Surgical Resection and Postoperative Chemotherapy for Optic Nerve Glioma with Intracranial Extension in a 10-year-old Male: A Long-term Follow-up Case Report

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Disclosures: The authors report no conflict of interest.

ABSTRACT

Objective: To present a long-term follow-up report of a 10-year-old male with optic nerve glioma who underwent surgical removal and postoperative chemotherapy.

Methods: Case report.

Results: A 10-year-old Filipino boy was referred to a tertiary institution for a five-year history of progressive right eye proptosis with vision loss. Pertinent findings included right eye proptosis, lagophthalmos, and limited elevation and adduction. He also had several hyperpigmented lesions on the abdomen and upper torso. Vision on the right was no light perception, with a relative afferent pupillary defect, exposure keratopathy, and optic nerve pallor. Vision on the left eye was 20/20 with a temporal visual field defect. Cranial and orbital computed tomography (CT) scan showed a circumscribed enhancing mass within the right intraconal space with widened right optic nerve canal. Additional magnetic resonance imaging (MRI) revealed a heterogeneously enhancing mass diffusely involving the intraorbital and intracanalicular segments of the right optic nerve suspicious for optic nerve glioma. He underwent excision of the orbital portion of the mass via lateral orbitotomy. Histopathology showed pilocytic astrocytoma. Eight cycles of chemotherapy with carboplatin and vincristine was completed. Significant improvement of globe position and resolution of ocular exposure was achieved postoperatively with residual right ptosis. These findings remained stable at six years after treatment.

Conclusion: Optic nerve gliomas with intracanalicular and chiasmal extension can be managed with surgical removal of the orbital component and postoperative chemotherapy. This can result in improvement of proptosis and long-term remission.

Keywords: optic nerve glioma, neurofibromatosis 1, orbitotomy, pilocytic astrocytoma, chemotherapy

Philipp J Ophthalmol 2025;50:112-116

Optic nerve gliomas (ONGs) are the most common optic pathway tumors affecting children.[1-3] It may involve the optic nerve alone and can extend posteriorly to the chiasm, or optic tracts and optic radiations.[1-5] ONG is seen frequently in association with Neurofibromatosis Type 1 (NF-1) and can also occur sporadically.[1,3-5] Gradual, painless, and unilateral proptosis associated with loss of vision and a relative afferent pupillary defect (RAPD) is a common presentation.[1,3-5] Cranial and orbital computed tomography (CT) and magnetic resonance imaging (MRI) with contrast help confirm the diagnosis, with MRI being the best modality to show the degree of involvement along the visual pathway.[1,4] Management options include observation, chemotherapy, radiation, and surgical removal.[1-5] Currently, chemotherapy is considered the initial treatment of choice for progressive optic nerve glioma.[1,3] Cosmetic surgical resection is indicated for blind eyes with disfiguring proptosis and definite documented radiologic tumor enlargement.[1-5] In this report, a 10-year-old male presented with vision loss and progressive proptosis with corneal exposure, and contrast-enhanced MRI of the brain and orbits showed heterogeneously enhancing mass diffusely involving the intraorbital and intracanalicular segments of the optic nerve with suspicious optic chiasmal involvement. A multidisciplinary team consensus recommended surgical resection via lateral orbitotomy versus transcranial approach and postoperative chemotherapy.

CASE PRESENTATION

A 10-year-old Filipino boy was referred to a tertiary institution for right eye proptosis with vision loss. When the patient was four years old, the mother noted outward deviation of the boy's right eye with proptosis. No blurring of vision, headache, dizziness, vomiting, or decreased sensorium were noted. One year later, there was notable progression in proptosis which prompted the mother to bring the boy to the local hospital. At this time, the patient had complete loss of vision in the right eye. CT scan was advised but due to financial constraints was not done. Patient was lost to follow-up for five years during which there was progression in the proptosis of the right eye. Neonatal, past medical, family, personal and social history were unremarkable.

On examination, the right eye had no light perception (NLP) with an RAPD, exposure keratopathy, and optic nerve pallor (**Figure 1**). There was right eye proptosis, lagophthalmos, hypotropia, exotropia, and limited supraduction and adduction. The left eye had a visual acuity of 20/20, with temporal defects seen on automated perimetry and temporal pallor of the optic disc (**Figure 2**). Systemic examination showed hyperpigmented lesions (café-au-lait spots) on the abdomen and upper torso.

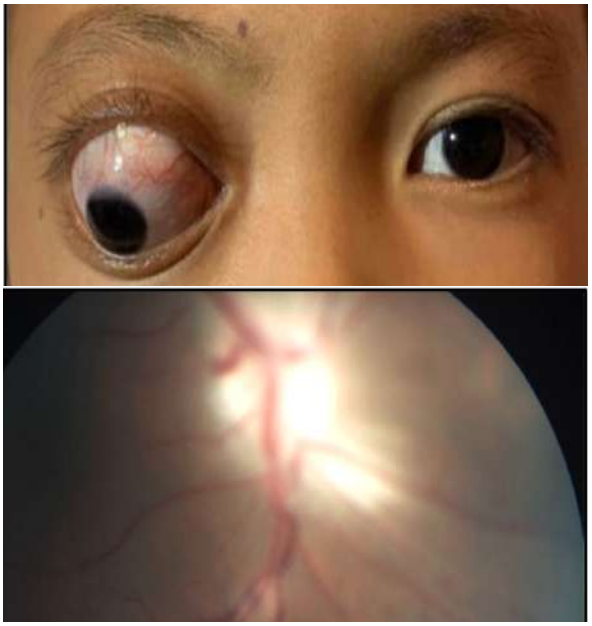

Figure 1. On presentation, the right eye demonstrated (A) proptosis, hypotropia, exotropia, and (B) diffuse optic disc pallor.

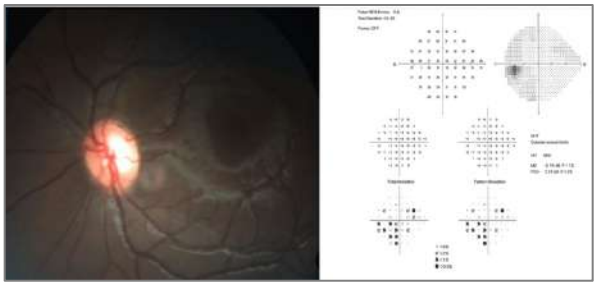

Figure 2. Left eye showed (A) temporal optic disc pallor and (B) temporal defects deeper in the inferotemporal quadrant on automated perimetry.

Cranial and orbital CT scan done showed a 2.4 x 2.3 x 3.2 cm circumscribed enhancing mass within the intraconal space of the right orbit, with widened right optic nerve canal (**Figure 3**). Additional MRI of the brain and orbits revealed a 4.6 cm heterogeneously enhancing mass diffusely involving the intraorbital and canalicular segments of the right

optic nerve with suspicious chiasmal involvement; optic nerve glioma was considered (**Figure 4**). Excision biopsy was suggested upon clearance of the institution's tumor board.

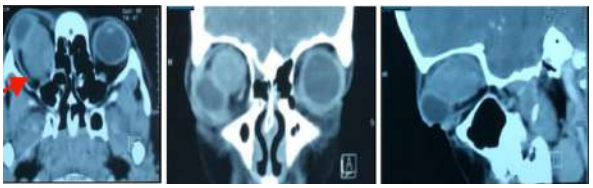

Figure 3. Orbital CT scan revealed a right intraconal orbital mass resulting in proptosis with anterior and inferior displacement of the globe, along with globe indentation.

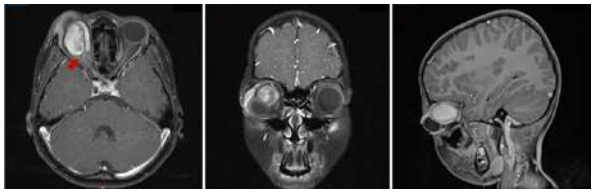

Figure 4. Contrast-enhanced cranial and orbital MRI showed a heterogeneously enhancing mass diffusely involving the intraorbital and canalicular segments of the right optic nerve (red arrow).

Multi-disciplinary discussion involving neurosurgery, radiology, radiation oncology, orbit specialist, neuro-ophthalmology, and pediatric hematology-oncology services reviewed the case. Options were discussed in the management, whether or not to unroof the orbit and perform the excision transcranially for complete excision, or perform surgical removal of the orbital segment and conduct chemotherapy postoperatively to address intracranial tumor residual.

Eventually, the patient underwent excision of the orbital portion of mass via the classic Stallard-Wright incision lateral orbitotomy, with the distal end of the mass behind the globe resected first, followed by excision of the posterior proximal end at the orbital apex. The lesion measured approximately 3.8 cm in horizontal length. Post-operatively, there was immediate reduction of proptosis, with note of ptosis and extraocular muscle limitation. Histopathology revealed a pilocytic astrocytoma WHO Grade 1 (**Figure 5**).

The patient completed eight cycles of chemotherapy with carboplatin and vincristine. Six months after surgery and completion of chemotherapy, the patient remained well. Vision remained NLP on the right, and 20/20 on the left eye. The right pupil was noted to be 4 mm in size. Funduscopy of the right eye revealed a pale optic nerve surrounded by peripapillary atrophy. Three large chorioretinal scars were seen at the superior, inferior and nasal peripheral retina, with the largest scar measuring 10 disc diameters (DD) in size. Follow-up MRI scans of the brain were done at six months and one year postoperatively, and these demonstrated no evidence of recurrence of the right optic nerve glioma and stable mild focal enhancement at the tip of the right optic nerve stump from postoperative granulation changes (**Figure 6**).

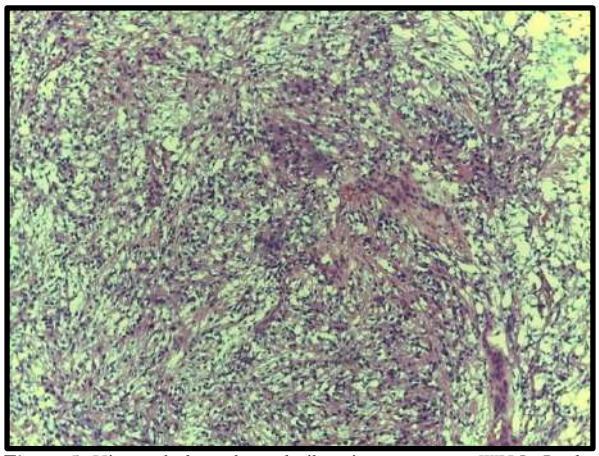

Figure 5. Histopathology showed pilocytic astrocytoma WHO Grade 1 displaying biphasic pattern of piloid and stellate astrocytes, associated with microcystic changes admixed in different proportions throughout the tumor.

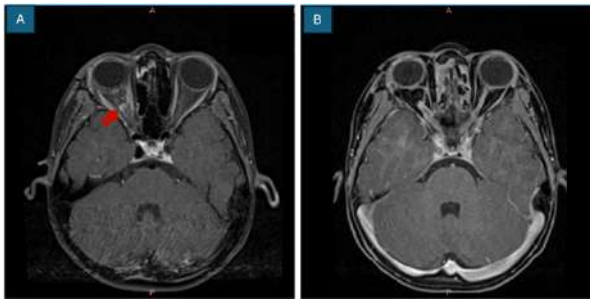

Figure 6. Repeat MRI with gadolinium at (A) 6 months and (B) 1 year after excision of optic nerve glioma. There was resolution of proptosis and a small foci of enhancement seen at tip of right optic nerve (arrow) which represented postoperative granulation changes. Stable imaging findings were noted one year after surgery.

Six years postoperatively, on teleconsult with the patient, there is stable globe position and ptosis, and no clinical signs of recurrence (**Figure 7**). Near vision for the left eye was J1. He has graduated from elementary school and remains well in his teens, reflecting sustained effectiveness of the medical management in the long term.

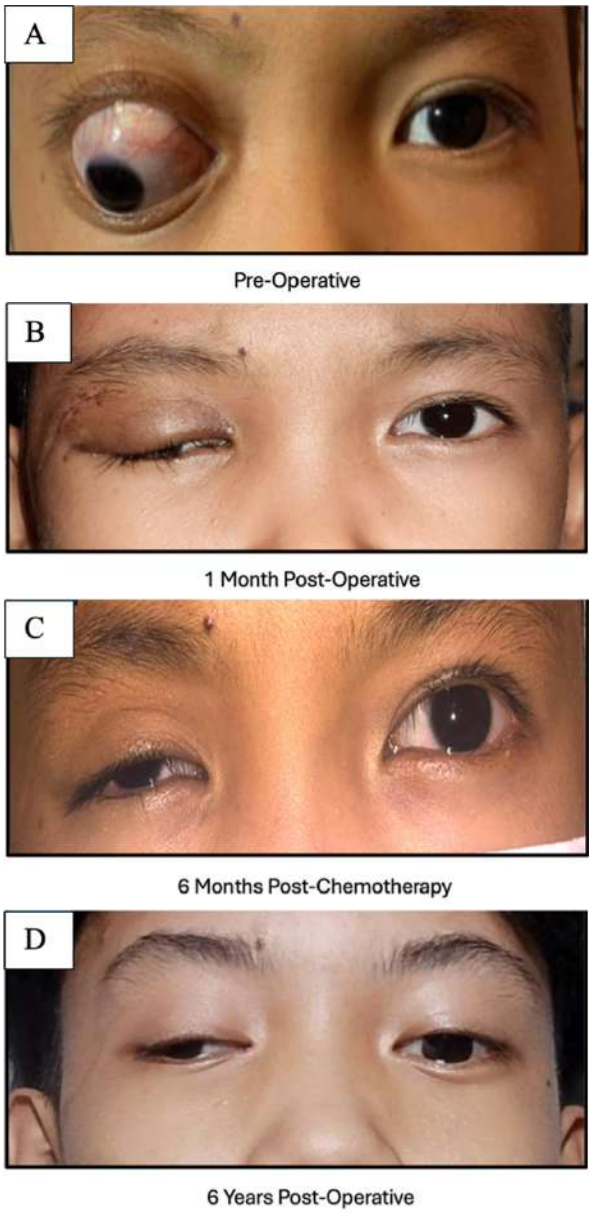

Figure 7. Pre-operative and post-treatment photograph series of the patient. Proptosis, hypotropia, and exotropia of the right eye were observed preoperatively (A) with improvement of globe position and ptosis at one month after surgery (B). Globe and eyelid position appear stable with acceptable cosmesis at six months after chemotherapy (C) and stability up to six years after treatment (D).

DISCUSSION

Unilateral proptosis with progressive blurring of vision in pediatric patients may suggest an orbital mass and warrant neuroimaging. In this case, the patient is a 10-year-old boy who presented with unilateral, slow, and progressive proptosis and vision loss, which are common presentations of optic nerve glioma.[1,3-5] ONGs are often associated with neurofibromatosis type-1 (NF-1), a common autosomal dominant phacomatosis. [1,3-5] The presence of optic nerve glioma and multiple café-au-lait spots in this case suggests an association with NF-1.[5] A contrasted CT scan showed a circumscribed enhancing mass within the intraconal space of the right orbit, with widened right optic nerve canal. ONG usually presents as fusiform enlargement of the optic nerve with additional features that include widening of the optic canal, seen in this case, as well as variable contrast enhancement, and rarely eccentric enlargement of the nerve and cystic degeneration. MRI has been described to be the best neuroimaging modality as it evaluates the intracranial disease better than CT and avoids radiation.[1,4] An orbital MRI with contrast revealed the extent of the enhancing mass that went beyond the orbital portion of the optic nerve, with suspicious extension towards the chiasmal region.

Management should be individualized since ONGs typically have variable clinical courses, with some having an indolent and stable course, while others demonstrate slow or rapid growth over the years.[1-5] ONG can usually be diagnosed through a comprehensive clinical and ophthalmologic examination and neuroimaging modalities. A biopsy is often not necessary because of the risk for vision loss.[3] Current management options include observation, chemotherapy, radiotherapy, newer molecularly targeted therapies such as bevacizumab and mitogen-activated protein kinase pathway inhibitors (e.g. selumetinib), and surgical resection/debulking. [1-5] Observation is appropriate in patients with indolent courses that do not cause vision loss, disfiguring proptosis, and some cases have even reported spontaneous regression. [1-5] Chemotherapy is considered the first-line treatment and is indicated for patients with documented clinical worsening, progressive vision loss, and progression on neuroimaging.[1,3,4] Surgical resection is recommended for those that cause severe vision loss, cosmetically disfiguring proptosis, and exposure keratopathy. [1-5]

In this case, a multidisciplinary team involving different specialties within the institution discussed the best approach in the patient's treatment. Given the patient's vision loss, severe proptosis, exposure keratopathy, and tumor extension beyond the orbit,

it was decided to perform an excision biopsy with debulking of the mass through an orbital approach instead of a craniotomy, since full excision may not be achieved completely with suspicious extension of the lesion towards the chiasmal region. A transcranial orbitotomy also carries a higher surgical morbidity.[3] Post-operative chemotherapy was given to address the residual tumor beyond the orbit. Radiotherapy was not recommended at the time due to associated risk for secondary malignancies, [1,3] and was considered to be a salvage therapy. The patient completed 8 cycles of vincristine and carboplatin, the most common first-line regimen for ONG with a reported 77% 3-year progression free survival (PFS) rate among patients with NF-1.[3] ONGs of childhood are most commonly benign pilocytic astrocytomas[1-5], similar to the histopathology seen in this case.

For ONG involving the intraorbital nerve without intracanalicular extension, surgical resection of the orbital segment of ONG can be done by anterior or lateral orbitotomy. On the other hand, when the tumor involves the intracanalicular and intracranial segments already, a transcranial approach has been advocated for complete tumor resection.[2-4] In this case, a lateral orbitotomy approach was done with resection of the orbital segment of the tumor *en-bloc*. Post-operatively, there was immediate improvement of eyelid closure and proptosis. Since the procedure involved removing the entire orbital portion of the tumor, with limited view during resection of the mass at the orbital apex, risk of damage to the third cranial nerve can result in eyelid ptosis, as was observed in this case. Modifications in the surgical technique that can minimize cranial nerve injuries at the orbital apex include resection after evacuation described by Mohammad which allows good visualization of the proximal end during tumor removal.[2] Farazdaghi *et al*. advocated for limited debulking of the tumor while leaving a minimal portion at most posterior apex.[3]

This report highlights that in cases of optic nerve glioma with intracanalicular and chiasmal extension, surgical removal of the orbital segment of the tumor and post-operative chemotherapy with vincristine and carboplatin effectively improve proptosis and can help patients achieve good quality of life. Communication with the patient and family helps ensure long-term follow-up and is recommended for continued surveillance of potential recurrences and general health.

ACKNOWLEDGEMENT:

The authors would like to express their sincere gratitude to Dr. Bernardita C. Navarro and the Tzu Chi Foundation for the referral and support in the patient's care as well as to the multidisciplinary Tumor Board committee of Cardinal Santos Medical Center particularly Dr. John Mark Carabeo and Dr. Claro B. Ison for the neuroimaging interpretation, and Dr. Alex Sua for the histopathologic report.

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