Holocord syringomyelia presenting as rapidly progressive foot drop

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A 14-year-old girl presented with rapidly progressive bilateral foot drop of 1-week duration with no sensory, bowel or bladder symptoms. Examination showed weak dorsiflexion of the feet, mild wasting of the left hand, dissociated sensory loss over left C4 to T8 dermatomes, diminished stretch reflexes and extensor plantar response. Nerve conduction studies showed low-amplitude compound motor action potential and normal latency and velocity for both peroneal and left ulnar nerves. The F wave was normal. Sensory nerve action potentials for median, ulnar and sural nerves were normal. Electromyographic examinations showed chronic neurogenic changes in first dorsal interossei, tibialis anterior and medial gastrocnemius muscles on both sides. MRI revealed a Chiari I malformation with syrinx occupying almost the entire spinal cord from C2 to the conus medullaris (holocord syrinx) [Figures 1 and 2]. The patient declined any surgical intervention.

Foot drop is a common problem that a patient can present in outpatient department. Although the most frequent cause is a common peroneal neuropathy at the neck of the fibula, other causes include anterior horn cell disease, lumbar plexopathies, L5 radiculopathy, partial sciatic neuropathy and rarely parasagittal lesion. In our case, isolated foot dorsiflexion weakness without any sensory finding localize to anterior horn cell, ventral root or motor nerve involvement. In our case, it is reasonable to assume that foot drop is due to the anterior horn cell dysfunction, as shown by the imaging and electrophysiology.

Syringomyelia has variable clinical presentations which include asymmetric weakness and atrophy of hands, upper and lower limb spasticity, areflexia of upper extremity, dissociated sensory loss in the neck and arms (classic cape like distribution) and increased tone and hyperreflexia of lower limb.[1] This is the first reported case of rapidly progressive bilateral foot drop.

Figure 1: Sagittal T2W MRI of the cervical spine shows a Chiari I malformation (thick white arrow) with syrinx extending along the entire spinal cord from C2 to the conus medullaris (white arrow)

Figure 2: Axial T1W MRI of cervical spine shows syrinx (white arrow)
as the presenting symptom of syringomyelia and Chiari I malformation without cord tethering.

Reference

1. Muhn N, Baker SK, Hollenberg RD, Meaney BF, Tarnopolsky MA.

Syringomyelia refers to the presence of cavities within the spinal cord or dilatation of the central spinal cord canal. Although most cases are associated with a concomitant Chiari I malformation it can also be associated with cord tethering, intramedullary spinal lesions or traumatic injury. Syringomyelia is occasionally an isolated or idiopathic finding. Many patients present with slowly progressive sensory symptoms (hypesthesia or dysthesia) primarily affecting the upper extremities. The classic presentation is numbness in a ‘cape-like distribution’ due to the disruption of decussating sensory fibres lying just anterior to the central canal.

Many patients, particularly children may present with non-sensory symptoms that can include muscle weakness and atrophy, scoliosis or brainstem dysfunction. The case reported in this issue provides an excellent example of how clinicians must remain alert to syringomyelia as a diagnostic consideration. Distal weakness and atrophy has been documented in about one-third of children with syringomyelia. Symptoms of lower extremity weakness may present abruptly and can mimic a compressive neuropathy. Progressive hand weakness and atrophy due to syringomyelia has also been reported to clinically resemble an ulnar neuropathy. Muscle weakness in such cases is likely the result of corticospinal tract disruption and/or anterior horn cells dysfunction. Chiari-associated syringomyelia has been linked with rapidly progressive flaccid paralysis and brainstem dysfunction including apnea, dysphagia and vocal cord paralysis. Scoliosis and back pain may be the presenting complaint in 20–40% of pediatric patients; although, careful examination can identify motor or sensory deficits in these patients. Neurosurgeons typically recommend patients with Chiari I associated syringomyelia to undergo suboccipital decompression with or without duroplasty as the initial treatment, reserving syrinx shunting for those cases in which the former modality of treatment fails. Spontaneous syrinx resolution is thought to be rare. The treatment of idiopathic syringomyelia represents a dilemma since syrinx shunting can carry the inherent risk of increasing neurological dysfunction. The majority of children with neurological symptoms attributable to syringomyelia will demonstrate complete symptom resolution within several months after successful posterior fossa decompression surgery. Dysesthesia and motor symptoms (weakness) are more likely to show clinical improvement compared to scoliosis or hypesthesia. Radiographic improvement is also commonly seen postoperatively although it tends to lag behind clinical recovery. Even children with holocord syringomyelia on MR imaging of the spine and active denervation on electromyography may nevertheless demonstrate rapid and complete postoperative recovery.

Clinicians must therefore consider syringomyelia on their differential diagnosis of patients presenting not only with sensory loss and dysthesia but also those with motor weakness, progressive scoliosis and bulbar dysfunction.


How to cite this article: Saifudheen K, Jose J, Gafoor VA. Holocord syringomyelia presenting as rapidly progressive foot drop. J Neurosci Rural Pract 2011;2:195-6.

Source of Support: Nil. Conflict of Interest: None declared.