ORIGINAL ARTICLE

Subacute progressive ascending myelopathy following spinal cord injury: MRI appearances and clinical presentation

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Study design: Retrospective Case Review.

Objectives: To describe the clinical presentation and course of patients with magnetic resonance imaging (MRI) features of subacute progressive ascending myelopathy (SPAM). A rare complication of spinal cord injury.

Setting: National Spinal Injuries Centre, Stoke Mandeville Hospital, UK.

Materials and methods: A retrospective review of the case notes and MRI studies of 11 cases with typical MRI features of ascending myelopathy presenting to a tertiary Spinal Injuries centre over a 15-year period.

Results: Eleven patients were identified with MRI features typical of SPAM, a median of 13 days (mean 24, range 4–86 days) following cord injury. The median number of cord segments involved above the initial insult was 6 (mean 6.2, range 4–11). MRI appearances include extension four or more segments cephalad to the initial cord injured segment, cord expansion and increased intramedullary T2 signal with a rim of cord sparing peripherally. Cord changes and neurological deficits improved over time but did not return to the initial injured level on MRI. One patient died (mortality 9%).

Conclusion: SPAM is a rare cause of neurological deterioration following spinal cord injury but may remain subclinical. SPAM most commonly presents as neurological deterioration but may present with shoulder pain, respiratory deterioration or remain subclinical. There are characteristic MRI appearances. It can be fatal.

Spinal Cord (2008) 46, 140–144; doi:10.1038/sj.sc.3102056; published online 3 April 2007

Keywords: subacute progressive ascending myelopathy; MRI; imaging; spinal cord injury; complications

Introduction

Neurological deterioration within 30 days of a spinal cord injury is well described and has a reported incidence of 1.8–6%.^{1,2} It most commonly relates to intervention including early surgical fixation, traction or Stryker frame rotation but may also be secondary to sepsis and vertebral artery injury. Ascending cord injury involving at least four segments in the early period following spinal cord injury was first described by Frankel in 1968. Subacute progressive ascending myelopathy (SPAM) represents a rare, poorly understood neurological complication of spinal cord injury^{3–8} and may develop following an insult to any part of the spinal cord.⁴ Patients typically present with an ascending neurological deficit within 3 weeks of the initial spinal cord insult. MR imaging characteristically demonstrates cord changes ascending at

least four vertebral levels above the initial injury site.⁴ Clinically, the neurological deterioration may extend at least two segments above the level established at the initial insult. Most patients make a good recovery but there is a reported mortality of up to 10%, particularly, if the cord involved ascends to the level of the brainstem.⁵

The cause for this neurological deterioration is not understood. Hypotheses include vascular injury, further mechanical insult, infection and immune-mediated injury, including apoptosis. In the series described to date, a number of interventions have been carried out including decompression, anticoagulation, steroids and osmolar treatments with no benefit.^{3,4}

Materials and methods

The National Spinal Injuries Centre at Stoke Mandeville is a tertiary referral centre with average 175 new admissions of spinal cord injury per annum. Two-thirds of these injuries are post-traumatic.

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Received 23 October 2006; revised 16 January 2007; accepted 11 February 2007; published online 3 April 2007

Eleven cases of patients with spinal cord injury with increased cord signal on magnetic resonance imaging (MRI) extending at least over four segments above the initial injury level were identified through the weekly spinal injuries clinico-radiological meeting between 1990 and 2005.

A retrospective review of case notes and imaging was conducted to examine clinical presentation, indication for MRI, clinical outcome and MRI findings.

MRI studies included those from referring hospitals in addition to those carried out at the NSIC. MR systems used included a 0.2T Hitachi MRP20 and Siemens Symphony 1.5T. All patients had fast spin echo (FSE) sagittal T1-, T2- and axial T2-weighted sequences, with three having axial T1-weighted sequences performed. Only one patient had axial gradient echo sequences.

All MRI studies were reviewed by a neuroradiologist and a radiologist with an interest in spinal cord injury.

Results

Clinical presentation – initial spinal cord injury

The 11 patients comprised nine male and two female patients aged between 18 and 50, with a mean age of 33 years. Ten had traumatic spinal cord injury; seven following motor vehicle accident (MVA) (Figures 1 and 2), two from falls, one following a shotgun injury and one patient with a cervical arteriovenous malformation repair. All patients were well before the initial spinal cord injury, with no significant premorbid history. Clinical assessment of the extent of the initial spinal injury in all cases was undertaken by a Consultant Spinal Surgeon. In all patients, the initial spinal cord insult resulted in a complete injury with each patient documented as ASIA grade A. Nine injuries occurred in the cervical cord and two in the thoracic cord (Table 1). MRI evaluation occurred within 24 h for all patients. Intraspinal haemorrhage was present in five cases, but no significant epidural haematoma was demonstrated. Eight injuries were considered unstable and underwent surgical stabilization within 48 h. Five patients received early high-dose intravenous methylprednisolone. All patients were clinically and neurologically stable before onset of SPAM. Five patients developed a low-grade fever transiently after the initial injury, but only one required antibiotic intervention for a confirmed urinary tract infection. There were no recorded hypotensive episodes in the period following spinal cord injury.

Clinical presentation – SPAM

Following a latent period, clinical deterioration was identified in nine of the 11 patients (Table 1). In three patients, there was a recorded rise in neurological level; one patient complained of dysphagia and three patients suffered respiratory deterioration requiring ventilation. Three patients complained of new shoulder pain with altered arm sensation. Two patients had MRI features consistent with SPAM but their symptoms remained subclinical throughout; one

Figure 1 Patient 6: a 23-year-old male sustained a C5/6 fracture dislocation managed by anterior plate and screw fixation. Sagittal T1- (a) and T2-weighted images (b) acquired on day 3 demonstrate two focal intramedullary haematomas. Breathing difficulties requiring ventilatory support prompted the second scan 12 days after the injury. Sagittal T1- (c) and T2-weighted (d) images on this occasion demonstrate dramatic cord expansion and signal change cephalad to the injury, extending almost to the obex.

had routine postoperative MRI studies and another had a preoperative assessment MRI following transfer from another unit.

MRI findings – SPAM

All patients demonstrated abnormal signal within the cord (Figures 1 and 2) extending at least five vertebral segments above the site of the original injury. The maximum ascension was 11 vertebral segments. The cord changes returned high signal on T2-weighted images and minimal change on T1-weighted sequences (Figure 1). There was mild cord expansion with preservation of a normal rim of cord beyond the cord changes. In five patients, these cord

Figure 2 Patient 5: sagittal T2-weighted image (a) through the thoracic spine performed 2 days after an MVA demonstrates a T4 burst fracture in a 24-year-old male. There are retropulsed bone fragments compressing the cord at the T4 level, associated signal change in the cord extending 2.5 vertebral segments above the injury and a moderate sized intraspinal haematoma. The development of bilateral shoulder pain and weakness prompted a repeat MRI scan on day 86. A Sagittal T2-weighted image (b) demonstrates extensive intramedullary high signal and cord expansion extending to the C2 level. An axial T2-weighted image (c) at the C5 level demonstrates high signal centrally within the cord surrounded by a thin rim of preserved normal cord signal.

changes ascended into the medulla oblongata (Figures 1 and 3). These features were consistent within all 11 patients.

Clinical course

All patients received supportive treatment only. Progressive myelopathy was implicated in one death. Three weeks after the initial spinal injury, a serious chest infection developed in a patient who failed to be weaned from a ventilator and the patient died subsequently.

Recovery from SPAM was observed in the remaining 10 patients. This recovery took between 3 and 14 months. Significantly, in all 10 patients recovery of neurological function was incomplete. Recovery to the original, pre-SPAM, spinal cord injury level did not occur in any of our patients. Improvement was found to be limited to a level close to, but cephalad to the initial injury site.

Follow-up MRI

Imaging in the surviving patients following recovery from SPAM demonstrated a common pattern of characteristic findings (Figure 3) with resolution of the previously

described changes and near normalization of the cord cephalad to the primary spinal injury site. A focal small region of myelomalacia at and approximately one vertebral segment cephalad to the injury site is present on the MRI in all patients on recovery.

Discussion

The syndrome of delayed neurological deterioration by four or more neurological segments following traumatic spinal cord injury is rare but well recognized.^{3,4,8} It was first described in 1968 by Frankel who identified eight patients, an estimated incidence of 1.5% of all spinal cord injuries. He postulated thrombosis or inflammation as the cause. Other smaller series or single case reports identify a pattern, including presentation in the first 3 weeks after injury, a low-grade fever in many cases and characteristic MRI changes of cord expansion and an increased signal on T2-weighted sequences with a medullary distribution.

Neurological deterioration is the typical presentation in the series published to date on ascending myelopathy. Our cases were identified from an imaging perspective and although neurological deterioration was the most common presentation, other presenting features included shoulder pain, dysphagia and increased ventilatory requirement. Two patients remained subclinical throughout with routine imaging identifying the characteristic cord changes. This partly explains the broader temporal presentation in our group compared with other series. Although in our series deterioration most commonly occurred within the classical 3 week period, both subclinical cases were identified at 8–10 weeks.

In patients who have been stabilized hypotheses for this syndrome include arterial occlusion, including fibrocartilage embolism, venous thrombosis,⁹ prolonged hypotension causing ischaemia to more sensitive grey matter,¹⁰ infection, altered apoptotic cascade and an unregulated immune response.⁵ A wide range of treatments have been proposed including anticoagulation, decompression, methylprednisolone and mannitol. There is no evidence for the efficacy or superiority of any particular intervention. The condition is potentially fatal and the mortality in our group and other series was approximately 10%, usually as consequence of respiratory failure subsequent to brainstem involvement. Whether the initial injury was considered stable or unstable does not seem to affect outcome.

The nonspecific pattern of cord involvement on MRI does not provide firm clues as to the aetiology of this condition.

Possible aetiology

A vascular injury to the spinal cord, either arterial or venous, is an attractive possibility. An episode of profound hypotension often accompanies spinal cord injury, although no patients in our cohort had such an episode recorded. Blood pressure measurements were recorded in the initial period following the injuries. It is possible that the frequency of recording was not sufficient to pick up a profound hypotensive episode; however, there were no clinical entries in the

Table 1 Clinical summary

	Age/sex	Insult	Treatment	Initial neurological level	Days to MRI with SPAM changes	Clinical signs	Cephalad extension of MRI changes (vertebral segments)	Outcome
1	43/F	MVA bifacetel dislocation	Surgical reduction at 24 h.	C4//5	10 days	Ascending neurology to C4	C6 to Foramen Magnum (6)	Recovered
2	18/M	MVABifacetel dislocation	Traction	C3/4	14 days	Dysphagia and respiratory deterioration	C7 – medulla (8)	Died 28 days after cord injury
3	49/M	MVA bifacetel dislocation.	Surgery at 4 days.	C6	69 days	Neck and shoulder pain	C6 – C2 (4)	Recovered
4	39/M	MVABurst fracture C6.	Surgery at 23 days methyl prednisolone.	C5	16 days	None	T5 to foramen magnum. (11)	Recovered
5	24/M	MVA burst fracture T4.	Surgery at 24 h	T4	86 days	Shoulder pain	T4 – C2 (8)	Recovered
6	23/M	MVA fracture dislocation C5/6	Surgery at 24 h.	C5	12 days	Respiratory deterioration.	C5 – foramen magnum (5)	Recovered
7	37/M	Gunshot injury T4	None	T4	36 days	Respiratory deterioration	T4 – medulla (10)	Recovered
8	30/M	FallC6 burst	Traction	C5	7 days	Ascending neurology to C3	C5 – foramen magnum (5)	Recovered
9	32/M	Following surgical repair of AVM	None	C5	8 days	Shoulder pains and altered arm sensation	C5 – foramen magnum (5)	Recovered
10	21/F	Fall hyperflexion C6 injury	Surgery at 3 days	C6	4 days	Ascending neurology to C4	C6 – medulla (7)	Recovered
11	23/M	MVA	Surgery at 24 h	C4	13 days	None	C6 – foramen magnum (6)	Recovered

Abbreviations: C, cervical; F, female; M, male; MVA, motor vehicle accident; T, thoracic.

Figure 3 Patient 3: an MRI scan (a) was performed on day 69 following a C6/7 fracture dislocation injury and anterior surgical stabilization to investigate an insidious onset of neck pain and bilateral shoulder weakness. The sagittal T2-weighted image demonstrates intramedullary high signal and marked cord swelling extending to level of the obex. At the level of the injury, there is cystic myelomalacia, probably representing resolving haematoma, whereas central high signal and moderate cord swelling is evident caudal to the level of the injury. Neurological improvement to the early post injury status occurred over 10 months and a repeat MRI scan (b) was performed 14 months post-injury. The sagittal T2-weighted image demonstrates normalization of the cord appearances cephalad to the level of injury with severe cord atrophy and syrinx formation caudally.

case notes to suggest any was missed. The cord distribution affected in patients with SPAM is the central part of the cord. Certainly, the central grey matter of the spinal cord is considered the most sensitive to changes in vascular supply. Although limited to a few cases, in both spinal angiography⁴ and post-mortem studies,⁶ no evidence of arterial or venous occlusion was found. Furthermore, the clinical and radiological pattern of recovery seen in surviving patients argues strongly against an irreversible vascular injury.

It is possible that injuries to the spinal cord result in significant disruption to cerebrospinal fluid (CSF) flow dynamics.^{11,12} Abnormal CSF flow is one theory considered to predispose to syrinx formation within the central cord.¹³ Five patients in our cohort developed intraspinal haematomas following the initial spinal cord injury, potentially producing a mass effect and disruption to normal CSF flow, however, no patients in the group went on to develop a significant syrinx. It is possible that temporary changes in CSF flow may contribute in the development of SPAM. Unfortunately, decompression surgery performed in SPAM patients in other studies failed to produce clinical improvement.

There is some similarity in both the cord appearance and delayed clinical deterioration with other insults to the spinal cord. Patients have developed similar clinical and imaging features following episodes of virally infective myelitis¹⁴ or electric shocks.¹⁵ Viral organisms identified include Herpes Simplex, Epstein–Barr and Coxsackie B4 viruses. The reporting of a mild febrile illness and a CSF lymphocytosis in some cases, although nonspecific, lends support for a possible

infective aetiology. CSF analysis in four patients in our cohort and in other studies failed to identify an infective aetiology.

Altered or unregulated immune response, perhaps to a viral trigger, remains an attractive, but currently unproven hypothesis.^{16,17,18} In SPAM patients, apoptotic cells have been demonstrated at sites within the spinal cord remote to the site of the initial cord injury.^{5,19,20} Certainly, apoptotic cells and an altered immune environment are present following spinal cord injury, but their role in SPAM remains unclear.

Lastly, I think it is important to acknowledge a few limitations of the imaging used in this case review. First, as a tertiary spinal injuries centre, initial imaging was performed at other hospitals in nine of the 11 cases. MRI scanners and imaging protocols clearly differed between each hospital. The universal use of gradient echo imaging to look for acute haemorrhage may have improved the initial imaging assessment, but in practice a good T1 sequence remains a reasonably sensitive tool.

Conclusion

Subacute ascending myelopathy is a rare, potentially fatal complication of spinal cord injury. Onset is most common within 3 weeks of cord injury but may occur up to 3 months after the initial insult. Our observations suggest that MRI changes may occur without a detectable neurological deterioration. Incomplete recovery occurs in the majority of cases.

MRI has a central role in identifying ascending myelopathy, and should be considered in all spinal cord injury patients developing atypical pain, increased ventilatory requirements and low-grade fever, in addition to ascending neurological signs.

Acknowledgements

We thank to Pauline Bates in Medical records and Debbie King in MRI.

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