Oligodendroglioma with 35-Year Survival

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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. suggested that the average duration of life with an oligodendroglioma is 8 to 14 years, while Horrax and Wu considered 7 years to be the average, with 1 patient known to have survived for 35 years. Other reports, however, suggested that these tumors are more aggressive, as that of Shenkin et al. 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 4 1/2 cm. to the opposite side. Encephalogram showed a 4 1/2 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 Weil).

From this time on until patient's death from bronchopneumonia 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 3.3 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. medially 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. Posteriorly, 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 oligodendroglioma, in proportion to its incidence, is more likely to be calcified than any other glioma. The histologic demonstration of the oligodendroglioma 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 hallucinations...
nations throughout this patient's history with the hindsight of the neuropathological findings, we may now consider certain other symptoms as having been produced organically. This patient experienced both illusions and hallucinations of olfactory sensation (peculiar and often foul odors) which we may explain by involvement of the temporal lobe. At other times he complained of strange somatic sensations, bloating and gas in the upper abdomen, for example. Such sensations are known to occur with lesions of temporal lobe, Sylvian region and insula.2, 4 Finally, this patient suffered from defects of memory such as loss of recent memory, but more frequently from falsification of memory including "déjà vu" phenomena. Here again there is good evidence, as in the reports of Penfield and Roberts,4 and others, that involvement of the temporal lobe is responsible.

The tumor tissue at autopsy, and that at operation 9 years prior to death, are typical of the oligodendroglioma in all respects. There is no evidence of atypism or anaplasia in either of these tissues, such as might be interpreted as indicating more rapid growth. It has been our experience that anaplastic areas in oligodendrogliomatous tissues do indicate more rapid growth, but to a lesser degree than is true of such changes in any other brain tumor. The vascularity of the tumor varied, and it was apparent that the dark tan-gray color which may be noted in some oligodendrogliomatous tissues, was related to the high vascularity of these tissues. Those portions of the tumor in which the vessels were much less numerous, were lighter in color, and more difficult to distinguish grossly from an astrocytoma.

There were no areas in the tumor in which the appearance of the tissues with ordinary stains would lead us to suspect the presence of some other type of neoplastic tissue. It will be recalled that in many instances, oligodendrogliomatous tissues may be present in the same mass of tumor in which astrocytomatus or ependymomatous tissues may be recognized. This is a matter of considerable importance, since in such mixed tumors, the evolution of the tumor, and the clinical prognosis, may reflect the more aggressive character of these other neoplastic tissues. Indeed, it is possible that in some instances in which an oligodendroglioma appears to have grown quite rapidly, and in which the tumor has assumed the gross and, in some respects, the microscopic appearance of a glioblastoma multiforme, these features may reflect the overgrowth of astrocytomatus tissues present in addition. If this be true, the principle that oligodendrogliomatous tissues grow slowly would remain valid.

Leptomeningeal spread, quite common with the oligodendroglioma, was present in this instance as well. Though such leptomeningeal spread in other instances may be so massive as to mask the primary lesion, in this case this spread was relatively slight, except in areas in direct continuity with the primary tumor.

Summary

A case is reported in which an oligodendroglioma arising in the left temporal lobe of a 24-year-old man, grew slowly until death, 35 years later. The clinical and pathologic features are summarized.

References