Cerebral Medulloepithelioma*

Case Report

RICHARD R. VAN EPPS, M.D., DENNIS R. SAMUELSON, M.D., AND WILLIAM F. MCCORMICK, M.D.

Division of Neuropathology, Department of Pathology, and Department of Neurology, University of Iowa College of Medicine, Iowa City, Iowa

We have recently studied a brain tumor that terminated with extracranial metastases which we consider a "pure" example of one of the rarest neuroectodermal tumors of man, a medulloepithelioma.

Case Report

A 5-year-old boy was admitted to the University of Iowa Hospital on March 28, 1966, because of headache, vomiting, and diplopia. He had been in good health until early that month when he began complaining of frontal headache. Two weeks before hospitalization he developed double vision.

Examination. The neurological examination was normal except for a right sixth nerve palsy and minimal truncal dystaxia. The optic fundi were normal. There was no evidence of systemic malignancy. Urinalysis, blood studies, intravenous urography, chest x-ray, and skin tests for histoplasmosis and tuberculosis were normal or negative. X-rays of the skull showed separation of the sutures and a normal sella turcica. An electroencephalogram showed excessive diffuse slow activity in the right occipital area. Ventriculography, angiography, and radioisotopic brain scan demonstrated a mass in the right temporal area.

Operation. A right temporal craniotomy was performed, exposing an area of fluctuation suggesting a cyst. This area was cannulated with a ventricular needle, and necrotic, liquefied tissue was removed. Subtotal resection of a large temporal lobe neoplasm was carried out; it was deep-seated, cystic, nodular, and relatively vascular. Its exact origin and relationship to the lateral ventricle was not established.

The child tolerated the operation well, and later received midline radiation totaling 4556 r through opposing lateral temporal fields. He was discharged on May 27, 1966, free of neurological signs.

Pathological Examination. The specimen removed at operation consisted of 14 gm of soft, yellow, tan, and pink tissue. Microscopically, the tumor had a marked papillary pattern with extensive tubule formation. The neoplasm did not appear to form glial fibers; the more compact, less papillary, tubular areas seemed to be differentiating into small neurons (Figs. 1 and 2). These tubules were lined by tall columnar cells with long elliptical nuclei (Fig. 3) and many mitoses (Fig. 4). There were no cilia nor blepharo-plasts. The Armed Forces Institute of Pathology and the U. S. Naval Hospital, San

Fig. 1. Photomicrograph of biopsy specimen showing a highly organized papillary neoplasm forming tubules. (H. & E., X 200).
Diego, confirmed a probable diagnosis of medulloepithelioma.

Postoperative Course. Careful search was made for a possible primary tumor elsewhere in the body, especially in the thyroid and kidney; no evidence was found.

The patient remained symptom-free for 2 months after completion of radiotherapy. Then, several small rubbery masses appeared around the craniotomy scar. Within another 2 months vomiting, headache, lethargy, and ataxia returned. The burr holes were bulging, and there was a left homonymous hemianopsia, early papilledema, and mild left facial paresis. Right carotid angiography and pneumoencephalography both demonstrated a large mass in the right cerebrum involving the temporal, posterofrontal, and parietal regions; the tumor was considered inoperable. The patient died on September 19, 1966, approximately 6 months after the onset of symptoms.

Autopsy Findings. A 2.5 cm mass of lymph nodes was found in the right postero-superior cervical region. These nodes were grossly necrotic, and their normal parenchyma had been largely replaced by neoplasm resembling the brain tumor removed several months earlier (Fig. 5). Several soft, well-circumscribed nodules were present in the craniotomy scar and attached to the periosteum of the skull. These nodules could be easily peeled from the skull and did not appear to invade bone. They proved to be neoplasm similar to that seen in the brain.