Hyperpathia in the central cervical cord syndrome

ANTHONY HOPKINS AND PETER RUDGE

From the Department of Neurology, St. Bartholomew's Hospital, London, and the Institute of Neurology, Queen Square, London

SUMMARY Seven patients are described with hyperextension or flexion injury to the cervical cord. They illustrate the symptoms and signs previously associated with damage to the centre of the cord, in that weakness is greater in the upper limbs than the lower. We do not believe that the relative sparing of the legs can be accounted for on the basis that corticospinal fibres passing to the lumbar anterior horn cells lie laterally in the pyramidal tract, as has previously been suggested by Schneider, Cherry, and Pantek (1954) as there is no evidence that there is such laminination in man. Severe pain in the shoulders and arms was a major symptom in six of the patients, even in those with relatively minor injuries. The nature of this pain was initially often not recognized.

Schneider, Cherry, and Pantek described in 1954 a number of patients with 'the syndrome of acute central cervical spinal cord injury'. These patients had suffered severe hyperextension injuries to the cervical spine. The main clinical signs were of weakness greater in the upper limbs than in the lower, retention of urine, and variable impairment of sensation below the lesion. From a consideration of the stresses involved, and from pathological evidence, Schneider and his colleagues have suggested that the clinical signs can be accounted for by destruction of pathways lying close to the centre of the cervical cord. However, we have come to the conclusion that the explanation of the physical signs given by Schneider and his colleagues is not wholly correct. We report seven patients with this syndrome seen in the past few months.

CASE 1

St. B.H. 491347 A man aged 71 years got up at night to micturate, and fainted after this. He struck his face in falling. He was unconscious for only a few minutes. When he recovered, he found that his fists were involuntarily clenched, and his arms were flexed across his chest. He could not move his right leg. When he arrived at hospital, facial lacerations were noticed. He complained so bitterly of pain in the forearms that radiographs of them were taken and found to be normal. Over the next 36 hours the abnormal posture of his arms resolved.

He was first seen by one of us three days later. The cranial nerves were normal. Tone in the upper limbs was normal as was power in the proximal muscles. There was severe weakness of wrist extension on both sides, and of finger extension on the left. The finger extensors on the right, all the finger flexors, and small muscles of both hands were completely paralysed. The triceps tendon reflex was absent bilaterally. There was a marked weakness of the hip and knee flexors, and of dorsi-flexors in the right leg, with an extensor plantar response. Sensory examination showed areas of hyperpathia\(^1\) in the C\(_5\) dermatome on the right, and in C\(_6\)–D\(_1\) dermatomes on the left. In these areas there was patchy hypalgesia and analgesia. Hypalgesia was also present over the left trunk and leg. Light touch was impaired over the right side of the trunk and leg. The cerebrospinal fluid (CSF) was normal. Radiographs of the cervical spine showed severe degenerative changes, but a myelogram was normal.

The patient was given a light collar and mobilized, with considerable improvement in the strength of his right leg. Over the next weeks he developed wasting of the forearm extensors and small muscles of the right hand. Electromyography of the first dorsal interosseous muscle showed fibrillation and positive sharp waves. The hyperpathia disappeared, but the areas of hypalgesia persisted.

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\(^1\) Hyperpathia is defined in this paper as the unpleasant response to a noxious or non-noxious stimulus that is usually delayed, overshoots the normal, and has a prolonged after-reaction (Noordenbos, 1959).
CASE 2

St. B.H. 490653 A 61 year old railway worker fainted after fasting and struck his face on a desk. He was helped from the ground by colleagues but he found that he could not move any limb. On admission to a local hospital he complained of pain and paraesthesiae in both forearms. The pain was of such severity that the arms were examined radiologically and found to be normal. He was transferred to a neurological unit six days later where he was found to have retention of urine.

Examination showed a periorbital haematoma and some flattening of the curvature of the cervical spine. Tone in the upper limbs was normal. Power was normal in the shoulder girdle muscles, but slightly reduced in the triceps and wrist extensors bilaterally. There was gross reduction of power in the finger extensors and small muscles of the hands, much greater on the right. The triceps tendon reflexes were depressed bilaterally. There was a severe weakness of the hip and knee flexors and dorsiflexors of the foot of the right leg. Both plantar responses were extensor. The only sensory abnormality was hypalgesia on the left below D_{10} dermatome.

The CSF protein was 214 mg/100 ml. Radiographs of the cervical spine showed gross degenerative changes. Iophendylate (Myodil) introduced by the lumbar route showed a complete extradural block at C_{7} vertebra. Dye introduced cisternally demonstrated an almost complete block at the lower border of C_{4} vertebra, but enough contrast medium trickled past to show indentations of the column at the two discs immediately below.

At laminectomy of C_{4-5} vertebrae (Mr. Campbell Connolly), the cord was displaced backwards over a bony bar at the lower border of C_{4} vertebrae. No attempt was made to remove this bar, but the dural ligaments were divided at three levels, and the dura mater left open. Postoperatively there was early improvement in the power of the right leg, and normal micturition returned. Four weeks after the operation, all the small muscles of the right hand, and the abductor digitii minimi of the left hand were still very weak and wasted but the other muscles had recovered. The hypalgesia had largely disappeared. Electromyography of the weakest small hand muscle was normal.

CASE 3

St. B.H. 494645 A kitchen porter, aged 55 years, was seen by a policeman to be intoxicated, standing unsteadily in a shop doorway. He was observed to fall suddenly on his face, making no movement to break his fall. The following morning, he was complaining of pain in his right shoulder and chest. He was ‘unable to move from the bed’ because of this pain and weakness of the legs. He was admitted to hospital. On examination, there was laceration and bruising of his nose. The cranial nerves were normal. He was unable to move his right arm, and the left arm and both legs were generally weak. A right hemianalgesia was noted up to the neck. During the first five days there was hesitancy of micturition.

He was first seen by one of us six days later. There was marked limitation of extension and lateral flexion of the neck. Tone was increased in all four limbs. There was no weakness in proximal muscles of the arms, but gross weakness of wrist and finger extension, finger flexion, particularly of the lateral fingers, and total paralysis of all the small muscles of the hands on both sides. The right upper limb was weaker than the left. Power in the legs was normal. All reflexes were very brisk, and the plantar responses extensor. Analgesia was present in the right C_{6}, C_{7}, and C_{8} dermatomes, and patchy areas of hypesthesia were located in these. Vibration, light touch, and passive movement were all normally appreciated. Below C_{8} dermatome there was patchy hypalgesia down the whole of the right side.

The CSF protein was 82 mg/100 ml. on the day of admission, and two weeks later 40 mg/100 ml. Radiographs of the cervical spine showed considerable degenerative changes. At myelography, the contrast medium was held up at the lower border of C_{6} vertebra with the neck in extension, but passed cranially in flexion. Transverse bars were also present at C_{4/5}, C_{5/6}, and C_{6/7} intervertebral spaces.

He was treated by mobilization in a light plastic collar. Surgical decompression was not advised in view of his continuing improvement.

CASE 4

Q.S. A61474 A man aged 44 years was thrown from his motor scooter. He did not lose consciousness, but was unable to move any limbs. His incisor teeth were injured, so it is probable that the injury to his neck was one of extension. At the hospital, he found that, although he could see his limbs extended, they felt as if they were flexed so tightly that he was ‘trussed up like a chicken’. He complained of very severe pain in the upper limbs, such that the house officer had radiographs taken of them to exclude bony injury. Some hours later he developed a sensation which he described as a ‘severe netterlash’ over the shoulders and upper arms. He was unable to pass urine. At about the same time after the injury his right leg began to recover, and 24 hours later the left. The retention of urine and sensation of netterlash improved after three days. Power began to recover in the arms about 10 days after the accident, but he...
noted impairment in thermal sensation in the right leg at this time. Subsequently there was a steady improvement.

He was first seen by us one year after the accident. At this time he still complained of unsteadiness of gait, weakness of the arms, and impairment of thermal perception in the right leg. In the upper limbs there was bilateral wasting of supraspinatus, biceps, and deltoid muscles. Tone was increased in the pronator muscles of both arms, and in the left biceps. There was weakness of abduction and external rotation of the shoulders, extension of the elbows and fingers, flexion of the fingers, and of the small muscles of the hands. The left upper limb was the more affected. Tone was increased in both legs, more on the left, but the legs were not weak. All reflexes were brisk, and the plantar responses were extensor. Pain and thermal sensation were impaired on the right from the third cervical to the sacral segments, most markedly in the lower segments. The CSF protein was 60 mg/100 ml. Radiographs of the cervical spine showed that C₂ and C₃ vertebrae were congenitally unsegmented. At lumbar myelography the flow of dye was partly obstructed at the lower border of C₂ vertebra. The cord was flattened at this level, and the root sheaths did not fill. It was felt that operation had little to offer at this stage.

CASE 5

St. B.H. 494045 A 59 year old man was struck on the back of the neck, while kneeling, by a piece of timber measuring 130 x 7.5 x 5.0 cm, which fell approximately 3 m. The blow was slightly to the left of the centre of the neck. Immediately after the blow he could not move any limb. His legs had recovered sufficiently after five minutes to allow him to be helped to an ambulance. Over the next few days the power returned in the left arm but he had a persisting weakness of the right arm and hand, which for some weeks was attributed to a radial palsy.

When seen by one of us six weeks after the injury the cranial nerves and the left arm were normal. There was wasting of the back of the right forearm and marked weakness of the right finger extensors and small hand muscles. Power in the lower limbs was normal. All tendon reflexes except the left biceps and supinator reflexes were brisk, and both plantar responses were extensor. Sensory examination showed a slight impairment of thermal and pain sensation over the left side below D₄ dermatome. Electromyography of the weak arm and hand muscles demonstrated a reduced interference pattern of normal motor unit action potentials without fibrillation.

CASE 6

Q.S. A59862 A car dealer aged 32 years fell down some basement steps while on his way to a party. He was already intoxicated. The pattern of the fall is not known, but there was an occipital laceration, and he was unconscious for a few minutes. The next morning he complained bitterly of a sensation like sunburn bilaterally in both arms, and over the upper chest and neck up to the jaw. The sensation was so unpleasant that he refused to wear his pyjama jacket, as it caused him further discomfort. There was apparently some alteration with the nursing staff on this account, and he left the ward. No abnormal physical signs were recorded. Over the next three weeks, this discomfort faded, leaving an area of burning paraesthesia only on the left side of the neck. Five weeks after the injury, he noticed that his finger tips were becoming less sensitive, and the fingers felt stiff. If he flexed his neck, paraesthesia ran round his waist and into his thighs.

He was first seen by us 11 weeks after the accident. There were no abnormal signs in the cranial nerves. Tone was increased in the pronator muscles of the upper limbs, and there was slight weakness of triceps, wrist and finger extensors, and of all small muscles of the hands, more marked on the left. The tendon reflexes were brisk, more so on the left. The lower limbs were normal. The threshold to discrimination between two points on all finger tips was about 10 mm and there was slight hyperpathia over the upper chest anteriorly. The CSF protein was 60 mg/100 ml. Radiographs of the cervical spine and myelography were normal.

CASE 7

Q.S. A61640 A painter aged 45 years fell 4 m from a trestle on to his shoulders, and on to the back of his neck. He did not lose consciousness but found he could move neither arm nor leg. Within a few minutes he noticed paraesthesia in the right arm, and he was then able to call for help on a telephone which happened to be close by on the floor. When the ambulance arrived he could walk to it. At the hospital he complained of severe pain in shoulders and arms. These areas were examined radiologically and found to be normal. There were no abnormal neurological signs, and he was sent home.

Over the next three weeks he found that he could not bear the touch of his shirt on his skin. He sat 'as still as a robot' to avoid the movement of the shirt. This symptom gradually resolved, and he returned to work. Two months later he developed tingling in the arms, buttocks, and thighs on forward flexion of his neck, and some paraesthesia in his finger tips. He was first seen by us at this stage.
Neurological examination was normal apart from brisk reflexes in the left arm. The CSF, radiographs of the neck, and myelography were normal. His symptoms subsequently resolved.

DISCUSSION

Schneider et al. (1954) suggested that, in hyperextension injuries, the lesion responsible for the greater weakness of the upper limbs, the urinary retention, and the variable sensory loss lay close to the centre of the spinal cord. In this and subsequent papers, pathological evidence has been forthcoming (Schneider, Thompson, and Bebin, 1958; Schneider, Gosch, Norrell, Jerma, Combs, and Smith, 1970).

Many studies have shown that a sudden compressive force applied directly to the surface of the exposed spinal cord of animals does result in central haemorrhagic necrosis of the grey matter. The lesion increases in diameter, and extends towards the periphery, as the force is increased (Allen, 1914; Freeman and Wright, 1952; Albin, White, Acosta-Rua, and Yashon, 1968). Pantek (Schneider et al., 1954) proposed a model to explain this distribution. A grid was superimposed on the surface of a rubber cylinder representing the cord, and the cylinder compressed from front to back. Study of the displacement of the grid-lines showed that the maximal displacement occurred near to the centre of the 'cord'. The strain occurring with slow compression is, however, not necessarily closely related to the forces acting in acute injuries.

If it is accepted that the lesion lies in the centre of the cord, can the clinical findings of weakness mainly affecting the upper limbs and retention of urine be explained by the anatomy of the cord? Schneider and his colleagues accepted Foerster's statement (Foerster, 1936) that the lateral corticospinal tracts are laminated so that the fibres passing to the cervical segments lie medial to those going to the more caudal segments (Schneider et al., 1954, Fig. 21). They state 'from our experience with cases 2 and 8, the fibres to the internal rotators of the hip seem to lie most lateral in the pyramidal tract, and those for finger movements most medial'. Nathan and Smith (1956) in a review of the literature, could find no evidence for such lamination and there is considerable evidence against it in higher primates caudal to the level of the pons (Barnard and Woolsey, 1956; Liu and Chambers, 1964). All the data have recently been reviewed by Noback and Harting (1971).

If there is no somatotopic organization of the corticospinal tract as it descends the length of the spinal cord, how can a central medullary lesion account for severe weakness of the hands, as is seen in our patients, with relative sparing of the lower limbs? One explanation is that, although somatotopic organization is absent as the corticospinal tract descends the length of the cord, some segmental organization must take place as fibres pass medially to their termination. They form axosomatic or axodendritic synapses chiefly on interneurones in the nucleus propius, basal parts of the posterior horn, intermediate zone and central horn (equivalent to Rexed's laminae 4–9). These fibres as they pass medially must be more susceptible to a central medullary lesion. A lesion of this type in the cervical region would thus damage a large proportion of the corticospinal fibres destined for these segments. However, as the fibres passing to the lumbar neurones are randomly distributed in the descending corticospinal pathways in the cervical region, the tracts would have to be destroyed completely by an injury at this level before every fibre to a group of lumbar anterior horn cells was interrupted. Thus a central lesion in the cervical cord would not be expected to cause as much weakness in the legs as in the arms. Furthermore the focal wasting in the upper limbs seen in cases 1, 2, 4, and 5 suggests that in these cases the lesion extended into the anterior horn. This would accentuate the disproportion of the weakness between upper and lower limbs.

Of particular interest was the unpleasant, painful, burning or stinging sensation which occurred over the chest or upper limbs. This has been mentioned in passing by Schneider et al. (1958) and by Symonds (1953) but we believe it to be an important clinical point, affecting six of our seven cases. In a number it led to limbs being examined radiologically in a search for fractures as the pain was so severe. The abnormal sensation was frequently accentuated by contact with clothing or sheets. The sensation slowly resolved over a few weeks. Hyperpathia could not then be demonstrated, although part of the areas remained hypalgesic in some cases.
From the distribution of the hyperpathia we think that this symptom is due to damage to the spinothalamic fibres at their decussation in the cord. We do not think that it is caused by damage in the roots or in the spinothalamic tracts, since in no patient did the hyperpathia extend to the segments below D5. It is difficult to envisage the dorsal roots as low as this level being involved by traction in cervical injuries. Some patients, including those with hyperpathia in the upper limbs, did have abnormalities of pain and temperature sensation, but never hyperpathia in the lower limbs. Presumably the spinothalamic tracts were involved in these cases and this was the cause of the sensory impairment in the lower limbs. The fact that hyperpathia did not occur, however, suggests that an alternative site of damage to the spinothalamic system was the cause of this symptom in the arms and upper chest. The fibres destined for the spinothalamic tract decussate obliquely over several segments. A long central cervical cord lesion extending into the eighth cervical segment could damage fibres from the upper thoracic segments and thus cause hyperpathia confined to the cervical and upper dorsal segments. The hyperpathic state is a temporary one but when it resolves there is often an area of persistent hypalgesia in part of the previously hyperpathic region. This presumably indicates permanent damage to some of the decussating spinothalamic fibres. In cases 6 and 7 further symptoms developed after an interval of several weeks. Both noted paraesthesiae in the fingers and developed Lhermitte’s phenomenon. It may be that gliosis was the cause of these new symptoms, although the resolution of them in case 2 is difficult to explain on this basis. It is possible that the paraesthesiae are associated with a stage of recovery of conduction in some fibres.

All our patients had the clinical picture of central cord damage due to compression. The injury in four of the seven patients reported was caused by hyperextension (cases 1–4) and all had severe degenerative disease of the cervical spine or a congenital anomaly. These four had partial or complete obstruction to the flow of dye at myelography. Symonds (1953) drew attention to the interrelation of trauma and cervical spondylosis in compression of the spinal cord. The mechanism of the injury to the cord is however unclear. Barnes (1948) and Taylor and Blackwood (1948) had shown that the anterior longitudinal ligament could be ruptured in hyperextension and the latter authors proposed that movement of the upper segment of the vertebral column posteriorly upon the lower could cause narrowing of the canal. Taylor (1951) postulated an alternative hypothesis on the basis of myelographic investigations in cadavers. He suggested that the ligamentum flavum buckled forward in hyperextension and narrowed the canal, especially opposite spondylotic spurs. Either mechanism could compress the cord. Subsequently, Schneider et al. (1954) placed great emphasis on hyperextension as the cause for the cord injury and favoured Taylor’s hypothesis. However, our case 5 shows that a direct blow can produce exactly the same syndrome. Further, from cases 6 and 7 it seems that even minor flexion injuries can produce elements of the syndrome, of which burning pain is the most striking. The neurological damage was not initially recognized in either of these patients. Similarly, one of Symonds’s (1953) cases developed the same syndrome after flexion injury. Reversible prolapse of the intervertebral disc does not seem to have occurred in our cases as there was no disc calcification or focal thinning of the disc.

Finally, we would like to emphasize that the syndrome of ‘acute central cervical cord injury’ is commoner than hitherto recognized and can follow relatively minor extension or flexion injuries of the neck.

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REFERENCES


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