

FAMILIAL GLIOMA

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Much remains to be learnt about the genetic process of carcinogenesis and about the environmental factors that can alter DNA and thus lead to malignancy. New insights in the fundamental role of DNA changes in carcinogenesis will lead to improved and more specific ways of prevention and treatment of malignant disease in the entire body.

Case Report

A 8-year-old girl was admitted with history of left sided focal seizures and headaches for 6 months. On examination, she was conscious with normal vision, but had papilledema, left upper motor neurone facial weakness and mild left hemiparesis. CT scan showed a ring enhancing lesion with mural nodule in the right fronto-parietal region (*Fig. 1*) she underwent craniotomy and intra-tumoral decompression. The histology showed astrocytoma grade III. Her chromosomal analysis did not show any abnormality. She was yet to complete her radio-

therapy when last seen 2 weeks since operation.

Her family history had revealed that her elder brother had, at the age of 7 years, undergone cranial surgery in another city two years ago. He had complained of headaches and failing vision for 3 months. The CT scan, had shown a parasagittal, enhancing, globular lesion (*Fig. 2*). The histology was glioblastoma. In spite of radiotherapy, he expired 4 months later. The children were born of a nonconsanguineous marriage, and had an uneventful antenatal period. The family tree was reconstructed (*Fig. 3*). The third brother 5 years old underwent CT scanning (plain and contrast) which fortunately did not show any tumour.

Discussion

Genetic studies of gliomas have been directed in 3 ways-(a) defined genetic syndromes(1,2); (b) familial aggregation of gliomas; and (c) analysis of chromosomes or DNA from sporadic gliomas using standard metaphase banding technique. Of the numerous karyotypic abnormalities, increase or decrease of chromosome 7 is the most common(3,4). A report in 1947 concluded that there is difference in the incidence of intracranial neoplasms between the relatives of normal families(5). Till 1982 there were 21 reports of verified gliomas in the two generations and two reports in three generations(6). In 1984, four other families were reported(7). The maximum number of relatives affected was ten. These tumors occur predominantly in the frontal and temporal regions and are frequently deep seated. High grade astrocytomas and glioblastomas account for the majority. In relatives of patients with glioma, the possibility of familial occurrence is 7.9%; this occurrence is more frequent than that expected on a

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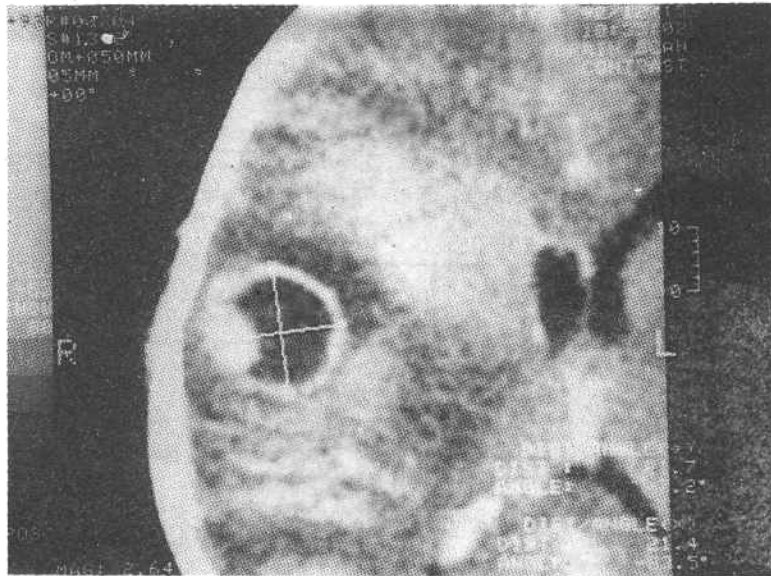


Fig. 1. Post-contrast CT scan showing "ring enhancing lesion" with mural nodule in the right frontoparietal region.

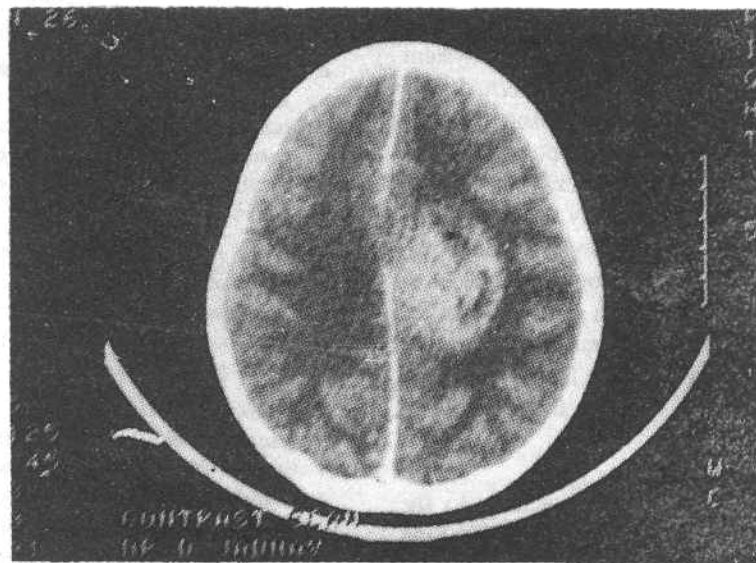


Fig. 2. Post-contrast CT scan showing parasagittal, enhancing, globular lesion.

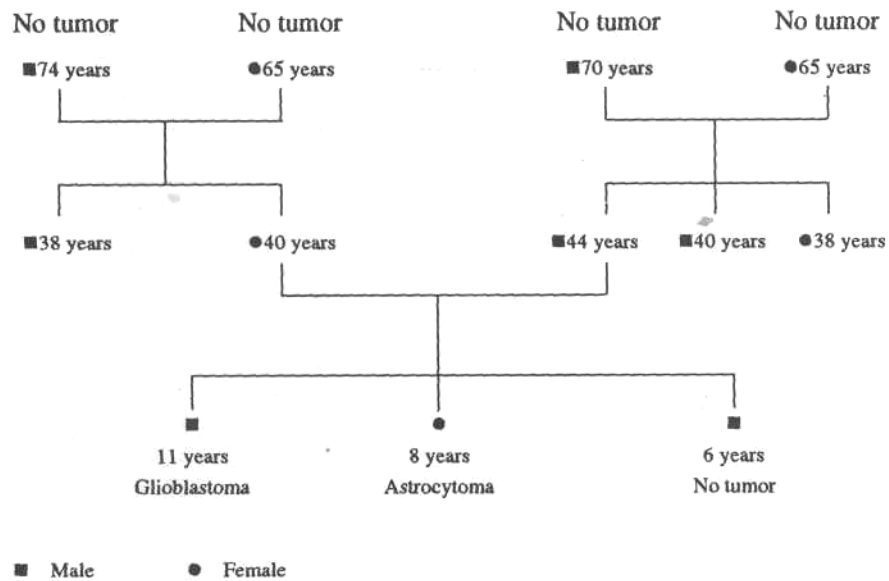


Fig. 3. Family tree-traced back for two generations.

change basis. The risk of occurrence of gliomas in two siblings on the basis of chance alone varies from 0.8×10^5 in a sibship of two of 36×10^3 sibship of ten(6). More than 5% of central nervous system tumors are genetic but when inherited may well be transmitted as an autosomal dominant trait although polygenetic inheritance cannot be excluded. However, some studies have failed to show any chromosomal abnormality(8), and "endogenous factors" have been postulated by some authors. A report in 1989 discussed the genetic influence in the etiology of gliomas(9). The first report from India has been from Hyderabad in 1992(10) and this may be the second.

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FAMILIAL ADENOMATOUS POLYPOSIS COLI

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Familial adenomatous polyposis coli is genetically transmitted disease, as a Mendelian dominant trait with high degree of penetrance, characterized by multiple

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adenomatous polyps in colon and rectum. Less than 12% of these patients are prone to develop adeno-carcinoma upto 20 years of the age in the rectal stump after the ileo rectal anastomosis(1,2). The youngest patient encountered in literature was 8 years old(3). The best way of screening them is by mapping the long arm of chromosome number 5(4).

Coffey(5) was the first person to perform the total proctocolectomy and ileostomy for the treatment of this disease while Ravitch *et al.*(6) performed ileoanal anastomosis combined with total proctocolectomy.

The other mode of treating these patients are subtotal colectomy with ileorectal anastomosis alongwith regular follow up with proctoscopy for development of cancer in retained rectal mucosa(3,7). Recently, Stevan *et al.* (8) utilized laproscope for total proctocolectomy as well as ileoanal anastomosis while others performed ileopouch anal anastomosis alongwith total proctocolectomy(9,10). Thus the ideal op-