ACUTE POLYNEURITIS CRANIALIS WITH TOTAL EXTERNAL OPTHALMOPLEGIA AND AREFLEXIA

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A case of acute neurological disease presenting with complete external ophthalmoplegia, bulbar palsy and areflexia is described here in order to stress the fact that, whereas the condition is very alarming, the ultimate outcome is very good.

Case-Report

Mrs. S. C., aged forty-one gave birth to a female child on March 19, 1970. She had a manual removal of placenta without anaesthesia and the baby needed resuscitation. Ten days later, on March 29, 1970, she noticed dizziness and unsteadiness in the morning. She also noticed that her voice was low pitched and hoarse. When she ate her breakfast she could not swallow properly and fluids regurgitated through her nose. The following day she complained of double vision and also tingling and numbness in her hands and feet. Her voice became so hoarse that she was barely able to speak. During the next twenty-four hours there was further deterioration with progress of all the symptoms, but the double vision disappeared. On examination on 3rd April, 1970 she was found to be alert, fully orientated, her mental functions and special senses were normal. Although the pupils reacted briskly to light, there was complete loss of movement of the eyes in all directions with slight bilateral ptosis and the eyes were fixed in the forward-gaze position. Mild bilateral facial weakness was also present. The palatal arches did not move and the gag reflex was absent. She was unable to cough. There was no weakness of trunk or limb muscles. However, all the tendon reflexes were absent and the plantar responses were flexor. She had mild tenderness of the muscles of the upper arm. Sensation was normal and her gait was slightly ataxic.

Investigations: A full blood count, serum electrolytes, a radiograph of chest and skull were normal; E.S.R. was high (57 mm. in first hour). A lumbar puncture showed clear cerebrospinal fluid (C.S.F.) under normal pressure. C.S.F. protein was high (Figure) with normal cells (less than 1/c.mm.) and a slightly abnormal Lange’s colloidal gold curve (0123321000). The Wasserman reaction in blood and C.S.F. was negative. Tests and culture of bacteria and viruses from throat, urine, faeces, blood or C.S.F. were negative.

Progress and Treatment: Intravenous injection of 10 mg. edrophonium hydrochloride on two occasions did not improve the ophthalmoplegia. A week after the onset of illness, paraesthesiae, dysphonia and dysphagia began to improve. However mild bilateral facial weakness of lower motor neurone type progressed and became very marked in another four days; Bell’s phenomenon became positive on both sides and she was unable to close her eyes properly. At this time the eye movements also began to improve. She was able to swallow fluids without regurgitation through her nose and the tendon reflexes returned in the lower limbs. Thereafter all the signs gradually cleared up. A definite improvement of the facial weakness
on the left side was noticed on 15th April, 1970. On the same date there was no
dysphagia, the eye movements had returned to a great extent and all the tendon
reflexes were present. The E.S.R. had come down to normal in ten days and C.S.F.
protein was also less but not back to normal in the third week (Figure). Electromyography and nerve conduction studies were normal. The patient did not receive
any form of medication.

**Figure**

![Graph showing protein levels in cerebrospinal fluid during the course of illness.](image)

Protein levels in cerebrospinal fluid during the course of illness.

**Discussion**

There were several clinical features in this patient which are worthy of emphasis—for example, complete bilateral external ophthalmoplegia, areflexia without loss of
power or sensation and the striking symmetry of neurological signs. There were no
prodromal symptoms and the condition progressed for the first seventy-two hours
followed by spontaneous improvement in the second week although the facial
weakness became quite evident later on. The cerebrospinal fluid protein was high
in the second week of the illness coming down but remaining high in the third week.

As there were no signs of a pyramidal or a sensory tract lesion it is likely that
the lesion in this case was in the course of cranial nerves i.e. it was polynévritis
cranialis. In the absence of mental symptoms, headache and neck stiffness, and with
normal cells in cerebrospinal fluid, it is unlikely that the patient had encephalitis
or meningitis. Wernicke's disease was not considered as there was no evidence of
malnutrition. No toxin was found and in particular botulism and diphtheria were
excluded. Absence of response to edrophonium hydrochloride excluded acute
myasthenia gravis.

Areflexia with bilateral facial weakness and cyto-albuminologic dissociation in
the cerebrospinal fluid suggested that the patient had a "Guillain-Barré type of
neuropathy". The predominance of cranial nerve palsy indicated that the brunt of
the disease was borne by the cranial nerves. Marshall (1963) in his review of cases
of Landry-Guillain-Barré syndrome observed that some of the patients also had involvement of the cranial nerves and two of these had complete external ophthalmoplegia. One of the cases of polyneuritis reported by Dreifuss, Hurwitz and John (1957) also had complete external ophthalmoplegia.

There are many similarities between the cases of ophthalmoplegia associated with peripheral neuropathy and cranial nerve palsy described by Fisher (1956), and Gibberd and Kelly (1964) and the case described here for example, the illness progressed in all the cases from forty-eight hours to seventy-two hours; there was spontaneous improvement in all the cases indicating a good prognosis; the C.S.F. protein was usually high in the second or third week (Figure) and came down subsequently. Pupillary reaction to light was only sluggish and not completely lost in the three cases with total external ophthalmoplegia reported by Fisher (1957), and one of the three cases reported by Gibberd and Kelly (1964) also had external ophthalmoplegia without internal ophthalmoplegia in the early stage of the disease.

Gibberd and Kelly (1964) considered mumps infection in their cases as antibody responses to mumps virus were present in these cases but they felt that it was unlikely, because none of their patients had been in contact with mumps and that they did not have the usual manifestation of mumps. In the present case there was no evidence of infection with mumps virus and hence it supports the view of Gibberd and Kelly (1964) that the condition is not due to the virus.

**SUMMARY**

A case of complete external ophthalmoplegia associated with bulbar palsy and areflexia is described. Although the cause remains obscure, it appears that the syndrome is a form of Guillain-Barré type of neuropathy affecting mainly the cranial nerves (polyneuritis cranialis).

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