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Clinical Case of the Month

Ascending myelopathy in the early stage of spinal cord injury

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A 30-year-old healthy woman was involved in a road traffic accident. She sustained a fracture dislocation of T11/12 with a complete Frankel A paraplegia below T11. She had no associated injuries. High Dose Methylprednisolone was administered according to the NASCIS III protocol (48 h) together with low molecular weight Heparin and gastroprotected medication. Complete transection of the spinal cord and an anterior haematoma from T11 to T12 were confirmed on X rays, CT's and MRI scans. Posterior surgical stabilisation was performed using Isola instrumentation, starting 8 h post injury. Her post surgical period was uneventful except for some episodes of low blood pressure (85/60 mmHg) from which she had no symptoms. On the 12th post operative day, while in the physiotherapy department, she complained of right scapular pain. This occurred every time she was sat up and was associated with paraesthesia of both upper limbs. Two days later she deteriorated neurologically and her level ascended initially to T8 and then to T3. MRI of the spine with and without gadolinium showed spinal cord oedema between C3 and T1. There was no evidence of haemorrhage or syringomyelia. The authors discussed this case making different hypotheses. They are mainly the following: (1) Gradually ascending ischaemia due to a vascular disorder; (2) Double spinal trauma; (3) Ischaemia related to repeated hypotensive episodes; (4) Low grade intramedullary tumour; and (5) Thrombus of the Radicularis Magna artery. The case has been recognised as being very rare and interesting. In the conclusions, the presenting author stresses the importance of adopting MRI-compatible instrumentation for the surgical stabilisation of the spine, and careful monitoring of blood pressure during the acute phase of spinal cord injury. Dr Aito agrees with Mr El Masry about the opportunity of forming a group of clinicians in order to discuss protocols to cope with this devastating complication.

Keywords: spinal cord injury; paraplegia; ascending myelopathy

Introduction

Dr S Aito

Ascending myelopathy is a clinical condition in which ascending paralysis manifests itself during the period following a spinal cord lesion. This clinical condition has been often described in the literature as occurring in the late period after the injury.[1–5] Most authors refer to this occurring at least 2 months after injury, describing cases which manifested several years after the lesion. It is mostly due to the development of syringomyelia that ascends from the lower thoracic segments up to the cervical spine.[6–13] Rare cases of ascending myelopathy during the early period after the S.C.I. (within the first 3 weeks) have been described. Mostly they involve a few segments above the original lesion.

Frankel[14] reported, in 1969, eight patients out of 808 admissions presenting with such a clinical condition. Only one of them ascended from T12 to C7, 17 days after the injury.

Yablon[15] in 1988 described 14 cases of ascending myelopathy during the first 4 weeks after injury, of which one ascended four levels, two ascended three levels, eight ascended two levels and three ascended one level. These cases were due to oedema and MRI studies showed diffuse intrathecal haemorrhage.

The case we are going to present is a rare case of ascending myelopathy in the acute stage of a T12 lesion without any evidence of syrinx cavity that involved the cervical segments.

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Case presentation

Dr S Aito

On 9 January 1998 a 30-year-old healthy midwife was involved in a road traffic accident. She sustained a fracture-dislocation of T11/12 with a complete Frankel A paraplegia below T11. She had no associated injuries. High Dose Methylprednisolone was administered according to the NASCIS III protocol together with low molecular weight Heparin (Nadroparine) and gastroprotected medication. Complete transection of the spinal cord and an anterior haematoma from T11 to T12 were confirmed on X rays, CT's and MRI scans. Posterior surgical stabilisation was performed 8 h post injury using Isola instrumentation (Figure 2).

Her post surgical period was uneventful except for some episodes of low blood pressure (85/60 mmHg) from which she had no symptoms. Prior to the accident she was hypotensive with a systolic pressure of about 90 mmHg. Overall feeding and hydration were commenced on the 5th post operative day and the patient started to mobilise early post operatively, as per our protocol, and continued anticoagulation.

On the 12th post operative day, while in the physiotherapy department, she complained of right scapular pain. This occurred every time she was sat up and was associated with paraesthesia of both upper limbs. The scapular pain disappeared immediately when she lay down, but the paraesthesia persisted for a few hours. Two days later she deteriorated neurologically and her level ascended to T8 and then to T3. MRI of the spine with and without gadolinium showed spinal cord oedema between C3 and T1. There was no evidence of haemorrhage or syringomyelia (Figure 3).

Unfortunately the level of the initial spinal cord injury could not be well visualised because the instrumentation was not MRI compatible. On January the 23rd there was a level to pinprick

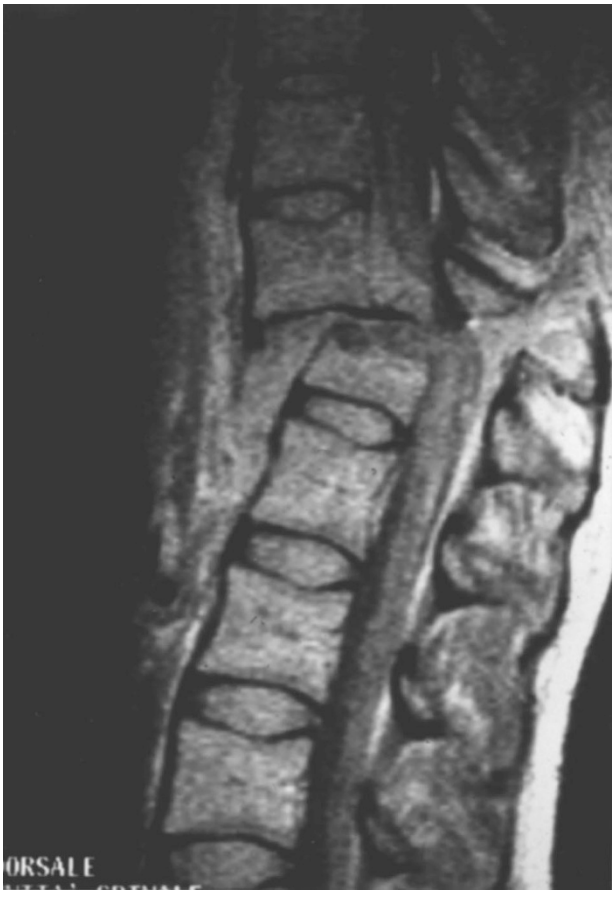

Figure 1 MR of thoraco-lumbar spine immediately after trauma showing T11 – 12 fracture-dislocation and cord transection

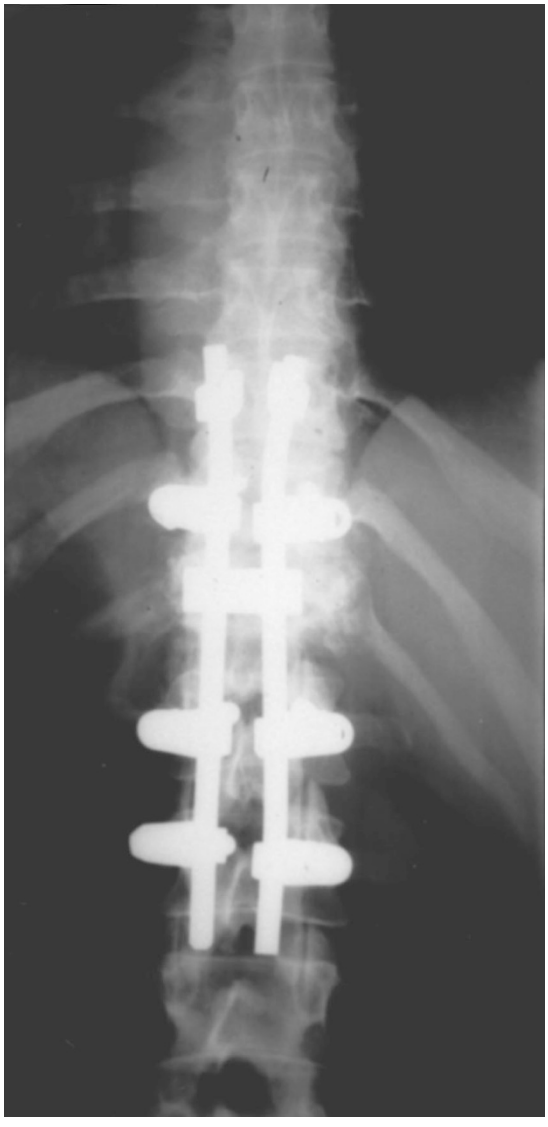

Figure 2 X ray of thoraco-lumbar spine after surgery showing posterior stabilisation using Isola instrumentation

sensation at T5 and light touch at T8. Neurophysiological examination on this day revealed no abnormalities in the upper limbs.

On January the 30th the patient started to experience bilateral dysaesthesia in the dermatomal distribution of C8, T1 and T2, which occurred in the sitting position and tended to disappear when she lay down and her sensory level ascended to T3. Electromyography and motor-evoked potentials (MEP's) performed on February the 2nd revealed normal findings in the upper limbs and confirmed a level at T3. On February the 26th sensory-evoked potentials (SEP) of the medial and ulnar nerves were normal; MEP's of the hypothenar muscles were also normal.

There were however MEP alterations in the responses of the intercostal muscles from T2, as well as signs of motor denervation of the small muscles of the hands supplied by T1. Alterations of the simple skin reflex (SSR) of the hands (Spinal Center situated between T2 and T4) were detected. At this point the pinprick sensation level was T3.

An MRI on March the 17th (2 months after injury) revealed swelling of the cord between C4 and T1 (Figure 4). An MRI performed on June the 5th (5 months after injury) revealed a hyperintense area of the cord at T4 and T5 and an altered signal with clean borders which did not enhance after gadolinium at the level of C7/T1. The spinal cord appeared small (Figure 5).

Other investigations such as Colour Coded Doppler Ultrasonography of the neck and vertebral vessels, MRI of the brain, lumbar puncture and dynamic X rays of the cervical spine were carried out during the early stages of deterioration and were all normal.

The patient was treated with bedrest and steroids for 21 days. Her level settled at T3, however, she experiences recurrent paraesthesia in the territory of T1/T2.

Comment

Mr WS El Masry

Thank you for asking me to comment on your patient. The incidence of this most interesting condition, which was first described by Frankel is

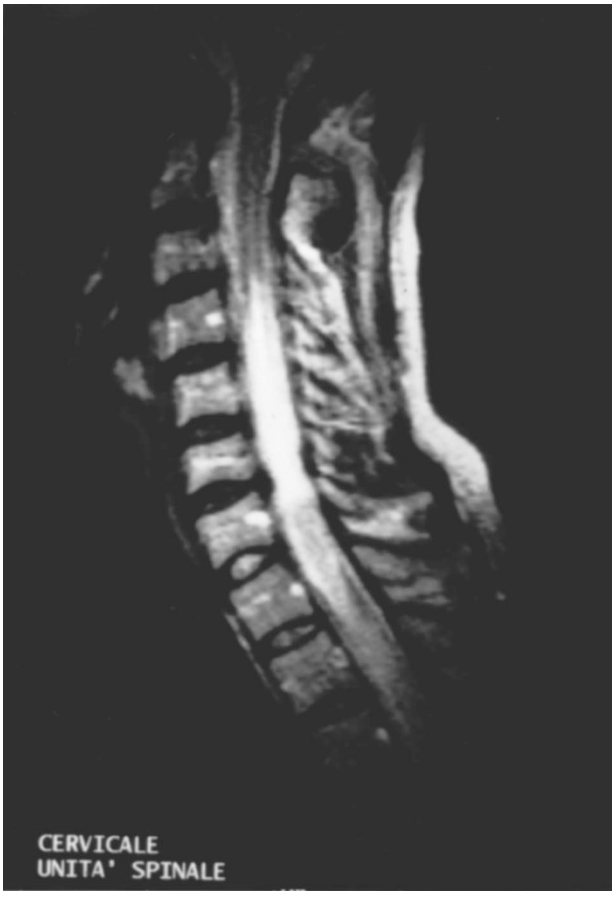

Figure 3 MR of spine performed on January the 23rd (15 days after injury) showing spinal cord oedema between C3 and T1

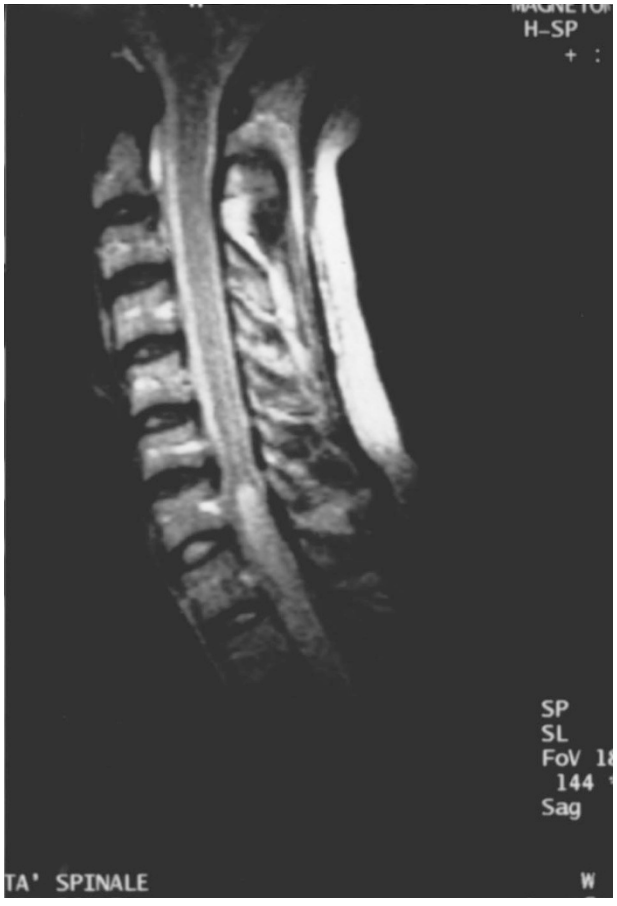

Figure 4 MR of spine performed on March the 17th (2 months after Injury) showing swelling of the cord between C4 and T1

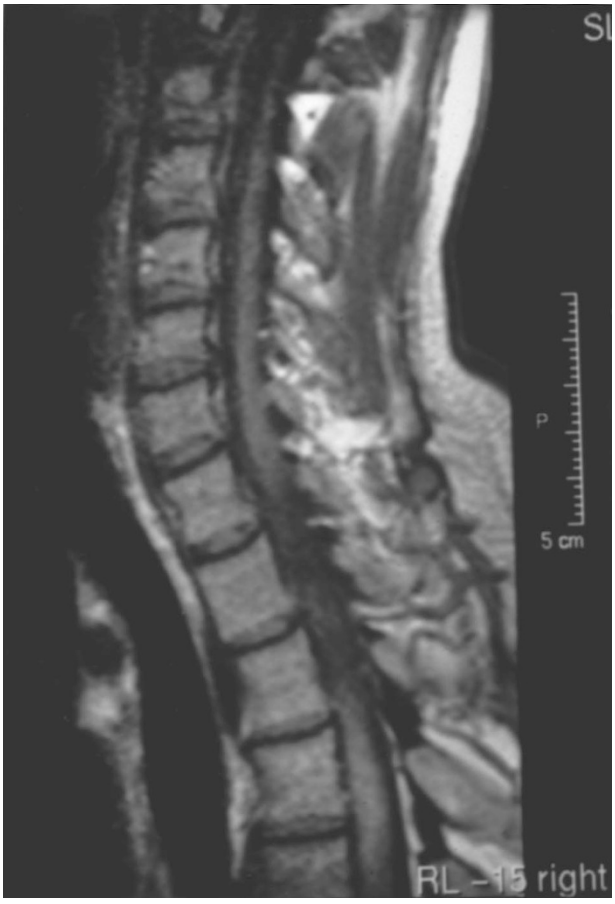

Figure 5 MR of spine performed on June the 5th (5 months after injury) showing well-defined ovoid-shaped lesion at C7–T1

rare and/or perhaps under-reported. The pathophysiology of ascending myelopathy is rather speculative and possibly multifactorial. The first symptoms this lady experienced were those of scapular pain and paraesthesia in the upper limbs suggesting a cervical cord pathology. I understand that initially there was generalised oedema of the spinal cord. On slide 3 the oedema seems to have involved the spinal cord up to the level of C3. Although it is tempting to implicate a second cervical cord injury, in my opinion this is unlikely since all the ligaments of the cervical spine seem to be intact. The question this poses is: can a spinal cord injury occur without any MRI and CT scan evidence of injury to the bony vertebra and/or the ligaments? To my knowledge no one can confidently answer this question. This lady's symptoms were positional and were associated with hypotension, a combination suggestive of a vascular phenomenon. The level of the lesion however did not ascend to T5 until 2 days later (14th day post accident). How this ascent of level occurred is fascinating. Although there is a small possibility that an expansion of the lesion in the cervical cord could have occurred this is unlikely as one would have expected more impairment of motor power and more sensory changes in the upper limbs especially in the hands. Haemorrhage within the cervical cord (possibly related to anticoagulant therapy) could have explained the cervical radiological lesion but not the neurological change. Considering however the practice of liberal administration of anticoagulant therapy to these patients, one would have expected to see many more patients with this condition and more frequently. In any case we need to have some information on the level of anticoagulation prior to and at the time of the neurological deterioration. Although the aforementioned mechanism cannot be entirely excluded, the offending physiology is likely to have started distally at the site of the fracture and progressed cranially. One of two mechanisms are likely to be implicated or they may both have acted in combination. We have demonstrated a fundamental change of the blood flow of the spinal venous system following a spinal injury; with possible reversal of the pressure differential which could result in slow drainage of the cord.[16] This could possibly explain the oedema of the cord but cannot be the only factor otherwise we would have also expected to see more patients with this presentation. Alone this mechanism would not explain the localisation of the lesion in the cervical spine. The other mechanism is possibly one of an ascending thrombus resulting in ascending ischaemia of the cord.

In all similar reported cases the trauma and initial radiological changes are seen in the lower thoracic spine, in the territory of the artery of Adamkiewicz. It would not be unreasonable to hypothesise that the original trauma and/or the subsequent trauma of manipulation/realignment and/or the possible interference with the blood supply of the spine during surgery could have resulted in a thrombus in the lower thoracic main feeder artery which extended cranially and resulted in gradual ascending ischaemia.

Figure 5 could be suggestive of ischaemia in the upper thoracic cord as there appears to be no enhancement to the cord signal by gadolinium. The other possibility is a generalised myelitic reaction of the cord tissue to some autogenous or external substance or organism which would easily cross the disrupted blood brain barrier.

What is certainly clear is that neither high dose methylprednisolone nor surgical stabilisation seem to have provided immunity against an ascending myelopathy in this patient. Unfortunately vital information could not be obtained because the internal fixator was not MRI compatible which should always be borne in mind during surgical stabilisation.

Fortunately the incidence of this devastating condition is very small and the upper limbs usually eventually recover. In view however of our ignorance

of the actual pathology, our inability to halt the ascent of the neurological level, the devastating physical and mental effects of this condition and the small incidence which does not allow any one clinician to develop wide experience; it would be very worthwhile to consider forming a small group of clinicians to discuss protocols of investigations, documentation and management, and centrally reporting patients with this rare condition. Perhaps this protocol could be published in Spinal Cord.

Comment

Dr B Fromm, Dr M Freund and Prof HJ Gerner

In addition to the case presentation listed above, the following information regarding the mechanism of injury was obtained: the injured person was driving when she came off the road, and the car overturned at least once. She was not wearing a seatbelt. The vehicle was equipped with head rests, but no airbags.

The patient was reported to be completely paraplegic below T11 with no clinical signs of head or neck injuries. On admission, radiographs of the entire spine, a CT-scan of the spine and a thoracolumbar MRI were performed. The lateral MR images of the thoracolumbar region made available to us (Figure 1) show a two column injury with anterior and posterior disruption and flexion spondylolysis of T11/12, suggestive of a flexion-distraction injury according to the AO classification,[17] probably with signs of rotation. A further classification necessitates additional radiographs.

The MRI images of the cervical and upper thoracic region 2 weeks after the injury (Figure 3) – after the patient had deteriorated neurologically – show a diffuse swelling and oedema of the spinal cord with its p.m. between C4 and T1, and a disc protrusion at the C6/7 level. A control MRI performed 26 days after the injury (Figure 4) shows a decrease of the spinal cord oedema which now appears most pronounced between C7 and T1/2. A further MRI study done nearly 4 months after the primary injury (Figure 5) shows a gliosis at T2-4 with cyst formation at C7/T1. The appearance of these lesions suggest a traumatic origin.

A literature research (Medline 1986 – 1998) for 'ascending myelopathy' revealed ten articles, most of which were not related to trauma of the spinal cord. The only study linking ascending myelopathy to acute trauma[15] demonstrated ascending neurologic deficits of between one and four segments in tetraplegics and in spinal segments directly adjacent to the original levels of the neurological deficit. Using MRI, Gebarski[18] demonstrated ascending paralysis to occur without cyst formation, a condition named myelomalacia.

The only article reporting spinal cord injuries at the thoracolumbar region ascending during the acute phase after a spinal cord injury to lower cervical or high thoracic levels was written by Frankel[14] in 1969, after a paper he read at the 1968 IMSOP meeting which was discussed by prominent paraplegiologists during the conference. He reported eight patients with ascending neurologic deficits 2 – 18 days after their primary injuries, five of whom sustained a fracture at T12 and had a primary level of paralysis at T12-L4, and ascended to the C7-T7 levels to reach their final neurological deficits between T5 and T11. In this study only patients with a rise of neurological level of at least four segments were included, eight out of 808 admissions during a 14 year period. As for the reasons of the neurologic deterioration, the author mentioned ascending thrombosis, ascending haematomyelia or an inflammatory or necrotising lesion which he named ascending myelitis. In the discussion following the presentation of this paper, Sir Ludwig Guttmann linked the aetiology of these cases to vascular catastrophies like haematomyelia or spinal arterial thrombosis, and AG Hardy added six or seven more cases of his own, also originating from Th12/L1 bony injury levels.

These reports date back to pre-MRI times, and many of the diagnoses mentioned above can be ruled out today by serial MRI images, as performed in the case of this 39-year-old woman. Spinal artery thrombosis or embolism produce a typical MRI picture of a rugged, moth eaten appearance of the anterior columns of the spinal cord. An ascending haematomyelia produces different MRI signals as well.

The MRI appearance of the lesion in this case appears like spinal cord oedema due to primary trauma at the lower cervical and upper thoracic region, healing by scar tissue (post-traumatic gliosis) and post-traumatic cyst formation. The mechanism of the actual trauma, i.e. the car overturning and the injured person not wearing a seatbelt and being thrown out of the vehicle could imply a severe flexion-extension of the head and neck resulting not only in a fracture-dislocation at the thoracolumbar level, but of a spinal cord injury at the cervicothoracic junction as well. The resulting spinal cord trauma could have been aggravated by a primary stenosis at the C6/7 level due to the bulging disc as seen on the MRI imaging (Figure 3), forming a fulcrum in hyperflexion with the spinal cord being unable to escape due to the narrowness of the spinal canal. A detailed contribution to this kind of injury with spinal cord lesions and negative radiology is given by Braakman and Penning.[19]

In summary, the theory of an ascending myelopathy in this 39-year-old lady appears unlikely in our opinion; we would favour a two level injury mechanism probably enhanced by pre-existing degenerative disk disease at the C6/7 level in combination with a severe hyperflexion/extension injury. The MRI pictures and the trauma mechanism would support this theory; why however she was asymptomatic for 2 weeks remains unclear.

Comment

Prof N Di Lorenzo
This case appears to be extremely complex and difficult to clinically define. Different hypotheses can be formulated, and summarised as follows:

(a) A concomitant lesion at cervical level: this does not account for the delay of the symptomatology and its progressive ascending feature.
(b) A concomitant neoplastic mass at the cervical level; in my opinion this explanation can be ruled out by neuroimaging evolution, but is not likely to fit the common neoplastic features.
(c) A tethered cord: although this option cannot be ruled out, it is difficult to correlate an adhesion at the thoraco-lumbar level with a following damage in such a localised segment at the cervical level.
(d) A cervical ischaemia related to repeated hypotensive episodes during the post-operative period.

The last hypothesis is the more likely to occur, assuming that the thoraco-lumbar trauma might have reduced the blood supply to the spinal cord in the territory of the artery of Adamkiewicz, especially during the sitting position. This also accounts for the neuro-imaging of the cervical segments (centromedullary oedema in the thoraco-cervical tract evolving later in a more localised but smaller cervical spinal cord lesion) and for the evolution of the ascending neurological symptomatology.

Comment

Dr G Pellicanò
MRI performed immediately after spinal trauma showed a T12 fracture and posterior displacement of the vertebral body with spinal cord transection. An epidural haematoma was found, spreading to the paravertebral spaces. Due to the neurological level at T3, MRI was performed with evidence of spinal cord enlargement at the C7-T1 level; a focal lesion inside the spinal cord was revealed at that level with similar signal intensity with the cord on SE T1 images and slightly hyperintensity signal on SE T2 images. A high intensity signal area was seen in the cord on SE T2 due to cord oedema with cord swelling, extending from C3 to C7. No lesion enhancement was found after Gd-DTPA intravenous injection.

MRI performed on March the 17th showed reduction of the extension of the high intensity area inside the cord due to oedema reduction; the lesion was more clearly recognisable as an intramedullary one.

On MRI performed on June the 5th, there was a well-defined ovoid-shaped intramedullary mass with a similar signal intensity to the cord on SE T1 images and hyperintensity on SE T2. The lesion extended from the inferior half of C7 to the inferior margin of T1. Also in this case no lesion enhancement was found.

The lesion found in the cord at the cervico-thoracic junction seems to have no direct relationship with the spinal trauma. Due to the intensity signal characteristic, intramedullary location and no enhancement after Gd-DTPA injection, the lesion may be likened, first, to a low grade intramedullary tumour, possibly an astrocytoma. Moreover, it may be thought that the severity of trauma could have broken the tumour balance as demonstrated by the presence of spinal cord oedema and its regression after therapy. No osseous anomalies were found at intramedullary mass level.

Comment and conclusions

Dr S Aito and Dr M D'Andrea
As far as we are concerned, in a retrospective review of 1100 patients treated in our Centre since 1978, we have found seven patients with ascending myelopathy within the early 3 weeks after trauma. Two patients suffering cervical fractures had neurological deterioration during the period under skull traction, two patients with unilateral cervical dislocation during manipulative attempts for reduction, three patients with cervical fractures after surgical stabilisation (anterior approach with arthrodesis and plate), and one patient with a thoracic fracture in ankylosing spondylitis. The upward neurological involvement was restricted to 1–2 levels and all the patients recovered to the initial level within 8 weeks.

This case of ascending myelopathy presents very interesting features as the other authors have pointed out.

It is our impression that the most likely pathogenic factor involved may be a blood flow dysfunction due to a thrombus of the artery of Adamkiewicz with subsequent ascending ischaemia.

A further hypothesis is a tethered cord at the level of the injury (T11–T12), evolving in ascending oedema, with concomitant recurrent episodes of hypotension, leading to a decreased blood supply to the spinal cord cranially.

This second hypothesis can be proposed mainly because of the correlation between posture and neurological involvement.

We do question a two level injury mechanism considering the lack of X Ray and MR signs of ligamentous injuries of the cervical spine, the delay of the neurological symptoms following the trauma, and mainly for the absolute absence of a clinical cohort (pain, limitation or range of movement of the cervical spine).

The hypothesis of an ascending myelitis seems to be very unlikely in view of the rarity of this entity, especially with such a wide involvement of the rostral levels, the lack of responsiveness to high doses of methylprednisolone, and the absence of improvement of the neurological level 1 year after trauma.

The report of Dr Pellicanò (low grade intramedullary tumor) who denies an apparent direct relationship of the lesion at the cervicothoracic junction with spinal trauma is certainly related to the observation of the last images, performed 5 months after injury. We think that this hypothesis does not take into consideration the evolution of the complication, the initial oedema, and, most of all, the symptomatology: why should the neurological deterioration have appeared and disappeared, initially, according to the changing positions (sitting or lying)? Anyway further investigations will be made in order to exclude such a pathology.

More information will be available when we will remove the spinal instrumentation and obtain an MRI of the thoracolumbar tract. However it is relevant to remark the importance of adopting MRI-compatible instrumentation and careful monitoring of blood pressure during the acute phase after a spinal cord injury.

Finally we absolutely agree with Mr El Masry about the opportunity of forming a group of clinicians in order to discuss protocols to cope with this devastating complication.

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