MULTIPLE OCCURRENCES OF GLIOMAS IN A FAMILY

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Intracranial tumors are said to account for over 1 per cent of all deaths. However, the occurrence of gliomas in several members of one family has rarely been recorded. It is our purpose to report the occurrence of 2 verified gliomas and 1 presumed glioma in 2 non-identical siblings and a nephew.

Koch described similar multiplicity in two German families. In one of his families gliomas (glioblastomas) developed in a mother, daughter and son. The Schie family included A. with a proven left temporal astrocytoma. An uncle of A. had a left posterior cerebral tumor without microscopic identification. This patient expired with evidence of intracranial hypertension at the age of 17 years after morbidity of 1 year. Another uncle of A. died at the age of 65 years following a history of tumor with terminal intracranial hypertension. The lesion was presumed to be a right cerebral tumor but was not confirmed.

Riese, Meredith and Zfass reported the development of gliomas in a brother and sister at about the same age (55 years); these tumors were similar histologically. In Leavitt’s report of cerebellar tumors occurring in identical twins, one was proven to be a medulloblastoma and the other had caused similar clinical findings. Bing reported the occurrence of cerebral neoplasms in 3 members of a family without other data. Klemme has identified gliomas in 2 members of a family. Astrocytomas in 2 members of a family were treated at the New York Neurological Institute during the experience of one of the authors (A.H.H.).

CASE REPORTS

Case 1. W.L., a 52-year-old white male, was admitted to Johns Hopkins Hospital in July, 1936, under the care of Dr. Walter Dandy. Approximately 2 months previously he had had slurring of speech and for the last 2 weeks he had had right facial numbness. Twitching attacks followed by transient aphasia had occurred. No headaches were noted.

Examination. The patient was right-handed, with right facial hypesthesia and paresis, slurring of speech and absent abdominal and cremasteric reflexes.

Operation. A left craniotomy was performed by Dr. Dandy, whose operative record states: “In Broca’s area there was an infiltrating tumor which had produced reddening of the surface of the convolutions and also widening and flattening of them. This region was quite hard. It was in such a position that its removal would have almost certainly caused motor aphasia, although this is sure to come with the progress of the tumor.” A small piece of tissue just anterior to Broca’s area was removed for diagnosis, and a large temporal decompression was performed.

Course. Death occurred soon after.

Pathological Diagnosis. The surgical specimen contained no neoplastic cells. Although lacking histological verification the tumor was identified as a glioma by Dr. Dandy.

Case 2. E.L., a 64-year-old white male and brother of Case 1, was admitted to the Nix

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Memorial Hospital in San Antonio, Texas, on Sept. 11, 1952. Eight weeks previously he had suffered a sudden coma of 6 hours' duration with residual, rapidly receding, hemiparesis. After a 4-week interval of relatively good health there was onset of progressive aphasia, confusion, vomiting and headache.

**Examination.** He was a right-handed man who was overreacting and jocose. He had a right hemiparesis, sensory aphasia, astereognosis of the right hand and blurred disc margins.

Roentgenograms of the skull were normal. Electroencephalograms discovered a left temporoparietal focus. Initial pressure on lumbar puncture was 296 mm.; the fluid contained 61 mg. per cent total protein.

**Course.** The patient refused a craniotomy and expired on Oct. 13, 1952.

**Autopsy.** A large infiltrating left frontal tumor, 6.5 X 6 cm. in size, was encountered. Dr. J. W. Kernohan's histological diagnosis was Grade III astrocytoma (Figs. 1 and 2).

**Case 3.** A.L., a 46-year-old paternally related nephew of Cases 1 and 2, entered a hospital in Phoenix, Arizona in October, 1950, under the care of Dr. John Green. He presented a 3-month history of personality change, headaches and left-sided paresthesias.

**Examination.** The patient was right-handed, with definite evidence of increased intracranial pressure, left astereognosis and dysesthesias.

Spinal fluid pressure and protein were elevated. Ventriculography suggested a right intraventricular tumor.

**Operation.** At craniotomy a right paraventricular tumor was encountered and grossly removed.

**Course.** The patient expired after 2 months' hospitalization.

**Histological Diagnosis.** The tumor was a malignant glioma varying from a protoplasmic astrocytoma to a glioblastoma multiforme in character (Figs. 3 and 4).

**DISCUSSION**

Though the nervous system is host to many hereditofamilial disease processes, it is generally held that no genetic influence on gliomas exists in the human family. The familial nature of various degenerative diseases and certain neoplastic involvements is well recognized. Generalized neurofibromatosis, multiple enchondromata, von Hippel-Lindau disease, and glioma (more properly neuroepithelioma) of the retina show hereditary influence. Bailey (page 6) made the statement, “No glioma of the brain has even been supposed to be hereditary. . . .”

Brain tumors are common, with figures showing that they make up to 1.8 per cent of all tumors and account for 1 per cent of all deaths. Gliomas comprise some 40 per cent of brain tumors. With such a moderately common affliction multiple occurrences of gliomas among members of the same family should appear occasionally by coincidence. However, the occurrence of 2 proven gliomas and 1 presumed glioma in two generations of the family reported herein is a distinctly unusual coincidence. The generation in which 2 gliomas occurred included a total of 6 full siblings and a paternally related half sibling. The generation to which Case 3 belongs includes many members with the majority under the age at which most gliomas occur.

It is our belief that cases of multiple incidence in a family should be reported in order to contribute, if possible, to understanding of the pathogenesis of this distressing condition. It is our impression, also, that if all such incidences were reported the frequency might prove to be more than coincidence. Koch, for instance, presented evidence that led him to believe that there is an hereditary influence on a certain group of gliomas. We have cited, in addition to the cases presented, two other instances of multiple gliomas in a family which have not reached the neurological literature. There may be many more unreported, and the total may be significant of more than coincidence.
Fig. 1. Case 2. Photomicrograph of astrocytoma (grade III).

Fig. 2. Case 2. High power view of tumor in Fig. 1.
Fig. 3. Case 3. Photomicrograph of malignant glioma varying between protoplasmic astrocytoma and glioblastoma multiforme.

Fig. 4. Case 3. Higher magnification of tumor in Fig. 3.
SUMMARY

1. Three cases of gliomas in two generations of a family are described.
2. References to the few accounts of similar multiplicity are cited.
3. Reporting of such multiplicity is suggested as a possible contribution to understanding the pathogenesis of gliomas of the brain.

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REFERENCES