Spinal Cord (2009) 47, 822–825

www.nature.com/sc

CASE REPORT

Progression of spinal cord atrophy by traumatic or inflammatory myelopathy in the pediatric patients: case series

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Study design: Case series.
Objective: To present spinal cord atrophy in pediatric patients who had spinal cord injury developed after trauma or acute transverse myelitis, and had no motor recovery later.
Setting: Department of Rehabilitation Medicine, Tertiary National University Children's Hospital, Seoul, Korea.
Methods: Case series.
Results: Two pediatric patients with paraplegia due to acute transverse myelitis and one pediatric patient with paraplegia due to traumatic myelopathy were included in this case series. Their initial MRI (magnetic resonance imaging) findings were spinal cord swelling and high signal intensity in T2-weighted image. After several months, they had no motor recovery and showed no change of neurological level, though they underwent steroid and physical therapy. A follow-up MRI revealed spinal cord atrophy.
Conclusion: If a pediatric patient with traumatic or inflammatory spinal cord injury does not show motor recovery after several months, spinal cord atrophy must be considered.
Spinal Cord (2009) **47,** 822–825; doi:10.1038/sc.2008.175; published online 27 January 2009

Keywords: spinal cord injury; pediatrics; myelitis; cord atrophy

Introduction

Pediatric spinal cord injuries, unlike injuries in adults, show significant gains in functional status irrespective of the completeness of injury.[1] However, some pediatric patients show no motor recovery or even the progression of neurological injury. In such cases, chronic spinal cord injury sequelae must be considered. If children with spinal cord injury show poor motor recovery later, it is important to consider the possibility of spinal cord injury sequelae, such as, spinal cord atrophy, with follow-up spinal cord MRI (magnetic resonance imaging).

We report three pediatric cases, in which spinal cord atrophy developed after traumatic spinal cord injury or acute transverse myelitis and no motor recovery occurred.

Case 1

A 2-year-old girl with paraplegia caused by a traffic accident was referred to our department of rehabilitation medicine from pediatric neurosurgery for rehabilitation therapy. Initial X-ray findings showed no abnormality, and physical examination revealed a complete loss of muscle power below the L2 level with increased deep tendon reflex, also called as spinal cord injury without radiologic abnormality. Sensory impairment level was below T4 bilaterally. She had no voiding or defecation sense, and impaired perianal sensation and anal contraction. Laboratory data showed WBC (white blood cells) 6240 mm^{-3}, RBC (red blood cells) 4.48×10^6 mm^{-3} and hemoglobin 12.0 g/dl. T2-weighted magnetic resonance images showed spinal cord swelling and high signal intensity from the T2 to T8 levels (Figure 1). Physical and neuromuscular electrical stimulation therapies were started, and maintained at twice per week on an outpatient basis.

At 5 months after the accident, her neurological status was unchanged, and no motor recovery was evident. Nerve conduction and electromyographic studies disclosed no evidence of nerve root injury. A follow-up MRI revealed severe diffuse spinal cord atrophy from T3 to T7 (Figure 1). She has been trained for ambulation with walker and hip-knee-ankle-foot orthosis.

Case 2

A 5-year-old girl experienced sudden onset of bilateral lower extremities weakness after developing symptoms of an upper respiratory tract infection. She was diagnosed as having acute transverse myelitis based on brain and spinal cord imaging findings. Cerebrospinal fluid tapping revealed an IgG index of 0.19, and an elevated WBC level 16 480 mm^{-3},

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Received 25 August 2008; accepted 22 November 2008; published online 27 January 2009

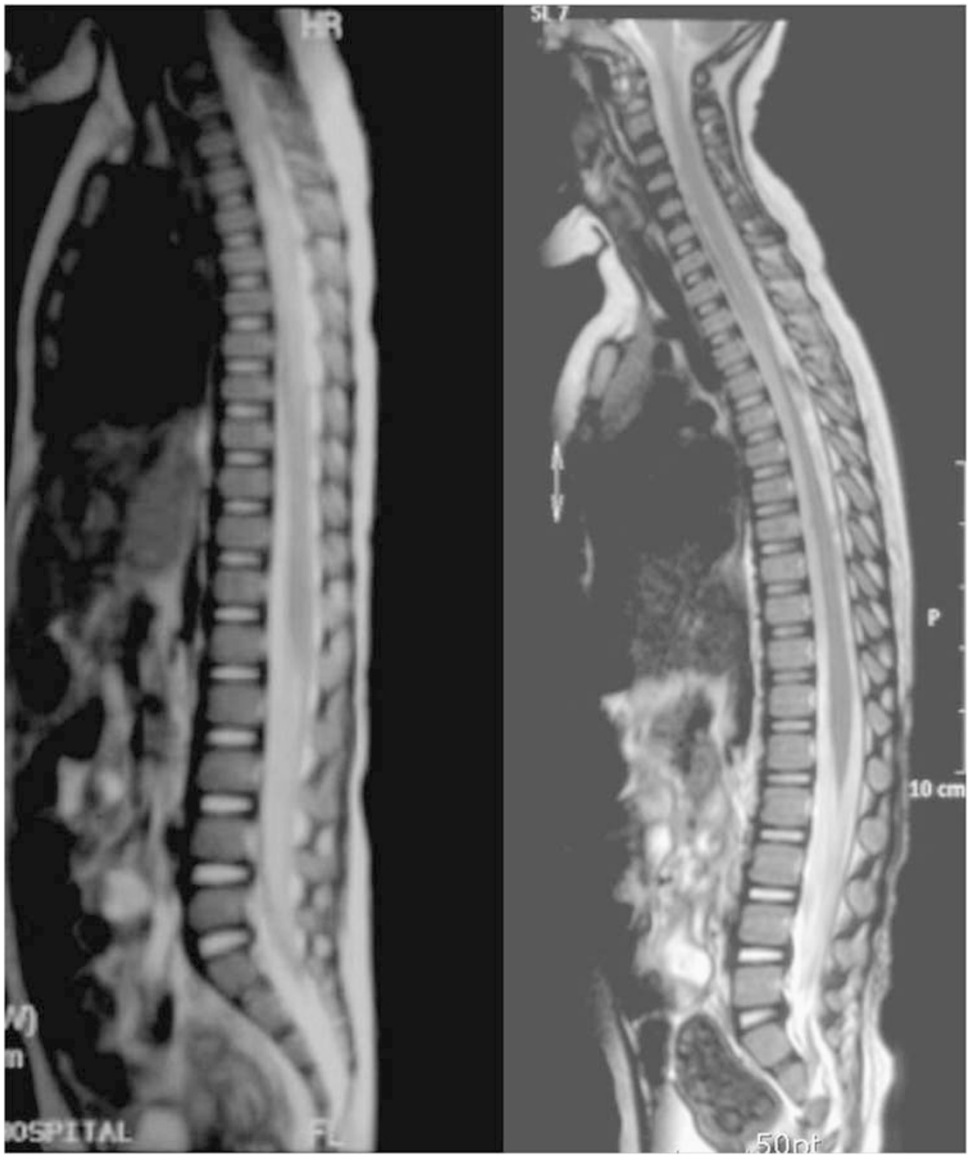

Figure 1 Case 1. Sequential magnetic resonance images in a pediatric patient with traumatic spinal cord injury. The spinal cord swelling and high signal intensity in T2-weighted image were found at the initial image (left), and spinal cord atrophy at the follow-up image at 5 months post-onset (right).

though this normalized to 7550 mm^{-3} after several days. Blood and urine laboratory data were within normal range. Motor power in both lower extremities was nil and sensory impairment was present below T11. Deep tendon reflex was reduced, and perianal sensation and anal contraction were absent. Her ASIA (American spinal injury association) impairment scale was T10 ASIA (A). Initial T2-weighted magnetic resonance images revealed spinal cord swelling, high signal intensities at T4-L1, and syringomyelia at T2 and T3 (Figure 2). Intravenous immunoglobulin therapy was administered, and when the disease stabilized, she was transferred to the rehabilitation department for daily physical therapy.

At 2 months post-onset, she was discharged without any motor or sensory recovery. Spinal cord imaging immediately before discharge showed moderate spinal cord atrophy (Figure 2).

At 3 months post-onset, spinal cord atrophy progressed to a severe stage and the spinal cord appeared like a thread (Figure 2), and no motor recovery was evident. Nerve conduction and needle electromyographic studies at 4 months post-onset revealed no evoked complex motor action potential at bilateral peroneal or tibial nerves and profuse abnormal spontaneous activities in lower extremity muscles with intact sensory nerve action potentials. She underwent gait training with walker and bilateral hip-knee-ankle-foot orthosis.

Case 3

This case involved a 7-year-old girl who manifested sudden onset paraplegia. She was diagnosed as having acute transverse myelitis based on MRI and cerebro spinal fluid studies. She also received steroid and physical therapy during

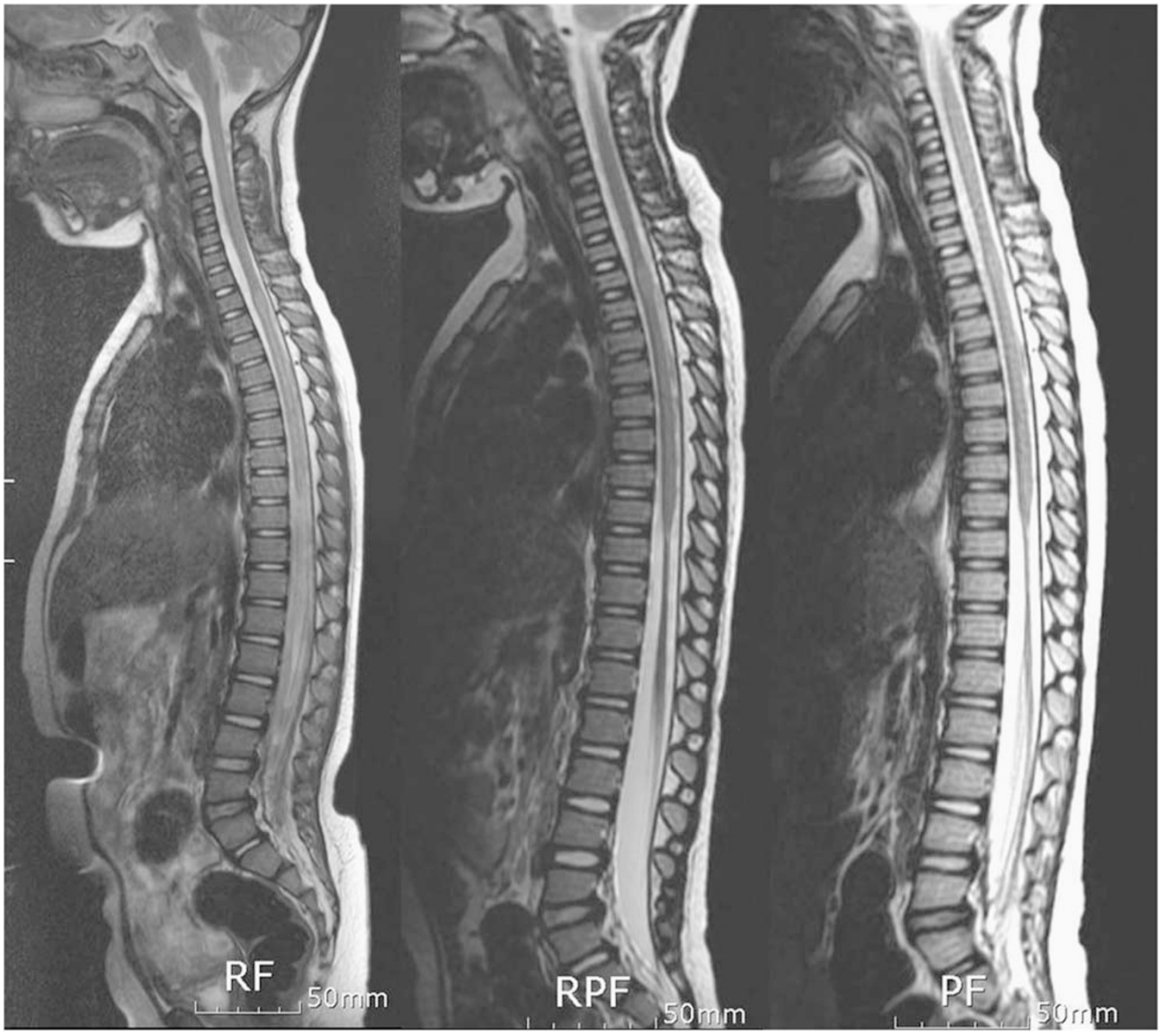

Figure 2 Case 2. Sequential magnetic resonance images in a pediatric patient with acute transverse myelitis. The spinal cord swelling and high signal intensity in T2-weighted image were found at the initial image (left), and moderate at 2 months post-onset (middle) and severe (right) spinal cord atrophy at 3 months post-onset.

admission. No treatment effect was found and both lower extremities showed no voluntary movement or sensation below T8 (right)/T9 (left), which included a lack of perianal sensation. Initial T2-weighted magnetic resonance image findings at day 1 post-onset were high signal intensity and mild cord swelling below T2 (Figure 3). Deep tendon reflex was absent and no spasticity was found. An electrodiagnostic examination performed at 1 month post-onset revealed abnormal spontaneous activities in muscles innervated by lumbosacral roots.

Sequential MRI revealed cord segmental atrophy progression from mild at 1 month post-onset to moderate at 2 months and severe at 19 months (Figure 3). Lower extremity motor power had not recovered, and sensory levels were unchanged even after 1 year post-onset. She underwent gait training with bilateral forearm crutch and knee-ankle-foot orthosis. However, she showed exaggerated lumbar lordosis during standing and ambulation because of the absence of hip extensor power.

Discussion

Spinal cord atrophy is defined as an abnormal narrowing of the spinal cord in the sagittal plane in two segments or more beyond the limits of vertebral injury,[2] and represents an epiphenomenon related to the effects of dynamic inflammation within the central nervous system, which leads to demyelination, axonal injury, neuronal loss, Wallerian degeneration and possibly iron deposition.[3] Spinal cord atrophy represents axonal loss,[4] and invariably reflects a poor prognosis.[5]

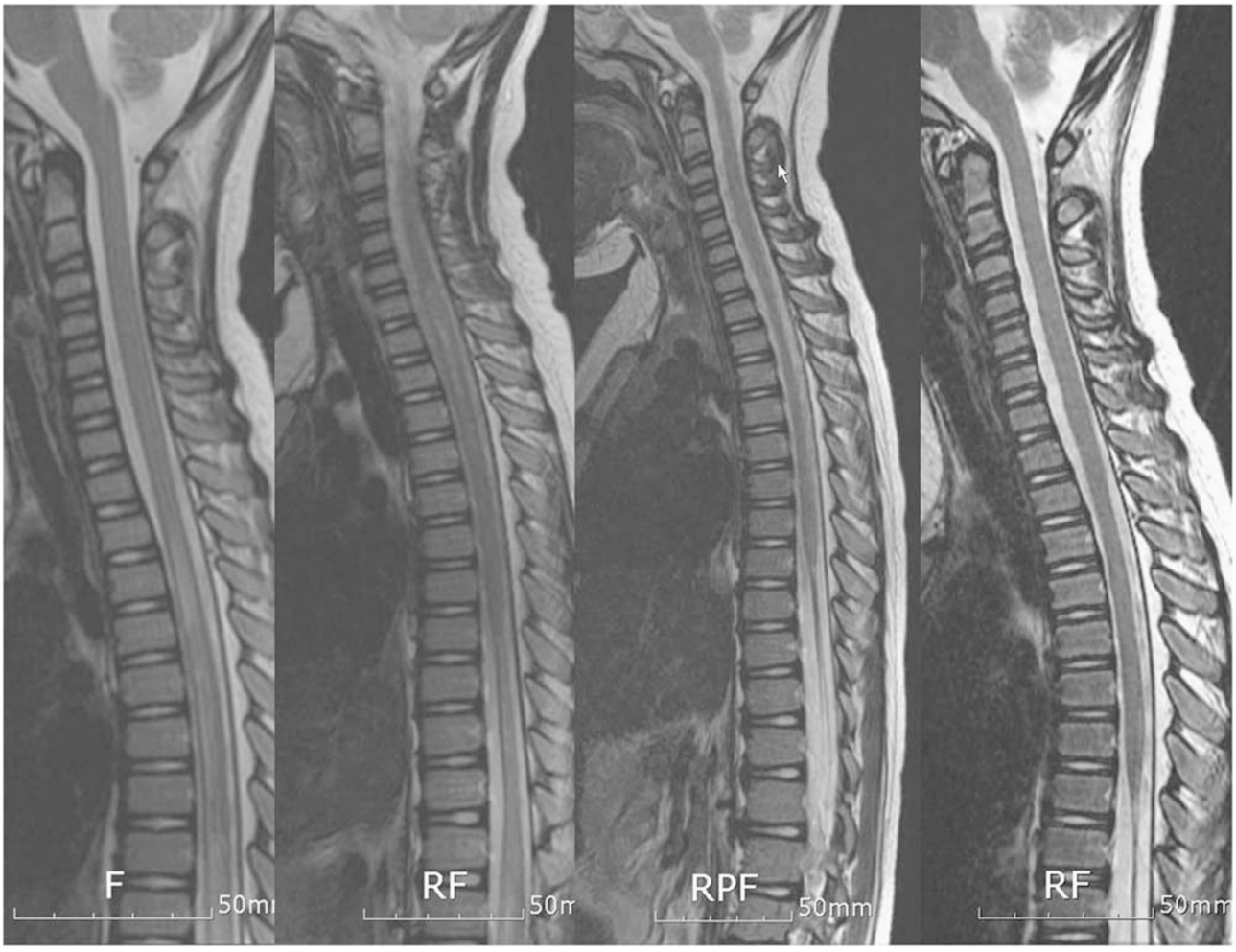

Figure 3 Case 3. Sequential magnetic resonance images in a pediatric patient with acute transverse myelitis. The spinal cord swelling and high signal intensity in T2-weighted image were found at the initial image (left), and mild (middle left) at 1 month post-onset, moderate (middle right) at 2 months post-onset and severe (right) spinal cord atrophy over 1 year post-onset.

Spinal cord atrophy is rarely reported in the pediatric patients. Shen *et al*.[6] reported a case of acute transverse myelitis with cord swelling in the acute stage and diffuse cord atrophy during follow-up MRI at 2 months post-onset. In this study, cases 2 and 3 showed cord atrophy at 2 months post-onset similar to Shen's case.[6]

If spinal cord atrophy appears during the disease course, prognosis will be poor, although one reported case showed no lower-limb deficits despite severe spinal cord atrophy after a cervical operation and postoperative radiotherapy.[7] Nevertheless, spinal cord atrophy must be considered if a pediatric patient with a traumatic or inflammatory spinal cord lesion does not show motor recovery after several months.

Acknowledgements

We specially thank Dr John Roberts for helping us to edit English writing.

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