ON A CASE OF MULTIPLE NEUROMATA.

BY

THOMAS F. CHAVASSE, M.D., C.M. (Edin.)

SURGEON TO THE BIRMINGHAM GENERAL HOSPITAL.

Received February 9th—Read June 8th, 1886.

MARGARET E—æt. 30, admitted into the General Hospital, Birmingham, July 18th, 1885.

History.—Four years ago was treated as an out-patient for what was then considered to be an enlargement of the cervical glands on the right side. An abatement in the size of the tumour apparently resulted, and no further increase in size took place until three months before admission. At first some large glands also existed on the left side of the neck, but these gradually disappeared. The patient now sought advice at the hospital because the tumour was growing, and caused pain down the arm on the affected side.

On admission.—There is a tumour as large as a duck's egg in the right posterior triangle of the neck, movable and seeming to all intents and purposes of a lymphomatous nature. The patient was short and stout, and slightly anæmic, but the general health appeared normal, and the various functions of the body were naturally performed.

Operation, July 24th.—A longitudinal incision was made over the growth, and, on reflecting the skin and fascia, a large nervous cord was found running over its upper

surface and required to be dissected off. The tumour itself extended deeply, dipping down behind the clavicle, and at its upper part was found attached by a pedicle, the thickness of the little finger, to the vertebral column.

During manipulation this pedicle was torn across close to the spine, and the step was followed by a gush of blood. A rounded aperture into which the tip of the finger could be inserted was left by the removal of the pedicle, and had to be plugged by a strip of boracic lint to stop the hæmorrhage. The wound was then drained and its edges approximated.

The evening temperature was 101° F., and the patient complained of violent headache.

July 25th.—Patient was semi-conscious, but could be roused, when she complained of her head aching. The arms and legs were constantly tossed about. There were twitchings of the facial muscles, and the urine passed involuntarily. Pupils slightly contracted and sluggish; the temperature varied between 102° and 103.4° F.

Next day the patient was quite sensible and remembered nothing of the previous day. On dressing the wound and removing the plug about two drachms of a clear-looking fluid escaped.

July 28th.—There was a rigor, followed by delirium and marked rigidity of the neck and head. This condition continued until July 31st, when muscular tremors and slight clonic spasms became marked. The pupils were widely dilated, and coma supervened, and the patient died in the evening.

Post-mortem August 1st.—Body very bloodless; rigor mortis slight.

Neck.—An incision three inches long existed parallel to the edge of the sterno-mastoid muscle, in the right posterior cervical triangle. This opened into an irregular cavity, one and a half inches in diameter, that led backwards and inwards to the spinal column, and at its very bottom was a round intervertebral foramen (fifth) empty of its nerve and containing pus. All the structures bound-

ing this cavity were thickened and adherent from inflammatory exudation.

Nervous system.—Brain weighed 52 oz. The membranes were smooth and shining; no sign of meningitis either on the vertex or at the base. The ventricles were distended with a thin clear fluid, and their walls were softened. This was the only abnormal change found in the brain-substance.

The *spinal cord* and plexuses were removed entire. The posterior surface of the cord was deeply congested, and thinly coated with a soft layer of dirty, yellowish-brown fibrin. This began at the fifth cervical nerve, and extended down the cord, but it did not pass to the front or ascend to the brain. There was a considerable quantity (half an ounce approximately) of thin semi-purulent fluid in the cavity of the arachnoid, which escaped when the latter was opened. The pia mater on the front of the cord was deeply congested, but there was an entire absence of lymph. The cervical enlargement was soft and pulpy, especially opposite the sixth nerve, and, on section, the substance was discoloured, the white matter being of a greyish tint, and the grey matter less defined than usual. The fifth cervical nerve was discoloured and thickened on the right side from inflammatory changes. The sixth nerve had been torn off, the root giving way inside the dura mater, so that the ganglion went with the torn portion. The ends of the anterior and posterior roots were found within the dura mater. All the nerves that could be examined were found to be irregularly enlarged. Surrounding them were various sized tumours contained within the nerve-sheath, and apparently having the nerve running through them like an axis. Most of them were fusiform, a few globular, and, on section, they appeared white, glistening, semi-translucent, and extremely firm. The nodulation began as soon as the nerve left the dura mater, and was first seen in the ganglion of the root which, all down the cord, was greatly enlarged.

Inside the dura mater the nerves were quite normal.

The trunks of the nerves were much increased in size by a sort of diffusion of the tumour, so that, for example, the sciatic was one and a quarter inches broad and proportionally thick, and the anterior crural half as large again as the normal sciatic. Even the small nerves, *e. g.* the genito-crural, were affected, and on them, the nodules were much larger in proportion to the diameter of the nerve, than was the case with the larger ones. The sympathetic nerves were similarly affected, and the fine filaments in the rectum could be easily traced by means of the nodules. They could also be seen beneath the mucous membrane of the tongue and the pharynx.

The pneumogastrics were equal to a penholder in size. The phrenic nerves appeared like a string of dahlia roots. The various thoracic and abdominal viscera were healthy.

Similar cases of so-called multiple neuromata appear to be somewhat rare. Lebert has collected seventeen cases, and Prudden,¹ of New York, has extended these to forty-one. From such records, the clinical histories being extremely meagre in seven, the following facts may be deduced :

I. The male appears to be more prone to this development than the female. The sex is recorded in thirty-two instances, and of these twenty-four were men, eight were women.

II. The middle period of life is most liable to the affection, but in some of the cases Prudden's opinion is that the tumours were undoubtedly congenital.

III. The duration of the disease has not been determined. In twenty-three cases, where the age at death is stated, the fatal termination occurred, on an average, between thirty-three and thirty-four years.

What Lebert terms the second stage of development, and this appears to be the period of pronounced swellings, is stated to be, five or six months.

¹ American Journal of Med. Sc., July, 1880.

IV. Clinically no constant symptoms are manifest in cases of multiple neuromata.

In twenty-six instances where the history is fully enough reported, twelve had no symptom pointing to a nerve lesion.

In three there was more or less paralysis, but this by no means in proportion to the size and number of the tumours.

Pain was only experienced in thirteen patients; this varied much; in some it is described as being spontaneous, in others it was elicited by pressure or atmospheric changes. This absence of pain seems remarkable, considering that both the mixed and sensory nerves were covered with tumours. Typhoid fever appears to be badly borne in this class of case, five deaths being attributed to it in a mortality of twenty-seven.

Three patients died of phthisis; in one recorded by Dr. Wilks,¹ the writer thinks that it is possible that the condition was due to the lesion of the pneumogastric inducing the pulmonary changes.

In many cases, nutrition of the body is reported to have been interfered with, yet on post-mortem examination no organic disease of the viscera was found. Most authors agree that the prognosis in this disease is unfavorable.

V. The tendency of the condition is to appear in several members of a family. Nicaise² thinks there is sufficient evidence to show that it is frequently congenital and hereditary. Hitchcock³ has reported cases in which the mother, her son and daughter all exhibited multiple neuromata. Generisch⁴ cites an instance in which the patient, whose mother had suffered from numerous tumours diagnosed as neuromata, died of pneumonia. At the post-mortem, tumours of various sizes were found on nearly all the nerves of the body. Four years

¹ 'Transactions of the Pathological Society of London,' vol. x.

² 'International Encyclopædia of Surgery,' vol. iii.

³ 'American Journal of Med. Sciences,' vol. xliii, 1862.

⁴ Virchow's 'Archiv,' Band 49, 1870.

afterwards the brother of the preceding case died of tetanus, and neuromata were then found to exist everywhere. Both vagi and the phrenics were affected. The roots of the spinal nerves were normal.

Bruns¹ reports a case in which death was caused by hæmorrhage from the carotid artery. Many tumours, some the size of a pigeon's egg, were found on the nerves. The patient's brother had congenital elephantiasis with plexiform neuromata about the head and neck, and the mother is said to have had wart-like tumours in the skin.

VI. Operative interference is badly borne. This is exemplified by the following cases :

(A) One of the tumours removed from the left radial nerve. Death in five weeks from pyæmia.

(B) Amputation of right leg for ulceration and gangrene of toes. Died in two days of pneumonia.

(C) Tumour near clavicle the size of a hen's egg and another small one near the lip were enucleated and did not return. According to the statement of the patient the tumours in the other parts of the body increased in number more rapidly after the operation.

(D) Removal of tumour, six and a half by three and a half inches in size, from the right ulnar nerve, the nerve itself being severed in the operation. The wound healed ; a year later disarticulation at the shoulder-joint was performed for a return of the growth. The stump did not heal. Death from exhaustion seven months later.

(E) Removal of tumour the size of a clenched fist. Vagus divided. Died on the tenth day, hæmorrhage taking place from ulceration of the carotid.

(F) Attempt made to remove a tumour from the back. Died of pyæmia.

(G) A portion of the lesser sciatic nerve excised in an endeavour to check the growth of many tumours corresponding to the branches of the nerve. Wound healed by suppuration. After four months many of the swellings disappeared and the rest gave no trouble.

¹ Virchow's 'Archiv,' Band 50, 1870.

VII. The tendency of the disease to become malignant.

This appears to be rare. In Hitchcock's third case the tumour removed from the ulnar nerve, after existing for upwards of twenty years, presented on section the characters of a doubtful neoplasm. A year later, after amputation of the limb for its recurrence, it was certified to be of an encephaloid nature, and the patient died a few months later with a return in the cicatrix.

In Genersich's¹ case, multiple tumours having existed for some time, ten weeks before death a rapidly growing neoplasm appeared in the right buttock. Examination after death showed that some of the tumours were fibromata, some sarcomata, and others myxomata.

In both these cases there was a hereditary tendency to neuromata, and the mother of the second case had carcinoma of the mamma.

VIII. The position of the tumours.

In twenty-seven cases the peripheral nerves were affected.

In ten cases special groups of nerves were implicated.

In sixteen cases the sympathetic and in twenty-two the vagi were involved.

As a rule, however, special nerves appear to be unaffected and the nerves of the hands and the feet are free from the lesion.

Microscopic Examination of the Tumour itself.

The tumour is composed of anastomosing and branching bundles of white fibrous tissue, which intersect one another at varying planes. Between the fasciculi are embedded numerous fusiform, oval, and round cells, resembling the embryonic connective-tissue corpuscles. No elastic fibres are to be seen and no well-defined stellate or branching connective-tissue cells. There is no evidence of fully developed nerve-fibrils in any of the sections. (See Woodcuts on page 524.)

¹ Virchow's 'Archiv, Bd. 49, 1870.

a. Spindle-cells and connective tissue cut transversely, with some round-cells ; *b*, spindle-cells in delicate connective tissue.

a. Spindle-cells and fibrous tissue cut longitudinally, with a few round-cells ; *b*, spindle-cells and connective tissue cut transversely, with some round-cells.

LITERATURE.

BARKOW.—Acad. Cæs. Leop., Nova Acta, Bd. 14, 1828, p. 514.

BRUNS.—Virchow's Archiv, Bd. 50, 1870, p. 80.

COURVOISIER.—Die Neurome, 1886. (Contains a full bibliography of Neuromata.)

- CZERNY.—Archiv für klin. Chirurg., Bd. 17, 1874, p. 357.
- GENERSICH.—Virchow's Archiv, Bd. 49, 1870, p. 15.
- GERHARDT.—Deuts. Archiv für klin. Med., Bd. 21, 1878, p. 268.
- GUNSBURG.—Comptes Rendus de l'Académie des Sciences, tom. 17.
- HELLER.—Virchow's Archiv, Bd. 44, 1868, p. 338.
- HEUSINGER.—Virchow's Archiv, Bd. 27, 1863, p. 206.
- HITCHCOCK.—American Journal of the Medical Sciences, vol. 43, 1862, p. 320.
- KOSINSKY.—Centralblatt für Chirurgie, July 18th, 1878.
- LEBERT.—Mem. de la Société de Chirurgie de Paris, tom. 3, 1853, p. 249.
- NICAISE.—The International Encyclopædia of Surgery (Ashhurst), vol. iii.
- ODIER.—Manuel de Médecine Pratique, 1811.
- PRUDDEN.—American Journal of the Medical Sciences, vol. 80, 1880, p. 134.
- SCHIFFNER.—Med. Jahrbüch. Oester. Staats, Bd. 4, 1818, p. 77 ; Bd. 6, 1820, p. 44.
- SERRES.—Comptes Rendus de l'Académie des Sciences, tomes 16, 21, 22.
- SIBLEY.—Medico-Chirurgical Trans., vol. 49, 1866, p. 39.
- SMITH (Robert W.).—A Treatise on the Pathology, Diagnosis, and Treatment of Neuroma, Dublin, 1849.
- SMITH (Thomas).—Trans. of the Pathological Society of London, vol. 12, 1860, p. 1.
- WEGENER.—Berliner klin. Wochenschrift, 1870, p. 24.
- WILKS.—Trans. of the Pathological Society of London, vol. 10, 1859, p. 1.
- WOOD (William).—Trans. of the Medico-Chirurgical Society of Edinburgh, 1829, vol. iii, Part 2.

(For report of the discussion on this paper, see 'Proceedings of the Royal Medical and Chirurgical Society,' New Series, vol. ii, p. 125.)

DESCRIPTION OF PLATE XIV.

Multiple Neuromata. By T. F. CHAVASSE, F.R.C.S.

a. Smallest splanchnic.

b. Genito-crural.