Commentary

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.[1] 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.[2] 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[3] 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.[2] Symptoms of lower extremity weakness may present abruptly and can mimic a compressive neuropathy.[4] Progressive hand weakness and atrophy due to syringomyelia has also been reported to clinically resemble an ulnar neuropathy.[5] 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[6] and brainstem dysfunction including apnea, dysphagia and vocal cord paralysis.[7,8]

Scoliosis and back pain may be the presenting complaint in 20–40% of pediatric patients;[7,8] although, careful examination can identify motor or sensory deficits in these patients.[7] 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.[9] Dysesthesia and motor symptoms (weakness) are more likely to show clinical improvement compared to scoliosis or hypesthesia.[9] Radiographic improvement is also commonly seen postoperatively although it tends to lag behind clinical recovery.[9] Even children with holocord syringomyelia on MR imaging of the spine and active denervation on electromyography may nevertheless demonstrate rapid and complete postoperative recovery.[4]

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.

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