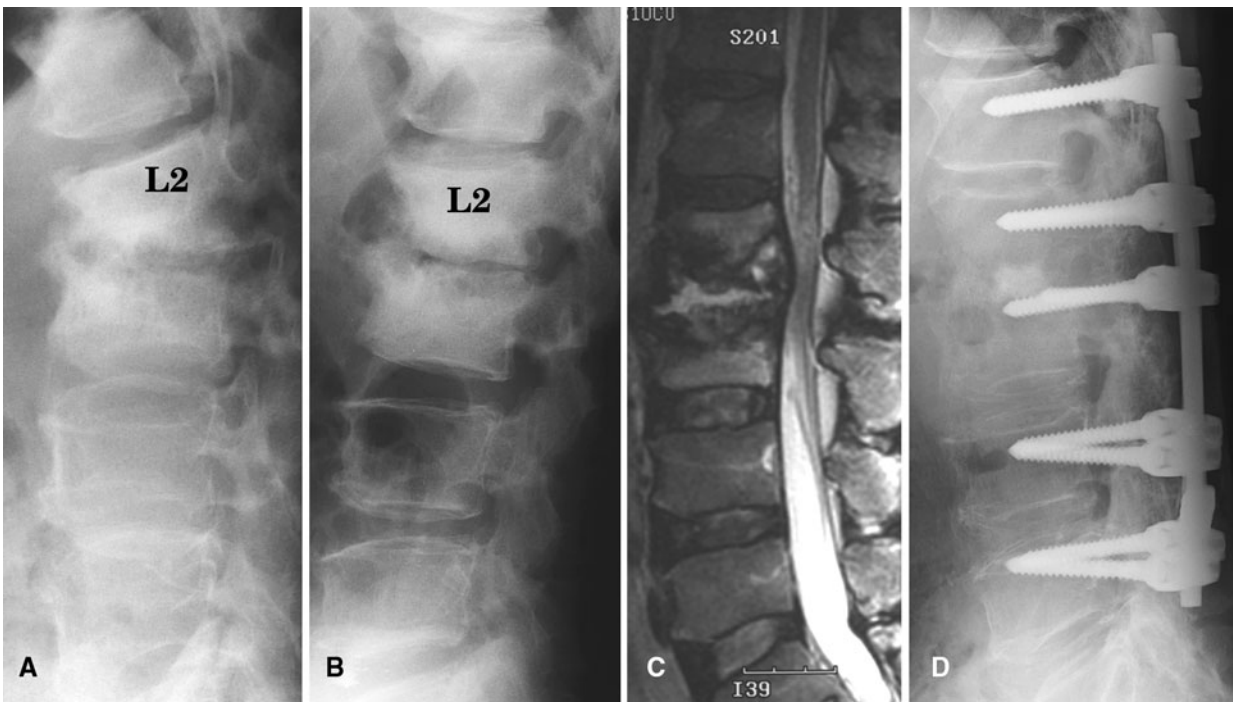

Fig. 1 **a** Lateral radiograph in a sitting position and **b** in extension in the decubitus position demonstrates gross instability of the segment. **c** Sagittal T2-weighted MR image demonstrates destruction of the endplates of L2/3 and a fluid-filled cavity at the lesion. **d** Lateral radiograph in a sitting position 4 years postoperatively shows the arthrodesis with an interbody bone graft at L2/3

Discussion

A search of the English literature revealed 83 cases of Charcot spine following spinal cord injury [11]. Although 19 of these 83 cases had a spinal cord injury above the sixth thoracic level [1, 2, 5, 6, 9, 10, 13–18] (cervical spinal cord injury: 11 cases, thoracic spinal cord injury: 8, complete spinal cord injury: 14, incomplete spinal cord injury: 2, not available: 3 cases), AD associated with Charcot spine is quite rare, with only 5 cases identified in our review of literature [9, 14, 17, 18]. Table 1 summarizes the detailed profiles of these five patients and the present case.

AD may occur in spinal cord-injured subjects with a lesion above the sixth thoracic level, because a lesion above that segment involves an interruption of the connection between the brain and the splanchnic vascular bed, entailing an inability to dilate the vascular bed by central command when needed [7]. AD is provoked by peripheral afferent stimulation below the lesion reaching the isolated spinal cord, and the symptoms are usually associated with disturbance of bowel or bladder function. Theoretically, it is possible that all afferent stimuli below the lesion may induce an AD reaction, but there is no literature supporting a causative relationship between Charcot spine and AD reaction.

Thumbikat et al. [17] suggested, in their case report of L4/5 Charcot spine, that spinal instability exerting pressure on the presacral plexus of nerves and retroperitoneal viscera would induce the AD reaction. Based on the progression of spinal instability caused by Charcot spine, adhesions with surrounding tissues and cicatrization of the dura mater at the lesion of Charcot spine could be severe and sometimes result in transection of the dural tube and neural elements [9]. In our case, continuity of the stiffened dura mater surrounded by severe scar formation at the lesion was observed, and manual motion of the lesion reproduced hypertension at the time of surgery. Although only some patients with Charcot spine following spinal cord injury above the sixth thoracic level develop AD symptoms, and triggering factors of AD in patients with Charcot spine are unclear, instability of the Charcot spinal lesion should be considered a possible cause of AD reaction.

Although the most common complaints of patients with Charcot spine include audible noise on posture change, low back pain independent of the insensitive area, and sitting imbalance [11], all reported cases of Charcot spine with AD, including the present case, complained of symptoms of an AD reaction, such as headaches due to hypertension and sweating, as chief complaints. The symptoms of AD reaction may be one of the clinical manifestations that suggest the occurrence of Charcot spine in patients with spinal cord injury, though Charcot spine is a very rare cause of AD. It is important to note that AD is usually seen in paraplegics and tetraplegics with a lesion above the level of the sixth thoracic segment. On the other hand, the lesions of Charcot spine are typically located in the thoracolumbar spine caudal to T10. Furthermore, previous reports on the incidence of AD in patients with spinal cord injury revealed that AD was more prevalent during the first few years postinjury [4, 8, 12] and decreased in subsequent years (complete tetraplegia) or after 15–20 years postinjury (complete paraplegia) [12]. On the other hand, patients with Charcot spine usually require more than 10 years to develop symptoms caused by this condition after spinal cord injury [11]. Physicians should check radiographs not only at the site of injury, but also at the thoracolumbar and lumbar spine, when paraplegics and tetraplegics with a lesion above the level of the sixth thoracic segment complain of symptoms of AD reaction, especially when the patient had begun to complain of the symptoms after more than 10 years postinjury.

Selmi et al. [14] reported two patients with AD caused by Charcot spine, and the symptoms resolved with 6–8 weeks of bed rest. Although bed rest might prevent the afferent stimuli caused by the instability of Charcot spine, prolonged bed rest would have adverse effects and be stressful to patients, especially in paraplegics who have an active wheelchair life. Considering the cause of Charcot spine, the potential for progression and the fact that

Table 1 Literature review of patients with Charcot spine presenting autonomic dysreflexia

Case	Age/sex	Neurogenic status:level	Interval (year) SCI-Diag.	Lesion of Charcot spine	Treatment	Reference
1	50/M	Complete:C8	20	T11/12	PIF(T11/12) + PSF(T8-L3)	Mohit et al. [9]
2	53/M	Complete:C8	30	T11/12	Bed rest: 6 weeks	Selmi et al. [14]
3	61/M	Complete:C5	30	L4/5/S	Bed rest: 8 weeks	Selmi et al. [14]
4	60/F	Complete:T5	8	L4/5	PSF(L3-S)	Thumbikat et al. [17]
5	25/N.A.	Complete:C8	10	T7/8, L5/S (post T9-L4 PSF)	PIF(T7/8, L5/S) + PSF (T4-S)	Vialle et al. [18]
6	50/M	Complete:C8	30	L2/3	PIF(L2/3) + PI (L1-L5)	Present case

N.A. not available, *SCI* spinal cord injury, *PSF* posterior spinal fusion, *PIF* posterior interbody fusion, *PI* posterior instrumentation

surgical stabilization resulted in amelioration of AD in other reported cases as well as in the present one, we recommend surgical treatment of the affected lesion for patients with Charcot spine presenting with AD, though there are only a small number of cases with this condition.

Conclusion

Surgical stabilization of the affected lesion should be considered a treatment strategy in patients with Charcot spine presenting with AD.

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

References

1. Devlin VJ, Ogilvie JW, Transfeldt EE et al (1991) Surgical treatment of neuropathic spinal arthropathy. J Spinal Disord 4(3):319–328
2. Glennon TP, Madewell JE, Donovan WH et al (1992) Neuropathic spinal arthropathy after spinal cord injury. Spine 17(8):964–971
3. Harrison MJ, Sacher M, Rosenblum BR et al (1991) Spinal Charcot arthropathy. Neurosurgery 29:273–277
4. Helkowski WM, Ditunno JFJ, Boninger M (2003) Autonomic dysreflexia: incidence in persons with neurologically complete and incomplete tetraplegia. J Spinal Cord Med 26(3):244–247
5. Kalen V, Isono SS, Cho CS et al (1987) Charcot arthropathy of the spine in long-standing paraplegia. Spine 12(1):42–47
6. Kapila A, Lines M (1987) Neuropathic spinal arthropathy: CT and MR findings. J Comput Assist Tomogr 11(4):736–739
7. Karlsson AK (1999) Autonomic dysreflexia. Spinal Cord 37:383–391
8. Lindan R, Joiner E, Freehafer AA et al (1980) Incidence and clinical features of autonomic dysreflexia in patients with spinal cord injury. Paraplegia 18(5):285–292
9. Mohit AA, Mirza S, James J et al (2005) Charcot arthropathy in relation to autonomic dysreflexisia in spinal cord injury: case report and review of the literature. J Neurosurg spine 2:476–480
10. Montgomery TJ, McGuire RA (1993) Traumatic neuropathic arthropathy of the spine. Orthop Rev 22:1153–1157
11. Morita M, Miyauchi A, Okuda S et al (2008) Charcot spinal disease after spinal cord injury. J Neurosurg Spine 9(5):419–426
12. Noreau L, Proulx P et al (2000) Secondary impairments after spinal cord injury: a population-based study. Am J Phys Med Rehabil 79(6):526–535
13. Pritchard JC, Coscia MF (1993) Infection of a Charcot spine. Spine 18(6):764–767
14. Selmi F, Frankel HL, Kumaraguru AP et al (2002) Charcot joint of the spine, a cause of autonomic dysreflexia in spinal cord-injured patients. Spinal Cord 40:481–483
15. Sobel JW, Bohlman HH, Freehafer AA (1985) Charcot's arthropathy of the spine following spinal cord injury. J Bone Joint Surg Am 67(5):771–776
16. Thomason K, Emran IM, Chan D (2007) Shortening osteotomy for the treatment of spinal neuroarthropathy following spinal cord injury. A case report and literature review. Eur Spine J 16(3):318–321
17. Thumbikat P, Ravichandran G, McClelland MR (2001) Neuropathic lumbar spondylolisthesis: a rare trigger for posture-induced autonomic dysreflexia. Spinal Cord 39:564–567
18. Vialle R, Mary P, Tassin JL et al (2005) Charcot's disease of the spine: diagnosis and treatment. Spine 30:315–322