CASE REPORT

Rapidly progressive foot drop: an uncommon and underappreciated cause of Chiari I malformation and holocord syrinx

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SUMMARY

We describe a case of a 16-year-old boy presented with rapidly progressive right foot drop without any predisposing illness or antecedent events. Nerve conduction study was non-contributory and needle electromyography localised proximal lesion which prompted spine neuroimaging. MRI of whole spine and brain revealed Chiari I malformation along with holocord syringomyelia. The patient underwent suboccipital craniectomy and C1 laminectomy with duroplasty decompressing the foramen magnum. He responded to surgical intervention without further complications.

BACKGROUND

Foot drop is common but distressing. It can be due to either peripheral nervous system (PNS) disorder or central nervous lesion (central nervous system, CNS). Clinical interview, comprehensive neurological examinations, neurophysiological study and sometimes neuroimaging, that is, MRI of lumbar spine and nerve roots would disclose the aetiology.

CASE PRESENTATION

A 16-year-old boy developed rapidly progressive right foot drop over a span of 2 months leading to inability to clear ground with compensatory high-steppage gait followed by mild wasting of anterior

compartment of right leg, which was unaccompanied by any sensory complaints. Nervous system examination revealed height and neck ratio of 13.2 (normal >13), mild wasting of anterior compartment of leg with hypotonia around right ankle joint (figure 1). Medical research council (MRC) grading for the power strength of the right lower limb revealed the following: hip flexors, extensors, adductors and abductors were 5/5, tibialis anterior 0/5, tibialis posterior and peroneus longus 1/5, gastrocnemius 3/5, extensor hallucis longus 0/5 and toe flexors 3/5. A deep tendon reflex of right knee was diminished (1+) and ankle jerk was absent. Right plantar response was mute and left was flexor. He had 50% sensory loss of all modalities over dorsum of right foot, medial aspect of leg and thigh (L 3, 4 and 5) distribution. Left leg and upper extremity examination were completely normal except brisk deep tendon reflexes (3+) of left knee and ankle without clonus.

INVESTIGATIONS

Nerve conduction study (NCS) showed normal value, that is, right common fibular nerve motor

Figure 1 Right foot drop while attempting to dorsiflex at ankle.

Figure 2 Sagittal T2-weighted MRI of craniovertebral junction and cervical spine showed tonsillar herniation (white arrow) and central cavitory lesion (yellow arrow) suggestive of syringomyelia.

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Figure 3 Sagittal T1-weighted MRI of craniocervical junction and cervical spine showed tonsillar herniation (white arrow) and central cavitory lesion (yellow arrow) suggestive of syringomyelia.

responses—distal amplitude 3.0 mV, latency 2.7 ms, velocity 43.1 m/s vs left common fibular nerve amplitude 4.6 mV, distal latency 3.4 ms, velocity 50 m/s and absent bilateral F wave responses. Tibial nerve motor responses as well as sural, superficial peroneal and saphenous sensory response were also normal. Needle EMG revealed fibrillation potentials and positive sharp waves in the right tibialis anterior, peroneus longus, medial gastrocnemius, gluteus medius and lumbar paraspinal muscles confirming the lesion to be proximally located. MRI of whole spine including craniocervical junction (C-V junction) was performed. Sagittal T1-weighted and T2-weighted images of the C-V junction, cervical spine and dorsolumbar spine showed holocord syrinx (extending from the cervicomedullary junction to the conus medullaris) along with the Chiari I malformation (CM-I; cerebellar tonsils extend 10 mm below foramen magnum) without any bony deformity (figures 2–5). Other investigations including routine blood parameters, C reactive protein, rheumatoid factor and antinuclear antibody (ANA) were normal.

DIFFERENTIAL DIAGNOSIS

Foot drop can be due to either involvement of PNS or central structures. PNS disorders include common fibular neuropathy (most common), L5 radiculopathy, lumbosacral plexopathy, partial sciatic neuropathy, sarcoidosis, leprosy, hereditary neuropathy, mononeuritis multiplex and anterior horn cell disease. Central causes include either involvement of parasagittal structures like high-grade glioma, low-grade astrocytoma, metastasis, meningioma, abscess, tuberculoma, stroke or spinal cord

Figure 4 Sagittal T2-weighted sequence of MRI of dorsolumbar spine showed central cavitory lesion involving whole cord up to conus medullaris (yellow arrow).

involvement such as syringomyelia.¹ In the present case, the presence of upper motor neuron sign in left lower limb, that is, brisk deep tendon reflexes in left knee and ankle suggested a central cause. Though nerve conduction study was non-contributory, needle EMG confirmed the lesion to be proximally located. MRI of spine including C-V junction showed holocord syrinx along with the CM-I.

TREATMENT

The patient underwent suboccipital craniectomy, C1 laminectomy and duroplasty decompressing foramen magnum. Besides surgical treatment, he was offered neurorehabilitation and ankle-foot orthosis for the foot drop.

OUTCOME AND FOLLOW-UP

There was no postoperative complication. After 4 weeks of surgery, moderate recovery of right foot drop was noticed. The power of right foot dorsiflexors and invertors were MRC grading of 3/5 (preoperative power strength of dorsiflexors was 0/5 and invertors was 1/5). Powers of the rest of the muscle groups were same as preoperative state (figure 2). Follow-up post-operative MRI of spine (after 4 months) showed marked reduction of syrinx with placement of cerebellar tonsil above the foramen magnum (figures 6 and 7).

Figure 5 Sagittal T1-weighted sequence of MRI of dorsolumbar spine showed hypointense central cavitory lesion involving whole cord up to conus medullaris (yellow arrow).

Figure 7 Post operative Sagittal T2-weighted sequence of MRI of dorsolumbar spine revealed marked reduction of central cavitory lesion as compared with previous scan (yellow arrow).

Figure 6 Post operative Sagittal T2-weighted sequence of MRI of craniocervical junction with cervical spine revealed marked reduction of central cavitory lesion as compared to previous scan (yellow arrow).

DISCUSSION

CM-I was first described by Hans Chiari, an Austrian pathologist in the late 19th century. It is the most common type of Chiari malformations and as many as 70% of syrinx is associated with it. It is simply the abnormal cerebellar tonsillar herniation below the foramen magnum (5 mm or more in young adults).² Apart from tonsillar herniation, other associated features of CM-I are compression of cerebellar cisterns, decreased posterior fossa (PF) volume and dysplasia of the posterior skull base.² The common mechanisms of tonsillar herniation are cranial constriction, spinal cord tethering, cranial settling, intracranial hypertension and intraspinal hypotension.³ Patients with CM-I usually develop first symptom in adolescence or early adulthood and few of them have associated preceding trivial neck or head trauma.⁴ The common presentations of CM-I are occipital headache, neck pain and less frequently visual disturbances, neurootological symptoms and cranial nerve involvement.⁵ Patients with CM-I are usually associated with syringomyelia leading to motor, sensory, cerebellar and bladder disturbances. The presentations of syringomyelia are multisegmental with features of upper motor neuron in lower extremities and lower motor neuron findings in upper extremities.

The present case appears to be one of the rare early manifestations of syringomyelia with rapidly progressive unilateral foot drop without any preceding illness or history of neck or head trauma. There was no clinical evidence of short neck or low hair line as well as craniobulbar, cerebellar, bowel, bladder and upper limb involvements. He was initially misdiagnosed to be foot drop secondary to peripheral nerve involvement. Besides right foot drop and sensory changes in L2, L3 and L4

dermatomes, brisk deep tendon reflexes in left knee and ankle suggest involvement of corticospinal tract. Though nerve conduction study was non-contributory, needle EMG in proximal group of muscles including lumbar paraspinals suggest involvement of either lumbar roots or anterior horn cell. Further investigations with MRI of spine and C-V junction showed holocord syrinx along with the CM-I. Posterior fossa decompression and C1 laminectomy was the standard method which was opted here. The patient showed good clinical as well as neuroimaging response. To our knowledge very few case reports were described in literatures regarding syringomyelia as a central cause of foot drop.^{6–8} All the case reports described are in young children (4.5 years, 5.5 years and 14 years) and all the patients had definite abnormal nerve conduction studies. In contrast, our patient had higher age (16 years) of presentation with only isolated unilateral foot drop. The nerve conduction study was normal contrary to others.^{6–8} Eventually the needle EMG and spine neuroimaging support the diagnosis. Hence it can be concluded that, when foot drop is associated with atypical clinical findings or electrophysiological suggestion of proximal lesion, neuroimaging corresponsive to the clinical localisation can substantiate the true aetiological diagnosis.

Learning points

- ▶ Syringomyelia represents the uncommon and underappreciated cause of foot drop and hence one should keep in mind as an important differential diagnosis.
- ▶ Detailed structured clinical interview and comprehensive neurological examination may disclose the true aetiology.
- ▶ Electrophysiology and neuroimaging substantiate the clinical examination.
- ▶ Early diagnosis and treatment may halt the progression of the course with possible improvement of the disease per se.

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REFERENCES

- 1 Goia E, Hamilton L, Chan J, *et al.* Unilateral foot drop as an initial presentation of a brain tumor in a child. *J Child Neurol* Published Online First: 1 Mar 2013. doi:10.1177/0883073813479172
- 2 Elster AD, Chen MY. Chiari I malformations: clinical and radiologic reappraisal. *Radiology* 1992;183:347–53.
- 3 Milhorat TH, Nishikawa M, Kula RW, *et al.* Mechanisms of cerebellar tonsil herniation in patients with Chiari malformations as guide to clinical management. *Acta Neurochir (Wien)* 2010;152:1117–27.
- 4 Meadows J, Kraut M, Guarnieri M, *et al.* Asymptomatic Chiari type I malformations identified on magnetic resonance imaging. *J Neurosurg* 2000;92:920–6.
- 5 Milhorat TH, Chou MW, Trinidad EM, *et al.* Chiari I malformation redefined: clinical and radiologic findings for 364 symptomatic patients. *Neurosurgery* 1999;44:1005–17.
- 6 Muhn N, Baker SK, Hollenberg RD, *et al.* Syringomyelia presenting as rapidly progressive foot drop. *J Clin Neuromuscul Dis* 2002;3:133–4.
- 7 McMillan HJ, Sell E, Nzau M, *et al.* Chiari I malformation and holocord syringomyelia presenting as abrupt onset foot drop. *Childs Nerv Syst* 2011;27:183–6.
- 8 Saifudheen K, Jose J, Gafoor VA. Holocord syringomyelia presenting as rapidly progressive foot drop. *J Neurosci Rural Pract* 2011;2:195–6.

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