the lower edge of the incision because it is apt to retract beneath the bladder, and the unwary might sew the upper edge of the incision to the inside of the uterus instead of to the lower edge of the incision. A continuous interlocking suture is used to oversew the muscular layer which has already been approximated by the mattress sutures. The incision in the peritoneum covering the lower segment is closed with a continuous inverting suture. In a case potentially infected a drainage tube may be used for a few hours in the peritoneal cavity. The two sides of the incision in the parietal peritoneum are brought face to face, as advocated by Blair-Bell in order to minimize the risk of bowel or omentum becoming adherent to the suture line.

The rest of the incision is closed in the usual way, finishing with a subcuticular suture or Michel's clips. In a fat subject it may be advisable to employ deep silkworm-gut sutures, but they may carry infection from the skin to the deeper layers of the wound, and they tend to spoil the appearance of the scar in the abdominal wall by leaving permanent marks, in spite of protection by fine rubber tubing.

Convalescence

Unless it is required to sterilize the patient the uterus is not brought outside the abdominal wall, for when the uterus is not eventrated there is less flatulent distension after operation. If the stomach is seen at operation to be distended with gas a stomach tube may be passed before the patient leaves the operating theatre. I have been using the operation for some years, and am of opinion that it is not only a better operation to perform than the "classical" procedure, from the point of view of craftsmanship, but it is followed by an easier convalescence.

BIBLIOGRAPHY

- Bonney, V.: *Lancet*, 1933, i, 796.
Bailey, K. V.: *Ibid.*, 1934, i, 672.

PROGRESSIVE FAMILIAL HYPERTROPHIC NEURITIS (DEJERINE-SOTTAS)

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In 1893 there was recorded by Déjerine and Sottas a distinct entity in the group of familial disorders to which the name "hypertrophic neuritis" was given.¹ Cases were also described by Marie (1906), Boveri (1910), Yokomari (1915), and others, but in 1929 de Brugn and Stern² could find only twenty cases recorded in the literature. In the following paper at least four more cases are added, probably the first to be reported from Australia.

Family History

Briefly stated, the family consists of a woman aged 55 years and three of her sons, aged 28, 24, and 21 years, respectively. The notes of these cases are given in full. In the previous generation the mother of the female patient described here had wasting of the peripheral portions of all four limbs, and one other member of the family, at present in Queensland, is said to have the same disability, which is known as a "family weakness": neither of these two has been examined by me.

The members of the family so far examined have marked wasting and loss of power in the lower limbs extending symmetrically as high as the junction of the lower and middle thirds of the thighs. The electrical reactions are those of "reaction of degeneration": fibrillary tremors are present in the muscles, and a secondary foot deformity of the "pes cavus" type occurs in each case. In the upper limbs there is wasting and weakness of the hands and forearms with the develop-

ment of a claw-hand. The muscles of the arms, shoulders, trunk, neck, and upper two-thirds of the thighs show no change. There is loss of sensation to pin-prick and cotton-wool of glove and stocking type extending as high as the knees and elbows. The peripheral nerves are thicker than normal, especially the ulnar, and the trunks of the brachial plexus are also enlarged.

While the above signs are uniform throughout this family group some members have other abnormal neurological signs. Nystagmus, abnormal pupil reactions, and defects in speech are present in some, while others have loss of sensation to heat and cold and trophic changes in the hands.

Case I

The patient, a widow aged 55 years, has had weakness and wasting of her hands for over twenty years; even before this time she had noticed her hands to be "clumsy," particularly in cold weather. For about the same period she has had some difficulty in walking; the legs are weak, and she is liable to fall over if walking upon irregular ground. For at least thirty years her feet have been deformed. Occasional "numbness" in the left leg has been the only other symptom noted. She has always been in good general health, has brought up a family of five children, and still cooks and does all the work of a large house.

She is a rather thin, short woman of nervous type. The hands are small and the grip is weak, the muscles of the thenar and hypothenar eminences being almost completely atrophied and the forearms wasted to a marked degree. The muscular development of the arms and shoulders is normal. The supinator jerks are absent, but the biceps and triceps jerks are overactive. There is no tremor or fibrillation. In contrast to the normal thighs the legs are thin, and there is a very gross bilateral "pes cavus." The knee-jerks are active, and the hamstring jerks overactive, but the ankle-jerks and plantar reflexes are absent. The muscles of the calves show fibrillary tremors.

There is loss of sensation to pin-prick, cotton-wool, and to heat and cold of the glove type to the elbow and of the stocking type to the knee. Vibration sense and sense of position are normal and there is no ataxia. Cranial nerves are normal, the mental condition is good, and the sphincters are not affected. The peripheral nerves all seem larger than normal and are insensitive to pressure.

Case II

The first son, aged 28 years, a cook by occupation, has a long-standing deformity of his feet. For five years there has been progressive wasting and weakness of the hands and forearms, and recently he was compelled to give up his work as a cook owing to inability to lift heavy utensils. He is unsteady on his feet, and the muscles of the calves of his legs have gradually wasted. He frequently burns his hands at his work and only realizes this when blisters develop. His speech has become slurred, especially during the past few months.

The patient is a healthy-looking adult, with extremely well-developed shoulders, trunk muscles, and thighs (see Fig.). The small muscles of the hands are wasted, with the development of a bilateral claw-hand. All movements of the hands and wrists are weak, while at the elbow and shoulder power appears normal. Fibrillary tremors are present in the thenar eminence on the left side and in both forearms. The biceps and triceps jerks are active, the supinator jerks being greatly reduced on both sides. In the lower limbs there is bilateral "pes cavus," with wasting of the muscles of the calves and distal thirds of the thighs, the muscles being replaced by a firm

elastic type of fibrous tissue. The upper two-thirds of the thighs appear to be normal, in fact are well developed: the movements at both knees and ankles are very weak, there being fibrillary tremors in the muscles of the left calf and thigh. The knee-jerks and ankle-jerks are absent; the plantar reflexes cannot be elicited. The muscles of the legs show complete reaction of degeneration in the peroneal nerve group and partial reaction of degeneration in the tibial muscles.

Partial anaesthesia to light touch, to pin-prick, and to pain, heat, and cold of glove and stocking type extends from the periphery as high as the elbows and the knees. Vibration sense and muscle and joint sensibility appear to be normal. The hands are scarred with multiple burns, some only partly healed, and the subcutaneous tissues are thickened as in Morvan's disease. The only abnormalities in the cranial nerves are a bilateral horizontal nystagmus and pupillary reactions which are sluggish to both light and accommodation. The speech seems slurred, but the vocal cords move normally. The ulnar and peroneal nerves are very thick and completely insensitive even to firm pressure or pinching.

The Wassermann reaction is negative and the sugar tolerance curve of "flat" type, the maximum reading half an hour after 50 grams of glucose being 110 mg. of glucose per 100 c.cm. of blood.

Case III

The second son, aged 24 years, at the age of 12 met with a motor accident and two years later was operated on for a "pes cavus." For the past ten years his legs have progressively wasted and become more weak, until he is now unable to work. He frequently "trips" when walking, and his legs are more troublesome in cold weather. His hands have been clumsy for at least five years, and he is now unable to button his shirt during the winter.

The patient is a man of rather slow mentality, walking with the aid of a stick. The small muscles of the hands have almost completely atrophied and the forearms are thin; the grip is weak. The deep reflexes are active in both arms. No fibrillation can be detected in any of the muscles. The feet show a very marked bilateral "pes cavus," with scars of previous operations and some oedema of the subcutaneous tissues. The muscles of the calves are almost completely replaced by elastic fibrous tissue, and there is very little movement or power in the ankles. The knee-jerks are present but reduced; the ankle-jerks and plantar reflexes are absent.

There is partial loss of sensation to cotton-wool and pin-prick of glove type to the elbow and of stocking type to the knee. Sensations to heat and cold and to vibration, and muscle sensibility, are all within normal limits. There is a horizontal nystagmus on looking to the left, but otherwise the cranial nerves are normal.

Case IV

The third son, aged 21 years, a tailor's cutter by trade, four years ago noticed that his feet were abnormally thin and the veins on the surface very prominent. A year later "cramps" began in the muscles of the calves and thighs and the legs became more and more thin. For two years the fingers have been "numb" and useless during the winter; the hands are developing a claw-like appearance and are weaker than previously. For almost one year he has noticed some trouble with his speech, the words being "difficult to get out" and often slurred. Examination of the eyes reveals a bilateral rotary nystagmus and pupils that react slowly to both light and accommodation. There are no other abnormalities in the cranial nerves.

The hands and forearms are extremely thin, the small muscles of the palm being almost completely atrophied. There is marked loss of power in all movements of fingers and wrists. The biceps and triceps jerks are overactive, the supinator jerks being reduced. There is no fibrillation in the muscles, but a fine general tremor is present in both upper limbs. The malleoli are prominent, there is bilateral "pes cavus," and the muscles of the legs and the lower third of each thigh are very wasted. The knee-jerks are overactive; the ankle-jerks and plantar reflexes are not elicited. Glove and stocking anaesthesia to level of elbows and knees includes complete loss of sensation to cotton-wool and partial loss to pin-prick. Sensations to heat and cold

and to vibration, and muscle and joint sensibility, are normal. The nerves throughout the body are all abnormally easily palpable, especially the trunks of the brachial plexus and the peroneal nerves.

One son, one daughter, and two grandchildren of the widow (Case I) have been examined, but no abnormality of their nervous systems has been discovered.

Comment

Although the nerves have not been examined histologically it is probable that the four cases described above fall into the group of Déjerine-Sottas's disease—progressive familial hypertrophic interstitial neuritis. Although the onset of the condition is later than in the cases originally described by Déjerine and Sottas,³ others have been recorded in which the onset was delayed until adult life.⁴ The clinical picture of peripheral nerve thickening, symmetrical peripheral wasting of muscles, fibrillation, glove and stocking anaesthesia, and bilateral "pes cavus" is that of Déjerine-Sottas's syndrome.⁵ The limitation of the wasting to the forearms, legs, and lower third of the thigh,⁶ and the lack of progress of the condition beyond a certain degree of disability, are also characteristics.

REFERENCES

- ¹ Nattrass, F. J.: *Journ. Neurol. and Psychopathol.*, August, 1921, ii, No. 6, 159.
- ² De Brugn, R. S., and Stern, Ruby O.: *Brain*, 1929, lii, 84.
- ³ Schaller, W. F.: *Arch. Int. Med.*, 1912, x, 399.
- ⁴ Russell, W. Ritchie, and Garland, H. G.: *Brain*, 1930, ciii, 376.
- ⁵ Spiller W. G.: *Osler's Modern Medicine*, 1910, vii, 113.
- ⁶ Ironside, Redvers: *Oxford Loose Leaf Medicine*, vi, 998 (a).

J. E. Blair and F. A. Hallman (*Proc. Soc. Exp. Biol. and Med.*, December, 1935, p. 382) record their observations on the antihæmolysin titre in patients suffering from various staphylococcal infections. The sera were titrated in terms of the international standard antitoxin. Of sixty normal and Wassermann sera, fifty-four had a titre of 0.5 unit or less, the remaining six having 1 unit. Of twenty-five sera from patients with staphylococcal infections other than osteomyelitis, fifteen had a titre of 0.5 unit or less, while the remaining ten sera were ranged over a titre extending up to 22.7 units. Of eighty sera from patients with staphylococcal osteomyelitis, thirty-nine had a titre of 0.5 unit or less, fifteen had 1 unit, ten had 2 units, and sixteen had over 2 units. It has usually been found that sera from patients with osteomyelitis have a high antihæmolysin titre. In the present series, however, approximately two-thirds of these sera fell within the normal range of 1 unit or less. The fact that low titres are often found in patients with staphylococcal infections does not necessarily indicate inability to produce antibodies. This is shown by the observations made on thirty-five patients with osteomyelitis who were treated with staphylococcal toxoid. In every instance the patient responded to the antigenic stimulus by the development of a serum titre ranging from three to thirty times the original titre.