Surgical Management of an Optic Nerve Glioma with Perineural Arachnoidal Gliomatosis Growth Pattern

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Abstract: We describe a vision sparing surgical approach for optic nerve glioma. A 7-year-old girl experienced declining academic performance and social withdrawal attributed to progressive disfiguring proptosis. Three years earlier, she had undergone a limited biopsy, a course of chemotherapy, and orbital radiation therapy for a right optic nerve glioma with perineural arachnoidal gliomatosis (PAG). Because of marked proptosis, another surgery was performed via a lateral orbitotomy. After cutting a window in the thickened dura of the optic nerve, rouge colored spongy tissue was suctioned from the subarachnoid space. Small, more solidified areas were excised with unipolar cautery. Care was taken to avoid identifiable blood vessels and the optic nerve, and approximately 60%–70% of the tumor was removed. The dural window was approximated with interrupted sutures. Postoperatively, there was 9 mm reduction in right proptosis and visual acuity improved to from 20/70 to 20/60. This case illustrates the possibility of debulking optic nerve gliomas without sacrificing vision. It should be stressed that this technique is only applicable to gliomas with PAG and the durability of the surgical benefit is unknown.

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Optic nerve gliomas adopt one of the 2 growth patterns. More commonly, there is intraneural glial proliferation as neoplastic growth occurs within individual nerve fascicles. The other pattern is characterized by perineural arachnoidal gliomatosis (PAG) with invasion of the leptomeninges and relative sparing of the optic nerve (1,2). Although PAG is more often seen in patients with neurofibromatosis type 1 (NF1) (3,4), recent reports have documented cases of PAG occurring in patients without NF1 (1,5). Gliomas may exhibit mostly intraneural or PAG growth characteristics, or a combination of both. Theoretically neoplastic infiltration, without optic nerve invasion, should be amenable to surgical excision without vision loss. In this report, we describe a young girl, with moderately reduced visual acuity, who underwent surgical debulking of PAG without further loss of vision.

CASE REPORT

In 2008, a case was published of a 4-year-old girl with a right optic nerve glioma, associated with PAG (1). Initially, she presented with a 3-month history of rapidly progressive, painless right proptosis and visual acuity improved to from 20/70 to 20/60. This case illustrates the possibility of debulking optic nerve gliomas without sacrificing vision. It should be stressed that this technique is only applicable to gliomas with PAG and the durability of the surgical benefit is unknown.

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A lateral orbitotomy was performed via an extended eyelid crease incision. The expanded dura of the optic nerve was encountered after incision of the periorbita and blunt dissection through a thin layer of orbital fat. A large fusiform section of dura was excised. This comprised roughly 90% of the length and 25% of the circumference of the optic nerve. The underlying tumor was rouge colored and spongy. A large portion was easily removed with suctioning using a Cavitron Ultrasonic Surgical Aspirator (CUSA; Tyco Healthcare Radionics, Burlington, MA). Unipolar cautery was used for intermediate portions of the tumor. Nothing more than suction and simple grasping with forceps was used as dissection approached the optic nerve. Injury to identifiable blood vessels was avoided. Approximately 60%–70% of the tumor was removed. The dura was stiff and when closed, it did not approximate the remaining optic nerve tumor. Presumably, the space between the optic nerve and disc was filled with cerebrospinal fluid (CSF). Closing the dural window was done to reduce the theoretical risk of tumor spreading outside of the optic nerve sheath.

Postoperatively, visual acuity measured 20/60 in the right eye, with only 2 mm of proptosis (Figs. 2B and 3). Ocular motility improved to near normal. MRI disclosed significant decrease in the size of the optic nerve glioma (Fig. 4). The only lasting negative impact of surgery was injury to parasympathetic pupillary innervation with mydriasis of the right pupil and partial loss of accommodation. Two years later, the patient’s examination was stable. The patient is no longer under the authors’ care. However, 3 years after surgery, tumor growth has been documented and repeat surgical intervention is being considered.

**DISCUSSION**

Management options for optic nerve glioma include observation, chemotherapy, radiotherapy, and surgery. Many of these tumors never grow and, in a minority of cases, spontaneous regression may occur (6). Accordingly, most advocate observation. Although controversial, chemotherapy is considered by some to be effective (7). Radiotherapy increases the risk for developmental abnormalities of the central nervous system and secondary tumors. Generally, it is used in older patients and those with more aggressive tumors (8).

Surgical excision of optic nerve glioma usually is reserved for individuals with limited visual potential because of the expected vision loss (9). There are exceptions. Debulking of gliomas without loss of vision has been described where growth patterns appear separate from axons of the optic nerve. Wisoff et al (10), found that in 16 patients with intracranial optic pathway gliomas with an exophytic growth pattern, none lost vision as a result of surgery. Ahn et al (11) reported that in a series of 16 patients with optic nerve glioma who underwent surgery, 4 did not lose vision. PAG represents another growth pattern where a portion of the neoplasm might be excised without axonal injury.

In our patient, an additional consideration was the effect radiation might have had on the tumor prior to surgery. The MRI appearance of the tumor was different before surgery (Fig. 4A) compared with initial presentation (Fig. 1). This might, in part, represent tumor necrosis or other effects of radiation and might have contributed to the ease with which the tumor was removed. Another consideration is the role of CSF. Optic nerve sheath decompression has been reported to be beneficial in treating optic nerve glioma.

**FIG. 1.** Preoperative magnetic resonance imaging (MRI). Contrast-enhanced fat suppressed T1 axial (A) and T2 coronal (B) scans show the right optic nerve of relatively normal caliber visible within a tumor of meningeal thickening and infiltrate consistent with perineural arachnoidal gliomatosis.

**FIG. 2.** A. Appearance of patient with marked right proptosis before surgery. B. After surgery, there is marked improvement in proptosis and position of the right eye.
Although in our patient there was no appreciable egress of CSF when the optic nerve sheath was opened. This is a single case and should be interpreted with caution. Any surgical maneuver within the subarachnoid space puts the optic nerve at risk. Neuroimaging and avoidance of sharp dissection are essential. The blood supply to the optic nerve should be preserved by avoiding identifiable blood vessels. Risk of thermal injury has been described with the use of CUSA. In our case, CUSA aspiration and unipolar cauterity were used as the optic nerve was approached. Also, the long-term impact of such surgery is unknown. Finally, although our patient’s visual acuity was preserved, visual field testing was not performed in this young child and we cannot state with certainty that, following surgery, some field loss did not occur.

In summary, the dramatic improvement of proptosis with preservation of vision suggests that surgical debulking is a reasonable consideration in patients with orbital optic nerve glioma with PAG. Vision-sparing surgery only seems likely with PAG, where neoplastic infiltration primarily involves the meninges. Further experience is needed to determine if and how often this result can be reproduced.

REFERENCES