MENINGEAL MENINGIOMATOSIS

REPORT OF CASE

ALFRED UHLEIN, M.D., EDWARD M. GATES, M.D.,* AND ROBERT G. FISHER, M.D.*

Section on Neurosurgery, Mayo Clinic and Mayo Foundation, Rochester, Minnesota

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In 2,023 cases of intracranial tumor, Cushing,10 according to his own classification, described 271 cases of meningioma representing 13.4 per cent of his total cases. Baker,4 in 757 cases of intracranial tumor found 122 cases of meningioma, or a percentage of 16.1, while Courville6 found, at autopsy, 71 meningiomas in 561 intracranial tumors, an incidence of 12.7 per cent. This type of tumor, for the most part, represents a benign lesion and frequently is operable. However, it is important from both an academic and surgical viewpoint to realize that this type of lesion may occur in multiple sites; a number of case reports of multiple meningiomas have appeared in the literature in the last fifty years.1,2,7,12,15,16,17,19,21,22 Frequently, a number of tumors are found in such conditions as von Recklinghausen’s disease with no explicable cause. Cases of multiple meningiomas, according to the literature, are comparatively rare. The case to be reported represents one that we have elected to classify as “meningeal meningiomatosis.”

REPORT OF CASE

The patient was a 24-year-old boy who had had a normal birth and development. About April 1, 1947, he complained of episodes of backache in the sacral region. The mother noted that when she picked the child up by grasping him under the arms he would cry out with pain. These pains and the backache occurred periodically each day but he continued to play normally. However, he would awaken at night screaming with pain, which lasted but a short time, and then he would fall asleep.

In May, 1947, the mother noticed that the boy began to walk “like a little old man who needed a cane.” He complained of pains in the right knee and thigh, and would awaken from his sleep nearly every 2 hours during the night crying with pain in the back and leg. He would not bend the right leg or thigh beyond an angle of 45° because of the severe pain that such motion caused him. Roentgen studies of the spinal column conducted by the family physician were reported as negative.

In June, 1947, the patient was no longer able to walk upright and had to crawl to get about his home. He was hospitalized at that time in the vicinity of his home. Repeated lumbar punctures were attempted but fluid was not obtained. Fluid obtained on cisternal puncture was reported to have contained 15 cells/cc., with 20 per cent polymorphonuclear leukocytes and 80 per cent lymphocytes; protein 50 mg./100 cc.; globulin test positive; cultures failed to show any growth of organisms. While the patient was in the hospital, there developed right facial paralysis and weakness of the right leg which was followed in a few days by weakness of

* Fellow in Neurosurgery, Mayo Foundation, Rochester, Minnesota.
the left leg. He experienced considerable pain in his back and legs at that time. Passive motion of his legs elicited rather severe pain. A diagnosis of brain tumor was made and the patient was brought to the clinic on July 4, 1947.

*Examination.* The boy was lying in bed crying with pain. He was normally developed and well nourished. Temperature was 99.8°F. There was ptosis of the right upper lid. Questionable nystagmus was noted on left lateral gaze. Ocular movements were normal. The pupils were equal and their responses to light and convergence were normal. His speech did not show any abnormalities. There was complete paralysis of the muscles of facial expression on the right side. Motor power was normal in the left upper extremity and left side of the trunk. The muscles of the right upper extremity and right half of the abdomen were weaker than normal. There was definite weakness of the muscles of the right lower extremity; the muscles of the left lower extremity were weak but the weakness was not as great as that of the right. The triceps reflexes of both arms were absent. The quadriceps surae and triceps surae reflexes were bilaterally absent. No Hoffmann or Babinski reflexes were obtained. Kernig’s sign and Lasègue’s sign were present in both extremities. A moderately stiff neck was noted. The anal sphincter was completely relaxed and paralyzed. The boy was incontinent of both feces and urine.

Sensory examination gave normal results except for complete loss of all modalities in the region of the buttocks, perineum and posterior and anterior aspects of the thighs. Pin prick over the legs produced intense pain characteristic of hyperesthesia. Funduscopic examination did not disclose any papilledema or lesions of the retina. The results of laboratory studies of the urine and blood were essentially normal. Flocculation studies for syphilis gave normal results. Roentgenograms of the entire spinal column, head and thorax failed to show any abnormalities. Lumbar punctures were attempted on several occasions but were always unsuccessful since no spinal fluid could be obtained.

An EEG made on July 9, 1947, revealed generalized delta activity that was most marked posteriorly. At that time deafness in the right ear was noted. The diagnosis of intracranial neoplasm with metastasis to the lower part of the lumbosacral region was made.

*Operation.* Because of the severe and unbearable pain which could not be relieved by medication, a lower dorsolumbar laminectomy with decompression was carried out by one of us (A.U.) on July 17, 1947. During the operation it was noted that the filum terminale was consolidated into one large tumor mass. The specimen taken at this time was reported by the surgical pathologist as being a highly malignant tumor, possibly a malignant meningioma.

*Postoperative course.* was satisfactory and the patient was relatively comfortable. Since it was felt that this type of tumor might respond to roentgen therapy, a palliative course of treatment was given to the cerebellar region. However, the disease continued to progress in spite of this and evidence of acute increased intracranial pressure became apparent. In order to relieve this acute situation a ventriculostomy with continuous ventricular drainage was performed. The boy remained comfortable until the final day of his illness when convulsions developed; death occurred on the 36th hospital day.

*Necropsy.* A careful examination of all the body tissues and organs was made to determine whether the neoplasm of the central nervous system was a metastatic or a primary growth. No evidence of metastatic lesions could be found anywhere outside of the central nervous system. The brain and spinal cord weighed 1,175 gm. (Fig. 1). There was a well-healed scar, 8 cm. long, over the spinous processes in the midline extending from the level of the 9th thoracic spine to the level of the 1st sacral vertebra. A small right occipital trephine opening where a ventricular catheter had previously been in place was noted. The calvarium over its entire extent was unusually thin.

The dura was tense and adherent to the skull in the occipital region around the foramen magnum, where there were multiple soft, purplish-colored pedunculated tumors. In the region of the right petrous bone surrounding the internal auditory foramen and growing into the foramen, a flesh-colored soft pedunculated mass engulfed the auditory and facial nerves. A similar condition obtained about the left auditory foramen, auditory nerve and facial nerve. About the posterior clinoid region extending to the anterior border of the foramen magnum
FIG. 1. a. Gross appearance of the brain and spinal cord. b. Many tumors are present along the course of the cord. c. Horizontal section through the brain.
another soft flesh-colored pedunculated mass was noted. The venous sinuses were all unobstructed.

The arachnoid over the brain and cerebellum was thin, delicate and translucent except in multiple areas over the lobes of both cerebellar hemispheres. In these small areas the lesions appeared slightly elevated, opaque and pedunculated. The arachnoid throughout the entire length of the spinal cord and the filum terminale was surrounded by tumor growth. The growth consisted of multiple pedunculated masses which had grown freely from anterior and posterior nerve roots into the cord substance, forming thick plaques encircling the cord. The filum terminale was a solid mass of tumor tissue.

Examination of the cerebrum disclosed small fleshy pink tumors in the choroid plexuses of both anterior and posterior horns of the lateral ventricles. The ventricular system was slightly dilated. A small pink friable mass had infiltrated the posterior limit of the right side of the thalamus. The 3rd ventricle was anatomically open. The 4th ventricle contained a pedunculated flesh-colored tumor which almost completely filled the chamber (Fig. 2a). The cerebellopontine angles were free of tumor tissue. The right anterior inferior surface of the cerebellum was deeply grooved by a pedunculated friable neoplastic mass.

Multiple cross sections of the brain stem revealed tumor tissue that had grown into the anterior portion of the pons and medulla. The spinal cord, on cross section, at no level was found free of neoplasm (Fig. 2b, c, d and e). In the lumbar region the cord was compressed and infiltrated to the extent that nothing remained but the posterior horns of gray matter and the posterior fasciculi. The filum terminale was a solid sheet of tumor which surrounded and compressed the nerves.

Microscopic Examination. The dural masses were found to be composed of a dense collagenous stroma in which were imbedded islands of oval-shaped, pale, pink-staining cells with elliptical nuclei. The cells were highly pleomorphic and grew in no characteristic pattern. Tumor giant cells were present but not in any great numbers. In some sections, the nuclei of these neoplastic cells were hyperchromatic and showed abnormal mitotic figures. In general, a fair amount of chromatin material was present about the periphery of the nuclei. The tissue was well vascularized, with the vessels running in the collagen network. Some adventitial and intimal proliferation could be found in the network of vessels.

In routine sections of the cerebral lobes in which tumor tissue was not suspected grossly, such tissue was found microscopically. These sections showed the masses to be arising from the arachnoid cap cells and streaming off along the pia-arachnoid down deep into the sulci and infiltrating the cortex (Figs. 3 and 4). This was a common picture seen throughout the entire cerebrum and in the cerebellum. The picture was again repeated when sections of the pons and medulla were examined. Microscopic study of the cerebellum revealed small bits of tumor growing between the folia and into the cerebellar tissues. The choroid plexuses were engulfed in neoplastic cells.

Sections of the spinal cord showed the same type of tissue and the same ingrowth of tumor tissue from the pia-arachnoid. The picture was consistent with that of highly malignant meningiomatosis of meningeal origin. No primary neoplastic site could be found. It was felt from studying these sections that the entire arachnoid had, in various and multiple locations, undergone malignant changes, giving rise to many primary malignant tumors which were highly invasive. The term "sarcoma of the meninges" was avoided for it was felt that insufficient evidence exists to justify classification of the meningiomas as of mesodermal origin. Thus until such time as it could be shown that the meninges arise from mesoderm, it was felt better to adhere to a different term for these arachnoid tumors; accordingly, in this case the term "malignant meningeal meningiomatosis" was used to describe the condition.

The tissues were seen by Dr. J. W. Kernohan who concurred in the diagnosis and in the justification for the term "meningeal meningiomatosis."

COMMENT

In the preoperative study of this patient, the clinical diagnosis was intracranial tumor; it was thought that this tumor was the primary source for a metastatic lesion
Fig. 2. Sites where tumor tissue was found. 

a. Fourth ventricle ($\times 2\frac{1}{2}$). 
b. Cervical cord ($\times 5\frac{1}{2}$). 
c. Thoracic cord ($\times 5\frac{1}{2}$). 
d. Lumbar cord ($\times 5\frac{1}{2}$). 
e. Filum terminale ($\times 5\frac{1}{2}$).
in the spinal column. The degree of malignancy was determined at the time of the exploratory laminectomy but the degree of involvement of the nervous system by such a diffuse process was not determined until the postmortem examination had been completed. The pathologic classification of a case of this type is difficult and again brings up the question of the origin of the meninges. If one assumes that the meninges are of mesodermal origin, then the findings in this case might represent an instance of primary sarcoma in the meninges, with secondary metastasis to other parts of the leptomeninges. The other assumption is that this case represents diffuse transition of normal meningeal tissue to neoplastic tissue.
Schultze, in 1880, reported a case of “multiple pial sarcoma” and Nonne, in 1902, substantiated this idea and described tumor cells in the pial vessels which secondarily invaded the meninges. Fried described a rare condition under the title of “diffuse sarcomatosis of the meninges” in which there had been a general change in the nature of the leptomeninges. Bailey, in 1929, stated that the meninges were of neuro-ectodermal origin but recognized that intracranial sarcomatous tumors may have leptomeningeal origin. Bailey and Bucy, in discussing the origin of meningeal tumors, stated that tumors such as we have described are of connective-tissue, not gliomatous, origin. These authors assumed that meningeal mesenchyme derived from neuro-ectoderm or mesoderm might give rise to the various types of meningioma.

Globus, Levin and Sheps stated that the pia is of mesodermal origin and is derived from a primitive endomeninx. Also they said that the endomeninx, with the ectomeninx which is the periosteum, and the dura had a common precursor in the skeletoneural intertissue which was derived from the mesenchyme. Cushing and Eisenhardt stated that the term “multiple meningiomas” referred to several dis-
crete tumors, but that if the neoplastic nodules were numerous and coalesced, then the condition was meningiomatosis or diffuse meningioma. Previously Connor and Cushing cited reports of 2 cases in which diffuse tumors of the meninges were apparent only on microscopic examination and the condition in these cases was called "true sarcomatosis" or "endotheliomatosis." It was their impression that the tumors originated in the adventitia or perivascular sheaths of the cortical blood vessels. Brown and Kernohan, in 1941, limited the term "meningiomatosis" to an arachnoid type of lesion and assigned purely mesodermal tumors to the category of sarcoma. In their case, the cells had morphologically benign characteristics. They reasoned that the process of meningiomatosis resembled a neoplastic transformation of multcentric origin arising on an incompletely differentiated arachnoidal membrane. Since there is no further evidence to justify classification of the histopathologic features seen in our case differently, we have used the title "meningeal meningiomatosis."

SUMMARY

The case of a 22-year-old boy is reported in which a diffuse transitional process in the meninges resulted in multiple tumors of the leptomeninges with malignant characteristics. The term "meningeal meningiomatosis" is used to describe the pathologic characteristics observed.

REFERENCES

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THIRD VENTRICULOSTOMY PATENT AFTER FIFTEEN YEARS

IRA COHEN, M.D.*
New York, N. Y.

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When obstructive hydrocephalus in adults is due to a lesion that is not removable, one of two palliative procedures may be employed. The obstruction may be bypassed by a tube leading from the lateral ventricle into the cisterna magna, as described by Torkildsen. Or the accumulation of excess fluid in the ventricle may be prevented by opening a communication between the 3rd ventricle and the basal cisternae. This procedure, first described by Dandy in 1932, was devised for congenital hydrocephalus in older age groups. Among others its use has been described by Stookey and Scarff and by White and Michelsen. The latter reported on its use in 11 cases; of these, the longest survival up to the time of reporting was 2 1/2 years. In Dandy’s report in 1945 there was 1 patient living 23 1/2 years after operation and 3 living between 10 and 20 years.

It is a fair assumption that these long survival periods are dependent on the continuous patency and functioning of the artificial stoma, though the verification of such a conclusion is lacking. In a case reported by Sweet the spontaneous ventriculostomy found at necropsy had, to judge by the history, functioned for 12 years. The following case report yields additional proof of the long-range patency of a ventriculostomy.

CASE REPORT

In 1931, a young woman aged 17 years was admitted to the Neurological service of the Mount Sinai Hospital, complaining of adiposity of 2 years’ duration and visual difficulty for 4 months. She had always been plump and became definitely stouter with increase in food consumption. The first visual difficulty was that of black spots before both eyes. Then vision in the left eye became impaired, followed by loss in the right eye. Two months prior to admission vision was so poor that she was unable to distinguish objects or colors. For 2 months she had complained of tinnitus in the left ear with some loss of hearing in both ears, slight vertigo and occasional headache. There was nothing in the history to suggest polyuria, polydypsia, nor menstrual disturbances.

Examination. She was noted to grope her way about the ward because of visual loss. Her

* 4 East 95th St., New York 28, N. Y.