MENINGEAL MENINGIOMATOSIS

REPORT OF CASE

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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,8 in 757 cases of intracranial tumor found 122 cases of meningioma, or a percentage of 16.1, while Courville9 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

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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