Prolonged Survival with Extensive Astrocytoma*

Case Report

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Infiltration of the cerebrum, cerebellum, and brain stem by an astrocytoma unassociated with the stigmata of neurofibromatosis or "glioblastosis cerebri" is rare. We have recently studied the brain of such a patient who moreover survived for nearly 30 years after the onset of signs and symptoms and 12 years after subtotal resection of a cerebral astrocytoma.

Case Report

The patient, a right-handed woman, was first seen at the University of Iowa Hospitals in 1954 at 28 years of age. Her chief complaints at that time were uncontrollable seizures and mental deterioration.

History. She had been in good health until 1938 when, at 12 years, she began to have grand mal seizures. A year later she was admitted to another hospital because of persistent headaches and failing vision. Physical examination at that time revealed optic atrophy and marked loss of visual acuity in the right eye. Her left eye was normal, and no papilledema was present. The remainder of the neurological examination was normal. Skull x-rays were reported to have shown in increased convolutional markings and flattening of the posterior clinoids; this was not considered significant evidence of increased intracranial pressure, and the patient was discharged with no definite diagnosis.

A few months later she was admitted to the same hospital and a right temporal burr-hole and "drainage of a cyst" was done. (Several letters of inquiry to that institution have failed to clarify the nature of this lesion.) Following this procedure she was maintained on Dilantin and phenobarbital, but continued to have seizures about once weekly. In spite of her convulsive problem she was able to complete high school and 2 years of college with average grades. She never menstruated and her physical development was obese and juvenile. Her condition was stable until 1951, when mild dystaxia was noted. Two years later she gradually became more apathetic and careless about her personal appearance and the seizures became more frequent. In 1954 she was referred to the University of Iowa Hospitals.

Examination. On admission the right temporal trephination site was flat. There was a mild left hemiparesis but no sensory disturbances. Speech was slow but not dysphasic. Pupils were equal and reactive but vision was poor on the right, and the right optic disc was pale. There was no papilledema. The patient was dystaxic and walked with a broad-based gait. There was no nystagmus. Pelvic examination revealed an infantile uterus. A right carotid angiogram and ventriculogram showed a right temporal mass and a porencephalic cyst extending from the right temporal horn to the old burr-hole site.

Operation. A right frontotemporal craniotomy was performed, and an extensive, infiltrating, vascular tumor was subtotaly resected from the frontotemporal region. Residual tumor extended into the thalamus. The dura was closed and the bone flap replaced. The histological diagnosis on tissue removed was a well-differentiated astrocytoma. Postoperatively the patient had left hemiplegia and seizures continued at the same rate. No x-ray therapy was given. She was followed at regular intervals and her condition slowly deteriorated. She was put in an institution and became bedfast.

Second Admission. She was admitted again to the University Hospitals in December, 1966, with pneumonia. She was responsive only to noxious stimuli, and had bilateral optic atrophy but no papilledema. The craniotomy site was bulging. There was a spastic tetraplegia. Cerebellar function was

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impossible to evaluate. Her general condition deteriorated and she died on the third day.

Autopsy Findings. The pertinent findings were limited to the central nervous system. No extracranial neoplasm was found. Severe generalized osteoporosis and bilateral pulmonary edema and atelectasis completed the anatomical findings. The dural sinuses were patent. There was a 4-mm brownish-white, rubbery nodule on the right leaf of the dura adjacent to the superior sagittal sinus. This lesion had the gross appearance of a small meningioma.

The cerebral hemispheres were prominently asymmetrical due to a large area of encephalomalacia and tissue loss from the right hemisphere. The left cerebral hemisphere had a maximal rostral-caudal dimension of 18 cm whereas the right hemisphere was only 16.5 cm. There was an extensive area of yellowish-orange discoloration and thickening of the leptomeninges with marked loss of the subjacent parenchyma involving the inferior frontal and superior temporal gyri on the right, beginning 1.5 cm from the frontal tip and extending caudally for a distance of 9.5 cm. There was a defect in the inferior frontal gyrus which communicated with the lateral ventricle. This defect measured 2.6×2 cm. The right temporal lobe was expanded and measured 6.5 cm in width (compared to 5.5 cm on the left). The leptomeninges over the base were opacified and filled with a soft, grayish tissue which appeared to be neoplasm (Fig. 1). There was obscuration of the optic chiasma, the proximal portion of the optic nerves, and the optic tracts. The left olfactory bulb and tract were expanded and appeared to be infiltrated by neoplasm. The rostral brain stem was partially encased in this neoplastic infiltration, which was quite gelatinous and was thickest about the right cerebellopontine angle. There was slight hemorrhagic discoloration of the leptomeninges over the right cerebellar hemisphere. There was opacification of the leptomeninges over the cerebellar vermis posteriorly. Multiple plaques of rubbery, grayish-white tissue were present over the ventral surface of the right temporal lobe. The third and fourth cranial nerves were completely encased within the mass of neoplastic tissue.

Serial coronal sections of the cerebral hemispheres revealed an extensive, focally hemorrhagic and cystic neoplasm largely replacing the right temporal lobe (Fig. 2), encasing the optic chiasm, filling the basilar cisterns, and producing multiple discrete nodules on the ependymal surfaces of the lateral and third ventricles (Fig. 3). The third ventricle was largely replaced by neoplasm. This neoplasm varied from firm, glistening white, and almost cartilaginous tissue to very soft and mucinous. The lateral ventricles were greatly dilated and the right lateral ventricle communicated freely with the subarachnoid space through the defect noted on the uncut specimen. There were areas of hemorrhage up to 1 cm within the neoplasm in the right temporal lobe. The neoplasm extended into and markedly enlarged and distorted the mesencephalon. The various nuclear components of the basal ganglia on the left appeared well preserved; those on the right were largely effaced and replaced by the neoplasm. The neoplasm extended from the middle of the frontal lobe to the mid-parietal region, but did not grossly involve the occipital lobe of the rostral 5 cm of the frontal lobe. The nodules along the ependymal surfaces of the lateral ventricles varied up to 1 cm in size; some...