Choroido-Ependymal Cysts of the Spinal Roots

Case Report

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The case presented herein concerns a disease of the spinal roots which is, as far as we know, unique. The patient was operated on at our hospital in 1960 and was observed and treated until her death early this year. The delay in reporting the case was because we wanted to watch the developments of the disease after operation. Despite incomplete pathological evidence* (no necropsy having been done) it is presented now in view of its exceptional features.

Case Report

Ser. No. 30573-1960. M.N. was a woman aged 32. In 1948, at the age of 14 she suffered marked loss of hearing in both ears followed about 15 days later by loss of visual acuity; in 2 weeks her sight diminished from normal to near blindness. This was followed by lethargy and violent headache. On admission to another hospital, about a month after the onset of the symptomatology, the patient was found to have bilateral papilledema of 2D with severe limitation of visual acuity (she could count fingers at a distance of 50 cm.) and bilateral loss of hearing with bilateral vestibular hyperexcitability. Ventriculography on Feb. 20, 1942 disclosed enormous dilatation of the lateral ventricles, especially on the right side, whilst the 3rd ventricle was incompletely injected because of a walnut-sized filling defect visible in the lateral view at the level of the foramina of Monro. An operation was performed at once and a thin-walled cyst full of faint yellow fluid, facing the right foramen of Monro, was completely removed through the right anterior horn.

Microscopical sections showed that the wall of the cyst consisted of a very thin, practically nonvascular, membrane lined with monostratified low columnar or cuboidal epithelium (Fig. 1). Diagnosis: Neuro-epithelial cyst of 3rd ventricle.

The postoperative course was uneventful and the patient's disorders disappeared except that she became totally blind.

In the spring of 1958, 16 years after operation, when the patient was 31, she began to have sciatic pains, first on the right side and then on the left, and rigor of the dorsolumbar spine. The pain was not continuous but it was aggravated by exertion and by the erect position. In 1959 the pain worsened and spread. After various treatments and tests at other hospitals, she was admitted to the Istituto Neurologico di Milano on March 4, 1960.

Received for publication December 20, 1963.

* We are most grateful to Prof. K. J. Zülich of Köln, Germany, for his invaluable help in the interpretation of the microscopical preparations.

Fig. 1. Photomicrograph of wall of cyst of the 3rd ventricle lined with monostratified epithelium. Hematoxylin and eosin, X 430.

Examination. There were many flat nevi and café-au-lait spots on the skin of the trunk. Positive neurological findings were bilateral blindness and nystagmus in all positions of gaze and bilateral secondary optic atrophy. The spine was rigid and tender on active and passive movements; paravertebral pressure and percussion were painful, especially in the lower dorsal and lumbar tracts; there was accentuation of the lumbar curve. There was slight loss of strength in the lower limbs, perhaps greater on the left. Lasègue's sign was present bilaterally. There was a bilateral band of hyperesthesia at D8-L1. Tendon reflexes of the upper limbs were weak symmetrically; abdominal reflexes and tendon reflexes of the lower limbs were absent (this was observed at once after the intracranial operation).

In the roentgenograms of the skull the Burr holes for ventriculography and right frontal flap were still visible. The sella was somewhat enlarged with the dorsum slightly decalcified. Films of the spine demon-
Fro. ~. Myelography. Several clear-cut defects are in the cervical (a), middorsal (b) and lumbar (c) tracts.
In the lumbar tract there are irregularities in the inner profile of the pedicles.

Myelography was performed with 6 cc. of Myodil by lumbar route. The contrast medium rose slowly up the lumbar tract because of the presence of many filling defects. There was a filling defect at the level of the right pedicle of D4 and others in the cervical tract (Fig. 2). Radiologic diagnosis: von Recklinghausen's disease with multiple spinal localizations.

Course. With the appearance of the skin and the result of myelography, von Recklinghausen's disease was diagnosed. The patient's parents were informed that no lasting benefit would result from operation, but they insisted on it in order that she might be given at least transient relief of pain.

Operation (March 11, 1960). Laminectomy of L3 and L4 was performed under endotracheal general anesthesia with N2O and ether. On incision of the dura mater the expected solid neurofibromatous nodule was not found but instead a thin-walled cyst containing clear, colorless fluid, adherent to a root of the cauda equina. The cyst was removed completely with resection of the root. This was followed by laminectomy of D12 and L1 and again at this level 3 cysts, identical to but smaller than the first, were found adhering to the roots of the cauda equina. The walls of these cysts were excised as completely as possible but the roots were not touched in order not to cause gross neurologic deficit.

Microscopical Examination. The walls of the cysts, which were all identical in aspect, consisted of a slender supporting framework of connective tissue, poor in vessels, lined with a single layer of cuboidal or columnar cells (Figs. 3 and 4). At some points there were ramified papillary structures with a vascular connective-tissue axis lined with cells identical to the ones described previously. These papillae closely resembled the choroid plexuses in structure (Figs. 5 and 6). There was no mitosis or other evidence of cellular malignancy, though there were some vesicular cells with very pyknotic nuclei, perhaps evidence of "ablaufenden Mitosen".

Course. The early postoperative period was disturbed by cramp-like pain in the thighs, which yielded to cortisone treatment and the patient was discharged on April 19, 1960 in fair health but without substantial modification of the objective signs, except for attenuation of the Lasègue's sign. The patient enjoyed fair health for 3 months and then her condition worsened again. She was treated both as an outpatient and as an inpatient until October 1962. During this period there were successive attacks of pain in the upper part of the chest on the right side, cervicothoracic root pain on both sides but mainly on the right, signs of a lesion of the left anterior and posterior lumbosacral roots, dysesthesia to painful stimuli in the right trigeminal region and finally cerebellar signs (a tendency to fall backwards and to the right). In view of the clearly systemic nature of the disease, no further operations were advised. The repetition of cortisone treatment and roentgen therapy...