Spinal Cord Injury without Radiographic Abnormality (SCIWORA): a case report and literature review

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Abstract

Spinal cord injury without radiographic abnormality (SCIWORA) is a rare post-traumatic myelopathy, more frequently seen in paediatric population and elderly. Clinical manifestation can range from transient isolated paraesthesia to quadriplegia. Due to its rarity in adults and the broad spectrum of neurological manifestations, SCIWORA actually represents a challenge for emergency physicians. Early diagnosis and timely intervention are crucial for the patient's prognosis avoiding permanent neurological deficits. Magnetic resonance imaging (MRI) plays a pivotal role in the diagnosis, management and prognosis of SCIWORA, being the golden standard technique to identify spinal cord injuries. To highlight the importance of in-depth neurological examination and early diagnosis of SCIWORA, we describe the case of a 53-year-old woman who presented to our emergency department following a forward fall down the stairs with a minor head injury.

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Highlights

- *SCIWORA is a post-traumatic myelopathy without radiological abnormalities (X-ray and CT scan) more common in paediatric population and elderly.*
- *"Adult SCIWORA" is a real challenge for emergency physicians for its broad spectrum of neurological deficits ranging from transient isolated paraesthesia to quadriplegia.*
- *The pathophysiology is not completely clear and includes hyperextension, traction or spinal cord damage, edema or vascular lesions.*
- *MRI is the gold standard technique to diagnose SCIWORA.*
- *Standard guidelines for the treatment have not been established yet.*

Case Report

A 53-year-old woman with no relevant clinical history presented to our attention after a forward fall down the stairs with a minor head injury. She reported a facial trauma with a nose wound. On examination her vital signs were stable, and GCS was 15/15. She denied neck and head pain, but she complained of paraesthesia in both her arms, thumb and index fingers occurred immediately after the trauma and still present on arrival. Initial neurological examination revealed bilateral reduction in sensitivity to light touch over the C5-C6 dermatomes. The muscle tone was normal in all four limbs. She was promptly immobilised with a rigid neck collar. The neural injuries were classified according to the American Spinal Injury Association (ASIA) Impairment Scale.[1] The neurological level was at C5-C6 with an ASIA grade C, compatible with an incomplete spinal cord injury.

The patient underwent a brain CT scan, that was normal, and a CT of the cervical spine, thus excluding soft tissue swelling, loss of alignment or fractures (Figure 1A). A cervical spine MRI was performed, showing the presence of a cervical myelopathy with increased signal intensity on T2 weighted and Short-Tau Inversion Recovery (STIR) sequences within the cord at the level of C5-C6, associated to an inversion of the physiological lordosis at the C5-

C6 level (Figure 1 B-D). T1 weighted showed no increase in the signal pattern. These findings were considered consistent with localised contusion and edema of the spinal cord. On the basis of clinical presentation and MRI findings, a diagnosis of SCIWORA was done. After discussion with the neurological consultant, the patient was managed conservatively with rigid neck immobilization for 3 months, and iv methylprednisolone 500 mg for 3 days. She was discharged after 3 days of observation in the Observation Unit with a prescription of dexamethasone 8 mg i.m. daily for 7 days, then 4 mg i.m. for 4 days, and a planned neurological evaluation. After 2 weeks, she was re-evaluated in the neurology outpatient clinic with a complete neurological recovery. A follow-up MRI and an upper extremity electromyography one month after the trauma resulted both normal.

Discussion

SCIWORA is defined as a post-traumatic myelopathy demonstrable through MRI of the spinal cord, in absence of evidence of bone damage on X-Ray and/or CT imaging. It is well recognized in paediatric trauma[2] since the term SCIWORA was firstly coined by Pang and Wilberg in 1982 for children under 8 years of age, who had a traumatic spinal cord injury (SCI) without radiographic evidence of an adjacent spinal column injury.[3] SCIWORA is seen in 30 - 40% of all paediatric spinal injuries[4] and most of the reports on SCIWORA deal with the paediatric age group.[3,5-7]

The incidence of SCIWORAs in the adult population is rather underreported (<10%).[4] For this reason, "adult SCIWORA" is considered as a rare syndrome, more common in males (68.5%) with a peak in their reproductive age,[8-11] even if in the elderly population (> 60 years) the incidence is comparatively higher due to degenerative pathologies, including spondylosis, ossification of the posterior longitudinal ligament and cervical stenosis.[12,13]

The most common location injured is the cervical spine (89%), between C2 and C4 in children, then moving between C5 and C6 in adults.[14] The thoracic or lumbar spines are involved respectively in 9.5% and 1.5% of cases.[9,11,15-17] Rare cases of thoracic-lumbar involvement are also reported in the literature.[18,19]

The pathophysiology

The pathophysiology has not yet been completely clarified. SCIWORA mainly occurs as a result of several different trauma mechanisms, including hyperextension, traction or spinal cord damage following edema or vascular lesions. Falls, road accidents and sport injuries are the most common causes of SCIWORA, and head trauma is the most common associated injury.[10,20] In the majority of paediatric cases, SCIWORA is caused by high-energy injuries with hyperextension or flexion due to characteristic anatomic differences resulting in neural injury without bony or overt ligamentous breaks.[21,22] These differences include greater head-to-body ratio, more horizontal orientation of the facet joints, anterior wedging of the superior aspects of the vertebral bodies, more elastic ligaments and joint capsules, and reduced development of paravertebral muscles.[23,25] Flexion–extension injuries can also cause a temporary occlusion of the vertebral arteries or the anterior spinal artery with the result of a spinal cord infarction.[26] In the meta-analysis by Launary *et al.*,[27] the most common cause of injury was motor vehicle accident, followed by fall from height. In older children, especially adolescents, SCIWORA is most commonly due to sport-related injuries, such as diving, rugby, wrestling, and baseball.[28]

A different mechanism has been postulated in adults because of anatomical and biomechanical differences in the spine at various ages.[29] Gupta *et al*. described 15 adult patients with SCIWORA due to a disc prolapse in six patients, intramedullary haematoma/contusion in four patients, cord edema in four patients, and multiple disc prolapses with associated intramedullary haematoma in one patient.[12] Ligamentous injuries, traumatic disc prolapse, intramedullary lesions and extradural haematomas are the most common findings by MRI in "adult SCIWORA".[30-32] Considering SCIWORA in patients of late middle age and old age, Crooks and Birkett found that many of these patients have pre-existing cervical spondylitic changes resulting in narrowing of the sagittal diameter of the cervical canal. In these cases SCIWORA is most often associated with low-energy falls with hyperextension injuries that cause the cord to be pinched between the osteophytes and the inward bulging of the interlaminar ligaments producing an acute central cord syndrome.[33,34]

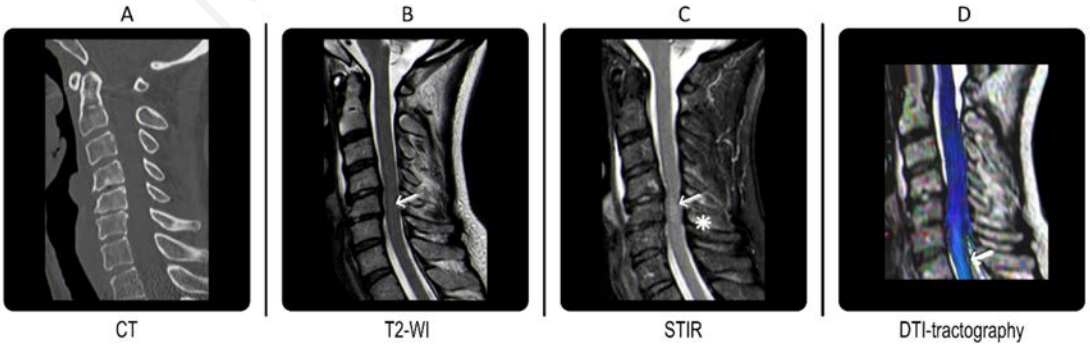

Figure 1. Imaging of spinal cord edema. Schematic diagram demonstrating normal CT findings (A) and the imaging of edema within the spinal cord on MRI (B-D). The sagittal T2-weighted image (B) and STIR (C) sequence show high intensity signal in the cord signal at level C5-C6 (arrow). Interspinous ligament edema was also observed (asterisk). The Diffusion Tensor Imaging (DTI) with tractography (D) highlights the involvement of the spinal cord fibers, which appears less represented in the lower portion of the cervical cord, under the medullary contusion (arrow). The lesser representation of the spinal cord fibers is attributable to edema. The fibers direction is color-coded to indicate tract orientation: red for left-right orientation, green for anterior-posterior orientation, and blue for superior-inferior orientation.

Neurological manifestations

Neurological deficit, that is usually more severe in the upper extremities than in the lower extremities, is the most typical clinical presentation in patients with SCIWORA.[13] The patient can complain of mild, transient spinal cord concussive deficits, such as paraesthesia in fingers, up to permanent, complete injuries of the spinal cord with quadriparesis.[11,35] When performing physical examination, emergency physicians should bear in mind that some patients experience symptoms only at the moment of injury, while in others neurological deficits can be acute or delayed ranging from a few minutes to 24h and even up to 4 days. This latency is associated with repeated microinsults to the spinal cord from striking against the unstable vertebrae. According to the Advanced Trauma Life Support (ATLS) guidelines, it is advisable to use the American Spinal Cord Injury Association (ASIA scale) to establish the level of neurological injury.[1] A retrospective study of 48 adult patients with SCIWORA showed ASIA grade D as the most frequent neurological deficit on admission, followed by grades C and grade B.[17,36]

Diagnosis

The early recognition and management of SCIWORA is crucial to optimize the patient's outcome avoiding catastrophic consequences. Emergency physicians must keep in mind that normal plain radiographs and/or CT scan of the adult cervical spine do not exclude neurological damage in the presence of an abnormal neurological examination.[37] Conventional X-rays are usually performed as the first-line imaging test. A lateral spine X-ray can identify 75% of fractures with sensitivity of 85%. The sensitivity increases to over 90% when antero-posterior (AP), lateral, oblique and open mouth or odontoid radiographs are taken.[38] In patients with post-traumatic cervical dystonia X-rays can be inconclusive and should be postponed until complete resolution of muscle spasm.[39] CT is most accurate in detecting bony pathology and can be used to visualize subtle injuries to the posterior arch or lateral mass of the vertebra, and injuries to the atlas and odontoid process that are poorly visible on standard X-rays. When a diagnosis of spinal fracture can be excluded by X-rays and CT scan, SCIWORA should be suspected in presence of neurological abnormalities. In these cases, MRI must be performed. MRI is the gold standard technique for the diagnosis of SCIWORA since it is the best modality for direct evaluation of the spinal cord and capable to identify the underlying causes by distinguishing extramedullary (disc hernia, canal stenosis, lesion of the anterior common vertebral ligament or posterior ligament complex and intracanal haematoma) and intramedullary (edema, contusion, and haemorrhage) lesions.[39,40] Spin-echo T1 (T1 SE), gradient-echo T2* (T2-weighted GRE*) and STIR-weighted MRI pulse sequences are preferred in patients with spinal injuries.[34] T1-weighted spin-echo MR images provide information about morphology and anatomy of the spinal cord and should be performed as first diagnostic step. The bleeding can be best identified on T2-weigted GRE sequences. An increase in the concentration of deoxyhaemoglobin in fresh haematoma causes a decrease in signal intensity on T2-weigted images and, in particular, on T2-weigted GRE images. A spinal cord edema is seen as hyperintense signal on T2-weighted images against a background of normal nervous tissue and is best visible on STIR images. Radiological features of post-traumatic disc herniation are similar to those of non-traumatic. MRI also allows identification of bone marrow edema in injured vertebrae that cannot be seen on CT scans.[41] Differential diagnosis should include embolism from vertebral artery occlusion associated with cardiovascular diseases such as endocarditis, cardiac arrhythmia, persistent foramen ovale, arteritis or bleeding disorder. Acute or chronic myelitis should also be excluded.

MRI can be also useful to determine the severity of cord lesion acting as a prognostic tool.[12,17,42] Small haematomas (measuring up to 1/3 of the spinal cord diameter) or edema have favourable prognosis and resolve over time in most cases. Anatomic transection of the spinal cord or large haematomas (greater than 1/2 of the spinal cord diameter) have poor prognosis and are manifested clinically as paresis or paralysis.[43]

Some patients diagnosed as SCIWORA have normal MRI findings. These cases should be better classified as SCIWNA (spinal cord injury without neuroimaging abnormality).[44] These patients have hypertense lesions on diffusion weighted imaging (DWI).[45]

Myelography or angiography have no role as diagnostic tools for SCIWORA.

Management

Treatment guidelines do not still exist. Conservative management includes neck immobilization and high dose steroids.[46] External immobilization of the spine for up to 12 weeks is the main therapeutic option for patients with spinal injury.[34] Increased-risk activities should be avoided for 6 months after diagnosis to prevent acute exacerbations of symptoms and reduce the risk of another injury. Administration of a bolus dose of methylprednisolone (30 mg/kg) within three hours of injury followed by a methylprednisolone infusion of 5.4 mg/kg per hour for 24 hours or administration of a bolus dose of methylprednisolone within eight hours of injury followed by an infusion for 48 hours results in a significantly better neurological outcome.[47] Methylprednisolone can be administered in the suspicion of SCIWORA based on the clinical findings and results of CT scan, before definitive radiological imaging by MRI.[48] Surgical treatment can be considered in selected cases with clear indication of extraneural abnormalities, such as ligamentous injury and/or cord compression which requires operative intervention.[24,35,46]

Prognosis

The prognosis for recovery is related to the patient's age, the mechanism of injury and its severity, and MRI findings.[42] Patients with SCIWORA with less severe injuries are likely to achieve complete recovery, while patients with incomplete neurological deficit (ASIA A–C)[1] on initial evaluation had residual weakness that needs longer follow-up to expect a complete recovery.[46] A delayed diagnosis may lead to poor neurological outcomes.[37]

Conclusions

Due to its rarity, SCIWORA represents a real challenge for emergency physicians. A proper neurological evaluation with the ASIA scale at admission, discharge and follow-up is mandatory in all patients after any trauma, immediately and even after 4 days after the accident, as early recognition and timely management of SCIWORA are crucial to prevent secondary spinal cord injuries and serious permanent complications.

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