Malignant optic glioma in adults

Case report

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Malignant optic glioma in adulthood is a rare tumor that causes early loss of vision and nearly always leads to death within a year. A case history is presented illustrating the clinical and neuroradiological characteristics of the malignant optic glioma in adults. A review of the literature is given.

KEY WORDS • brain neoplasm • optic glioma • astrocytoma

OPTIC gliomas in adults, contrary to those encountered in children, are usually highly malignant. This diagnosis should be considered in adult patients with progressive loss of vision in one or both eyes. With modern neuroradiological examinations, it may be suspected at an early stage, thus helping to exclude a potentially treatable cause of rapid visual failure. Few malignant optic gliomas have been described in adults. A case is presented here which illustrates the malignant nature and neuroradiological appearance of this tumor.

Case Report

This 59-year-old woman complained of rapidly deteriorating vision in her right eye over the preceding 2 weeks. Visual acuity was 3/60 in the right eye and 1.0 in the left eye. Visual field examination revealed a centrocecal scotoma in the right eye; the left eye showed a normal visual field. Fundus examination was normal. There was evidence of an afferent pupillary defect on the right side. Visual evoked responses of the right eye could scarcely be elicited. In the left eye there were normal responses. Optic neuritis was suspected. Three weeks later the vision in the left eye had decreased as well; on examination there was only light perception in the right eye, while in the left visual acuity was 1/300. Both pupils were dilated and responded only slightly to light.

Examination. On general physical and neurological examination no abnormalities were found. The erythrocyte sedimentation rate was 8 mm/hr. Results of a full blood count, electrolyte tests, and urea and blood glucose determinations were normal. Serological tests for syphilis were negative. The cerebrospinal fluid protein was slightly elevated (606 mg/liter). Slight atrophy of the dorsum sellae was seen on the skull x-ray film. A computerized tomography (CT) brain scan showed a suprasellar contrast-enhancing mass, which seemed to be separated from the sellar contents.

Operation. Bilateral carotid angiography showed no abnormal vessels. Neurosurgical exploration by a right frontotemporal craniotomy revealed thickened and discolored optic nerves and chiasm. A small biopsy specimen was obtained from the right optic nerve, and, on histological examination, showed an astrocytoma with mitotic activity (Fig. 1).

Radiotherapy (5470 rads in 6 weeks) did not improve the patient’s visual acuity. Six months after the initial symptoms, a CT brain scan showed the tumor to have invaded the hypothalamus and left temporal lobe. Shortly afterward she became comatose and died. There was no autopsy.

Discussion

About 1% of all intracranial neoplasms are optic gliomas, 90% of which occur in children before the age of 20 years. In most cases occurring in childhood, the disease runs a benign course with a well-known association with neurofibromatosis, and the tumor usu-
**TABLE 1**
Summary of 30 cases of malignant optic glioma in the literature*

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Sex &amp; Age (yrs)</th>
<th>Clinical Presentation</th>
<th>Radiation Therapy</th>
<th>Interval to Death (mos)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Collins &amp; Marshall, 1900</td>
<td>F, 46</td>
<td>visual failure, proptosis</td>
<td>NK</td>
<td>24</td>
</tr>
<tr>
<td>Pick, 1901</td>
<td>F, 40</td>
<td>visual failure, seizures</td>
<td>NK</td>
<td>12</td>
</tr>
<tr>
<td>Sourdille, 1904</td>
<td>F, 22</td>
<td>visual failure</td>
<td>NK</td>
<td>NK</td>
</tr>
<tr>
<td>Thomsen, 1920</td>
<td>M, 22</td>
<td>visual failure</td>
<td>NK</td>
<td>4</td>
</tr>
<tr>
<td>Cosmettatos, 1923</td>
<td>M, 40</td>
<td>visual failure, proptosis</td>
<td>NK</td>
<td>NK</td>
</tr>
<tr>
<td>Martin &amp; Gagel, 1931</td>
<td>M, 25</td>
<td>visual failure, headache</td>
<td>NK</td>
<td>9</td>
</tr>
<tr>
<td>Saebø, 1949</td>
<td>M, 56</td>
<td>visual failure, headache, ophthalmoplegia</td>
<td>NK</td>
<td>12</td>
</tr>
<tr>
<td>Condon &amp; Rose, 1966</td>
<td>M, 30</td>
<td>visual failure, headache, ophthalmoplegia, ptosis</td>
<td>NK</td>
<td>24</td>
</tr>
<tr>
<td>Mattson &amp; Peterson, 1966</td>
<td>M, 57</td>
<td>visual failure, headache, ophthalmoplegia, ptosis</td>
<td>yes</td>
<td>3</td>
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<tr>
<td>Hamilton, et al., 1973</td>
<td>M, 55</td>
<td>visual failure, painstill eye</td>
<td>yes</td>
<td>9</td>
</tr>
<tr>
<td>Gibberd, et al., 1973</td>
<td>M, 55</td>
<td>visual failure, painstill eye</td>
<td>yes</td>
<td>9</td>
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<tr>
<td>Hoyt, et al., 1973</td>
<td>M, 55</td>
<td>visual failure, painstill eye</td>
<td>yes</td>
<td>10</td>
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<tr>
<td>Manor, et al., 1976</td>
<td>M, 70</td>
<td>visual failure, headache, hemioparesis</td>
<td>no</td>
<td>3</td>
</tr>
<tr>
<td>Harper &amp; Stewart-Wynne, 1978</td>
<td>M, 75</td>
<td>visual failure, ptosis, ophthalmoplegia, hemioparesis</td>
<td>no</td>
<td>3</td>
</tr>
<tr>
<td>Spoor, et al., 1980</td>
<td>F, 69</td>
<td>visual failure</td>
<td>yes</td>
<td>10</td>
</tr>
<tr>
<td>Barbaro, et al., 1982</td>
<td>F, 60</td>
<td>visual failure, headache</td>
<td>yes</td>
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<tr>
<td>Rudd, et al., 1985</td>
<td>M, 75</td>
<td>visual failure, headache, ophthalmoplegia, glaucoma</td>
<td>yes</td>
<td>4</td>
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<tr>
<td>F, 44</td>
<td>visual failure, painful eye</td>
<td>no</td>
<td>4</td>
<td></td>
</tr>
</tbody>
</table>

* The first 10 cases were reviewed by Hoyt, et al.\(^9\) NK = not known.

ally appears to be a well-differentiated astrocytoma.\(^1,2\)
In adult patients, however, optic gliomas may have characteristics of a highly malignant tumor growing invasively, causing early loss of vision, and inevitably leading to death. Up to now, 30 adult patients with malignant optic glioma have been described\(^1,2,4-6,9,11,13,14\) (Table 1).
Hoyt, et al.\(^9\) defined the syndrome of malignant optic nerve glioma in adulthood; they considered that it occurred mainly in middle-aged men, causing rapidly

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*Fig. 1. Photomicrographs of sections of the biopsy specimen. Left: Section of the tumor showing fibrillary astrocytes. PTAH, × 230. Right: Section immunostained for GFAP showing a mitotic figure (open arrow) and two GFAP-positive cells (closed arrows). GFAP-hematoxylin, × 740.*
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deteriorating vision and death within a year. Table 1 shows that both sexes are affected (male:female ratio 1.3:1) and that the mean age at presentation is 52.1 years. The symptoms and signs depend on the location of the process. Tumor growth in the optic nerve gives rise to unilateral visual loss and an afferent pupillary defect. Papilledema or optic atrophy may be seen on funduscopy. Proposisis and ophthalmoplegia may result from expanding tumor growth into the orbit. Growth of the process toward the optic chiasm may lead to temporal hemianopsia in the contralateral eye. Tumors situated in the optic chiasm or optic tracts cause early visual loss and visual field defects on both sides. Infiltration by tumor growth into surrounding structures such as the hypothalamus and the medial temporal lobe may cause hydrocephalus by compression of the third ventricle, hypothalamic dysfunction, hemiplegia, or epileptic seizures. The mean interval until death is 8 months (Table 1).

The diagnosis of malignant optic glioma is seldom made before an exploratory craniotomy or even autopsy because it is such a rare tumor. Moreover, its symptoms and signs are not specific and are more likely to indicate one of the following diagnoses: an optic neuritis; a tumor arising in adjacent structures, such as a meningioma of the optic nerve sheath; a craniopharyngioma or pituitary adenoma; or an aneurysm of the internal carotid artery. Recently, with CT and magnetic resonance (MR) imaging, an optic glioma may be suspected at an early stage. Tubular thickening of the optic nerve and chiasm or a suprasellar globular mass with involvement of the optic tracts may be seen. There is usually contrast enhancement. Cyst formation is rarely seen. The skull x-ray film may reveal enlargement of the optic canals or deformation of the sella turcica. Bilateral carotid angiography usually shows no tumor blush, in contrast to a meningioma. Few malignant optic gliomas in adults have been described since the advent of CT and MR imaging.

In most cases occurring in childhood, histological examination reveals a well-differentiated astrocytoma (also called a pilocytic astrocytoma) without signs of malignancy. In the adult form, however, there is an invasive growing astrocytoma with malignant features. Perivascular growth causes focal tumor necrosis and hemorrhages. Early papilledema may be explained by venous obstruction in the optic nerve caused by subpial tumor growth. Debate still continues regarding the use of radiation therapy for optic glioma in childhood; irradiation has been given to several adult patients with malignant optic glioma (Table 1), but it does not seem to alter the course of the disease, nor has it prolonged life expectancy, which is nearly always less than 1 year.

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References


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