Acute inflammatory demyelinating polyradiculoneuropathy following varicella

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Summary: Four cases of acute inflammatory demyelinating polyradiculoneuropathy following varicella are described. The role of immunosuppression as a contributing factor in triggering an autoimmune disease of the peripheral nervous system following viral infection is discussed.

Introduction

Acute inflammatory demyelinating polyradiculoneuropathy (AIDP) or Guillain-Barré syndrome (GBS) is a rare complication of varicella infection, the calculated rate being one case per 15,000 cases of varicella.¹ AIDP constituted 7% of neurological complications of varicella studied by Miller *et al.*² Twenty six cases of AIDP in association with varicella have been reported up to 1984.¹ Four cases of AIDP following varicella were seen over a period of 8 years, during which period a total of 62 cases of AIDP were seen.

Case reports

Case 1

A boy aged 10 years was admitted for weakness of all four limbs of 4 days duration. Eight days before the onset of these symptoms he had fever associated with rash characteristic of chicken pox. On examination he had still crusted lesions of chicken pox. Muscle tone was reduced in all the four limbs and the power was graded at 2-3/5 in the limbs. Tendon reflexes were not elicitable. Perception for all modalities of sensation was preserved. Cerebrospinal fluid (CSF) analysis showed elevated proteins, 68 mg/dl. Electrophysiological studies showed abnormalities. The distal latencies were 15.1 millisecon (normal < 3.5) and 16.1 millisecon (normal < 4.4) in the median and common peroneal nerves respectively. The compound muscle action potential (CMAP) was polyphasic and dispersed, the amplitudes were much reduced 1.8 mV and 0.8 mV from abductor pollicis brevis and extensor

digitorum brevis (normal > 6.2 and 3.3). The motor conduction velocities (MCV) 15.3 m/sec (normal > 43.2) in the median nerve and 14.2 m/sec (normal > 40.3) in the common peroneal nerve. The sensory action potential of the median nerve measured 15 μ V (normal > 10.2) in amplitude and the peak latency was 3.8 millisecon (normal < 3.4). No proper sensory action potential could be recorded from the sural nerve (technical). He made a good recovery.

Case 2

A 62 year old man was admitted with weakness, numbness and paraesthesiae of all the four limbs. He developed these symptoms 8 days after chicken pox. He was normotensive and skin showed crusted lesions of chicken pox. Muscle tone was reduced in all the four limbs, power grading showed grade 2/5 power in all the group of muscles in the lower limbs and 3/5 in upper limb muscles. Tendon reflexes were not elicitable and plantars were down going. Perception for all sensory modalities was diminished below the elbow in upper limbs and below the knee in lower limbs, more so for position and vibration sensations. CSF protein was 60 mg/dl. During his hospital stay he had progression of the disease resulting in bulbar and respiratory muscle paralysis. He was put on respiratory assistance. He had wide fluctuations in blood pressure due to associated autonomic neuropathy and died.

Case 3

An 8 year old boy was admitted for weakness of all the four limbs of one month duration. The weakness appeared acutely, 10 days after chicken pox. He had bilateral facial paralysis. Muscle power grading showed grade 0/5 power in proximal muscles and grade 2/5 power in the distal group of muscles. The

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deep reflexes were absent. No sensory deficit could be detected. CSF analysis showed 120 mg/dl of protein. Electromyography of distal muscles showed occasional fibrillation potentials and positive sharp waves. Motor conduction velocities in the median and common peroneal nerves were 39.8 m/sec and 32 m/sec respectively. Distal latencies were 4.5 msec and 6.2 msec in the median and common peroneal nerves respectively. The amplitudes of CMAPs from abductor pollicis brevis and extensor digitorum brevis were 3.5 mV and 2 mV. Sensory action potential from median nerve measured 15 μ V and the peak latency was 2.1 msec. Sensory action potential of sural nerve measured 10 μ V and the peak latency was 3.6 msec. He showed gradual improvement and at one year he was able to walk independently.

Case 4

A 52 year old man developed weakness of all the four limbs and inability to close his eyes 8 days following chicken pox. He had bilateral facial paralysis, more on the right side. The power in the lower limbs was graded 3/5 and 4/5 in the upper limbs. Deep reflexes were absent. Perception for vibration and sense of position was decreased below the knees. Motor conduction studies showed 44.4 m/s in the median nerve and 23.4 msec in the common peroneal nerve. The CMAPs from abductor pollicis brevis and extensor digitorum brevis were of 4 mV and 2 mV amplitude respectively. Sensory action potentials of median and

sural nerves measured 14.5 μ V and 8 μ V respectively, the corresponding peak latencies were 2.7 msec and 3.8 msec. He made a good recovery.

Discussion

Symptoms of involvement of the nervous system following varicella develop between the fifth and twentieth day after the appearance of the rash, usually in the first half of the second week.^{2,3} There are no neurological features which serve to distinguish the syndrome of AIDP seen in association with varicella from that seen in other clinical settings. The prognosis is good.³ The electrophysiological findings were suggestive of a demyelinating type of neuropathy such as AIDP.⁴

Variable degrees of immunosuppression, particularly cell-mediated immunity are known to follow viral infections.⁵ Depression of tuberculin reactivity occurs in chicken pox.⁶ Lisak *et al.*⁷ suggested the role of immunosuppression as a contributing factor in triggering an autoimmune disease of the peripheral nervous system. It is possible that viral infections may reduce suppressor T cell activity thereby leading to proliferation of those few T and B lymphocytes which are normally present in humans and recognise nervous system antigen.⁸ Reduction of the suppressor T cell subpopulation in patients with AIDP⁹ is consistent with this hypothesis.

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