CURRENT TOPIC

Chinese paralytic syndrome or acute motor axonal neuropathy

Neil Gordon

When visiting China in November 1983 in a group organised by the British Paediatric Association we were shown a number of children in hospital with a diagnosis of the Guillain-Barré syndrome. We commented at the time on the frequency of this disease in China compared with its relative rarity in the UK.

Clinical features
Since then there have been a number of publications on the occurrence of epidemics of peripheral neuropathies in China and elsewhere. McKhann et al investigated 36 patients, aged 15 months to 37 years, with this syndrome in hospitals in Beijing and Shijiazhuang.1 Most were from rural areas, and in about half of them there was a history of an illness during the four weeks before the onset of the neuropathy. The earliest symptoms were of neck stiffness and weakness of the legs. The weakness ascended symmetrically to involve the arms and the muscles of swallowing and respiration, and there was loss of tendon reflexes. It was at its worst about six days after the onset.

Weakness of the bulbar muscles occurred in 61% of the patients, but only one had paralysis of the ocular muscles. Sensation was normal, but hyperhidrosis was noted over the head and parts of the trunk. Assisted respiration was needed by 31% of those affected. Cardiac arrhythmias and alterations of blood pressure have occurred. The concentration of protein in the cerebrospinal fluid was raised in 42% and the mean cell count was 3 cells/μl. There was severe reduction in motor evoked amplitudes from distal stimulation in those tested, but sensory action potentials were normal, except for two patients in whom they were absent.2 Electromyography showed denervation potentials in the limb muscles. Most patients recovered, at least partially, and the time from the onset of the illness to the start of recovery averaged 16 days.

Nature of the illness
The epidemiological evidence, and the clinical and electrophysiological findings, suggest that the disorder is different from the Guillain-Barré syndrome and from poliomyelitis. The latter findings are in favour of a reversible distal motor nerve terminal or anterior horn cell lesion.1 The aetiology of the Guillain-Barré syndrome is unknown, but the most favoured hypothesis is the generation of autoimmune responses because of the similarities between infective agents and host proteins.3

In Europe and North America the Guillain-Barré syndrome occurs sporadically, while in northern China this acute disease can reach epidemic proportions between June and October, affecting children and young adults, mostly from rural areas. There is a recurrence rate of less than 5% and the mortality of the disease is about 3–5%. In contrast to poliomyelitis there is no fever at the onset, no muscle tenderness, and very few cells in the cerebrospinal fluid. Most patients with poliomyelitis live in urban areas, and recovery from this disease is very variable; this is quite apart from the fact that the poliovirus has never been recovered from those with the Chinese paralytic syndrome. The Guillain-Barré syndrome is also different as it is a non-epidemic illness, and there are often sensory abnormalities. The electrophysiological findings help in the differential diagnosis, showing evidence of demyelination in this syndrome.

The seasonal incidence of the Chinese syndrome makes botulism and heavy metal poisoning most unlikely, although some type of intoxication with a seasonal incidence has not been excluded. Also there is no definite evidence of any viral infection. However there are similar disorders that occur in epidemics in other countries: in Bangladesh, India, Japan, and Latin America. There are differences, but they do share an involvement of the motor system neurone, and not of the myelin sheath as in the Guillain-Barré syndrome.

A boy aged 2-5 years, returning to the UK after a holiday in Bangladesh, showed clinical evidence of the Guillain-Barré syndrome, but investigations confirmed an axonal disorder similar to the Chinese paralytic syndrome, leading to the suggestion that the condition should be known as the Asian paralytic syndrome.4

In a record of acute non-inflammatory neuropathies occurring in India a number of patients showed the clinical and electrophysiological characteristics of the Chinese paralytic syndrome, which seemed to be of anterior horn cell origin, although fewer had absent tendon reflexes.5
In Japan a motor polyneuropathy has been reported with an onset after diarrhoea due to *Campylobacter jejuni*. The clinical picture may be similar to the Guillain-Barré syndrome, but as has been stated the pathology is that of an axonal degeneration of the motor nerves, and not a demyelinating disorder. Diarrhoea also occurred before a polyneuritis in Bangladesh, and serum antibodies to this organism were often high in the Chinese paralytic syndrome. Therefore there may well be a role for an infection with *C. jejuni* in the aetiology of the Chinese paralytic syndrome, as there appears to be in the Guillain-Barré syndrome.

Studies of patients in Mexico showed that at least half of them lacked the lesions in their peripheral nerves typical of the Guillain-Barré syndrome, and other differences included the amount of protein in the cerebrospinal fluid early in the disease, changes in the neurones, and in spite of resistance to passive flexion of the neck and to straight leg raising, the possibility of raising the patient by the shoulders without causing pain as long as the head was allowed to fall backward. Postmortem studies on 57 Mexican children showed that the disease in 32 was due to the poliovirus, and in 25 there were no inflammatory changes in the central nervous system. The findings in 10 of these were compatible with the Guillain-Barré syndrome, eight showed extensive chromatolysis of lower motor neurones and were categorised as cytoplasmic neuropathy, and seven showed argyrophil degeneration of the nuclei of the majority of the lower motor neurones, but no chromatolysis, and were categorised as nuclear neuropathy. The interval between the onset of weakness and death was longer than one week in 70% of the Guillain-Barré group, one week or less in 75% of the cytoplasmic neuropathy group, and less than one week in 86% of the nuclear neuropathy group.

A similar group of patients, possibly with cytoplasmic neuropathy, has been described in Spain.

It is suggested that there might be a number of toxins with a varying affinity for the peripheral portions, the cytoplasm, or the nuclei of the neurones. Poisonous berries growing in Mexico are one possibility. If there is a slightly raised cell count in the cerebrospinal fluid a mistaken diagnosis of poliomyelitis may be made.

There have also been reports in the USA of reversible motor neurone disease in adults, which may be of toxic or infectious origin.

**Conclusions**

In a recent review of this subject McKhann et al have entitled it 'acute motor axonal neuropathy'; most cases occurring in the summer months among children and young adults, most of them living in rural areas. It shares clinical and cerebrospinal fluid findings with the demyelinating Guillain-Barré syndrome, but the results of the electrophysiological and pathological investigations are different. These indicate an axonal neuropathy. There was a history of a febrile illness in 30% of the patients before the onset of the weakness; some had diarrhoea and some symptoms of an upper respiratory infection.

Recovery of the patients examined was on the whole good, but there was a significant mortality. Recurrence, although rare, can occur.

The cause of this disease is unknown, but now that it has been separated from the Guillain-Barré syndrome there will be opportunities for further research. Establishing the aetiology will be essential for effective epidemiological studies, and for prevention and treatment.

---

Chinese paralytic syndrome or acute motor axonal neuropathy.

N Gordon

Arch Dis Child 1994 70: 64-65
doi: 10.1136/adc.70.1.64

Updated information and services can be found at:
http://adc.bmj.com/content/70/1/64.citation

These include:
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/