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Introduction

Optic nerve gliomas (ONG) are low-grade, diffusely-infiltrative tumors that form from glial cells within the optic nerve. They are most common in children and have been associated with neurofibromatosis type 1 (NF1). As they grow, they can cause mass effect on the optic nerve leading to progressive vision loss. Management options include observation, chemotherapy, radiation, and surgery.

Methods and Materials

We present a case of ONG in an 11-year-old boy treated with a novel endoscopic endonasal approach for bony decompression of the optic canal and optic nerve sheath fenestration.

Results

An 11-year-old boy negative for NF1 presented with a 3-month history of intermittent right-eye visual obscurations increasing in frequency. MRI showed enlargement and signal within the right optic nerve consistent with intraorbital ONG. An endoscopic, endonasal, trans-sphenoidal, trans-tubercular approach was employed for bony decompression of the canalicular component of the right optic nerve 270-degrees around at the pre-chiasmatic sulcus and medial orbital wall and to fenestrate the optic sheath. Once the sphenoid sinus was accessed, we used an irrigated 4 mm coarse diamond bit to drill the tuberculum sellae extending laterally over the optic canal and all the way to the orbit. We used a Cottle elevator to fracture the lamina papyracea and thereby decompress the medial wall of the orbit. Once the medial wall of the optic canal was decompressed, we used a 2-mm coarse diamond burr to drill the roof and floor of the optic canal for 270 degrees of bony decompression. Then using an 11 blade we fenestrated the optic sheath along the length of the exposed optic canal. The patient did well and was discharged on postoperative day 2. 10 months postoperatively, he was undergoing adjuvant chemotherapy with carboplatin and vincristine chemotherapy in accordance with Children's Oncology Group (COG) protocol. He continued to do well, with no new deficits. Interval imaging showed decreased size of the mass.

Discussion

ONGs are slow-growing tumors that can cause progressive visual loss secondary to mass effect on the optic nerve. As they progress, they tend to grow from the orbit into the optic chiasm. They can be difficult to resect due to their eloquent location intrinsic to the optic nerve. Chemotherapy and radiation therapy are generally successful, with 5-year progression-free survival as high as 90% in some series. Our surgical approach focused on direct decompression of the optic canal and nerve sheath fenestration, with the goal to prevent new vision loss. Due to the facts that our patient did not have NF1, along with the progressive symptoms, our multi-disciplinary tumor board came to the conclusion that he was more at risk for disease progression than an incidentally found tumor in an NF1 patient might be and that a minimally-invasive endoscopic endonasal approach for bony decompression of the optic nerve could result in superior outcomes in terms of preventing progression of his visual symptoms. Our surgical approach provided a direct minimally-invasive approach for optic nerve decompression even in a pediatric population, and our patient continues to do well.

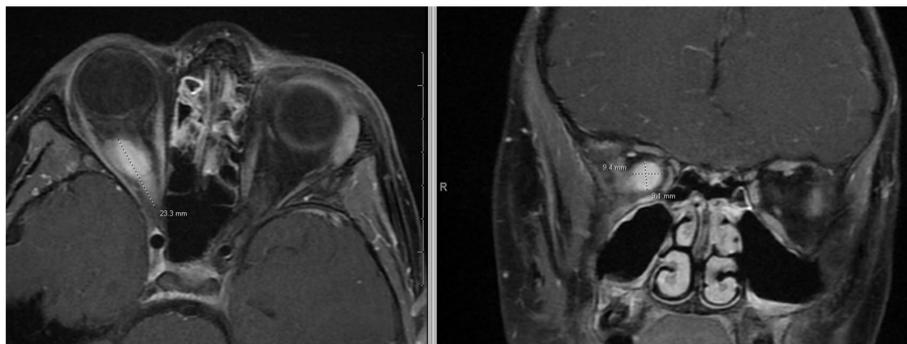


Figure 1. Axial and coronal T1 MRI with contrast sequences obtained before surgery showing right optic nerve glioma

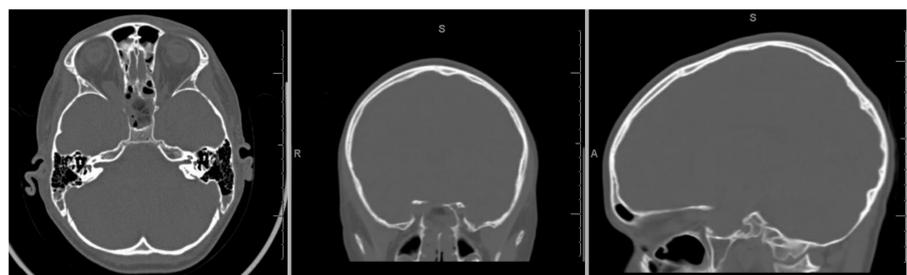


Figure 2. CT obtained after surgery showing bony decompression of the right orbit

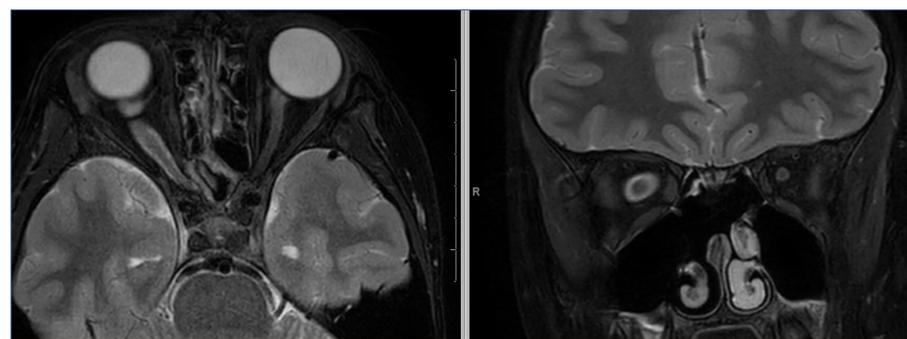


Figure 3. MRI of the orbits obtained 10 months after surgery showing right orbital decompression and decreased size of the optic nerve glioma after the patient underwent Children's Oncology Group (COG) protocol

Conclusion

As ONGs grow, they can cause mass effect on the optic nerve leading to progressive vision loss. Here we present a case of ONG in an 11-year-old boy treated with a novel endoscopic endonasal approach for 270-degree bony decompression of the optic canal and optic nerve sheath fenestration. As of 6 months postoperatively, our patient was doing well, with no new visual deficits, and decreased size of the mass on interval MRI. Our results suggest our surgical approach focusing on direct decompression of the optic canal and nerve sheath fenestration is a safe and effective strategy for managing ONGs and in particular for preventing symptom progression due to compression of the optic nerve against the optic nerve sheath or orbital wall.

To our knowledge, ours is the first report to explicitly show that this minimally-invasive technique is feasible and successful for the treatment of this pathology and to also show that this is feasible in a pediatric patient.

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