GLIOMAS OF THE OPTIC PATHWAYS IN CHILDHOOD

FRED D. FOWLER, M.D., AND DONALD D. MATSON, M.D.

Neurosurgical Service, Children's Medical Center, and the Department of Surgery, Harvard Medical School, Boston, Massachusetts

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A small series of cases of optic pathway gliomas is being reported for two reasons: first, because their proper treatment is still a controversial subject and second, because of the importance of early recognition in permitting possible surgical cure instead of palliation. Gliomas of the optic pathways make up a relatively small portion of neoplasms of the brain. Martin and Cushing reported an incidence of 0.84 per cent in a series of 826 intracranial tumors. Taveras et al. found 34 cases in a series of 2000 gliomas (1.7 per cent). If the various reported series of optic pathway gliomas are inspected, it is found that approximately 75 per cent of these lesions occur in children under the age of 12. In our clinic 13 cases of primary tumor of the optic pathways have been encountered among a series of 256 intracranial tumors in young children during the past 10 years. This is an incidence of 5.1 per cent in the total series and 7.0 per cent if only the gliomas are considered.

Much emphasis has been placed on the fact that these tumors have only a slight malignant potentiality. Hudson first pointed out that even after incomplete removal, patients may survive for 20 years or more without evidence of progression of the tumor. This has been confirmed by the experience of others. McFarland and Eisenbeiss have even stated that if such a tumor is only partially removed it probably will not continue to grow. As recently as 1954, Marshall said, “On theoretical grounds complete excision might be desirable, but recurrence after excision of the major portion is very uncommon.” Many of these statements have been made by ophthalmologists on the basis of an anterior exploration of the orbit, an approach that makes it difficult to visualize the limits of the tumor. Also, since it has been well established that many of these gliomas grow very slowly, the occurrence of intracranial symptoms 10–15 years after the original operation might not come to the attention of the original surgeon. Posner and Horrax have pointed out that even after known intracranial extension, these patients may survive for a long time, their only symptom being loss of vision.

Analysis of the 13 patients seen at the Children's Medical Center in the past 10 years with a verified diagnosis of optic pathway glioma reveals that on the basis of symptomatology and physical findings they fall into two groups, depending on whether the tumor is predominantly intraorbital or intracranial. This differentiation was first proposed by Dandy but has not been used by many subsequent authors. The following case histories illustrate some of the important differences between these two groups.
Intraorbital Glioma

Case 1. M.C., a 4\(\frac{1}{2}\)-year-old girl, was admitted to the Children's Medical Center because of increasing prominence of the left eye of 2 weeks' duration (Fig. 1). She had had no headaches and had noted no change in her visual acuity. Although there was some increased resistance to palpation of the globe, no tumor mass was palpable. The movement of the eye was unimpaired. Early papilledema was present in the left eye; the right fundus appeared normal. Visual acuity was diminished to 20/70 in the left eye and was normal in the right. Slight peripheral constriction was found in the visual field of the left eye, but the patient could not cooperate for detailed examination. There were no other abnormal findings and no evidence of von Recklinghausen's disease.

Roentgenograms showed the left orbit to be slightly larger than the right, but the optic foramen was normal. Films of the skull were otherwise negative. A left carotid arteriogram showed no evidence of displacement of the intracranial vessels and no abnormality of the orbital vascular pattern.

A left transfrontal craniotomy was performed. On exploring the orbit, a diffuse reddish swelling of the optic nerve was found. Further exploration demonstrated that this tumor lay entirely within the orbit. The nerve was first amputated from the globe. Intradural exploration of the chiasm was then performed and failed to show any abnormality of the intracranial portion of the optic nerve. The nerve was divided just in front of the chiasm, pulled through into the orbit, and removed. Since subsequent pathological examination showed that the tumor extended to the globe, the eye was enucleated 16 days later and the patient was fitted with a prosthesis.

Histologically, the resection extended 1 cm. behind the most proximal edge of the tumor and thus the neoplasm was considered to have been completely removed. The microscopic picture of this lesion was characteristic of an optic pathway glioma (Fig. 2). There was a matrix of fine neuropil fibrils in interspaces of a delicate reticulated syncitium. The tumor cells were small with oval or elongated nuclei, some of which were darkly stained. Many of the cells showed vacuolization of the cytoplasm.

It is now 8 months since operation and the patient is asymptomatic (Fig. 3).

Intracranial Glioma

Case 2. A.P., a 7-year-old child, had a history of a squint of her left eye which