CASE REPORT

Flail arms in a parachutist: an unusual presentation of hereditary neuropathy with liability to pressure palsies

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Parachuting is a popular sport but carries with it an appreciable injury risk. The case is presented of severe arm weakness after a parachute jump in a susceptible young woman.

Hereditary neuropathy with predisposition to pressure palsies (HNPP) is an autosomal dominant condition characterised by recurrent nerve injury to seemingly innocuous compressive forces. In this case report, an unusual manifestation of this condition in a previously well young woman after a parachute jump is described. This case highlights the importance of avoiding situations that may be harmful in this disorder. Some general principles of parachuting injuries are discussed.

CASE REPORT

An 18 year old woman with one week of bilateral upper limb weakness presented to the Austin Hospital in July 2000. One week before presentation, she had been parachuting in New Zealand. This was her first ever parachute jump.

It was a tandem jump, in that she was attached to the front of the instructor by a harness with very tight shoulder straps. During the descent, the parachute opened normally with not a particularly forceful pull. Half way down, as she dangled in the harness, she felt pain in her left shoulder.

When she got to the ground, after landing with a minor bump, she remained lying down as the harness was removed. She was then unable to move either arm and could not get up. After 5–10 minutes of rest (and considerable panic), the power in the right arm returned to about 80%. Unfortunately, the left arm remained weak and numb, with only a tiny improvement in strength.

There was no neck or back pain, nor any leg pain, weakness, or tingling. Bowel and bladder function were normal. She had previously been very well. There was no family history.

Examination disclosed mild right arm weakness of 4/5. The left arm was profoundly weak with shoulder power –4/5 but the remainder 1–2/5. Left arm reflexes were lost, while on the right, only the brachioradialis and a tiny biceps jerk remained. Lower limb and cranial nerve examinations were normal.

The clinical findings of bilateral upper limb weakness with hyporeflexia would at first glance appear to suggest a lower motor neurone lesion such as traumatic nerve root avulsion or a brachial plexus injury. Importantly, a spinal cord injury must be excluded, as in the initial stages of spinal cord injury profound hyporeflexia may be seen. This is known as the spinal shock stage. Immobilisation of the spine is vital until this can be excluded. With cord damage, autonomic instability may occur, so careful monitoring of vital signs is important.

This pattern of upper limb weakness and hyporeflexia and a paucity of lower limb signs is seen in the cervical “central cord syndrome”.¹ In cervical trauma, the central part of the spinal cord (which segmentally supplies the arms) is often damaged more than the peripheral long tracts to the lower limbs. Thus the legs are relatively spared.

Investigations found a normal full blood count, electrolytes, and liver function. Plain radiographs of the cervical spine were normal; however, plain films may miss important spinal pathology such as acute disc protrusion, spinal canal haematoma, and nerve root avulsion.

A magnetic resonance imaging scan of the cervical cord, nerve roots, and brachial plexus was normal. Electrodiagnostic evaluation two weeks from symptom onset found evidence of a sensorimotor neuropathy affecting upper and lower limbs (table 1). The reason for the delay in nerve conduction and electromyography is that signs of denervation (which would indicate a more severe injury with axonal disruption rather than neuropraxia) take seven to ten days to develop in affected muscles.

Prolonged distal latencies with slowing at common sites of compression (including the elbow, carpal tunnel, and the fibular neck) were found. Needle electromyography showed considerably reduced recruitment patterns, but no evidence of

Table 1 Motor nerve conduction

Nerve/segment	Latency (ms)	Amplitude (mV)	Velocity (m/s)	Normal velocity (m/s)
L Median wrist-APB	5.8	4.1		
L Median elbow-APB	11.8	3.4	41.8	>48
L Ulnar wrist-ADM	4.3	6.2		
L Ulnar below elbow-ADM	7.7	5.1	51.1	>48
L Ulnar above elbow-ADM	15.1	3.1	27.6	>48
R Median wrist-APB-ADM	7.0	4.6		
R Median elbow-APB	12.3	4.3	47.7	>48
R Ulnar wrist-ADM	4.9	9.8		
R Ulnar below elbow-ADM	7.8	9.4	61.6	>48
R Ulnar above elbow-ADM	13.8	6.2	31.9	>48
R Peroneal ankle-EDB	10.8	3.1		
R Peroneal fibular head-EDB	21.4	3.1	27.2	>40
R Peroneal knee-EDB	26.7	2.9	28.3	>40

L, Left; R, right; APB, abductor pollicis brevis; ADM, adductor digiti minimi; EDB, extensor digitorum brevis.

Table 2 Nerve injury in hereditary neuropathy with predisposition to pressure palsies

Nerve involved	Frequency (%)
Peroneal	36
Ulnar	28
Brachial plexus	20
Radial	13
Other	17

Taken from Mouton *et al*² (70 patients).

active denervation. Fluorescein in situ hybridisation testing showed a deletion on chromosome 17p11.2, as seen in most cases of HNPP.² The patient's father was found to have the same deletion. The patient made a good recovery, but return of strength took some nine months.

DISCUSSION

Recurrent painless neuropathies, caused by minor trauma, are characteristic of HNPP. In this autosomal dominant disorder caused by mutations in the peripheral myelin protein 22 gene (PMP22), phenotypic variation is quite variable² often with asymptomatic relatives. PMP22, a membrane glycolipid protein of peripheral nervous system myelin, is thought to play a role in myelin formation and integrity.³ The gene encoding PMP22 is on chromosome 17p11.2. This 1.5 Mb segment is deleted in most cases of HNPP, but frame shift, nonsense, and reciprocal translocation defects have been described.^{3,4} Interestingly, the PMP22 gene is duplicated in Charcot-Marie tooth disease type 1A.⁵

Bilateral brachial plexopathy is an unusual finding of HNPP;^{2,5,6} and this case was quite a remarkable first manifestation of this disease. Mononeuropathies are more common, as described by Mouton *et al*² (table 2). We postulate that the tight harness around the shoulders led to a compressive neuropraxic injury.

Conclusion

Parachuting injuries are common because of the rapid deceleration forces involved and the popularity of the sport. In a study during the 1980s of 110 000 sports jumps,⁷ Ellitsgaard reported serious injuries requiring medical treatment at a rate of 0.14% of jumps. Most of these were fractures or serious soft tissue injuries. Neurological injury is less common,⁷ but the consequences can be profound. It is important to take appropriate steps to prevent further cord injury (including spinal immobilisation) before transfer for further investigation at an appropriate facility. In the above parachute study, there were six deaths in the 110 000 jumps (0.005%).

Take home message

Parachuting has the potential to cause serious injury, and, if spinal injury is suspected, spinal immobilisation and transfer to an appropriate facility should occur immediately.

Our patient highlights the vulnerability of patients with HNPP to nerve damage. Practical advice, such as to avoid carrying heavy bags over the shoulders, resting on the elbows, or crossing the legs, is very important to prevent these problems.^{4,6} Tight harnesses such as in parachuting, rock climbing, and abseiling should be used with caution in such patients.

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