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Adult Spinal Cord Injury Without Radiographic Abnormality (SCIWORA). Two case reports and a narrative review

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Summary. The term SCIWORA (Spinal Cord Injury Without Radiographic Abnormality) indicates a clinically appreciable post-traumatic myelopathy in the absence of spinal column findings on radiographs and/or computed tomography (CT), but with pathologic findings at magnetic resonance imaging (MRI) in approximately two-thirds of cases. Affecting mainly children younger than 8 years, SCIWORA has been, however, also described in adult patients, but, due to the uncertainty of classification and frequent co-morbidity, the term "adult SCIWORA" has generated controversy, and some debate is still active. In this article, we report two different cases of adult SCIWORA involving cervical spinal cord, characterized by distinct and peculiar clinical features. A literature review and some clinical suggestions are also reported, mainly focused on the importance of a high level of suspicion in order to achieve a timely diagnosis and optimize the management and, consequently, the outcome of these trauma patients. (www.actabiomedica.it)

Key words: spinal cord injury, SCIWORA, SCIWORET, SCIWOCTET, cervical spine, whiplash injury

Introduction

Firstly described in 1974 (1), and then defined in 1982 in a pediatric population (2), Spinal Cord Injury Without Radiologic Abnormalities (SCIWORA) still represents a diagnostic challenge for emergency physicians (EPs).

The term SCIWORA indicates a clinically appreciable post-traumatic myelopathy in the absence of spinal column findings on radiographs and/or computed tomography (CT). In approximately two-thirds of cases, however, pathologic findings at magnetic resonance imaging (MRI) of the spinal cord are reported (3, 4). Affecting mainly children younger than 8 years (3), SCIWORA has been, however, subsequently also described in adult patients (5). Due to the uncertainty of classification and frequent co-morbidity, the term "adult SCIWORA" has generated considerable controversy (6), and some debate is still active.

Here we report two different cases of adult SCIWORA involving cervical spinal cord, characterized by distinct and peculiar clinical features.

Case report 1

A 58 year-old male with no relevant clinical history presented to the Emergency Department (ED) following an accidental fall from a truck trailer while working. He had fallen on his back, reporting a minor head trauma, and a whiplash injury.

On physical examination he presented a GCS of 15/15 and normal vital parameters; the patient was not complaining of pain anywhere. The thoracic and abdominal physical findings were normal, while the

neurological examination showed frank paraplegia associated with sensory loss in the upper limbs.

He felt hypoesthesia below the C4 dermatome level with sacral sparing, and preserved voluntary flexion of the first toe of left foot. The neural injuries were classified according to the American Spinal Injury Association (ASIA) Impairment Scale (7). The neurological level was at C4 with an ASIA grade C, compatible with an incomplete spinal cord injury.

The patient underwent a CT scan of spinal cord showing intervertebral osteochondrosis with C3-C4 spondylosis and degenerative C2-C3 fusion causing chronic spinal canal stenosis. No fractures, dislocations or loss of alignment were detected (Fig. 1, upper images). Based on these findings, initially a central cord syndrome was suspected, mainly due to misinterpretation of clinical manifestations.

A cervical spine MRI was then performed, showing C2-C3 disc protrusion causing significant compression and swelling of the spinal cord (Fig. 1, lower images). Based on clinical presentation and MRI findings, the diagnosis of SCIWORA was finally confirmed.

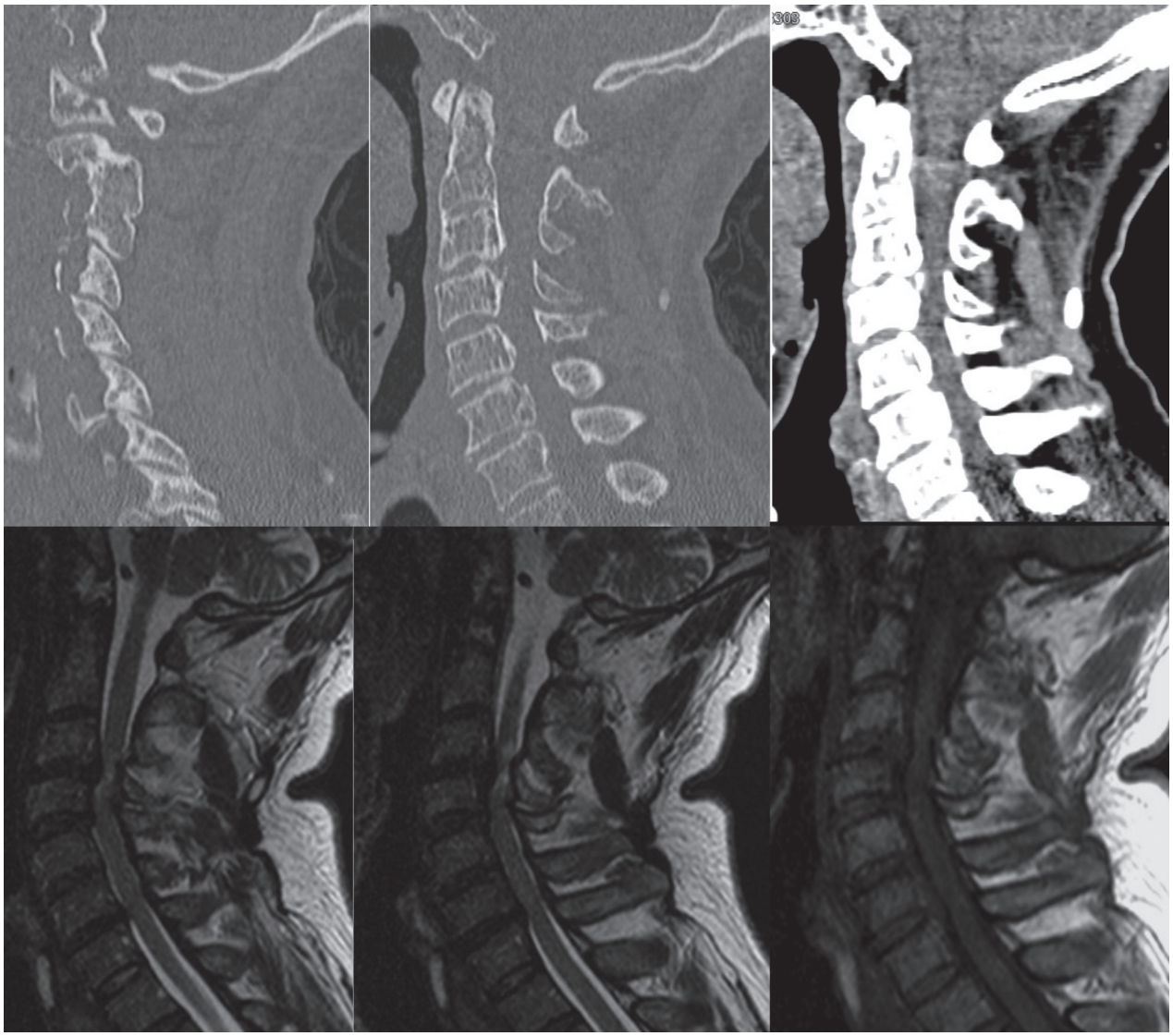

Figure 1. Upper images: sagittal CT scan of cervical spinal cord. Images show chronic spinal canal stenosis by C3-C4 spondylosis/degenerative and C2-C3 fusion. Lower images: T1 and T2 weighted magnetic resonance imaging. Images show C2-C3 disc protrusion and spinal cord swelling

The patient subsequently underwent an emergent C2-C3 discectomy using an anterior approach. After that, he was transferred to a Rehabilitation Medicine Department for rehabilitative treatment. After six months of hospital staying, partially in acute-care setting and partially in rehabilitation center, he did not show any neurological improvement.

Case report 2

A 46 year-old male with no relevant clinical history presented to the ED following a syncope. He reported a facial trauma with a zygomatic wound and a whiplash injury.

On physical examination he presented a GCS of 15/15, normal vital signs, and paresthesia and mild pain in both upper limbs.

Cervical spine x-ray showed no fractures or loss of alignment. CT scan of head and facial skeleton showed a complex fracture of the nasal bones thus needing surgical treatment.

The patient was then admitted to the Maxillofacial Unit. During the hospital stay, due to the persistence of paresthesia in the upper limbs, he underwent a neurological visit. The neurologist consultant suggested the execution of an MRI of the cervical spine.

This examination revealed chronic cervical spinal cord stenosis as a result of degenerative modifications due to intervertebral osteochondrosis/spondylosis, and presence of C2-C3, C4-C5 and C6-C7 cord contusions (Fig. 2).

Based on clinical presentation and MRI findings, the diagnosis of SCIWORA was thus confirmed.

The treatment was conservative with rigid neck immobilization for three months.

A follow up MRI after six months showed cervical spine C6-C7 cord contusion healing, and C2-C3, C4-C5 cord contusions improvement. These radiological findings were consistent with a complete neurological recovery.

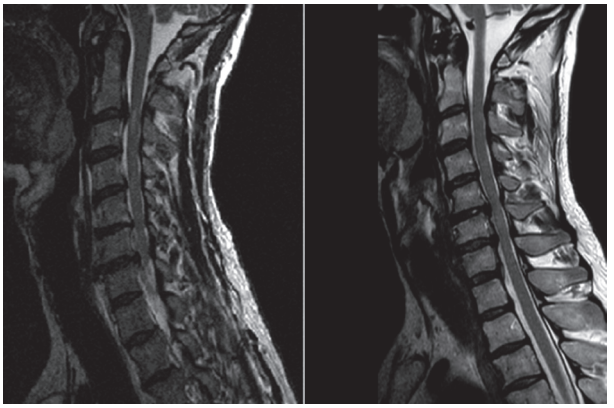

Figure 2. Left: sagittal T1 weighted magnetic resonance. Image shows chronic cervical spinal cord stenosis by intervertebral osteochondrosis/spondylosis and presence of C2-C3, C4-C5 and C6-C7 cord contusion. Right: follow up sagittal T weighted magnetic resonance. Image shows cervical spine C6-C7 cord contusion healing and C2-C3, C4-C5 cord contusion imp

Discussion

Traumatic spinal cord injuries have a significant impact on individuals, families, and society, with heavy repercussions in economic and social terms. The management and treatment of these patients displays a wide heterogeneity, largely dependent on the variety of presentation of these syndromes, the underlying pathophysiology, and the clinical characteristics of the patients themselves (8).

Spinal cord injuries (SCI) typically occur in males, with a peak in their productive ages (9).

The prevalence is increasing steadily due to improved survival in both the acute and chronic stages of the disease. Depending on the age of patients at the time of injury, on level, and on severity of injury, life expectancy ranges from 4 to more than 50 years after injury (9, 10).

SCI must always be suspected in patients with multiple injuries. Approximately 5% of patients with brain injury have an associated SCI, whereas 25% of patients with SCI have at least a mild brain injury.

Approximately 55% of SCIs occur in the cervical region, 15% in the thoracic region, 15% at the thoracolumbar junction, and 15% in the lumbosacral area. The cervical spine is the most vulnerable to injury, because of its mobility and increased vulnerability to trauma (9-11).

At variance, the thoracic and lumbar spines have greater stability due to the rib cage, and a greater articulation bone surface (1, 12-14). Road accidents and falls are among the most common causes of SCIs. The cervi-

cal canal is particularly vulnerable to trauma. From an anatomical point of view the spinal canal is wide in the most cranial part of the cervical tract, i.e., from the foramen magnum to the lower portion of C2. While spinal lesions at C1 level often cause the death of patient on scene, since they can lead to immediate apnea, the survivors, due to the aforementioned anatomical characteristics, are often neurologically unharmed. At variance, below the level of C3 the diameter of the spinal canal significantly decreases, thus resulting small when compared with the size of the spinal cord. As such, it is easier that a lesion of the bony part of the column below the C3 level could be associated with a SCI (11).

Spinal lesions, from a morphological point of view, can be distinguished in fractures, dislocations, spinal cord lesions without radiographic anomalies (SCIWORA) and penetrating lesions, and can be further divided into stable and unstable (9-11).

Originally, SCIWORA, was described in pediatric population, but it may also occur in adults, although the term "adult SCIWORA" has generated considerable controversy (15), since these patients usually have degenerative changes in their cervical spine.

The alternative terms "SCIWORET" (SCI without radiographic evidence of trauma) (16) and "SCIWOCTET" (SCI without CT evidence of trauma) (17) have subsequently come into use for describing SCIs with clinic-radiologic mismatch in adults (18). We could say that our understanding of SCIWORA, SCIWORET, and SCIWOCTET has evolved in parallel with MRI improving, and their definitions have changed accordingly.

Recent data show an incidence of SCI comprised between 13 and 19% in children (19, 20) and between 10 and 12% in adults (21, 22). SCIWORA is also more common in males (68.5%) than in females (31.5%) (20).

The highest prevalence in pediatric patients has been tributed to the different anatomical and biomechanical characteristics of the cervical spine that distinguish children from adults.

Among these, greater mobility of the cervical spine, greater head-to-body ratio, greater ligament laxity, incomplete ossification of the vertebrae, and reduced development of paravertebral muscles have been considered (12, 13).

As such, the involvement of the cervical spine is undoubtedly the most common (89% of patients having cervical or cervicothoracic involvement), particularly in the lower part. The thoracic or lumbar spines are involved respectively in 9.5% and 1.5% of cases (3, 20-23), and rare cases of thoracic-lumbar involvement are also reported in the literature (14, 24). The fulcrum of cervical movements is usually placed between C2 and C4 in children, then moving between C5 and C6 in adults (25, 26).

The pathophysiology of SCIWORA has not yet been clarified. It mainly occurs as a result of several different trauma mechanisms, including hyper-extension, traction or spinal cord damage following edema or vascular lesions.

Falls, car or motorbike accidents and sports accidents are the most common causes of SCIWORA. In particular, falls and traffic accidents are the leading causes in adulthood (23).

The rarity of SCIWORA in adult population poses a diagnostic challenge for clinicians who must rely on the clinic to identify spinal cord injuries in patients with neurological deficits but normal X-ray films and CT scans. The management of a patient with spinal cord injury begins with early recognition, an appropriate extra-hospital triage, the stabilization of vital functions, a proper immobilization and then the referral to a center specialized in the treatment of spinal lesions. (11-13).

SCIWORA patients can display a wide spectrum of neurological dysfunctions, going from transient spinal cord concussive deficits to permanent complete injury of spinal cord (13). According 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.

Conventional radiographs are usually performed as first-line imaging tests, since they can identify fractures with a sensitivity of 85% with lateral projection only, but up to over 90% when the three classical projection (i.e., anteroposterior, lateral, and open-mouth) are performed. Guided by clinical suspicion, since radiographs can miss up to 6% of fractures, CT scan is recommended, since it is more accurate in detecting minor lesions at the posterior arch or lateral mass of

the vertebrae and lesions of the atlas and/or the odontoid process, usually poorly visible on standard radiographs. When cervical lesions are found, it is recommended to extend the examination to the thoracic-lumbar tract to exclude concomitant occult lesions of the lower spine. When X-rays and CT scan exclude the presence of a vertebral fracture or dislocation, in a patient suffering from trauma with clinical signs of myelopathy, SCIWORA should be suspected and MRI should be performed. MRI is recommended to identify the underlying causes of the syndrome, since it is capable to distinguish extramedullary (disc hernia, canal stenosis, lesion of the anterior common vertebral ligament or posterior ligament complex and intra-canal hematoma) and intramedullary (edema, contusion and hemorrhage) lesions (8).

No definitive treatment protocol has been established yet, and a kaleidoscope of opinions regarding surgical and conservative management still exists among clinicians.

Nonsurgical treatment consists in neck immobilization and high dose steroids. The neck immobilization by cervical brace or collar for a minimum of three months and restriction of high risk activities for six months represents the mainstay of treatment (13-18).

Although there is no evidence enough to support the routine administration of high-dose intravenous methylprednisolone in SCIWORA patients, some studies suggest 24 hours of methylprednisolone as a treatment option, starting within 8 hours from injury for patients with acute, non-penetrating SCI (8-13).

The neurological recovery in SCIWORA patients is primarily related to several issues, such as the mechanism of initial injury, the diameter of the canal, the patient's age, the extent of the injury, the presence of disc-ligament lesions and the severity of the neurological syndrome. For all these reasons, surgery is not the gold standard of the treatment (18). Surgical treatment should be reserved for patients with clear MRI evidence of extra neural findings, including spinal cord compression, ligamentous injury and instability, along with worsening, or not improving, of neurological findings (13).

To date, the possible prognostic correlations between the findings obtained by MRI and neurological recovery are still controversial. Some data seem to suggest that severity of both initial clinical manifestations and MRI findings do not correlate with subsequent clinical course (16-27, 28).

Conclusions

Patients diagnosed with SCIWORA have a broad spectrum of neurological deficits, from mild and transient symptoms, such as isolated paresthesia in the fingers, to quadriplegia. Some patients show symptoms only at the time of injury.

Spinal cord injury often poses a challenge for EPs. As such, a high level of suspicion is compelling for EPs in order to achieve a timely diagnosis and optimize the management and, consequently, the outcome of trauma patients (29).

After the initial management on scene and the acquisition of a detailed history, when available from eyewitnesses, in order to determine the mechanism of the lesion, a clinical examination focused on the neurological findings is of pivotal relevance (30).

The clinical suspicion of SCIWORA must be high, particularly in patients with normal X-ray and CT scan findings, but with signs and symptoms suggestive of SCI. Before confirming a diagnosis of SCIWORA, however, EPs must exclude other neurological lesions with a similar clinical and anamnestic history, such as cervical burners, also known as stinger syndrome, and spinal cord syndrome.

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