RECURRENT HYPERTROPHIC NEURITIS.

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I HAVE recently observed, in a boy 18 years old, a clinical syndrome consisting of extensive flaccid paralysis of muscles with loss of reflexes and with electrical changes; sensory phenomena, mainly subjective; inco-ordination of movements; and hypertrophy and hardening of the nerve-trunks. Almost complete recovery has occurred, and there is evidence of two previous attacks of the same nature.

Clinical Features.—The patient is an apprentice joiner. He was admitted to the Royal Victoria Infirmary, Newcastle-upon-Tyne, on Nov. 5, 1920, under Dr. Beattie. He stated that a fortnight before admission he began to feel very tired and weak in the legs; the power in his legs and arms gradually diminished, and within forty-eight to seventy-two hours he had completely lost the use of his legs and could only move his arms a little. During this time he had a lot of pain in all his limbs and in his back, and after the onset of the symptoms he was unable to sit up or make any attempt to feed himself, and indeed was scarcely able to move at all. He retained complete control of the bladder and bowels, and had no difficulty in swallowing, no squint, and no other symptoms.

On admission the temperature was 98° and the pulse 80, and throughout the illness both remained normal. The patient was small for his age, and pale and of poor physique; but his sexual development was normal, and his intelligence equal to the average. His speech at first was distinctly blurred and slow, as though articulation was difficult. The tongue was furred, and the patient appeared acutely ill. The pupils were equal and reacted normally to light and accommodation, all eye movements were normal, and there was no ptosis, strabismus, nystagmus, or exophthalmos. The optic discs appeared normal. Examination of the remaining cranial nerves revealed no abnormality.

Motor.—Nearly all the muscles of the upper and lower limbs were wasted and showed partial or complete loss of voluntary power. In addition there was great weakness of the trunk muscles, and the patient was unable to sit up or raise the head from the pillow. In the upper limbs there was weak voluntary power in all muscles, but this diminished towards the periphery of the limbs, and the interossei, lumbricales, and the muscles of the thenar and hypthenar eminences

were much wasted and retained only very weak voluntary power. There was a coarse tremor of the upper limbs on attempting voluntary movement, and the rebound phenomenon and adiadochokinesis were marked. The tendon reflexes of the upper limbs were absent. In the upper arms all the muscles responded fairly well to faradism and quickly to galvanism; in the forearms all responded to faradism except the flexor sublimis and the flexor profundus digitorum, and the response to galvanism was fairly sharp; in the hand muscles there was no response to faradism, and the response to galvanism was slow (complete R.D.).

The abdominal reflexes were present and equal.

In the lower limbs the loss of voluntary power was complete, except for slight power of flexion of hip and knee. There was complete double foot-drop and no voluntary movement of the toes. The knee- and ankle-jerks were absent. There was a doubtful response to faradism in both quadriceps, but no response to faradism in the muscles below the knees. The anterior tibial group responded slowly to galvanism, and the peronei and calf muscles did not respond to either current. There was marked hyposensibility to the electric current, and strong currents were in all cases necessary to evoke a response.

Sensation.—The muscles all over the body, including those of the neck and face, were markedly tender to palpation. There was no objective sensory loss to pain, touch, heat, or cold in any part, but appreciation of the vibrations of the tuning-fork appeared to be defective. The patient complained of numbness and tingling in the periphery of all the limbs.

Nerve-trunks.—On palpation there was found to be very definite enlargement and apparent hardness of such nerve-trunks as could be felt, viz., the median, ulnar, and external popliteal on both sides. The enlargement was uniform and not nodular, and as far as could be detected the nerves were about one and a half times to twice their normal diameter. In addition, the trunks were distinctly tender on pressure. No enlargement of cutaneous nerves was observed, but this was not specially looked for. The Wassermann reaction of the blood was negative, blood-films showed no abnormality, and the cerebrospinal fluid obtained by lumbar puncture showed no abnormality on microscopical examination or culture.

Previous History.—Exactly a year previously (on Nov. 7, 1919) the patient was admitted to the Royal Victoria Infirmary with symptoms closely resembling the present attack. His history at that time was that he had been quite well till a month before admission, when he returned from work one evening complaining of ‘pins and needles’ in the feet. Later the same evening the pains extended

up the legs. He stayed in bed the next day, but got up on the following day; he was able to walk, but the pains recurred, and he returned to bed, and had remained there till admission. The pains spread to his back and to the upper limbs, and he gradually lost the power of all his limbs, and within a week was unable to move his legs at all. The sphincters were never affected.

The notes of his condition on admission state that the intelligence and memory were good and the speech normal; he slept well, but was not drowsy or apathetic. There was almost complete paralysis of both lower limbs, with wasting, flaccidity, and hypotonicity of the muscles. The knee-jerks, ankle-jerks, and plantar reflexes were absent. In the upper limbs there was a lesser degree of flaccid paralysis, more marked on the right side than on the left. The right supinator-jerk was absent, and the left less active than normal. The epigastric, abdominal, and cremasteric reflexes were present, and the sphincters were normal. There was no objective loss of sensibility to touch, pain, heat, or cold.

The patient remained in hospital nearly three months (until Jan. 29, 1920). He had gradually improved, and on discharge was able to walk and the paresis of the arms had disappeared. The diagnosis made was poliomyelitis. He continued to improve, and three months later resumed his ordinary work, and remained in full employment for six months, during which time he states he was in complete health, and had no pains and no weakness of any part of his limbs. At the end of this period the symptoms of the relapse appeared, leading to his re-admission.

The mother states that when the patient was 4 years of age he had a very similar illness, in which he gradually lost the power of his legs, which remained completely paralyzed for a month and then slowly recovered; the total duration of this attack before complete recovery was about six months. He had measles as a child, and an operation for removal of tonsils at the age of 5 years; otherwise he had been in good health, and the recovery from the attack of paralysis had been perfect.

Family History.—Father and mother healthy; three sisters, age 21, 19, and 15 years; one brother, age 13 years. All these healthy. Careful inquiry failed to elicit any history suggesting the same symptoms as those of the patient in his own or in previous generations.

Progress.—Evidence of gradual improvement appeared within a few days of admission, and was first noticeable in the arms. At the same time the general condition improved and the tongue became clean. Power of voluntary movement slowly appeared in the legs, and progressed steadily. The patient was kept in hospital nearly

three months (until Jan. 26, 1921), at the end of which time the upper limbs appeared normal and the lower limbs had approached complete recovery. The muscles had recovered the greater part of their normal bulk and tone, and on the left side voluntary power was present in all muscles, though it was considerably weaker than normal; on the right side there was still marked weakness of the anterior tibial muscles and peronei, but with the aid of a light toe-uplifting spring the patient could walk quite well in ordinary boots. There was no ataxia in Romberg's attitude. Since discharge the improvement has continued and the toe-uplifting spring has been abandoned. Recovery now appears to be almost perfect, and, further, the size and consistence of the nerve-trunks have become nearly normal, though in the opinion of most who have seen the case recently these trunks are still abnormally large; at any rate the greater part of the enlargement has disappeared. The knee- and ankle-jerks are still absent, and the electrical reactions are returning to the normal, the quadriceps and anterior tibial groups responding to faradism; faradic response in the calf muscles remains doubtful, as pain is produced by the current required.

The treatment adopted was the prevention of stretching of the paralyzed muscles by suitable orthopædic appliances, and gentle massage and galvanism.

Diagnosis.—The diagnosis of acute poliomyelitis made during the second attack was not unnatural, but in view of the subsequent course of the case must be abandoned. It is inconceivable that there could occur three attacks of acute poliomyelitis of such severity and extent as to produce almost general paralysis, in which on each occasion there was no residual paralysis.

All the appearances pointed to a multiple peripheral neuritis, but concerning the etiology of this there is no evidence. The two outstanding peculiarities of the case are: (1) The periodicity; (2) The hypertrophy of the nerve-trunks.

1. I have been unable to find any record of cases resembling the present case in the occurrence of prolonged attacks of extensive flaccid paralysis with loss of reflexes, pains, etc., followed by complete functional recovery. In family periodic paralysis the attacks are similar in many ways, but their total duration is only a few hours or days.

2. From time to time cases have been recorded of muscular paralysis associated with enlargement of peripheral nerves. In 1893 Dejerine and Sottas¹ described two cases under the name of 'progressive interstitial hypertrophic neuritis of infants', together with the pathological findings in one of these cases. The patients were brothers; the first symptoms were noted in infancy in one case, and

at the age of 14 years in the other, and the main features were as follows :—

a. Slowly progressive muscular atrophy, with fibrillary contractions, loss of deep reflexes, and partial R.D., earliest and most marked in the lower limbs, producing double talipes equinovarus, but also present in the upper limbs, giving rise to atrophy of the intrinsic muscles of the hands of the Aran-Duchenne type.

b. Lightning pains and marked sensory changes in the periphery of both lower and upper limbs, viz., loss of tactile sense, delay of pain sense, diminution of heat and cold senses, great alteration of muscle sense.

c. Inco-ordination of all limbs, with gait of tabetic type, and ataxia in Romberg's attitude.

d. Myosis, with Argyll Robertson pupils.

e. Absence of sphincter trouble.

f. Severe kyphoscoliosis.

g. Hypertrophy and hardening of all the nerves of the limbs accessible to palpation, some of the nerves appearing to be double their ordinary diameter.

h. Pathologically, an interstitial hypertrophic neuritis, most marked in the periphery of the nerves, but also evident in the nerve-trunks and posterior nerve-roots, the change consisting in a gradual replacement of the nerve elements by proliferating fibrous tissue, the myelin sheaths being affected early and the axis cylinders late. In addition, consecutive sclerosis of the columns of Goll and Burdach.

i. A very gradual onset and progressive course ; the case which was examined pathologically died at the age of 45, the disease having been in existence since infancy. The cases resembled the peroneal muscular atrophy of Charcot-Marie-Tooth, but were distinguished by the marked sensory changes and the enlargement of the nerves.

Pierre Marie² in 1906 showed to the Paris Neurological Society two cases belonging to a family of seven, all afflicted with the same disease. These cases resembled in many respects those described by Dejerine and already referred to. They showed flaccid paralysis of muscles, with atrophy, loss of tendon reflexes, and partial to complete R.D. ; talipes equinovarus and kyphoscoliosis ; marked diminution of cutaneous sensibility ; and palpable thickening of nerve-trunks and cutaneous nerves. They presented, however, certain well-marked differences. The affection was almost limited to the lower limbs, though there was definitely slight involvement of the upper as well. There were no lightning pains ; no Argyll Robertson pupil, but only diminished reaction to light ; and no Romberg's sign.

In place of the ataxia described by Dejerine, there was intention tremor closely resembling that of insular sclerosis; in addition there was an affection of speech recalling this latter disease; and finally there was a slight degree of exophthalmos.

In the discussion which followed, Dejerine maintained that, in spite of the differences noted, these cases belonged to his group of progressive interstitial hypertrophic neuritis.

In 1912 Schaller³ described a case in a man of 20, which had begun at the age of 13 years and slowly progressed. This patient showed flaccid paralysis of muscles of the upper and lower limbs, with partial R.D. and slow response to galvanism; diminished cutaneous sensibility and considerable pallanæsthesia; inco-ordination of hands and feet, most marked with the eyes closed; some adiadochokinesis of both hands, but no asynergia or 'mouvements démesurés'; the cranial nerves showed nothing abnormal. Associated with these signs there was hypertrophy of the nerve-trunks in the axillæ and upper arm, of the external popliteals, the superficial cervical nerves, and the cutaneous nerves of the forearms; the ulnar nerve at the elbow was about the size of a large goose-quill. A portion of a superficial cervical nerve was excised and sectioned, and the microscopical appearances were those of an interstitial hypertrophic neuritis similar to that described by Dejerine. There was no family history of the disease in this case, and considerable improvement followed the administration of arsenic by the mouth and galvanism to the muscles.

Dide and Courjon⁴ detailed later a further case in a man commencing at the age of 40 and without family history. The wasting affected the upper limbs only, and was at first unilateral. There was no objective sensory loss, but there were shooting pains, and pressure on the enlarged nerve-trunks aroused pain. There was intention tremor, but no dysmetria or adiadochokinesis.

In a further contribution⁵ these authors described five more adult cases of similar type.

From a study of these cases it will be seen that there are many points of close similarity between them and the case which I have described. The flaccid paralysis, with wasting, loss of reflexes, and electrical changes; some degree of sensory change; inco-ordination or intention tremor of the limbs; and the hypertrophy of the nerves, are common to all. The outstanding difference between my case and all the others is the apparently complete recovery and equally complete relapse on two occasions.

It is unnecessary to discuss the differences between this case and other conditions which are characterized by muscular paralysis with wasting. The hypertrophy of the nerves alone distinguishes it from the peroneal atrophy of Charcot-Marie-Tooth, from the muscular

dystrophies, myelitis, syringomyelia, or any of the recognized forms of peripheral neuritis. In von Recklinghausen's disease there are thickenings on the nerves, but of a nodular type quite different from an interstitial neuritis. In addition, all these conditions except peripheral neuritis are chronic and progressive.

The view of the case which I put forward is that it is a modified and recurrent type of hypertrophic interstitial neuritis as originally described by Dejerine and Sottas.

The late involvement of the axis cylinders, as described by these authors, suggests the possibility of recovery in an earlier stage by regeneration of the myelin sheaths.

The case is published in the hope of eliciting comment and further information.

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