THE LANDRY-GUILLAIN-BARRE-STROHL SYNDROME OR POLYRADICULONEUROPATHY WITH EXTRA-OCULAR MUSCLE INVOLVEMENT IN TWO CHILDREN

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Although the Landry-Guillain-Barre-Strohl Syndrome of a rapidly progressive Polyneuropathy was first described over a century ago, disagreement still exists regarding clinical findings and prognosis.

Landry in 1859 described an acute ascending paralysis in 10 of his patients, and because 2 of them died he considered the prognosis bad. In 1916, Guillain, Barre and Strohl noticed albuminocytologic dissociation of the cerebrospinal fluid in this condition. Guillain in 1936 published a resume of 10 cases and found the distal muscles weaker. He also noted facial weakness in 3 patients. He was convinced that the disease was not fatal and considered Landry's ascending paralysis a different disease. Barre in 1938 stated that proximal muscles were weaker than distal ones in the condition.

The aetiology and treatment of this condition too is still not settled. From about 1950, ACTH and steroids have been tried but opinions on their efficacy are still varied.

This paper is a report on 2 cases of the Landry-Guillain-Barre-Strohl Syndrome with extra-ocular muscle involvement seen in this Unit. The first case presented with diplopia and weakness of legs. The second case was interesting in that she presented with bilateral ptosis and weakness of legs. As far as we know, in our review of the English literature we did not come across any description where bilateral ptosis was the presenting feature.

CASE 1

L.C.C., a 9-year-old Chinese male was admitted on the 14th of April 1965 with the complaints of headache, fever and weakness of legs for 10 days and diplopia on the day of admission. The headache was in the frontal region and was persistent but not severe. He vomited once at the onset of the headache. The fever was mild without chills, rigors or sweating. His leg weakness was slowly progressive and on the day of admission he could just manage to walk unsteadily without support. He had been lying in bed most of the time for the 7 days prior to admission. Direct questioning only revealed slight pain in his right calf for the last 3 days before admission. No sensory changes were experienced, and diplopia without associated vertigo only developed on the day of admission. He did not complain of a sore-throat and there was no significant past history.

On examination, his general condition was considered fair. He had a temperature of 99·8°F. There was mild pharyngitis and tonsillitis. His lungs were clear and no abnormality was detected in his cardiovascular system. Blood pressure was 115/70 mm Hg. His abdomen was soft.

As regards the central nervous system he was fully conscious and alert. There was neck rigidity and a bilateral positive Kernig's sign. Power was impaired in both his lower limbs and also his left upper limb, but he managed to walk unsteadily without support. Deep tendon reflexes were all present, though somewhat sluggish in the lower limbs. Babinski reflex was negative; sensations were all intact. All the cranial nerves were intact except for a bilateral VI nerve palsy which was not marked. The pupils were equal and reactive to light and accommodation and fundoscopy revealed no abnormality. There was no nystagmus or facial palsy.

In the ward he continued to complain of diplopia at first and the bilateral VI nerve palsy was confirmed by the eye surgeon. The diplopia and VI nerve palsy disappeared spon-

TABLE I

Date	Cells	Chloride(mg%)	Total Protein (mg%)	Globulin	Glucose
14.4.65	2	680	70	+	+
19.4.65	2	700	60	+	+
18.5.65	2	700	160	+	+

taneously after a month's stay in hospital. In all, he was warded for just over 2 months. He was afebrile from the 4th day.

During the first few weeks, his leg muscle power deteriorated until he was unable to walk at all by the 5th week, and the knee and ankle jerks could not be elicited. The proximal muscles appeared to be more severely affected than the distal. However, he started to make a rapid recovery from the 6th week and by the 7th week, he was walking without support all over the ward and all his reflexes had returned. His sensations remained intact throughout.

Because his neck remained less flexible than normal, an X-ray of the cervical spine was taken and this revealed the cause—he had a hemivertebra of C_7.

In the investigations, only the cerebrospinal fluid findings were significant and they are shown in Table I. The albuminocytologic dissociation is obvious.

During his stay the patient was treated symptomatically and also had Vitamin B_1 injections and tablets. No steriods were given. He was discharged well at the end of 2 months.

Cerebro-Spinal Fluid Findings

CSF clear macroscopically in all 3 specimens. Smear showed no organisms, few lymphocytes in all 3 specimens.

CASE 2

A. L., a 6-year-old girl was admitted to the Unit with a history of coryza, cough and slight pain over both legs, 1 week prior to admission. 3 days later, she developed drooping of both upper eye lids followed by weakness of legs 1 day before admission. She also had headache and numbness of both hands.

On examination, the child was conscious, intelligent and cooperative. Her general condition was fair. The significant findings were ptosis of both eye lids (see Fig. 1) and weakness of both lower limbs especially the distal muscles. The lower limbs were hypotnic but more marked over the dorsiflexor of the ankles and intrinsic muscles of the feet. The deep tendon reflexes of both knees and ankles were absent.

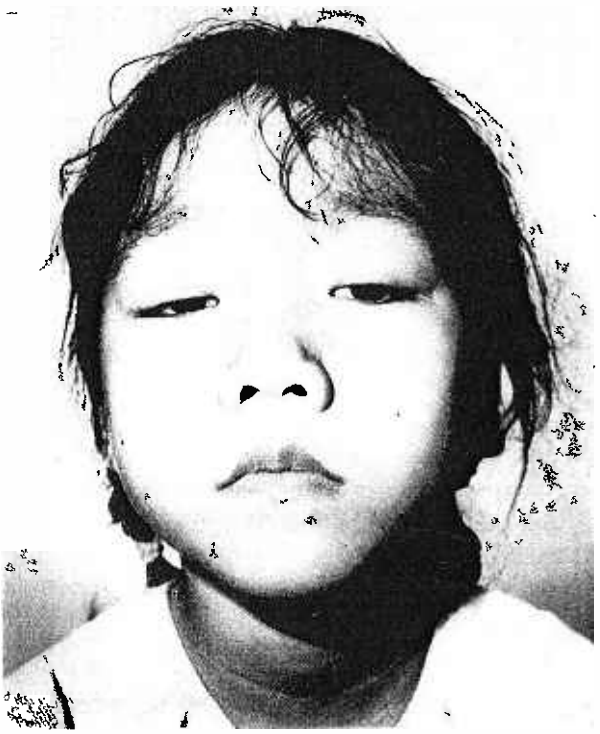

Fig. 1. Note bilateral ptosis of upper eye lids.

Lumbar puncture revealed a clear fluid with the following findings: Cell count = 3/c.m.; Chlorides = 700 mgm%; Globulin = +; Glucose = 70 mgm% and Total Protein = 100 mgm%.

In view of the possibility of myasthenia gravis, a neostigmine test was done on 2 occasions with no improvement of the ptosis although the child developed abdominal pain

and vomiting on the second occasion. A few days later 'tensilon' test was done with equally negative response.

The ptosis of her eyes showed spontaneous improvement on the 5th week of illness. By the 5th week there were no more ptosis (see Fig. 2). The weakness of both lower limbs lasted for 7-8 weeks.

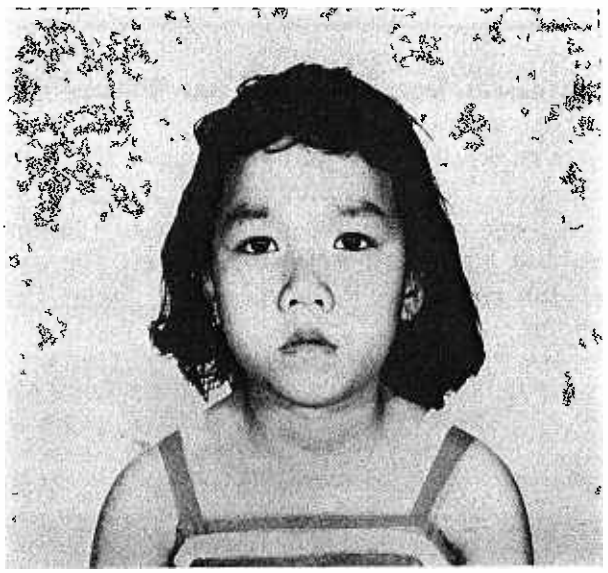

Fig. 2. Six weeks after admission, no evidence of ptosis.

During her stay in hospital no specific treatment was given. She was put on soft diet and multivitamin tablets daily.

DISCUSSION

The Landry-Guillain-Barre-Strohl Syndrome with extra-ocular muscle involvement has been described in the literature but its infrequency has often been stressed. In contrast the frequency of facial nerve involvement is often emphasised.

Taylor and McDonald[12] in describing the syndrome of polyneuritis with facial diplegia in 1932 stated that facial nerve palsy was fairly common but that other cranial nerves were rarely involved. In 1936, Gilpin, Moersch and Kernohan[5] analysed the cases seen at the Mayo Clinic for the last 15 years and found 35% with facial weakness.

Roseman, Ephraim and Aring[10] in 1941 from cases seen at Cincinnati General Hospital in the last 5 years, found in 16 selected cases with progressive flaccid quadruplegia that 10 had facial diplegia and 3 had unilateral facial involvement. The paresis was nuclear or infranuclear with involvement including the brow and eyelids. Four cases had paralysis of accommodation. Palatal weakness, dysphagia, voice changes, weakness of muscles of mastication, anosmia and deafness were mentioned specifically but not extra-ocular muscle weakness.

Also in 1941, Forster, Brown and Merritt[4] analysed 22 cases of polyneuritis with cranial nerve involvement seen at the Boston City Hospital in the last 10 years. All 22 had bilateral facial nerve involvement. The authors noted the conspicuous infrequency of involvement of the oculomotor nerves—only present in 2 of their cases with one involving the VI, and the other the III cranial nerve. Six of the cases had only VII nerve involvement; 5 had VII and X nerves affected. The others had VII and either V or IX, X, XI and XII nerve involvement.

Various authors[1,5,6,13] have quoted a figure of about 30% of cases having facial nerve palsy and have stressed on this being the most common cranial nerve lesion. They also state that dysarthria and dysphagia are fairly common complaints. Other cranial nerve involvements are unusual[8,12].

Jolly and Amarjit Singh[11] in 1958 reported that the facial nerve was the most frequent cranial nerve to be involved, followed by the IX and X nerves. In 25 of their own cases reviewed, only 2 had VI nerve involvement.

In 1959, 37 cases of polyradiculoneuropathy, being the cases seen at Vanderbilt Medical Centre from 1952—58, were reviewed[7] 20 patients had cranial nerve involvement—12 had facial nerve palsy; 2 complained of diplopia.

In another survey at the Mayo Clinic in 1964[13] of 97 patients seen from 1950—1963, 28 had facial palsy and 5 had extra-ocular muscles involved. Two others had involvement of palate and tongue.

We thus note that most authors have found that cranial nerve involvement is fairly common in the Landry-Guillain-Barre-Strohl Syndrome, the facial nerve being by far the one most commonly involved. Various authors have arrived at a frequency of about 30% with facial nerve involvement. The infrequency of other cranial nerve involvement and in par-

ticular extra-ocular muscle involvement has been stressed by various authors.

The mode of presentation in Case 2 was interesting in that it resembled that of a case of myasthemia gravis. Not only was there no response to neostigmine and tensilon test, but also she recovered completely spontaneously. The patient was followed up for 2 years by one of us (T.K.H.) and there was no relapse of her condition. Both cases had cerebro-spinal fluid findings showing normal cell count but raised total protein.

SUMMARY

Two cases of the Landry-Guillain-Barre-Strohl Syndrome with extra-ocular muscle involvement seen in this Unit are presented.

The frequency of cranial nerve involvement in this Syndrome is noted, the facial nerve being by far the commonest cranial nerve involved. The figure of about 30% of cases with involvement of the facial nerve is often quoted.

The infrequency of other cranial nerve involvement and in particular extra-ocular muscle involvement is stressed.

REFERENCES

1. Brown, J.R., and Baker, A.B. Am. J. Med. 2, 45-52 (Jan.) 1947.
2. Charous, D.I., and Saxe, B.I. New Eng. J. Med. 267, 1334-1338. (Dec.) 1962.
3. Fisher, Miller, New Eng. J. Med. 255: 57-65. (July) 1956.
4. Forster, F.M., Brown, Madelaine and Merritt, H.H. New Eng J. Med. 255, 51-56. (July) 1941.
5. Gilpin, S.F., Moersch, E.P., and Kernohan, J.W. Arch. Neurol. & Psychiat. 35, 937-963 (May) 1936.
6. MacNeal, P.S., and Bland, J.H. Am. Pract. & Digest Treat. 1, 337-346. (April) 1950.
7. Merrill, R.E., and Fredrickson, D. J. Pediat. 54, 816-822. (June). 1959.
8. Petch, C.P. Lancet 2, 405-408. (Sept.) 1949.
9. Peterman, A.F., Daly, D.D., Diou, F.R., and Keith, H.M. Neurology 9, 533-539. (1959).
10. Roseman, Ephraim, and Aring, C.D. Medicine 20, 463-494. (Dec.) 1941.
11. Singh, Amarjit, and Jolly, S.S. Indian J.M. Sc. 12, 347-350. (May) 1958.
12. Taylor, E.W., and McDonald, C.A. Arch. Neurol. & Psychiat. 27, 79-112. (Jan.) 1932.
13. Wiederholt, W.C., Mulder, D.W., Lambert, E.H. Mayo Clin. Proc. 39, 427-51. (June) 1964.