# CENTRAL NERVOUS SYSTEM MANIFESTATIONS OF CHICKEN POX – A REPORT OF TWO CASES

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## SYNOPSIS

**2 patients with neurological complications of chicken pox are reported. The first case developed acute cerebellar ataxia and the second a radiculo-myelopathy. Neurological complications are exceedingly rare, with myelopathy the least common. The prognosis is good and complete recovery is expected, although morbidity and mortality have been reported in patients with encephalitis. Treatment is mainly supportive, although corticosteroids has been shown to be effective in a patient with severe demyelination. The use of acyclovir in immunocompromised patients with varicella infection has been provened to be efficacious, but there has been no report of its use in patients with neurological complications.**

## INTRODUCTION

Varicella is a highly contagious and common infection occurring mainly in childhood, although about 20% of patients are adults. Complications following varicella infection is uncommon and central nervous system (CNS) involvement is considered exceedingly rare. Six clinical types of neurological complications have been described (1). These include the cerebral, cerebellar, meningeal, spinal cord, neuritic and extra-pyramidal types. In this report, we present two adult patients who developed neurological problems following chicken pox. One patient presented with acute cerebellar ataxia, and the other with acute radiculo-myelopathy, the latter being one of the rarest of CNS complications.

## REPORT OF CASES

### Case 1:

The patient, S.S., a 30 year old Sikh technician, was admitted to Middleton Hospital the day after he developed a chicken pox rash. On the 12th hospital day, he developed an unsteady gait with a tendency to fall to the right. There was no headache, although he did complain of mild giddiness. Clinically there was truncal ataxia, and he was unable to perform tandem walking. Sensorium was not disturbed. Nystagmus was absent. Finger-nose and heel-shin tests were both impaired bilaterally. The muscle tone was generally decreased and deep tendon knee reflexes were pendular. Investigations revealed a normal cerebro-spinal fluid (CSF) cytology and electroencephalography showed moderate slowing with increased theta activity bilaterally. Computerised axial tomography of the head was normal. His ataxia improved and his gait was fairly steady when he was discharged one month after hospitalisation. On review a month later, he had fully recovered.

### Case 2:

A.V., a 24 year old healthy man, was admitted to Middleton Hospital on the day he developed a chicken pox exanthem. On the 5th day of hospitalisation, he felt what he described as a "very cold" sensation over the lumbar region. The following morning he complained of weakness in both lower limbs. Initially he was still able to walk, but his weakness continued to progress over the next 2 days when he was unable to move his lower limbs at all. At the same time he also developed acute retention of urine which required urinary catheterisation. Bowel control was not affected.

On transfer to the Department of Neurology, he was found to be drowsy but well orientated and responsive to commands. He was afebrile and comfortable. There were numerous crusting lesions over the face and trunk. Signs of meningism were absent and cranial nerves were intact. Both limbs were completely flaccid. Lower lower limb movements, deep tendon reflexes and plantar responses were absent. The muscle power in all upper limb movements were grade 4 (Medical Research Council Grading System), and muscle tone and deep tendon reflexes were normal. Abdominal muscle tone was decreased and superficial abodminal reflexes were absent. Muscles of respiration were unaffected. There was diminished sensation to pin prick below the 9th thoracic dermatome. Propioception was lost in the lower limbs.

Cytological examination of CSF showed a mild pleocytosis of 60 cells/mm3, consisting mainly of lymphocytes. CSF protein was mildly raised to 60 mg% and globulin was present. A myelogram was performed to exclude a compressive myelopathy and this was found to be normal. Electromyography and nerve conduction studies revealed normal peripheral nerve conduction. Absence of the F and H reflexes in the lower limbs were consistent with radicular involvement.

On the day after transfer, the patient was started on a 10 day course of ACTH. The next day his sensorium improved and the day following his upper limb power returned to normal. The remaining neurological deficits were however slow to improve. At the time of reporting, and 3 months after the onset of illness, the lower limb power had improved to grade 3. He was ambulant with the aid of cruthes. Sensory level remained at the 9th thoracic dermatome, although subjectively sensation to pin prick had improved. Bladder dysfunction persisted and this was complicated by urinary tract infections.

## DISCUSSION

The incidence of neurological complications following varicella infections in healthy individuals is rare, and the estimated incidence vary between 0.01% and 0.1%. Most cases are mild and the mortality rate is low. The prognosis for complete recovery is good (2).

The commonest neurological complication following varicella infection is encephalitis. In a series reviewed by Miller et al, this accounted for 90% of cases, of which 37% showed signs of cerebellar ataxia (3). Polyradiculitis occurred in 7% and the remaining 3% developed transverse myelitis. The average latent period between the appearance of rash and neurological complication was between 1 to 2 weeks. This time interval was similar in the 2 patients reported here.

The clinical features and natural history of acute cerebellar ataxia following varicella infection are typically featured in the first case reported. The main exception being the age of the patient in that it occurs more commonly in children (4). The prognosis is excellent and virtually all patients recover.

Varicella myelopathy is the rarest clinical type of neurological complications. To date there have been 9 case reports of "pure" post varicella acute transverse myelitis (5-13). The case reported here showed evidence of radicular involvement as well, hence presenting as a radiculo-myelopathy. This mode of presentation is very unusual, although cases of Landry-Guillain-Barre syndrome after chicken pox have been reported (14). The prognosis of myelopathy is also good, and the complete or near complete recovery within a few months is the rule. The use of steroids has not been proven to be effective. In our patient, a course of ACTH was given, and although there appeared to be improvement in upper limb power, the effect was minimal.

The pathogenetic mechanisms underlying these neurological manifestations have been thought to be either related to the direct invasion of the central nervous system by varicella-zoster virus or an immune-mediated process similar to experimental allergic encephalitis. Evidence of direct viral invasion has been shown by Peter et al (15), where he was able to demonstrate the presence of varicella zoster antigen in the CSF cells by indirect immunofluorescent technique in 2 patients with post varicella acute cerebellar ataxia. However most of the pathological studies show a morphological picture that is more consistent with allergic encephalitis (3, 16). In addition, extensive demyelination reflected by low absorption in white matter on CT scanning is again in favour of an allergic encephalitis (17). It is therefore likely that both mechanisms are possible.

An understanding of the pathogenesis is of clinical importance as it would affect the mode of therapy. This is especially so in patients with severe life-threatening encephalitis, corticosteroid therapy has been shown to be effective in encephalitis associated with extensive demyelination (17). On the other hand, the use of an anti-viral agent, acyclovir, may be indicated in cases where direct viral invasion is present. This agent has been used only in immunocompromised patients with herpes zoster or primary varicella infection (18). There has been no report on the use of acyclovir in patients with neurological complications following varicella infection.

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