Brief Reports

Anterior Visual Pathway Tumors

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Anterior visual pathway tumors are rare in children and constitute less than 2% of all intracranial tumors(l-5) and 7-8% of all intracranial tumors in children(6-8). Only a few large series, and none from India have been published. We report 35 such patients admitted over a 10 yr period.

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Subjects and Methods

Thirty five children below the age of 15 yrs, diagnosed to have anterior visual pathway glioma, between 1982-1992, were retrospectively analyzed. During the same period, 5 adults were also treated for the same condition. In all the patients contrast enhanced CT scans were performed; in 5 patients MRI scans were also performed. All the subjects were subjected to surgery and 29 patients received postoperative radiotherapy. Postoperatively, all the patients were continued on anticonvulsant therapy and followed up.

Results

There are no significant difference in sex distribution. Twenty seven children were under 10 yr of age and 12 were younger than 5 yr. The mean age was 7.6 yr; for optic nerve glioma and chiasmal glioma the means ages were 6.5 yr and 11 yr, respectively.

Visual problems were the most frequent symptoms, recorded in 94.3%, followed by headache in 57.1% patients. Hypothalamic dysfunction and proptosis was recorded roughly in a third of the patients. Generalized seizures were seen only in 8.5% cases. Field defects were observed in a fifth of the subjects. Fundus examination revealed unilateral or bilateral primary optic atrophy in 64.6% patients and papilledema in 31.4%. Fifteen per cent patients were admitted in a state of altered sensorium. Six cases had associated neurofibromatosis type 1.

Radiological work-up showed that the sella turcica was enlarged and/or eroded in 22 patients. Suprasellar calcification was noticed in only four children. Optic canal view showed enlarged optic canal in 13 patients. Contrast enhanced CT scan showed a uniformly high attenuating orbital *(Fig.* 2) or suprasellar mass (Fig. 2) in 27 patients and mixed attenuating lesions in 8 patients. CT scan evidence of calcification was noticed in 6 and moderate to severe hydrocephalus *(Fig. 2)* in 13 patients.

All patients with hydrocephalus required a ventriculoperitoneal shunt. Patients were subjected to surgery through a right frontal craniotomy. In patients with optic nerve tumor, total excision was possible in 6 patients with optic nerve glioma. Rest 20 patients had partial excision or biopsy. In patients with chiasmal glioma, only partial excision was possible.All the patients received short term course of steroids and long term anticonvulsant therapy postoperatively. There was no operative mortality.

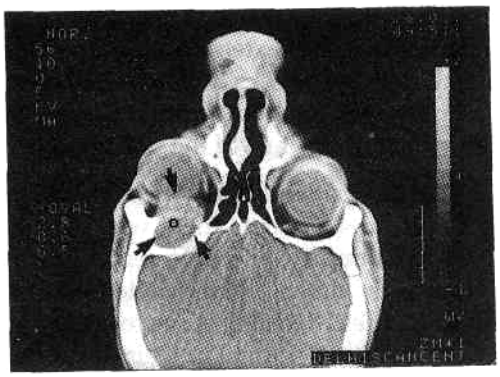

Fig. 1. Contrast enhanced CT scan (coronal cut) in a patient with optic nerve glioma shows high attenuating tumor behind the eye ball (arrows).

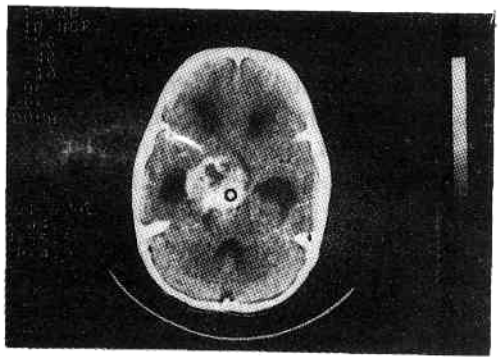

Fig. 2. *Contrast enhanced CT scan of a patient with chiasmal glioma shows high attenuating suprasellar tumor extending into the third ventricle with moderate hydrocephalus*

Histological examination revealed astrocytoma in 32 patients and mixed oligoastrocytoma in 3 patients. Excluding 6 patients with optic nerve glioma, who had total tumor excision, rest 29 patients

were subjected to external radiation using Cobalt 60. 3-6 weeks following surgery. The dosage ranged from 30-40 Gy, depending on the age of the patients.

Eighteen patients had a mean follow up of 1.5 yr. In 4 patients there was progressive visual improvement. In remaining 14, vision was either static or deteriorated. Eight of the 18 children were attending school and were normal in their day to day activity. Three patients died during follow up period. One patient expired following status epilepticus, and two other as a result of meningitis several months following surgery.

Discussion

Optic nerve glioma is exclusively a tumor of the pediatric age group. However, optochiasmal and chiasmal glioma do occur in adults. A high incidence of neurofibromatosis is associated with these tumors(l), and in the present study 17.2% children had neurofibromatosis type I. Unilateral or bilateral visual deterioration is seen in 90% patients(l,3); 94.3% of our patients had visual problems at presentation. In optic nerve glioma proptosis is a frequent finding, which was recorded in 30% of our patients.

Hydrocephalus is rare in anterior visual pathway tumors(3-5) and occurs due to obstruction of cerebrospinal fluid flow as a result of tumor extension into the third ventricle and blocking of the foramen of Monro. Fletcher *et al*.(3) reported hydrocephalus in 4 of their 22 patients with chiasmal glioma. In our study, 13 patients with chiasmal glioma had hydrocephalus.

Optic nerve tumors are usually low grade gliomas(l,3,5). The ideal treatment is total excision, because it not only provides a good long term result, but also avoids radiation in young children. On the other hand, total excision has a significant risk of visual deterioration in patients with good visual acuity (7-9). Total excision of a chiasmal glioma is not possible when the tumor is invading the hypothalamus and the third ventricle. Hence, in chiasmal glioma a conservative surgical approach is advocated(l,7,9,10). Partial excision followed by radiation is the mainstay of treatment in such cases(4,5,9).

Optic nerve glioma, on other hand is amenable to total surgical excision, when confined to orbit(l,5,6,8,ll). For optic nerve tumors infiltrating the chiasma, a partial excision followed by radiation is the accepted approach(l,2,5,6,8). Overall, long term prognosis is much better in patients with optic nerve glioma, as compared to chiasmal glioma.

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