Sultan Qaboos University Med J, November 2014, Vol. 14, Iss. 4, pp. e585–586, Epub. 14TH Oct 14
Submitted 3RD Jan 14
Revisions Req. 9TH Feb & 2ND Mar 14; Revisions Recd. 16TH Feb & 26TH Mar 14
Accepted 2ND Apr 14

INTERESTING MEDICAL IMAGE

Bilateral Wrist Drop at Presentation in a Child with Spinal Muscular Atrophy Type I

*Roshan Koul,[1] Rana Abdelrahim,[1] Susan Al-Nabhani,[2] Amna Al-Futaisi[1]

ارتخاء المعصمين في طفل لديه ضمور العضلات الشوكي من النوع الأول

روشان كول، رنا عبد الرحيم، سوزان النبهاني، آمنة الفطيسي

Figure 1: Photograph of a five-week-old male infant with spinal muscular atrophy type I, presenting with bilateral wrist drop (left wrist).

Spinal muscular atrophy (SMA) is a hereditary degenerative disease affecting the anterior horn cells of the spinal cord and cranial motor nerve nuclei, particularly the hypoglossal nerve, leading to progressive paralysis.[1] Based on clinical features, the disease is classified into three types: type I, type II and type III.[2] Data suggest that 98% of children with SMA have *survival of motor neuron* (*SMN*) gene deletion.[3] The diagnosis of SMA is based on clinical features, electromyography, muscle histopathology and genetic studies. With the advent of genetic tests, muscle biopsies are now rarely performed. SMA type I is not an uncommon disorder, with six to eight cases diagnosed every year at the Sultan Qaboos University Hospital in Muscat, Oman.[4] Children with SMA type I do not usually survive beyond two years of age.[5] Cases of wrist drop (hand drop) are uncommon among paediatric patients. A case of SMA type I in an infant, with bilateral wrist drop at presentation, is described below.

A five-week-old male infant presented to Sultan Qaboos University Hospital in April 2013 with decreased movements of the upper and lower limbs. The child had been born with no complications to consanguineous parents. Throughout her pregnancy, the mother had felt normal fetal movements. When the infant was just over one month old, the mother noted that his right hand was weak, with wrist drop. After a week, wrist drop was noted in his left hand and wrist as well. Over the following two weeks the infant's lower limbs also became weak. The infant's two older siblings were normal and the mother denied any history of trauma or pressure on the infant's arms.

On examination, the child was alert with normal cranial nerves, other than tongue fasciculations. He had weak respiratory muscles, generalised hypotonia

Departments of [1]Child Health and [2]Clinical Physiology, Sultan Qaboos University Hospital, Muscat, Oman
**Corresponding Author e-mail: koul@squ.edu.om*

and *areflexia*. The lower limbs were weaker (grade 1 of 5) than the upper limbs (grades 2–3 of 5), with the proximal limbs weaker than the distal. The patient showed obvious bilateral wrist drop [Figure 1]. Electromyography (EMG) revealed pure axonal motor neuropathy in the upper and lower limbs, with neurogenic EMG confirming the clinical suspicion of SMA. A radial nerve conduction test revealed bilateral motor axonopathy with preserved sensory action potentials and velocity, which is a feature suggestive of a lesion in the anterior horn cells and excludes radial neuropathy. The sensory nerve action potential and the conductions in the other nerves were normal. Although the diagnosis was clear from electrophysiology and the patient's clinical features, blood test results confirmed that the *SMN1* exons 7 and 8 were deleted on both alleles of the gene.

Comment

Wrist drop or hand drop is uncommon in childhood. The first case of unilateral wrist drop was reported in an eight-year-old boy in 1926.[1,6,7] Lesions of the anterior horn cells in the spinal cord, motor nerve roots, brachial *plexus* or the radial nerve can produce a unilateral wrist drop.[2,8] In rare cases, Nevo syndrome and cortical lesions can also result in wrist drop.[9] Bilateral wrist drop has been reported in cases of paediatric lead poisoning and infantile chronic immune-mediated demyelinating polyneuropathy.[10,11] Guillain-Barré syndrome and human immunodeficiency virus can also produce neuropathy and wrist drop.[8] Bilateral transient wrist drop was reported in a one-month-old baby after adhesive bandages were applied to both forearms in order to stabilise intravenous cannulas.[12] SMA and poliomyelitis are other conditions which can result in wrist drop.[11] The presentation of wrist drop in children with SMA type I is very rare. However, in view of the current case, SMA type I should be considered as a potential cause of wrist drop in infancy.

References

1. Dubowitz V, Ed. Muscle Disorders in Childhood. 2nd ed. London, UK: Bailliere Tindall, 1995.
2. Munsat TL, Davies KE. International SMA Consortium meeting (26–28 June 1992, Bonn, Germany). Neuromuscular Disord 1992; 2:423–8. doi: 10.1016/S0960-8966(06)80015-5.
3. Lefebvre S, Bürglen L, Reboullet S, Clermont O, Burlet P, Viollet L, et al. Identification and characterization of a spinal muscular atrophy-determining gene. Cell 1995; 80:155–65. doi: 10.1016/0092-8674(95)90460-3.
4. Koul R, Al Futaisi A, Chacko A, Rao V, Simsek M, Muralitharan S, et al. Clinical and genetic study of spinal muscular atrophies in Oman. J Child Neurol 2007; 22:1227–30. doi: 10.1177/0883073807306268.
5. Cobben JM, Lemmink HH, Snoeck I, Barth PA, van der Lee JH, de Visser M. Survival in SMA type I: A prospective analysis of 34 consecutive cases. Neuromuscular Disord 2008; 7:541–4. doi: 10.1016/j.nmd.2008.05.008.
6. Rao R, Balachandran C. Multiple grade II deformities in a child: Tragic effect of leprosy. J Trop Pediatr 2010; 56:363–5. doi: 10.1093/tropej/fmp138.
7. Sutherland GA. Case of wrist drop in a child. Proc R Soc Med 1926; 19:48–9.
8. Duman O, Uysal H, Skjei KL, Kizilay F, Karauzum S, Haspolat S. Sensorimotor polyneuropathy in patients with SMA type-1: Electroneuromyographic findings. Muscle Nerve 2013; 48:117–21. doi:10.1002/mus/23722.
9 Al-Gazali LI, Bakalinova D, Varady E, Scorer J, Nork M. Further delineation of Nevo syndrome. J Med Genet 1997; 34:366–70. doi: 10.1136/jmg.34.5.366.
10. Imbus CE, Warner J, Smith E, Pegelow CH, Allen JP, Powars DR. Peripheral neuropathy in lead-intoxicated sickle cell patients. Muscle Nerve 1978; 1:168–71. doi: 10.1002/mus.880010211.
11. Majumdar A, Hartley L, Manzur AY, King RH, Orrell RW, Muntoni F. A case of severe congenital chronic inflammatory demyelinating polyneuropathy with complete spontaneous remission. Neuromuscular Disord 2004; 14:818–21. doi: 10.1016/j.nmd.2004.09.003.
12. Shime N, Kato Y, Tanaka Y, Kim WC. Bilateral transient radial nerve palsies in an infant after cardiac surgery. Can J Anaesth 2001; 48:200–3. doi: 10.1007/BF03019736