FAMILIAL ASTROCYTOMA

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SUMMARY :

Familial astrocytoma occurring in two siblings at similar sites is presented. Documentation of such cases is important for determining their incidence and exploring possible genetic factors.

KEY WORDS :

Astrocytoma, Familial brain tumor, genetics.

INTRODUCTION

Brain tumours rarely occur in more than one member of a family (16,17). The genetic basis of glial tumour occurrence in association with hereditary syndromes such as neurofibromatosis, intestinal polyposis, tuberous sclerosis and von Hippel-Lindau is well understood (15). Aside from the states mentioned above, the role of genetic factors in the development of gliomas is controversial.

In an attempt to draw further attention to the subject, we present in this report, two siblings with familial astrocytoma occurring at similar sites.

CASE REPORTS

CASE I: A 21-year-old male patient was admitted to Gülhane Military Hospital, Dept. of Neurosurgery, after he experienced epileptic seizures. A right frontotemporal tumour was diagnosed and removed by subtotal resection in Fabruary 1980. The histopathological examination revealed astrocytoma grade II (figure 1). Radiotherapy was done. Although he was on anti-epileptic therapy, three years later, the patient's epileptic seizures recurred. On physical examination left hemiparesis was found. He was advised for a second operation which he did not accept.

CASE II: A 31-year-old male who is the older brother of case I, was admitted to clinic with a history of headache lasting for a year and left sided weakness which had been present for three months. Neurological examination revealed bilateral papilledema and left hemiparesis including the face. The CAT-scan demonstrated right frontotemporal mass and in November 1986, a subtotal resection was performed. Histopathological diagnosis was astrocytoma grade II. (figure 2).

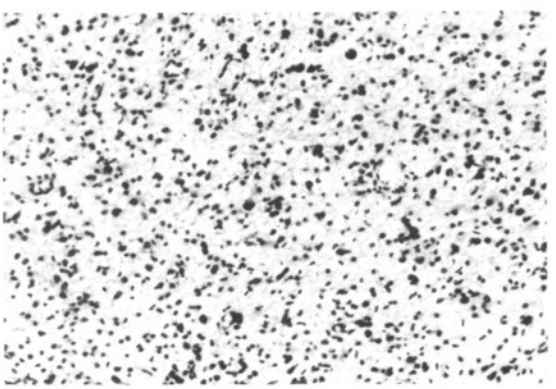

Fig. 1 Astrocytes containing hyperchromatic nucleus and pleomorphism (HE,X200).

There were two more healty siblings in the family and no cases in the pedigree going back two generations.

DISCUSSION

It has been accepted that, expcept for phacomatosis, genetic factors have no role in the development of gliomas (6,10). This consideration was based on the study conducted by Hauge and Harward (6) in which they compared members of 300 families in which gliomas, were detected and 250 families

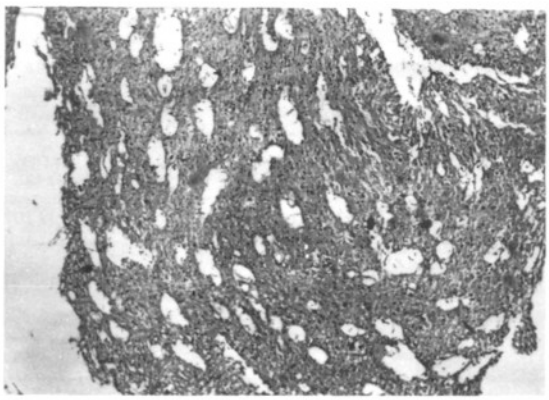

Fig. 2 Photomicrograph of astrocytes demonstrating hyperchromatic nucleus and diffuse microcytic degeneration zones (HE,X40).

with no members affected by gliomas, and no difference was found between the two groups.

In a later study conducted by Van der Weil (17), as glioma mortality was found to be four times greater than would be expected, attention has been directed toward this subject once more.

As the importance of precipitating factors in the genetic transmission of tumours is emphasized, familial occurrence is explained in one of three ways (4):

1. Autosomal transmission

2. Multifactorial transmission (two or more genetic and enviromental factors acting separately or together)

3. Transmission by chance

However, in chromosomal studies conducted in leukaemia, lymphoma and some carcinomas, chromosomal anomalies have been demonstrated by karyotyping and high resolution band techniques. But in studies, on gliomas a conclusion has not yet been reached and biochemical and immunogenetic studies are still being carried out (2,3). In spite of this, there are observations which suggest that genetic factors have a role (12):

1. Glioma cases reported in identical twins,

2. Its occurrence in association with familial diseases which show autosomally dominant transmission such as intestinal polyposis (Turcot Syndrome), neurofibromatosis, tuberous sclerosis and nevoid basal cell carcinoma,

3. The similarity of the histological characteristics of glial tumours occurring in a family and,

4. The occurrence of gliomas at similar sites and showing the same histological characteristics in siblings at similar ages (13).

Until today, along with the conditions mentioned above, hepatic nodular hyperplasia, xeroderma pigmentosum, presacral lipoma, atypical aplastic anaemia and intrinsic cellular immune deficiency have been reported to occur in association with familial gliomas (4,5). Among the types of glioma which have been reported are glioblastoma multiforme (5,8,12), astrocytoma (1,8,12), oligodendroglioma (13,14,16) oligodendrocytic-astrocytic mixed glioma (9,16), medulloblastoma (16) and pineoblastoma (11).

Although the difficulties of constructing a pedigree in this country are known, investigating the patients's relatives going at least one generation back and conducting studies in collaboration with the genetic departmens will be helpful in further classification of this condition. In 1983, in the Netherlands an international centre where familial glioma cases are being reported, was established and with the collection of cases and progress in tumoural genetic studies it is expected that the understanding of the aetiological factors in this group of cerebral neoplasms will become easier.

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