Spinal Cord Injury Without Radiological Abnormality (SCIWORA)

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Summary

SCIWORA or Spinal Cord Injury Without Radiological Abnormality; is a pre-MRI term that includes injuries to the spinal cord in the absence of radiological (plain radiographs, tomographs and CT scans) evidence of injury to the spinal column or cord. It occurs in skeletally immature spines because of the inherent plasticity of the bony structures in this age group. The prognosis is dependent on the extent of cord damage, and the role of active management is limited. A high index of suspicion is needed to establish a diagnosis. This diagnostic accuracy can be improved with the free availability of MRI scanning for the spines. We describe three cases of SCIWORA with a minimum follow-up of three years and a review of current literature.

Key Words: SCIWORA, Children, Spinal cord injury

Introduction

Spinal cord injury is a devastating event, especially in children. Because of the devastating nature of these injuries and the absence of a cure, prevention is of paramount importance. Nevertheless despite the best attempts to prevent these injuries, they still occur. It is usually obvious when radiographs of the injured spine show a fracture or malalignment of the spine that corresponds to the clinical level of neurological injury. However, there is a category of spinal injuries called SCIWORA, Spinal Cord Injury without Radiological Abnormality; that occurs almost exclusively in the children's spine, and remains a challenge to diagnose without special imaging such as MRI. The prognosis is dependent on the extend of cord damage, and the role of active management is limited. A high index of suspicion is needed to establish a diagnosis. This diagnostic accuracy can be improved with the availability of MRI. The usefulness of urgent MRI scanning is to exclude surgically treatable lesions such as epidural haematomas or localised

This article was accepted: 8 October 2002
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traumatic discs. Three such cases are presented highlighting problems of diagnosis and management.

Case 1

A three year old Malay girl was referred from a district hospital. She was a pillion rider on a motorcycle that collided with a car. She was admitted unconcious, with Glasgow Coma Scale of 8/15 and in hypovolemic shock. She sustained a posterior dislocation of the left hip, closed fracture mid-shaft right tibia, fracture right third to eight ribs with lung contusion and fracture of the frontal bone. She was resuscitated. The hip dislocation was reduced and a full length cast was applied to the right lower limb. CT scan of the brain did not show any intracranial bleeding. Cervical X-rays were normal. She regained full consciousness one week later but was unable to move her left lower limb. She had urinary and bowel incontinence. Repeat X-rays of the cervical and thoracolumbar regions were normal. A working diagnosis of sciatic nerve injury secondary to the posterior hip dislocation was made. Six weeks later, the full length cast in the right lower limb was removed. Neurological assessment showed that both lower limbs were hypertonic and hyperreflexic and she did not regain bladder and bowel control. Muscle power was grade 0 and ankle clonus was present bilaterally. Both upper limbs were normal MRI of the cervical spine showed a post-traumatic epidural haematoma located posterior to the spinal cord from the level of C7 to T6 (Figure 1). The cord was displaced anteriorly and compressed in this region. She underwent emergency decompression, two months after the initial injury. Intra-operatively the cord appeared compressed and atrophic. Post-operatively, the lower limb tone was flaccid and reflexes were absent. The muscle power remained at grade 0. A repeat MRI was done six weeks later. There was no recollection of the haematoma. The spinal cord appeared atrophic from the level of T1 to T6. Presently, it is three years since the initial trauma and she continues to be a paraplegic.

Case 2

A five year old Malay boy was hit by a bus while crossing a road. He lost consciousness and sustained a close fracture mid-shaft of right femur. Cervical X-ray and CT scan of the brain were normal. He was admitted and the fractured femur treated by skin traction. He also had difficulty in micturating and required a urinary catheter. He was unable to move his left leg and had no sensation from the T5 dermatome. Lower limb muscle power was grade 0. Abdominal, knee, ankle reflexes and anal tone were also absent. His upper limb was normal. Thoracolumbar spine X-rays were also normal. MRI was done nine days later showed an atrophic spinal cord from C7 to T2 levels. A repeat MRI done three months after the injury showed an atrophic spinal cord from C7 to T2 level (Figure 2). The smallest anterior-posterior transverse diameter was 1mm. There was no enhancement post-contrast media injection. These features were consistent with post-traumatic myelomalacia. It is now three years after the injury and there was no improvement in the lower limb neurology. He currently ambulates with a wheelchair.

Case 3

A four year old Malay boy was a passenger in a car which collided with a bus. Two occupants in the car died. He had loss of consciousness and was unable to move the left side of his body. The left upper and lower limb muscle power was grade 0 and hypotonic. Reflexes and sensation were also absent. Chest movement was absent on the left side. However, the right upper and lower limbs were normal. The anal tone was also normal. Cervical X-rays showed a pseudosubluxation of C1C2 and CT scan of the neck was normal. CT scan of the brain was normal. MRI done six days later showed a unilateral swelling on the left side of the spinal cord at the level of C1-C2. These features were consistent with Brown-Sequard syndrome. Two weeks after the injury, the neurological deficits had improved to grade 1 in the left upper limb with absent reflexes, the left lower limb was hypertonic with grade 2 to 3 muscle power and brisk reflexes. Four weeks later the power of the left lower limb had improved to grade 4 and tone was normal. He was able to stand, and walked with support. The left upper limb also showed improvement, with hypertonia and muscle power increasing to grade 3. Reflexes were brisk and sensation was normal. At three months, he was able to walk without support. However, he had a hemiplegic gait. MRI of the cervical spine was repeated and it showed minimal hyperintensity at C2 level. The cord at that level had irregular outline on axial scans consistent with myelomalacia. Nine months after the injury, he was able to run and jump. However, he still walked with a mild hemiplegic gait. Muscle power for the left upper and lower limbs remained at grade 4. Three years later he has had no further improvement.

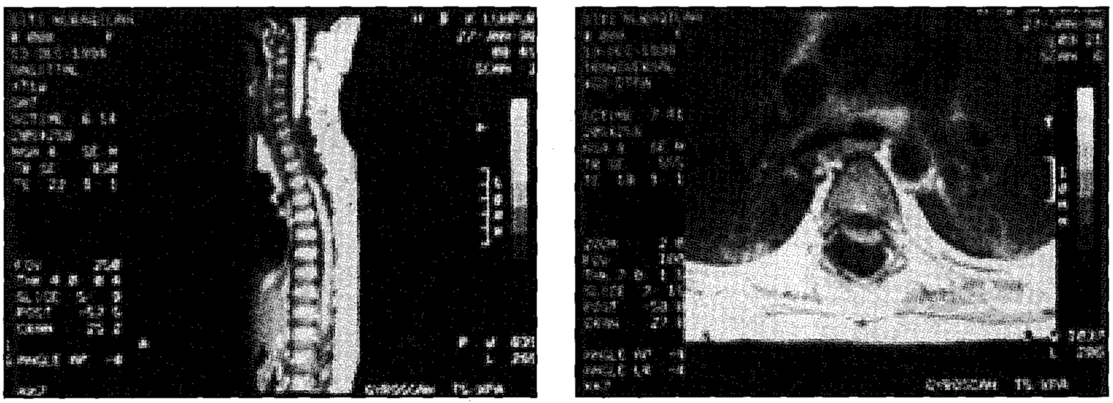

Fig. 1: MRI films showing the presence of an epidural haematoma from C7 to T6.

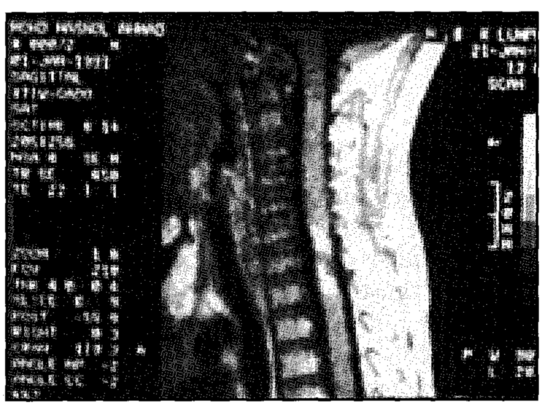

Fig. 2: MRI showing spinal cord contusion from C7 to T2 levels.

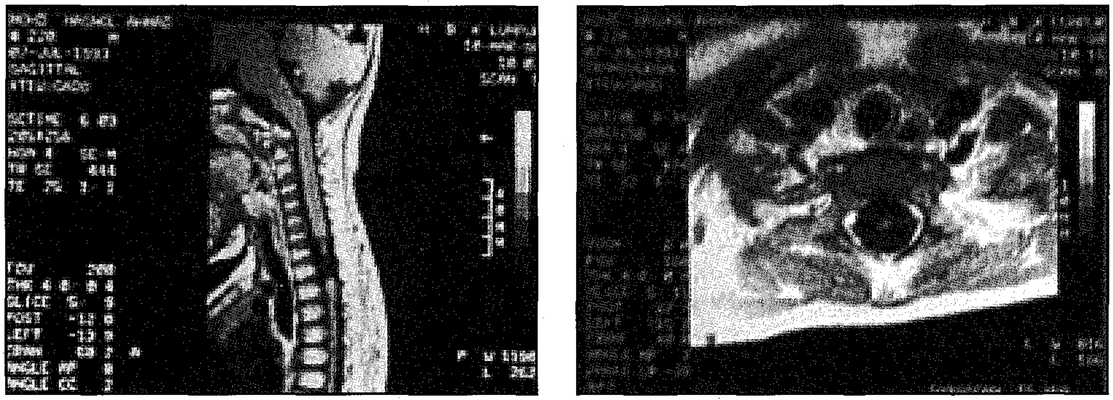

Fig. 3: MRI films taken three months after the injury showing spinal cord atrophy.

Fig. 4: MRI films showing a unilateral oedema of the spinal cord at the level of C1 to C2.

Discussion

Paediatric spine and spinal cord injuries are relatively rare and distinctly different when compared to those occurring in adults. Differences in anatomy, physiology and biomechanics translate into a lower incidence, different injury patterns and different functional outcomes that are unique to the paediatric age group. The incidence of paediatric spine and spinal cord injury is reported to range from 1% to 10% of all traumatic spinal injuries[1,4]. Four distinct injury patterns have been recognized - fracture of the vertebral body or posterior arch, fracture with subluxation, subluxation only and spinal cord injury without radiographic abnormality (SCIWORA). Pang and Wilberger introduced the acronym SCIWORA in 1982 in the time before MRI studies were readily available[3]. This terminology can be confusing since there are abnormalities that can be seen on MRI in patients with SCIWORA.

The incidence of SCIWORA is reported to range from 10% to 66% [1,4,5]. The most susceptible age group is between 0 and 9 years old[1,2]. Therefore, infants and younger children are more commonly affected than older children. There is a predilection for the cervical spine in this age group. The incidence of cervical spine involvement ranges from 20% to 81% [1,2,3,5]. The most common level of injury is C2. Motorvehicle accidents and falls were the most common aetiology of injury[1,3,5]. Other causes include birth trauma, sports related accidents, diving accidents and child abuse. Younger children are more likely to present with complete or severe lesions.

The most common injury mechanisms of SCIWORA are hyperextension, flexion and distraction of the head on the neck. Other mechanisms proposed include indirect spinal cord injury and vascular or ischaemic injury. Most of the existing knowledge on the biomechanics of spinal injury is based on the studies of the adult spine. During moderate degree of hyperextension, the cervical cord can be compressed by the inward bulging of the ligamentum flavum creating up to 50% narrowing of the spinal canal [2]. Exceptionally violent hyperextension will rupture the anterior longitudinal ligament and shear the intervertebral disk of its end plates causing a retrodisplacement of the upper body. Following this, immediate muscle contraction causes elastic recoil and spontaneous reduction. Reflex muscle spasm maintains the reduction giving a normal appearance on plain X-rays. Lloyd also has proposed a similar "Flexion-Recoil" theory in 1907.

The fulcrum for maximum flexion in a child is at C2-C3 and C3-C4, whereas the fulcrum in adults is at C5-C6. Therefore, it is not surprising that the upper cervical vertebrae in children are very mobile. In this case report, *Case 3* had signs of cord contusion at C2-C3 level.

In these three patients, the actual mechanism of injury was not established. From the pattern of injuries sustained, it may be hypothesized that *Case 1* and *Case 2* sustained a hyperextension type of injury while *Case 3* sustained a flexion type of injury. Pang and Wilberger found no correlation between the mechanism of injury and the type of neurological lesions in children with SCIWORA [3].

The diagnosis of SCIWORA can only be made after ruling out the presence of occult fractures that is missed on plain X-rays. Many authors recommend that CT scans should be used to rule out the presence of occult fractures. When available, MRI has a role in revealing most lesions including where surgery may be beneficial [4]. This fact is highlighted in *Case 1* where the presence of epidural haematoma is demonstrated on MRI.

Gross instability must be ruled out by carefully supervised flexion-extension films. If no immediate instability is found, the cervical spine should be immobilized in a well fitting hard collar. Conversely, if instability is demonstrated, either surgical fusion or halo-vest immobilization is recommended. Surgical management has not been of benefit in the majority of SCIWORA patients unless an epidural haematoma is demonstrated on MRI [4].

The incidence of delayed onset of SCIWORA was reported to be 54% (13 of 24 patients) by Pang and Wilberger [3]. Later, Pang and Pollack described 15 patients of 55 SCIWORA patients who experienced the onset of neurological symptoms as late as four days after the initial trauma [2]. They urge that children with trivial head and neck injuries be questioned specifically for transient neurological symptoms and spinal immobilization performed liberally when spinal cord injury is suspected.

The phenomenon of recurrent SCIWORA has been reported by Pollack and Pang [3]. Most of these patients were injured by mechanisms similar to their initial trauma after removing their cervical collars. The second injury was nearly uniformly more severe and associated with a poor neurological outcome.

For children with SCIWORA the long-term prognosis is poor. Younger children are more likely to sustain complete or severe spinal cord lesions. The younger the child, the more deformable the spine and the worse the neural insult. Neither age nor treatment will affect the final outcome and the only prognosticating factor is the initial neurological status. Complete lesions almost uniformly demonstrated no improvements shown in our patients in *Case 1* and *Case 2* who had complete spinal cord lesion and did not regain any useful motor function even after three years of follow-up. On the other hand, patients with incomplete lesions have a generally more favourable outcome.

In summary, spinal and spinal cord injuries in children is relatively rare, therefore a high index of suspicion is essential. The physician attending to a multiply injured child must be aware that children, especially below 9 years old, may have a spinal cord injury despite normal plain X-rays of the spine. It must be emphasized that any child, who suffered a high-energy trauma whether conscious or unconscious, must be treated with full spinal immobilization immediately and this should not be removed despite obtaining normal spinal X-rays or CT scans. Repeated neurological examinations must be performed in the conscious or recovering child in whom spinal injury cannot be ruled out. All these measures are essential in an effort to avoid a preventable life-long disability.

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