EPENDYMOMA OF THE CAUDA EQUINA WITH MULTIPLE VISCERAL METASTASES

REPORT OF A CASE

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Tumors of the central nervous system that metastasize outside the confines of the craniospinal axis have always been of interest because of their rarity. About 60 such instances have been recorded in the literature in the thirty or forty years since the histologic classification of brain tumors has become relatively standardized. All save 2 have been exclusively intracranial in origin. The exceptions were 2 ependymomas of the cauda equina—one of which invaded the muscles of the back and a second which developed intra-abdominal metastases. A third such ependymoma with metastases is described in the following account.

CASE PRESENTATION

In 1953, after a 3-year history of bilateral sciatica, a 28-year-old woman had an ependymoma of the cauda equina partly removed at another hospital. Following the operation she was given an intensive course of roentgen-ray therapy and remained free of symptoms for about 7 years. She then began to complain of low-back pain radiating down the posterior aspect of the right lower extremity, and urinary incontinence developed.

1st Operation. Exploration disclosed a large tumor that occupied the dural sac from the 2nd lumbar level to the sacrum. The tumor was resected, but it was necessary to leave small fragments attached to some of the nerve roots in order to preserve neural function. Much of the dura mater was involved by tumor and was excised.

Histology. The tumor has a papillary structure. Centrally there are small capillaries surrounded by acellular, eosinophilic material that is sometimes amorphous and sometimes fibrillar. The outer portions of these villi are covered by neoplastic cells in either single layers or pseudostratified formations (Figs. 1 and 2). The cells generally are uniform in size and have oval, vesicular nuclei. Mitotic figures were not seen. There are areas of necrosis, hemorrhage, and desmoplasia. Diagnosis: Ependymoma.

Course. Improvement with partial return of function followed operation. Over the next 3 years there were several episodes of thrombophlebitis in the lower extremities. Intermittent pain in the left buttok, radiating to the heel, developed subsequently.

Upon readmission to The New York Hospital in 1957, roentgenograms showed multiple round, nodular densities varying in size from 3 mm. to 3 cm., in both lungs. Roentgenograms of the spine showed that in addition to the operative defects there was erosion of the posterior aspect of the upper sacrum. An intravenous pyelogram showed mild right hydronephrosis. Barium enema and proctoscopy revealed normal findings.

2nd Operation. To determine whether the new lesions were metastases of the ependymoma or possibly a second neoplasm, the sacral tumor was biopsied.

Histologic Diagnosis. Ependymoma.

Course. Further irradiation to the sacrum was followed by some relief of pain, but 2 months later pain recurred in the back, pelvis, and thighs, and was accompanied by marked weakness of the lower extremities and severe dyspnea. At that time, 4 months prior to death, a left pleural effusion was present and multiple pulmonary metastases were seen on radiological examination. Additional irradiation was given to the chest and lower thoracic region, but without benefit.

On final hospitalization, because of pain and dyspnea, there were signs of a left pleural effusion, distension of the veins of the neck, ascites, en-
largement and tenderness of the liver, and edema of the ankles. The patient died Dec. 22, 1957, after an illness of 17 years.

Postmortem Examination. A complete autopsy performed 7 hours after death revealed that the laminae of the 2nd, 3rd, 4th and 5th lumbar vertebrae had been removed surgically. Their vertebral bodies were largely replaced by yellow-white tumor which extended laterally into the left psoas muscle and the roots of the lumbo-sacral plexus (Fig. 3). The tumor in the psoas muscle was continuous with a tumor mass that overlapped the lower lumbar and upper sacral vertebrae. The dura mater covering the lower half of the spinal cord was thickened and measured 1 cm. in places. Microscopically, the inner aspect of the dura mater at the sacral level was infiltrated by neoplasm similar to the tumor that had invaded and obstructed the iliac veins, the ovarian veins and the inferior vena cava up to the orifices of the renal veins. Both ureters were partly obstructed.

The superior and anterior mediastinum were filled with masses of tumor and the pleura was involved extensively. Scattered throughout the parenchyma of the lung were soft nodules of tumor measuring up to 7 cm. in diameter. Some of the hilar lymph nodes were also replaced by tumor.

The liver weighed 4060 gm. and contained many nodules of tumor (Fig. 4). The para-aortic lymph nodes were also replaced by tumor. Other organs, including the brain, were not remarkable.

Histologic Examination. The tumor in all areas is essentially the same in microscopic appearance. In sections of the lumbar vertebrae and adjacent tissues tumor involves the marrow, periosteum and psoas muscle and frequently surrounds the nerves of the lumbo-sacral plexus. Under low magnification the neoplastic cells appear to be of uniform size and shape, although higher magnification reveals nuclear pleomorphism and variability in intensity of staining. Most nuclei, however, are round and vesicular with a prominent nucleolus. Mitotic figures were found occasionally, and a few multinucleated cells are present. The cytoplasm is scant and lightly eosinophilic. Phosphotungstic acid-hematoxylin stains failed to reveal either blepharoplasts or foot processes (Fig. 5) and silver-carbonate preparations did not demonstrate neuroglial processes.

In most places the polygonal cells are arranged in low pseudostratified layers to form membranes and papillary structures (Fig. 6). At the center of these structures is loose, frequently amorphous, sometimes fibrillar, pale eosinophilic tissue that