Familial oligodendroglioma

Case report

NORBERT ROOSEN, M.D., CHRISTIAN DE LA PORTE, M.D., MICHEL VAN VYVE, M.D., CLAUDE SOLHEID, M.D., AND PAUL SELOSSE, M.D.

Department of Neurosurgery, University Hospital and Department of Neuropathology, Born-Bunge Foundation, University of Antwerp, Antwerp, Belgium

A case of familial oligodendroglioma, occurring in a mother and her daughter, is presented.

KEY WORDS • oligodendroglioma • glioma • familial brain tumor • genetics • congenital abnormality

In contrast to astrocytomas and glioblastomas, which have occasionally been reported as occurring in members of one family, the familial appearance of oligodendrogliomas has only been described once. We present two patients, mother and daughter, both of whom were operated on for an oligodendroglioma.

Case Reports

Case 1

This 37-year-old mother was admitted for evaluation of grand mal epilepsy. The neurological examination was normal. A right posterofrontal focus was noted in the electroencephalogram, and right carotid angiography demonstrated an avascular space-occupying lesion. A right frontal lobectomy was performed, and the tumor was excised. Histopathological examination revealed an oligodendroglioma with a homogeneous parenchyma and microcysts (Fig. 1). There were no mitoses or neovascularization. Nine years after the operation the tumor recurred, and the patient was submitted to a second operation. The tumor again proved to be an oligodendroglioma with essentially the same features as shown by the previous specimen.

Case 2

The 22-year-old daughter of the patient in Case 1 came under medical attention because of a grand mal seizure. A right parietal hypodense lesion was visualized on computerized tomography (CT) scanning. The patient was treated expectantly because of a lack of mass effect, a normal neurological examination, and control of her seizures with adequate antiepileptic therapy. Three years later, however, a slight left hemiparesis developed during pregnancy, and the epilepsy became resistant to therapy. Angiography and CT scanning confirmed progression of the tumor. A large biopsy of the tumor was performed. An oligodendroglioma with a metachromatic parenchyma, numerous microcysts, and a homogeneous isomorphous cellularity was diagnosed on microscopic study (Fig. 2). The tumor showed no mitoses or pathological hypervascularization.

Discussion

The familial occurrence of gliomas is a well known phenomenon in cases of von Recklinghausen's disease, tuberous sclerosis, and Turcot's syndrome, but has also been observed with no association to diseases. These were almost always astrocytomas or glioblastomas. We have found only one report of familial oligodendrogliomas: in 1962, Parkinson and Hall described two brothers, each of whom had a right frontal oligodendroglioma.

Studies have been carried out to determine if some increased predisposition to develop a glioma exists in close relatives of patients with astrocytoma and glioblastoma. Van der Wiel investigated the families of 100 glioma patients and found that the mortality resulting from glioma was four times higher in comparison with a control group. Metzel and Mohadjer found a prevalence of gliomas in the families of glioma patients 10 times higher than in the general population. However, in contrast to these results, Hauge and Harvold were unable to collect any evidence for a
on a genetic mechanism, but we may conclude that in certain families there may be an increased predisposition in close relatives to develop oligodendrogliomas. Further case reports will be of great value to show if this is true.

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References


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Address reprint requests to: Christian De La Porte, M.D., Dienst voor Neurochirurgie, Akademisch Ziekenhuis der Universiteit, Wilrijkstraat 10, B-2520 Antwerpen-Edegem, Belgium.