TWO CASES OF CHRONIC POLYNEURITIS IN CHILDREN

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Polyneuritis has been regarded as an uncommon disease in childhood. Formerly many cases have been due to diphtheria or lead poisoning but these conditions are now infrequent. In the past 25 years there have been numerous reports of the Guillain Barré syndrome, or acute infective polyneuritis affecting children. Hecht (1937) described seven personal cases and by 1941 Casamajor and Alpert had been able to find 38 recorded examples in children below the age of 12. Scheid (1946) reviewed the literature and concluded that this type of polyneuritis could not be regarded as rare during infancy and childhood.

Chronic and recurrent forms, however, appear to be very unusual. Thomson (1910) described a youth of 19 who developed an extensive flaccid paralysis a few days after he had fallen out of a sculling boat. The limbs, trunk and face were affected. Sensory symptoms were very slight. Recovery was slow but was complete after two years. At the age of 5 he had had a similar illness with recovery after 18 months.

Batten (1913) described recurring polyneuritis in a boy aged 8. The first episode lasted four months and chiefly involved the legs. Sensation was very slightly, if at all, affected. A second attack occurred four months later and the boy again made a good recovery. Batten also mentioned a girl aged 3 in whom the illness ran a relapsing course and ended fatally after 14 months. Details of a similar case have been given by Collier (1932). His patient was a girl aged 14 who died from bulbar paralysis five months after the onset of the illness.

Rabinowitz (1914) reported the case of a girl who had attacks at the ages of 9 and 12. On each occasion symptoms lasted some 12 months and there was considerable pain in the limbs. Nattrass (1921) published the case of a boy who had three attacks when aged 4, 17 and 18. There were sensory symptoms but no signs of impaired sensation.

Where palpable, nerve trunks were abnormally thick and hard. Brain (1933) mentions a boy aged 15 seen in his fourth attack, the first having occurred at the age of 4.

Case Reports

Case 1. A girl aged 13, the youngest child in a family of five, was admitted to the Royal Salop Infirmary in December, 1949. For one month she had noticed gradually increasing weakness of the arms and legs. She had experienced no pain and there had been no preceding febrile episode.

Examination revealed a symmetrical flaccid paralysis of the limbs, most pronounced peripherally. Tendon reflexes in the legs were absent and those in the arms much reduced. The superficial abdominal reflexes were present. There was slight muscle wasting but no tenderness. No impairment of any form of sensation could be demonstrated.

The cerebrospinal fluid contained 60 mg. protein per 100 ml. with a slight excess of globulin (Pandy test). A cell count gave 1 lymphocyte per c.mm. The blood count and sedimentation rate were normal, with no basophil stippling of the red blood cells. The urine was normal: no lead or porphyrins were detected. The Schick Test was negative. C. diphtheriae was not isolated from nasal and throat swabs.

The paralysis increased and spread to involve the trunk and the muscles of the neck, face and tongue. All tendon jerks were lost. Within two months of her admission to hospital she was completely helpless. No movement was possible in the limbs nor could she raise her head from the pillow. Muscle wasting became extreme, and the muscles showed loss of contraction to faradic current and gave only a very feeble response to galvanism.

There was great weakness of the facial muscles and masseters; the tongue was grossly wasted and showed pronounced fibrillation. As the illness progressed dysphagia became increasingly severe, phonation became very weak and she had difficulty in coughing and in clearing her throat of secretions. Eventually tube feeding had to be employed, and suction used to prevent
Two cases of chronic polyneuritis in children

Developed, muscles respirator was sensation. Of periods electrocardiogram barrassment from the experience prostigmin remains months involved. However she a no week Birmingham of history months was found was most absent could demonstrated. The muscles, nerves were complete paralysis of the limbs, and the trunk, facial muscles, masseters and tongue were involved. However she did not on that occasion experience dysphagia or choking attacks. There were no sensory symptoms nor signs of sensory loss. Improvement began after a year, and she was able to walk some 18 months after symptoms had first been noticed. She eventually returned to school, played games, and rode a bicycle. Recovery had appeared complete before her second attack began in November, 1949.

Case 2. A girl aged 8, an only child, was well until February, 1951, when she was noticed to be using both hands when turning door knobs, and to have difficulty in buttoning her clothes and in tying her shoelaces. A week later she began to walk clumsily and became unable to mount stairs. She had no pain and there was no history of any recent illness. She was admitted to the Birmingham Children’s Hospital on April 26, two months after symptoms had first been observed. There was found to be symmetrical weakness of all the limbs most pronounced at the periphery, with moderate muscle wasting. The muscles were not tender. Power in the trunk muscles was normal. All tendon reflexes were absent but the superficial abdominal reflexes were present. No definite defect of any form of sensation could be demonstrated. The cranial nerves were normal.

The blood count was normal, with no basophil stippling of red blood cells. The sedimentation rate estimated by a micro method was 3 mm. in one hour. In the cerebrospinal fluid there was 1 lymphocyte per c.mm. and 80 mg. protein per 100 ml. Throat and nose swabs were negative for C. diphtheriae. The Schick Test was negative. There were no lead or porphyrins in the urine. Radiographs of the long bones showed no evidence of lead deposition. The blood pyruvate level was 1 mg./100 ml. An electrocardiogram was normal.

She was treated with aneurne and given light massage and passive movements. She improved slightly and the cerebrospinal fluid on May 8, 1951, contained less protein (50 mg. per 100 ml.). She was allowed to go home on June 16, but three weeks later was re-admitted as her condition had considerably worsened. All power in the hands and feet was now lost and there was complete bilateral wrist and foot drop. A little power remained in the shoulders and hips. Some weakness of the trunk musculature had developed. There was considerable muscle wasting in the limbs. Electrical stimulation of the affected muscles gave a normal reaction to galvanism but only a weak response to faradism. Sphincter control remained normal. The C.S.F. was examined again on September 6, when the protein content had risen to 120 mg. per 100 ml. Improvement was slow but 14 months from the beginning of her illness she could use a spoon and fork, raise her arms above her head to brush her hair, and was beginning to walk with the aid of leg supports.

Discussion

In neither of these cases could the cause of the illness be found. In both a purely motor type of polyneuritis developed gradually and ran a prolonged course. There was no history of any preceding febrile disturbance, and no sensory symptoms. The cerebrospinal fluid contained an excess of protein, the cell count remaining normal. The protein increase was more pronounced in Case 2, rising to 120 mg. per 100 ml., and this case might be regarded as an instance of the Guillain Barré syndrome, or acute infective polyneuritis. Against this view are the absence of sensory symptoms, the pronounced muscle wasting and the long duration of the illness. The two cases resemble each other closely. They differ only in the severity and extent of the paralysis and seem to be examples of the same condition. The clinical features suggest that the cause may have been some slowly acting toxin rather than an infection.

Recurrence after two years took place in Case 1. Most examples of recurrent polyneuritis of unknown cause have occurred during adult life and the symptomatology has been very varied. The literature has been reviewed by Ungle (1933) and by Stucke (1947). Ford (1944) considers that this is a heterogeneous group of conditions and that no single explanation can be applied to all cases.

Collier (1932) referred to several cases in which second or third attacks of peripheral neuritis had occurred after intervals of months or years, and no causal agent had been found. In each the organism of diphtheria was eventually recovered from one or other of the paranasal sinuses, and he considered that diphtherial infection of the chronic carrier order was the most common cause of recurring neuritis. In neither of the present cases was the Klebs-Loeffler bacillus found. There was no accommodation palsy, and both the long duration and the severity of the paralysis appear to exclude a diphtheritic origin.

Harris (1922) described a case which is strikingly similar to Case 1. His patient, a girl of 15,
developed an extensive flaccid paralysis with involvement of the face and tongue. There was no sensory loss and the only sensory symptoms were slight paraesthesiae of the finger tips early in the illness. Recovery did not begin for 18 months but was complete two years later. She was well at the age of 31 and had not experienced any recurrence. Harris (1935) referred again to this case in a paper in which he reviewed the chronic forms of polyneuritis. He considered that there was a group showing common features of slowly progressive motor paralysis with muscle wasting, changes in the electrical reaction of the muscles, and few sensory changes. Both recurrence after a short period of complete recovery and neural hypertrophy were features which might or might not be present. He suggested that the cause might be an endotoxin, and that such cases might be classed under the term chronic progressive (endotoxic) polyneuritis.

The prognosis of polyneuritis in childhood is considered to be good. Death is rare and when it has occurred has generally done so early in the course of the disease as a result of a rapidly spreading paralysis of the Landry type. It is usual for recovery to be rapid and complete, but the two cases I have reported are a reminder that chronic and recurrent forms may occur in children.

Summary

Two cases of chronic polyneuritis are described. The cause was not determined. In one recurrence took place after two years. The literature is reviewed.

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References


