Medical Memoranda

Neurofibromata of the Vegetative Nervous System

The following case is reported not only on account of its personal interest but because I think it is not generally known that symptoms of visceral disorder in a case of Recklinghausen's disease may be due to neurofibromata lying on the conducting paths of the sympathetic and parasympathetic nervous system.

Bailey and Herrmann (1938) report two cases of neurofibromata involving the cranial nerves only. Ransom and Kay (1940) have collected no fewer than 18 cases of abdominal neurofibromata, in five of which tumours were found involving the small gut and its mesentery. Some of these cases had undergone malignant change. Klingenstein reports the case of a woman aged 29 suffering from severe melaena, which was found at operation to be due to an ileal neurofibroma; while Banerjee and Christeller (1926) mention two cases of ileal intussusception in which the intussusciens was a neurofibroma. Manzocchi (1946) reports a dramatic case of a volvulus of the ileum around a neurofibroma of the mesentery. Schwartzman *et al.* (1943), in reporting a large neurofibroma mass in the ileum of a boy aged 7, review the literature and mention that the condition may be inherited as a Mendelian dominant. Grill and Kuzma (1942) record a similar case which ended fatally without coming to operation: here a huge ileal mass caused death by intestinal bleeding. In this case, too, there was a strong family history of Recklinghausen's disease. Dr. Parkes Weber (1929) mentions the occurrence of visceral neurofibromata in the course of a very interesting paper dealing mainly with periosteal and subperiosteal manifestations of the disease.

CASE REPORT

A woman aged 54 was first seen in December, 1946. For two years she had had intermittent attacks of vomiting, associated with indigestion not amounting to pain and unrelated to meal-times. She was not losing weight, but her appetite was poor. Clinical examination revealed no abnormality other than the cutaneous lesions of Recklinghausen's disease affecting the whole surface of the body. She was admitted to hospital for further investigation. A barium-meal follow-through, a barium enema, a test meal, and blood investigation were all negative, though there was a faint trace of occult blood in the faeces. She also complained of haemorrhoids, and these I removed, together with two small neurofibromata near the anus which were pedunculated. Microscopy of the latter confirmed the already obvious diagnosis. On the clinical findings I did not feel justified in advising an exploration, and she was discharged from hospital.

She continued to have attacks of vomiting, however, and was admitted to another hospital, where she again went through the whole gamut of clinical investigations, including a cholecystogram, and with a similar negative result and a similar conclusion on the origin of her symptoms.

In March, 1949, Dr. George Varley, her medical adviser, referred her to me as she was starting to lose weight and nausea and retching were occurring almost every minute of the day.

A further clinical examination revealed no abnormality in the abdomen or chest; I nevertheless advised an exploration.

Operation.—Exploration on March 19 at once revealed the tumours described below, but first a thorough examination was

made of all viscera, solid and hollow, from the diaphragm to the pelvic floor. The oesophagus, stomach, and duodenum were quite normal, and exploration through the lesser sac was also negative. A careful dissection of the coeliac axis and an examination of the sympathetic chains down to and including the presacral region showed no other tumour masses, but the hypogastric plexuses were not explored.

Less than 1 in. (2.5 cm.) below the duodeno-jejunal flexure the tumour mass here illustrated was found. There were no adhesions about it and the double tumour appeared to arise from the ante-mesenteric border of the bowel, with a separate leash of vessels running to it, covered by peritoneum and joining the main mesentery at its root. The duodenum above was slightly dilated, but did not suggest gross mechanical obstruction. After division of the ligament of Treitz, the third part of the duodenum was mobilized sufficiently to bring it up into the incision so that a resection of the jejunum could be effected by end-to-end anastomosis with three layers of interrupted linen-thread sutures. About 4 ft. (1.25 m.) further down the jejunum a similar though much smaller mass was found; this was resected without difficulty, and a side-to-side anastomosis carried out, using continuous catgut. This tumour was not causing any obstruction.

After operation the patient had difficulty in passing water and had to be catheterized twice. This difficulty slowly diminished in the succeeding five days. Apart from this, convalescence was uneventful, and the patient has not vomited since the day of her operation, though she is still not perfectly well. Her appetite is poor and she is slow in regaining her normal vigour, but this is not surprising after five years of chronic nausea and vomiting. She also has some girdle pain at the level of the eighth to the tenth dorsal segments. On examination Dr. Louis Laurent found an extensor plantar response on both sides, but without sensory loss, and he thinks that this is probably due to an extradural compression by a neurofibroma, and that it may also explain the urinary difficulties.

Queckenstedt's test is normal, and Froin's test is negative. A barium follow-through is reported by Dr. Stevenson to show no obstruction either at the site of the recent anastomoses or at any other point. No further surgery on the condition is anticipated at present, as the symptoms are not disabling.

My thanks are due to Dr. L. Proger, Curator of the Pathological Museum of the Royal College of Surgeons, for the excellent preparation of the specimen and for the following pathological report: "The tumour is composed of spindle-cells with a varying amount of collagen. In places there is well-marked regimentation of nuclei characteristic of a neurofibroma."

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