Cerebral Medulloepithelioma*

Case Report

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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 blepharoplasts. The Armed Forces Institute of Pathology and the U. S. Naval Hospital, San

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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

Fig. 2. Junction of papillary component with more compact area. Neuronal differentiation found in these compact areas. (H. & E., X200).

Fig. 3. The neoplastic tubule formation closely resembles the primitive neural tube. Many mitoses are present. (H & E, X400).
brain and lymph nodes. No other pertinent abnormalities were encountered.

*Gross Neuropathological Description.* There was an extensive, bosselated neoplasm involving the right ventral surface of the dura from the torcular Herophili rostrally for approximately 15 cm (Fig. 6) and medially to the superior sagittal sinus. It was varied in color from gray to pink and in consistency from firm and rubbery to soft and fluctuant. The individual nodules tended to coalesce to form an irregular sheet which interdigitated with depressions left in the cortical surface of the brain. The largest of these subdural masses measured 7 cm; the majority measured between 2 and 3 cm. A few were filled with translucent, gelatinous material but most were solid throughout and smooth-surfaced. The torcular Herophili and posterior third of the superior sagittal sinus were invaded, but the straight sinus was free.

The cerebral hemispheres were prominently asymmetrical and edematous, more markedly on the left. In the right temporal lobe there was an operative defect and yellowish-orange discoloration of the superior,
middle, and inferior gyri. The caudal portion of the orbital surface of the frontal lobe was displaced into the right middle fossa; the right uncus, hippocampus, and cingulate gyrus were all herniated. The third nerves were not necrotic or excessively angulated. The cerebellar tonsils were enlarged and showed no evidence of necrosis.

There was opacification, fibrosis, and foci of yellowish-orange discoloration of the leptomeninges over the right temporal lobe. There were multiple depressions in the cortical surface of the right cerebral hemisphere (Fig. 7). The cortical depressions extended from the intercerebral fissure over the convexity to the Sylvian fissure and reached a depth of 7 cm. Although the underlying cortex appeared intact, there was some focal disruption of the posterior parietal and occipital lobes and loss of arachnoid. There were several small depressed areas as far forward as the tip of the right frontal lobe. The pineal gland was not enlarged, but was shifted slightly to the left of the midline. There was no gross tumor invasion in the base of the multiple depressions present in the right frontal lobe, but the cortex was destroyed in the base of the depressions in the right parietal and occipital lobes. In one of these areas a neoplasm appeared to be within the parenchyma; it varied from gray to pinkish-red and contained multiple cysts filled with a gelatinous material. This area measured 2.6×2.4×1.8 cm. No neoplasm was present within the ventricular system or basal ganglion. The choroid plexi in the lateral and third ventricles appeared normal.

Microscopic findings\(^5,6\) showed the tumor to be a medulloepithelioma, which had extensively invaded the dura leptomeninges (Fig. 8). No residual neoplasm was found in the right temporal lobe, choroid plexi, or pineal body, but small foci were present in the right occipital lobe.

**Discussion**

Bailey and Cushing\(^1\) identified medullo-
epithelioma as follows: "... the tumor tends to grow in bands of cells, having a striking resemblance to the medullary plate. These bands of cells fold themselves here and there to form a sort of medullary tube, and in places they have a definite internal and external limiting membrane." Willis\(^9\) has pointed out that "... since a teratoma with abundant neural tissue could easily be mistaken for a 'neuroepithelioma' this diagnosis can be accepted unreservedly only if sections from all parts of the tumor show that it contains embryonic neuro-epithelial tissue only." We believe our case meets these criteria.

Since Bailey and Cushing's original description,\(^1,5\) the existence of this variety of tumor has been questioned by several authors.\(^10,12,14,15\) It was not included later in Cushing's own reports. Davie\(^4\) described one case of a cerebral medulloepithelioma; this tumor was subsequently re-examined by Russell,\(^13\) who called it a teratoma. Cox\(^3\) found one questionable example of this tumor in his series of 85 gliomas. However, he included it only in a table, without description. Greenfield\(^9\) reported two cases, which others\(^16,18\) have felt were a papillary tumor of the choroid plexus and a teratoma respectively, Mabon, et al.,\(^10\) reviewed four medullo-

epitheliomas classified before 1932 at the Mayo Clinic. They rediagnosed them as a pinealoma, medulloblastoma, and two ependymomas, and concluded that "the medulloepithelioma subgroup of glioma is not an entity."

Treip\(^16\) described a rapidly growing tumor arising from the midbrain of an 8-month-old infant; this case appears to be one of the few accepted examples, although Willis still holds that it was clearly teratomatous.\(^18\) Willis\(^19\) described briefly two cases which he felt were acceptable examples of medulloepithelioma. One was a large tumor in the floor of the third ventricle and pituitary region, invading the sphenoid bone in a 21/2-year-old girl. The other was a large well-circumscribed tumor of the cauda equina in a 19-month-old girl; a similar tumor was removed at reoperation 1 year later.

Thus, of the slightly more than a dozen cases of medulloepithelioma reported, only four or five seem to have a surviving claim to that diagnosis.\(^17\) If the requirement of Willis, that the tumor be free of any teratomatous component, be strictly adhered to, even the generally accepted case of Treip\(^16\) can be challenged.\(^18,19\)

We believe the extracranial spread of the

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**Fig. 8.** Cervical spinal cord showing ventral sulcus with extensive neoplastic infiltration of leptomeninges. This type of leptomeningeal spread was very extensive over the brain stem and cerebellum as well. (H & E, ×75).
medulloepithelioma in our case is particularly interesting since we can find no other example reported. We do not regard this as an indication of malignancy, but think it is more likely the result of a disrupted barrier. We have encountered several other examples in the last 2 years.7,8

Summary

We have described a rare case of cerebral medulloepithelioma which was highly malignant and which terminally had extracranial metastases.

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