Oligodendroglioma with 35-Year Survival

LESLIE FREEMAN, M.D., AND IRWIN FEIGIN, M.D.
Veterans Administration Hospital, Lyons, New Jersey, and Department of Pathology,
New York University School of Medicine, New York, New York

This report serves to document the case of an individual with an oligodendroglioma thought to have persisted for 35 years, as indicated clinically by seizures of this duration. The oligodendroglioma is probably the least aggressive of the gliomas of the brain. Earnest et al.\(^1\) suggested that the average duration of life with an oligodendroglioma is 8 to 14 years, while Horrax and Wu\(^2\) considered 7 years to be the average, with 1 patient known to have survived for 33 years. Other reports, however, suggested that these tumors are more aggressive, as that of Shenkin et al.\(^3\) in which the average survival was recorded as 2 years.

Case Report

A 50-year-old white male, veteran of World War I and II, had a history of seizures for 35 years, beginning at the age of 24, which were relatively well controlled by luminal, and later on, when available, by Dilantin. He was first hospitalized in 1940 at age 38 for these seizures which were largely grand mal in type. Neurological examinations, including three spinal fluids during this 1940 hospitalization, revealed no abnormalities. However, roentgenogram of the skull showed extensive branch-like calcifications in the middle cranial fossa slightly anterior to and above the petrous portion of the left temporal bone, 3 cm. into the internal table. There was also a pineal displacement a ½ cm. to the opposite side. Encephalogram showed a 4 ½ cm. rounded calcified area in the region of the anterior and central portion of the left temporal lobe. Clinically the patient had had only four grand mal seizures in 2 years. However, in January 1941 he was hospitalized again for grand mal, petit mal seizures and dissociative states.

In 1942 he was inducted again into the army. Records show that he received treatment for grand and petit mal seizures. This state of affairs continued until 1948 when dissociative phenomena became worse with states of furor and psychotic behavior necessitating hospitalization. At this time electroencephalograms showed abnormal sleep records and spike focus in the left frontoparietal region. Neurosurgery in 1949 was unsuccessful because of excessive bleeding so that only a simple decompression was performed. In 1952 a left temporal craniotomy was performed and the calcified area first seen in 1940 was removed. The lesion extended to a depth of 5 cm. and surrounding brain substance showed the same tumor-like structure. The pathological diagnosis was oligodendroglioma (Dr. Arthur Wei).

From this time on until patient’s death from broncho-pneumonia in October 1961, he continued to have grand mal, petit mal and states of furor. At no time, however, were there any consistent positive neurological signs, or any signs of increased intracranial pressure in spite of many episodes of status epilepticus which were difficult to control by medication.

Autopsy. The significant changes were limited to the brain. The cerebral hemispheres were asymmetric, the left hemisphere measuring 8 cm. in width at a coronal level at which the right hemisphere measured 5 cm. in width. There was a large operative defect in the lateral aspect of the left temporal lobe over an area approximately 6 cm. anteroposteriorly and 3 cm. supero-inferiorly, leading into a cavity in the brain substance approximately 3.5 cm. deep. At the anterior margin of this defect, the tissues were abnormal, consisting of a gray somewhat soft neoplastic tissue (Fig. 1). This neoplasm spread over the inferior surface of the left temporal lobe and into the left Sylvian fissure, and large irregular soft masses of such tissue were present in the subarachnoid space at the base of the brain, overlying the left peduncle and portions of the cerebellum anteriorly. The gyri over the convexity were flattened.

On section of the brain, the tumor was seen to be larger than was apparent externally, measuring fully 7 cm. anteroposteriorly, 7 cm. mediolaterally and 6 cm. supero-inferiorly. Anteriorly, the tumor was uniform, homogeneously tan-gray in color, and was relatively sharply demarcated from the surrounding tissues. The latter appeared edematous. Posterolaterally, the tumor was lighter in color, and merged imperceptibly with the surrounding tissues. The tumor at the margins of the operative defect was discolored by blood pigments. The masses of tumor about the brain stem were homogeneous and light tan-gray in color and compressed the midbrain from the left. On palpation, fragments of calcific material were evident in various portions of the tumor. The left lateral ventricle was compressed, as was the third ventricle and aqueduct, while the right lateral ventricle was slightly dilated. Section of the cerebellum revealed a firm dense white zone posteriorly in the left hemisphere approximately 15 mm. in diameter, within which the folial markings were obliterated.

Microscopically, the tumor was found to be lightly cellular and, in many areas, highly vascular. The cells contained round or very slightly ovoid deeply chromatic nuclei which were essentially uniform in size, shape and staining characteristics (Fig. 2). The cytoplasm of most cells was unstained, and, in places, a distinct unstained perinuclear halo was sharply separated from the eosinophilic matrix present in small quantities among the cells, by a sharply defined, circular line of distinction. Some of the cells presented a pale pink cytoplasm immediately about the nucleus, the external margins of which were circular and sharply defined. The cells were arranged in broad sheets without distinguishing order. There were no mitoses and no zones of necrosis. Foci of calcification were present in many areas. The vascularity was very high in some areas, slight in others. The highly vascular

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zones were those which appeared dark on gross examination. The vessels revealed considerable endothelial hyperplasia. A few small cysts were noted in areas, these being filled with an eosinophilic material. Spread of the neoplastic tissue in the leptomeninges is clearly evident in some sections.

The cerebellar lesion was characterized by a loss of neurons in the granular and Purkinje layers, by marked increase in Bergman astrocytes and some rarefaction of the subjacent white matter.

Discussion

The assumption that this tumor had persisted for 35 years is based on the clinical observation of seizures starting at the age of 24, and persisting with varying degrees of severity for the 35 years that this individual lived thereafter. Seizures starting at this age probably are observed most commonly in the presence of a brain tumor. This impression is buttressed by the demonstration 14 years later, 21 years before death, of calcium deposits in the area in which the tumor was found subsequently. It is well known that the oligodendrogloma, in proportion to its incidence, is more likely to be calcified than any other glioma. The histologic demonstration of the oligodendrogloma by biopsy 9 years prior to death, and the subsequent confirmation at autopsy, complete the chain of evidence on which the assumption of long survival is based.

This case is somewhat remarkable also because of the almost complete absence of neurological symptoms and signs in spite of the large size of the tumor which involved most of the left temporal lobe, Sylvian fissure and parts of the brain stem. Of particular interest was the absence of any signs of increased intracranial pressure, even terminally. Much of this apparent paradox may be explained by the slow growth of the tumor which gave the brain tissue a chance to adjust to the changes even though the midbrain and ventricles were compressed.

However, when we reexamine what passed for psychogenically determined delusions and halluci-

Fig. 1. Coronal section of brain. The neoplasm is in the left temporal lobe. X0.4.

Fig. 2. Photomicrograph of tumor. The histologic characteristics are those of a well differentiated oligodendrogloma. Hematoxylin and eosin, X300.