Virchows Arch (1998) 433:487–491

CASE REPORT

Erich Schlüter · Karlheinz Heuser

Dramatic development of quadriplegia in a young man while engaged in heavy manual labour: a post-traumatic spinal cord injury?

Received: 30 January 1998 / Accepted: 23 June 1998

Abstract A healthy 16-year-old male suddenly developed paraesthesiae in his hands during heavy manual labour. He was able to walk to the nearest doctor's practice himself. The symptoms worsened dramatically, and within a short period of time cardiac arrest occurred. After resuscitation and intensive care, quadriplegia due to a high cervical cord lesion was diagnosed. The patient died 70 days later without any changes in the neurological symptoms. Post mortem revealed severe focal ischaemic lesions in the cervical spinal cord in a mixed arterial/venous distribution, but no evidence of direct traumatic changes in the spinal cord, the spine or the soft tissues. All other possible causes than trauma were excluded. The clinical development of the symptoms suggest that this spinal cord lesion should be classified as a secondary traumatic spinal cord injury caused by a subluxation of the cervical spine. The pathogenesis of post-traumatic ischaemic damage to the spinal cord appears to be related to localized hypercoagulability resulting in the formation of microthrombi. Impaired microcirculation in a limited area and for a limited period of time may have caused the irregularly distributed ischaemic necrosis.

Key words Spinal cord trauma · Quadriplegia · Hypercoagulability · Microthrombi · Microcirculation

Introduction

Sudden spinal cord injury in a young person who is not suffering from any other neurological disorder or cardiovascular disease may be due to trauma.

We describe a 16-year-old male patient who suddenly noticed paraesthesiae while loading heavy wooden panels. Shortly thereafter he developed quadriplegia. This severe neurological damage failed to show any improvement, and he died 70 days later with adult respiratory distress syndrome, bronchopneumonia, acute prostatitis and signs of circulatory failure.

E. Schlüter (✉)
Department of Pathology, University of Kiel, Michaelisstrasse 11, D-24105 Kiel, Germany
Fax: +49-431-597-3428

K. Heuser
Department of Neurology, University of Kiel, Kiel, Germany

Clinical history

The patient, a 16-year-old male, who had always been in good health, suddenly became ill while at work. He was loading a large number of wooden panels, each measuring 250 cm × 120 cm × 5.4 cm and weighing 90 kg. Suddenly he felt very intense pain in his neck and a prickling sensation in his fingers. At first he continued working. Immediately after loading the last board the patient became "more and more agitated – almost panicky", according to a fellow worker, and went to see the doctor. Accompanied by his colleague, he walked the approximately 300 m to the nearest doctor's office himself. By the time he got there he was suffering from dyspnoea, was breathing rapidly and had to lie down because of increasing paresis. Shortly thereafter he became unconscious and suffered cardiac arrest. The emergency doctor who was called in was able to resuscitate him with cardiac massage. The patient was subsequently transported to the nearest county hospital by rescue helicopter. For about 16 h he was in a coma. Five days later he was transferred to the Neurological Hospital of the University of Kiel with complete paralysis of the extremities. The paralysis gradually became spastic, and hypesthaesia and hypalgaesia developed in the right arm and caudally at T2 (right) and T5 (left). Although his posture sense was intact, the patient's vibratory perception was diminished on the right. He was conscious, but required continuous artificial ventilation. His blood pressure and heart rate were normal and stable. ECG, EEG, angiography and X-rays of the skull and the spinal cord failed to show any pathologic changes, nor did repeated MRI scans of the spine and the CNS with or without contrast medium (Fig. 1) reveal any disorders during the entire time he was in the clinic.

Five days after the primary event the sedimentation rate was 10/35 mmHg per hour and 10,100 leucocytes/mm^3 were found in the blood. The pressure of the CSF was 14 mm H_2O. The fluid was clear and colourless. The CSF contained 37 mg/dl protein and 0.001×10^2 leucocytes/l. The Queckenstedt test revealed a normal rapid rise of pressure. All other laboratory data, especially haematological data, were normal.

There were no changes in the quadriplegia before the patient's death. On the 70th day of treatment he died of heart failure.

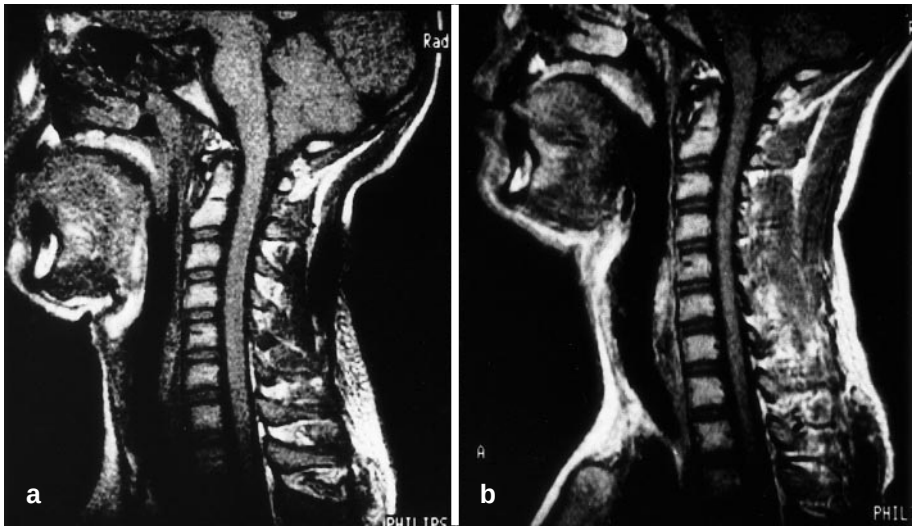

Fig. 1 MRI scans **a** without and **b** with contrast medium. They show no pathologic findings in the CNS

Pathological findings

Apart from the changes in the CNS, all the post-mortem findings could be attributed to long-term ventilation and prolonged circulatory failure. For instance, the typical changes associated with chronic adult respiratory distress syndrome (ARDS) were observed in the lungs, including oedema with a large number of macrophages, hyaline membranes, and interstitial fibrosis. There were also a few bronchopneumonic foci. As a result of chronic ARDS, right-sided ventricular hypertrophy and global heart failure had developed. In addition, there was acute prostatitis and signs of a terminal paralytic ileus – frequent complications in paraparesis or quadriplegia. Otherwise there were no pathological changes outside of the CNS. In particular, neither primary heart disease nor signs of trauma were identified in the spine, its ligaments, its muscles, or the associated soft tissue.

The most important findings were those in the spinal cord, which was cut into approximately 1-cm-thick slices and totally embedded in paraffin for histological examination. Microscopically, the upper 20 cm of the spinal cord showed multiple irregular areas of necrosis in the stage of resorption (Fig. 2). Predominantly the grey matter was affected. In some areas cysts had already formed. Along with accumulations of lipophages there was a proliferation of glial cells and glial fibres, especially in peripheral regions. Occasional capillaries containing hyaline thrombi were found; these were partially incorporated into the capillary wall and endothelialized (Fig. 3). The adjacent, nonnecrotic portions of the white matter were spongy and porous. This spongy porosity together with degeneration of myelin (wallerian degeneration) was particularly prominent in the posterior white columns of the upper 5 cm of the spinal cord. The necrotic areas were particularly numerous and extensive in the upper 10 cm of the spinal cord. The changes gradually decreased towards the thoracic part, becoming vir-

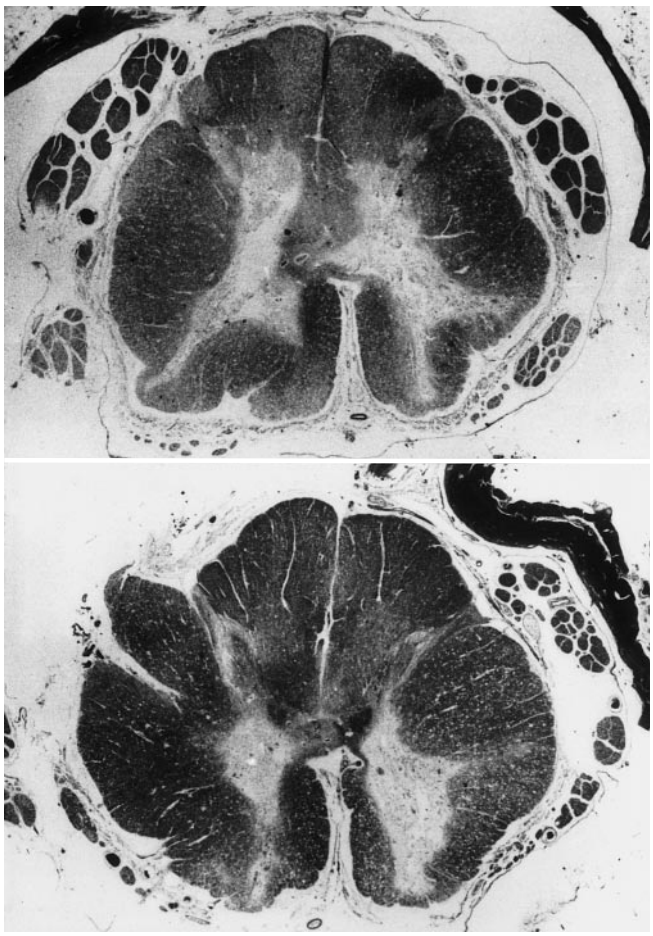

Fig. 2 Patchy areas of necrosis, predominantly in the grey matter of the upper 20 cm of the spinal cord. Klüver-Barrera, approx. ×8

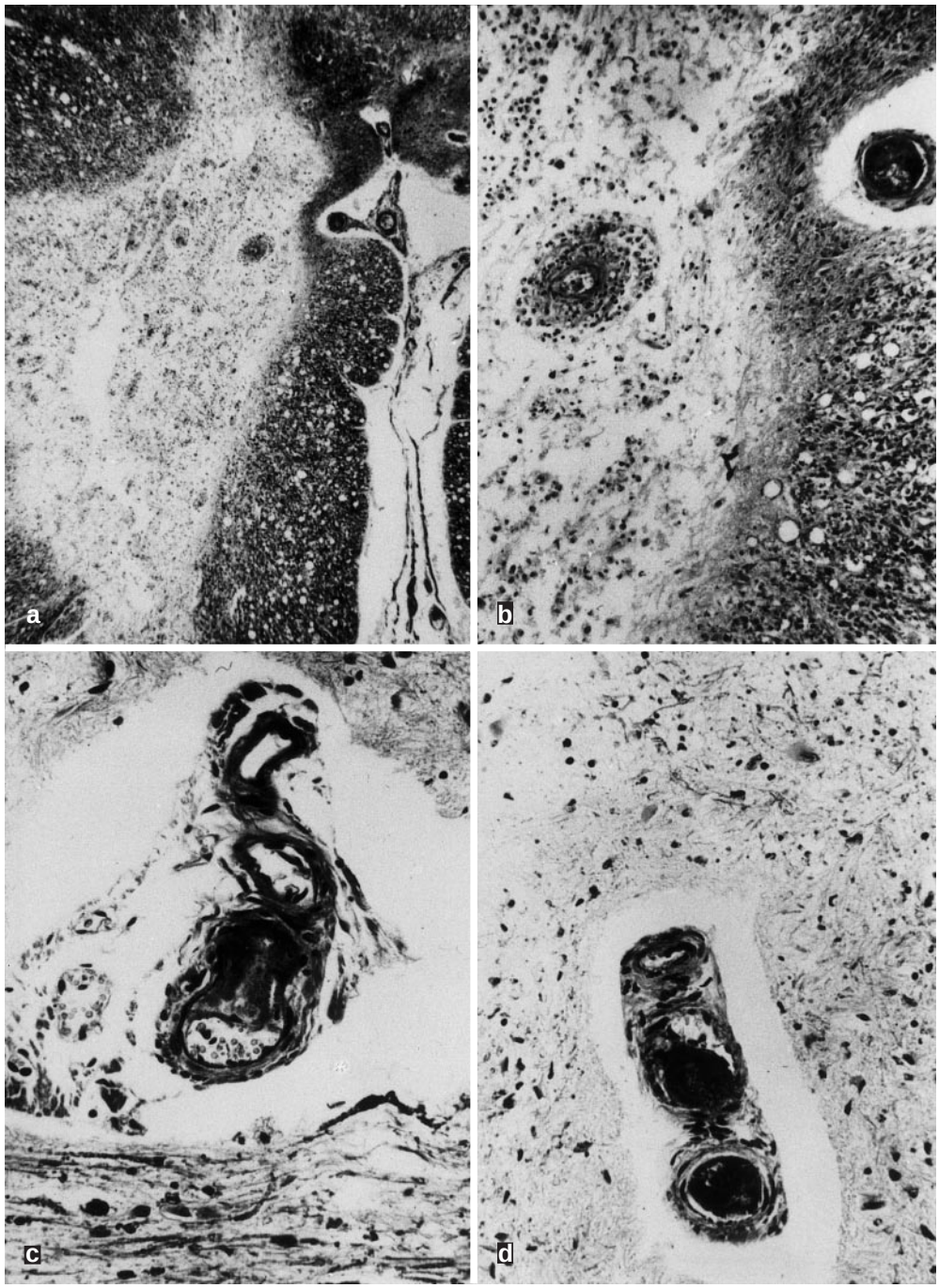

Fig. 3a–d Multiple old, incorporated, recanalized and endothelialized microthrombi in the neighbourhood of the old necrotic areas in the spinal cord. **a**, **b** Klüver-Barrera, **a** ×78.75, **b** ×312.5; **c**, **d** Martius scarlet-blue (MSB), **c** ×771.25, **d** ×771.25

tually undetectable below the first 20 cm of the spinal cord.

Discussion

The sudden onset of the disease while the young man was loading wooden boards, each weighing 90 kg, strongly suggests a mechanical injury such as a spinal fracture or a torn ligament. However, neither radiological nor post-mortem examination revealed any sign of traumatic damage to the spinal column; only ischaemic changes were found in the spinal cord. Therefore, the pathogenesis of this acute myelopathy was related to focal circulatory disturbances, even though it was evidently traumatic in origin.

The classification of acute myelopathies of vascular origin proposed by Castro-Moure et al. [4] distinguishes three main categories: (1) arterial lesions, (2) venous lesions and (3) mixed arterial and venous lesions.

In our case an arterial origin of the myelopathy could be excluded. Watershed necrosis due to general circulatory failure is unlikely because watershed necroses typically appear in the thoracic spinal cord (T4–T7) or in the

lumbosacral region. The histological findings exclude watershed necrosis, because there were unchanged nerve cells at the borders of the necrotic areas. The clinical symptoms also argue against this possibility, since the signs of spinal injury (the paraesthesiae) presented before the heart action ceased. Cardiac arrest, on the other side, is a well-documented complication of acute injury to the cervical spinal cord [9, 2, 17].

There was no occlusion of large vessels, including the aorta and the intercostal and the segmental arteries. There was no underlying disease predisposing to an embolic occlusion of spinal arteries (in particular no patent foramen ovale was found). Moreover, the distribution of the spinal injury was not related to well-defined spinal cord or segmental arteries.

The most frequent entity caused by occlusion of small-calibre afferent vessels, anterior spinal cord artery syndrome (ASAS), is generally located in the thoracolumbar region. Moreover, ASAS does not lead either to wallerian degeneration of the posterior columns or to impairment of the vibration sense, as seen in this case. Histologically there was no haemorrhagic infarct, ruling out venous occlusion as a cause of the spinal injury.

The necrotic areas correlate neither with a region of arterial supply nor with the drainage area of a particular vein, appearing rather to be related to a mixture of the two. This finding corresponds to the last category of acute myelopathies of vascular origin [4] following trauma. It also means that the ischaemic lesion in the patient's spinal cord was not due to a traumatic lesion of a single vessel, but to trauma to the spinal cord itself.

The clinical course fits in with this interpretation. Within a short period (approx. 10–30 min) after the acute onset of paraesthesiae in the hands, the symptoms worsened dramatically, with dyspnoea, increase in respiratory frequency, feelings of panic, paralysis of the legs, unconsciousness and cardiac arrest. All these symptoms can be observed in the presence of a spinal cord injury associated with a severe spinal shock. After successful resuscitation and intensive medical care a high cervical cord lesion with complete quadriplegia was diagnosed. It remained unchanged until the patient's death 70 days later.

Histologically, the necrotic areas of the spinal cord showed residues of old microthrombi. The assumption that these disseminated necrotic areas resulted from impaired microcirculation during circulatory shock is unlikely, because so-called shock necrosis is not found in the CNS. It is probably prevented by the so-called centralization of the circulation, but also by autoregulation of the CNS blood flow. Both mechanisms evidently guarantee that the blood flow in the CNS will remain constant even in the case of severe circulatory depression. Since there were also no signs of disseminated intravascular coagulation or so-called shock necrosis in other organs, a shock-related aetiology was ruled out. Instead we assume that the irregularly distributed ischaemia-related necrotic areas were the result of the occlusion of small vessels by microthrombi, which were formed due to the release of tissue thrombokinase (tissue factor) after tissue damage [14]. The predominant distribution of the necrotic areas in the grey matter is consistent with a high concentration of tissue factor in the grey matter [6] and, of course, with its high vulnerabilty to hypoxic/ischaemic damage.

The concept of spinal cord injury secondary to trauma was postulated by Allen as early as 1911 [1]. He had been able to demonstrate (in animal experiments) that permanent loss of neurological function following defined types of spinal cord trauma could be reduced by therapeutic measures. From this it was concluded that some types of trauma-related damage to the spinal cord do not develop until some time after the initial injury. In more recent animal experiments it was found that the microcirculation was reduced in circumscribed areas and for limited periods of time following spinal cord trauma. The necrotic areas that finally developed far exceeded the area of the primary traumatic lesion [21]. This suggested that the occurrence of necrosis is only a question of extent and duration of the reduced blood supply. Such secondary injuries to the spinal cord are referred to as posttraumatic infarct or infarction [20].

In our case the question remains as to the nature of the primary traumatic injury. Is spinal cord trauma conceivable without signs of traumatic injury to the bones, ligaments and muscles of the spine? It has been known for some time that often no obvious traumatic damage is found [19, 23].(Peters 1970) The assumed pathogenesis is subluxation or luxation, particularly in the cervical spine, where spontaneous repositioning occurs [22]. After such repositioning the spine may later show no demonstrable evidence of a trauma. The reason why the secondary traumatic injury developed so extensively in this case was probably that cardiac arrest occurred immediately after the beginning of spinal shock. Without the additional effect of the cardiac arrest, the reduced microcirculation would perhaps only have led to reversible functional disturbances, such as the initial vegetative hyperirritability [15].

Animal experiments have shown that it is possible to influence the microcirculatory changes during the first hours after injury by means of various therapeutic approaches [7, 8, 10, 12, 13, 15, 18, 21]. From these experiments it can be deduced that timely treatment would have been very effective. The most important measures that should have been considered are (1) treatment of the vegetative (sympathetic) hyperirritability syndrome with alpha-1 adrenoceptor blockers [15]; (2) treatment of the hypercoagulability in the phase of reduced microcirculation with heparin [16]; (3) an attempt to counteract the danger of general circulatory depression with the usual therapy for shock; and (4) an attempt to protect the neural parenchyma from further necrosis by means of calcium channel blockers [7, 18, 21] and by neutralizing free radicals with cortisone derivatives and vitamin E [3, 5, 7, 10, 11, 12, 13].

Nevertheless, it must be stressed again that during the entire time up to the patient's death no visible lesion was detected by any of the many repeated imaging proce-

dures, including CT and weighted and unweighted MRT. It was thus impossible to establish a clear aetiology and pathogenesis for this spinal cord lesion, and an exact diagnosis was not possible until after death.

How important immediate treatment would be in similar cases is obvious from the immense difference that must have existed between the primary traumatic injury and the secondary damage to the spinal cord. Initially the patient was able to walk the approximately 300 m to the doctor's office himself. The high spinal cord lesion evidently did not develop until after all the secondary damage had occurred.

References

1. Allen AR (1911) Surgery of experimental lesion of spinal cord equivalent to crush injury of fracture dislocation of spinal column. A preliminary report. JAMA 57:878–880
2. Bohn D, Armstrong D, Becker L, Humphreys R (1990) Cervical spine injuries in children. J Trauma 30:463–469
3. Braughler JM, Hall ED (1992) Involvement of lipid peroxidation in CNS injury. J Neurotrauma 9 [Suppl 1]:1–7
4. Castro-Moure F, Kupsky W, Goshgarian HG (1996) Pathophysiological classification of human spinal cord ischemia. J Spinal Cord Med 20:74–87
5. Cruse JM, Lewis RE, Bishop GR, Kliesch WF, Gaitan E (1992) Neuroendocrine-immune interactions associated with loss and restoration of immune system function in spinal cord injury and stroke patients. Immunol Res 11:104–116
6. Del Zoppo G J, Yu JQ, Copeland BR (1992) Tissue factor localisation in non-human primate cerebral tissue. Thromb- Haemost 68:642–647
7. Faden AI, Salzman S (1992) Pharmacological strategies in CNS trauma. Trends Pharmacol Sci 13:29–35
8. Francel PC (1992) Bradykinin and neuronal injury. J Neurotrauma 9 [Suppl 1]:27–45
9. Gilgoff IS, Ward SL, Hohn AR(1991) Cardiac pacemaker in high spinal cord injury. Arch Phys Med Rehabil 72:601–603
10. Hall ED (1992) The neuroprotective pharmacology of methylprednisolone. J Neurosurg 76:13–22
11. Hall ED, Braughler JM (1993) Free radicals in CNS injury. Res Publ Assoc Res Nerv Ment Dis 71:81–105
12. Hall ED, Braughler JM, McCall JM (1992) Antioxidant effects in brain and spinal cord injury. J Neurotrauma 9 [Suppl 1]: 165–172
13. Hall ED, Yonkers PA, Andrus PK, Cox JW, Anderson DK (1992) Biochemistry and pharmacology of lipid antioxidants in acute brain and spinal cord injury. J Neurotrauma 9 [Suppl 2]: 425–442
14. Harms D (1971) Grundlagen einer pathologisch-anatomischen Diagnostik der Verbrauchskoagulopathie. Blut 23:261–266
15. Krum H, Louis WJ, Brown DJ, Howes LG (1992) A study of the alpha-1 adrenoceptor blocker prazosin in the prophylactic management of autonomic dysreflexia in high spinal cord injury patients. Clin Auton Res 2:83–88
16. Kulkarni JR, Burt AA, Tromans AT, Constable PD (1992) Prophylactic low dose heparin anticoagulant therapy in patients with spinal cord injuries: a retrospective study. Paraplegia 30:169–172
17. Lehmann KG, Lane JG, Piepmeier JM, Batsford WP (1987) Cardiovascular abnormalities accompanying acute spinal cord injury. In humans: incidence, time course and severity. J Am Coll Cardiol 10:46–52
18. Nockels R, Young W (1992) Pharmacologic strategies in the treatment of experimental spinal cord injury. J Neurotrauma 9 [Suppl 1]:211–217
19. Peters G (1970) Klinische Neuropathologie. Thieme, Stuttgart, pp 217–218
20. Sandler AN, Tator CH (1976) Review of the effect of spinal cord trauma on the vessels and blood flow in the spinal cord. J Neurosurg 45:638–646
21. Tator CH, Fehlings MG (1991) Review of the secondary injury theory of acute spinal cord trauma with emphasis on vascular mechanisms. J Neurosurg 75:15–26
22. Unterharnscheidt F (1992) Traumatologie von Hirn und Rückenmark. In: Doerr W, Seifert G (eds) Spezielle pathologische Anatomie, vol 13/VII: Pathologie des Nervensystems. Springer, Berlin Heidelberg New York, pp 485–488
23. Weller RO, Swash M, McLellan DL, Scholtz CL (1983) Clinical neuropathology. Springer, Berlin Heidelberg New York, pp 96–99