THE TURKISH JOURNAL OF
PEDIATRICS
A HACETTEPE BULLETIN VOLUME 13 / NUMBER 4 / OCTOBER 1971

Syndrome of Acute Idiopathic Ophthalmoplegia with Ataxia and Areflexia in Childhood (Miller Fisher's Syndrome)

Kalbiye Yalaz, M.D.* / Kaynak Selekler, M.D.**

In 1956 Fisher described a syndrome characterized by opthalmoplegia with ataxia and areflexia.[1] Since that time forty similar cases, of which only six occurred during childhood, have been reported as Fisher's syndrome.[2-9]

Guillain recorded a form of Guillain Barré syndrome limited to the cranial nerves, "la forme mesencephalique Pure".[10] A relationship between Guillain-Barre's syndrome and Fisher's syndrome has been generally accepted.[9] Almost the only difference between these two syndromes is the rarity of the latter. We present the first case of Fisher syndrome from Turkey with a review of the literature.

Case Report

M. D. (Prot. 255491), this eight-year-old boy was admitted to the Hacettepe Medical Center for the evaluation of headache, ptosis, micropsia and ataxia. He had a history of rhinorhea, fever and cough which started 10 days prior to admission. He had double vision, ataxia and ptosis of the left eyelid 12 hours following a head trauma which occurred one week before admission. He had micropsia beginning two days before examination.

Past history and family history were non-contributory.

* Associate Professor of Pediatrics and Pediatric Neurologist, Hacettepe University Ankara, Turkey.
** Resident in Neurology in the same University.

Physical examination: Height 122 cm., weight 22 kg., head circumference 53 cm., temperature 36.5°C., pulse 80/min., respiration 20/min. Blood pressure 100/65 mm, Hg. postnasal discharge, hypertrophy of the tonsils and pain over the maxillary region were the only positive findings during the physical examination.

Neurological examination: He was an intelligent boy, answering questions properly. He had complete bilateral internal and external ophthalmoplegia. The pulpis were equal and reacted to light. Eye movements were totally absent in all directions. He had bilateral ptosis (Figure 1). Eye grounds were normal. Vision was described as normal. There was also left peripheral facial weakness. The rest of the cranial nerves were intact. Muscular weakness of the extremities or sensory deficit were not present. Deep tendon reflexes were absent. Superficial abdominal reflexes were normal. Planter responses were flexor. The gait was unsteady, dysmetria was evident on finger to nose and knee to heel tests. Light touch, pinprick, temperature and vibration perception were normal. Bowel and urinary control were also normal.

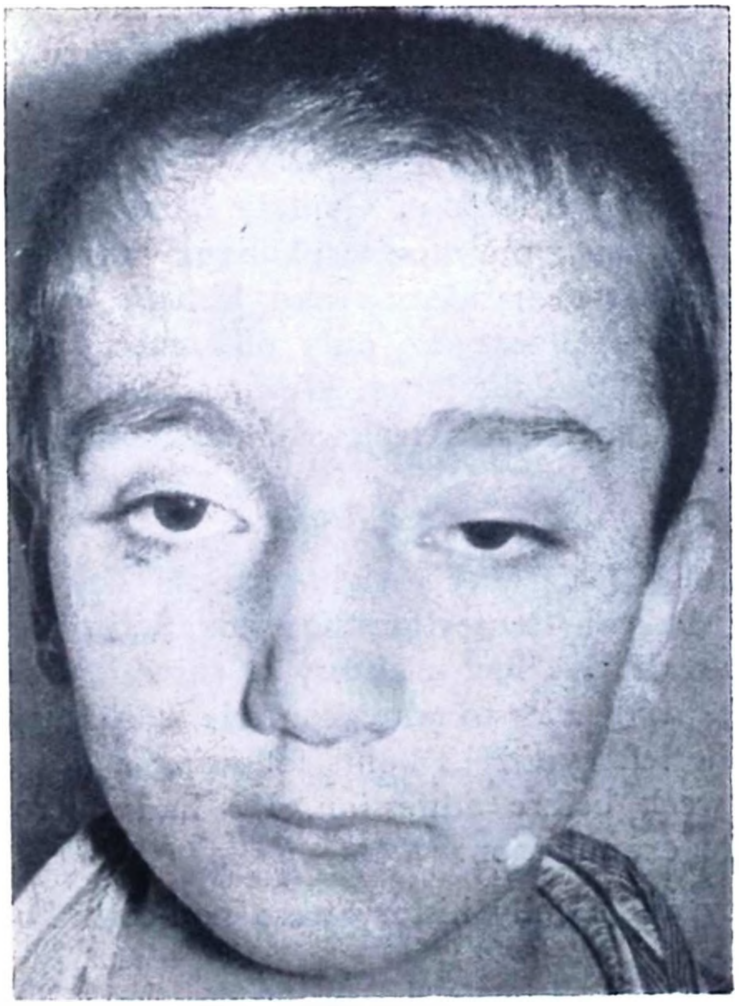

Figure 1

Patient had complete bilateral and external ophthalmoplegia and bilateral ptosis.

Laboratory Findings: Blood examination revealed the following information Hb: 15 gr%. WBC 11.800 mm.^{3} Sedimentation 6 mm/hr NPN 78 mg%. Serum electrolytes were normal. Total protein 6.7 mgr%; alb 4.5 mg%, glo 2.2 mg%, serum electrophoresis, alb. 54.6 %, α_1 1.85, α_2 20%, 10%, γ 26.6%. Immunoelectrophoresis, IgG increased, IgM increased, IgA normal. Thalluim was negative in the urine. Lumbar pucture pressure was 150 mm. H_2O and the fluid was clear. Cerebrospinal fluid protein was 93 mg. %, sugar, 32 mg. %; blood sugar 110 mg. % and lactic dehydrogenase was 10 units. Cerebrospinal fluid culture was negative and gastic washings for AFB were negative on three occasions.

X Ray Findings: Skull and chest X rays were negative. Bilateral maxillary sinusitis was apparent. Electroencephalogram was normal. Cerebral angiography and brain scannings were also normal.

Clinical Course: Tensilon (edrophonium cloride) test to exclude myasthenia gravis was performed with negative result. One week after

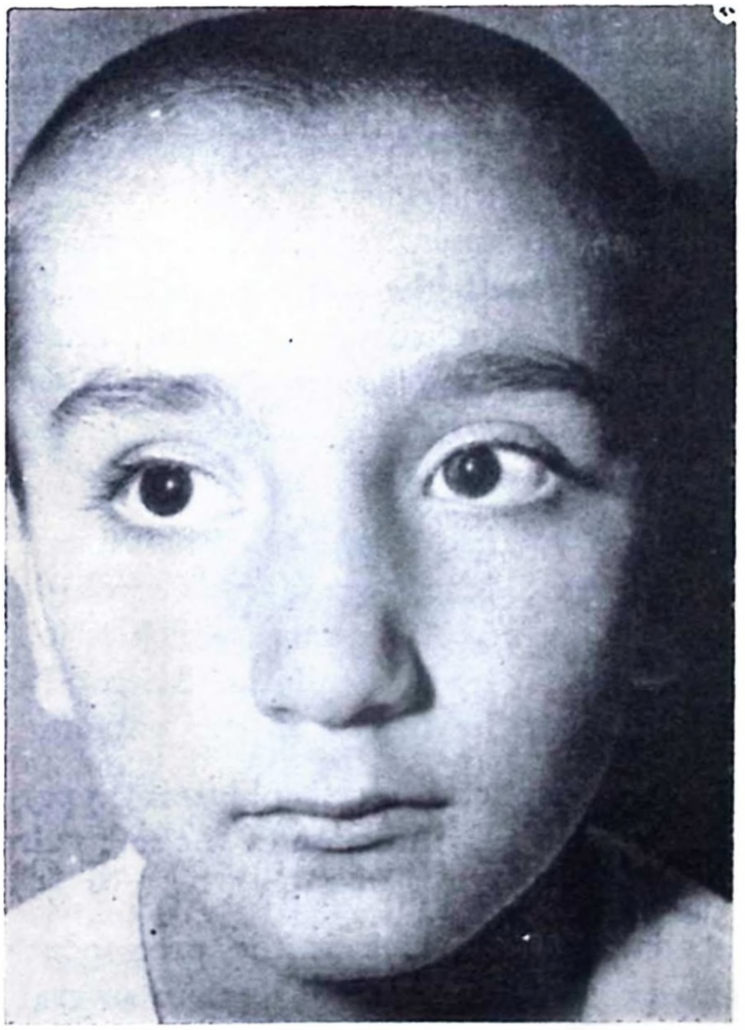

Figure 2

Patient recovered from external ophthalmoplegia and bilateral ptosis after three weeks.

admission left peripheric type of facial palsy was observed but disappeared during the next few days.

The patient recovered from the rest of the neurological symptoms in the next three weeks. Repeated LP revealed protein 147 mg% and 88 mg% with a week interval. However areflexia persisted for six months (Figure 2).

Discussion

The history of upper respiratory system infection, benign clinical course, increased protein level in the cerebro-spinal fluid are similar features in the Fisher syndrome and infectious polyneuritis. The etiology and underlying mechanisms are unknown. Various kind of infections[8-11] intoxication,[12] vaccination,[13] serum sickness, collagen disease, dysproteinemia leukemia, reticulosis, visceral carcinomaitosis,[4 5 11] are responsible in some cases for infectious polyneuritis. Similarity of the clinical and pathological pictures between infectious polyneuritis and experimental allergic neuritis in rabbits has suggested an autoimmune etiology.[14] According to recent studies idiopathic infectious polyneuritis is a cell-mediated immunologic disorder in which the peripheral nervous tissue, particularly myelin, is attacked by specifically sensitized lymphocytes.[15]

Association of upper respiratory infections maculo-papular skin lesions and gastroenteritis with the Fisher syndrome suggests reaction of the control nervous system to a viral agent. Short term stuporus has been reported.[1 7 9] Absence of deep tendon reflexes without evidence of muscular weakness is an important neurological finding. Babinski and clonus are negative.

The cranial nerves, specifically the third and fourth and sometimes the sixth may be the sites of the most severe neurological involvement. Sometimes diplopia is the initial symptom which is followed by ptosis, strabismus, partial or total opthalmoplegia. Micropsia was attributed to the deficit of accomodating muscle in our patient.[16 17]

Partial or total peripheral facial palsy is apparent during the course of illness frequently. Dysfunction of the soft palate and swallowing have not been recorded.

Acute cerebellar ataxia which is the prominent finding of the syndrome persists for periods of several days to several months. Other cerebellar dysfunctions such as nystagmus and dysrhythmia are less frequent.

Fisher's syndrome clinically resembles myasthenia gravis, Wernicke's encephalopathy, brain stem glioma, polyneuropathy due to diphteria, botusism, brain stem vascular insufficiency, cavernous sinus thrombosis, collagen disease porphyria, diabetes mellitus, sarcoidosis, neuropathy due to malignancy, brain stem encephalitis. History of upper respiratory system infection approximately two weeks prior to initial symptoms, generally increased cerebro-spinal fluid protein levels are the most important findings. Increase in cerebrospinal fluid proteins from 93 mg % to 147 mg. % with a week interval without cell reaction characteristic findings for the presented case. Increased IgG and IgM in immunoelectrophoresis probably implies and immunopathologic process of viral etiology.

A benign course, with rapid, fairly complete recovery within weeks to months after onset are the features of Fisher's syndrome. Histologically perivascular lymphatic infiltration around nuclei of the third, fourth, sixth nerves associated with nerve fiber breakdown the fourth cranial nerve has been recorded in a case of idiopathic polyradiculoneuropathy.[15] Swollen third and fourth cranical nerve nuclei, with chromatologysis of Nissl granules and eccentric displacement of the nucleus were histologic findings in another case with acute opthalmoplegia.[18] In the ataxic form of polyradiculoneuritis pathologic alterations in the cerebellum, pons and medulla have bot been found but degeneration of the fiber system of the column of Clarke was recorded.[19]

The benign course of the disease probably is an important indication against using steroids.

Summary

An eight-years-old boy with acute opthalmoplegia, ataxia and areflexia with rapid and fairly complete recovery within weeks after onset is described with a review of the literature.

REFERENCES

1. Fisher, M.: An unusual variant of acute idiopathic polyneuritis (syndrome of ophthalmoplegia, ataxia, and areflexia), New Eng. J. Med. 255: 57, 1956.
2. Charous, D. I.: The Landry-Guillain-Barré syndrome (Report of an unusual case, with a comment on Bell's palsy), New Eng. J. Med. 267: 1334, 1962.
3. Idris, J.: Facial Diplegia in the Guillain-Barré Syndrome, Brit. Med. J. 2: 84, 1964.
4. Munsat, T. L., Barnes, J. E.: Relation of multiple cranial nerve disfunction to the Guillain-Barré Syndrome, J. Neurol. Neurosurg. Psychiat. 28: 115, 1965.
5. Ford, F.: Disease of the Nervous System in Infancy, Childhood, and Adolescence, 5th ed., Springfield III., Charles C Thomas Co., 1066, pp. 406-407.

6. Patel, A., Pearce, L., Hairston, R.: Miller-Fisher syndrome (variant of Landry-Guillain-Barré-Strohl syndrome-ophthalmoplegia, ataxia, areflexia), Southern Med. J. 59: 171, 1966.
7. Bell, W., Van Allen, M., Blackman, J.: Fisher syndrome in childhood, Develop. Med. Child. Neurol. 12: 758, 1970.
8. Qaqundah, B. Y., Taylor, W. F.: Miller Fisher syndrome in a 22 month old child, J. Pediat. 77: 868, 1970.
9. Elizan, T. S., Spire, J. P., Andiman, M. R., Baughman, F. A., Lloyd-Smith, D. L.: Syndrome of acute idiopathic ophthalmoplegia with ataxia and areflexia, Neurology 21: 281, 1971.
10. Guillain , G.: Les Polyradiculonevrites avec dissociation albuminocytologique et a evalution favorable (Synrdome de Guillain Barré), J. Belg. Neurol. Psychiat. 38: 323, 1938.
11. Markland, L. D., Rilay, H. D.: The Guillain-Barré Syndrome in Childhood: A comprehensive review, including observations on 19 additional case, Clinical Pediatrics 6: 162, 1967.
12. Neubert, F. R.: Complete ophthalmoplegia in acute toxic polyneuritis, Brit. J. Ophthal. 42: 632, 1958.
13. Kisch, A. L.: Guillain-Barré syndrome following smallpox vaccination: Report of a case, New Eng. J. Med. 258: 83, 1958.
14. Waksman, B. H., Adams, R. D.: Allergic neuritis: An experimental disease of rabbits induced by the injection of peripheral nervous tissue and adjuvants, J. Exp. Med. 102: 213, 1955.
15. Asbury, A., Arnason, B. G., Adams, R. D.: The inflammatory lesion in idiopathic polyneuritis: Its role in pathogenesis, Medicine 48(3): 173, 1969.
16. Willanger, R., Klee, A.: Metamorphosia and other visual disturbances with latency occurring in patients with diffuse cerebral lesions, Acta Neurol. Scandinav. 42: 1, 1966.
17. Ehlers, N.: A case horizontal micropsia and ocular instability, Acta Ophth. 46: 75, 1968.
18. Bignami, A., Servi, M.: Acute internal and external ophthalmoplegia with muscle weakness, Lancet 1: 110, 1963.
19. Richter, R. B.: The ataxic form of polyrediculoneuritis (Landry-Guillain-Barré syndrome), J. Neuropath. Exp. Neurol. 21(2): 171, 1962.
20. Eiban, R. M., Gersony, W. M.: Recognition, prognosis, and treatment of the Guillain-Barré syndrome (acute idiopathic polyneuritis), Med. Clin. N. Amer. 47(5): 1371, 1963.