Optic pathway glioma

- Coexistence of Congenital Aniridia and Ptosis in a Patient with Neurofibromatosis Type I: A Case Report
- Growth hormone excess in children with neurofibromatosis and optic pathway glioma, an underdiagnosed condition: experience with long-acting treatment
- A Rare Case of Sporadic Optic Pathway Glioma in a 7-year-old Female
- Neuroimaging and Metabolic Features of Optic Pathway Glioma in a Child with Neurofibromatosis Type 1: Low-Grade Glioma Transformation in the Left Optic Radiation
- Impact of Optic Pathway Gliomas on Puberty and Growth in Neurofibromatosis Type 1: A 20-Year Experience From a Tertiary Center
- Importin-alpha transports Norrin to the nucleus to promote proliferation and Notch signaling in glioblastoma stem cells
- Recent Advancement of Neurofibromatosis Type 1: A Narrative Review
- Supra-sellar clear cell ependymoma in a 2-year-old female: A case report

---

In cases where the tumour is confined to the optic nerves, they can safely be referred to as optic nerve gliomas. Often optic nerve gliomas are either centred on or extend to involve the chiasm and optic radiations. In such cases, they are difficult to distinguish from hypothalamic gliomas and such a distinction is in most instances artificial. In such more posterior cases the term hypothalamic-optochiasmatic glioma is perhaps more accurate although it certainly does not roll off the tongue.

As such, generally, the term optic pathway glioma is favoured, recognising that there may be involvement of the hypothalamus [1)].

Most optic pathway glioma or hypothalamic gliomas are juvenile pilocytic astrocytomas, but their imaging characteristics are not specific with regard to their histologic features. Varying degrees of cystic change and enhancement are demonstrated. The tumors may appear smooth, fusiform, eccentric, or lobulated.

Optic pathway gliomas or suprasellar gliomas.

These tumors have sometimes been divided into optic pathway gliomas and hypothalamic gliomas (not to be confused with hypothalamic hamartomas). In cases where the tumour is confined to the optic nerves (Dodge stage 1), they can safely be referred to as optic nerve gliomas. Often, however, they are either centred on or extend to involve the chiasm and optic radiations. In such cases, they are difficult to distinguish from hypothalamic gliomas and such a distinction is in most instances artificial. In such more posterior cases the term hypothalamic-optochiasmatic glioma is perhaps more accurate although it certainly does not roll off the tongue.

As such, generally, the term optic pathway glioma is favoured, recognising that there may be involvement of the hypothalamus [2)].

Optic pathway/hypothalamic gliomas (OPHGs) are generally benign but situated in an exquisitely sensitive brain region, and may involve the optic nerves, the optic chiasm, and the optic tracts.

Martin and Cushing (1923) first noted the difficulty of determining the site of origin of optic nerve gliomas, since these tend to extend up and down the optic pathways, often making it impossible to decide even at postmortem examination whether the growth originated in the chiasm and extended to the hypothalamus, or vice versa.

Epidemiology

Gliomas of the optic chiasm and hypothalamus account for 10-15% of supratentorial tumors in children. Males and females are approximately equally affected.

Between 20 and 50% of patients with hypothalamic gliomas have a positive family history of neurofibromatosis type 1 (NF-1). Gliomas of the optic chiasm and hypothalamus in children with NF-1 usually have a more indolent course. Tumours may grow more slowly and occasionally regress spontaneously.

A retrospective, single-center, cohort study of 176 patients (93 boys), aged 6 years (range, 0.2-18 years), with hypothalamic-pituitary lesions showed craniopharyngioma (n = 56), optic pathway glioma (n = 54), suprasellar arachnoid cyst (n = 25), hamartoma (n = 22), germ cell tumor (n = 12), and hypothalamic-pituitary astrocytoma (n = 7). The most common presenting symptoms were neurologic (50%) and/or visual complaints (38%), followed by solitary endocrine symptoms (28%). Precocious puberty led to diagnosis in 19% of prepubertal patients (n = 131), occurring earlier in patients with hamartoma than in patients with optic-pathway glioma (P < .02). Isolated diabetes insipidus led to diagnosis for all germ-cell tumors. For 122 patients with neuro-ophthalmic presenting symptoms, the mean symptom interval was 0.5 year (95% CI, 0.4-0.6 year), although 66% of patients had abnormal body mass index or growth velocity, which preceded the presenting symptom interval onset by 1.9 years (95% CI, 1.5-2.4 years) (P < .0001) and 1.4 years (95% CI, 1-1.8 years) (P < .0001), respectively. Among them, 41 patients were obese before diagnosis (median 2.2 years [IQR, 1-3 years] prior to diagnosis) and 35 of them had normal growth velocity at the onset of obesity. The sensitivity of current guidelines for management of childhood obesity failed to identify 61%-85% of obese children with an underlying hypothalamic-pituitary lesion in this series [3].

These masses are, however, rare in adults and require tissue sampling for diagnosis [4].

Classification

Optic pathway glioma classification.

Histology

Most are juvenile pilocytic astrocytomas, but their imaging characteristics are not specific with regard to their histologic features. Varying degrees of cystic change and enhancement are demonstrated. The tumours may appear smooth, fusiform, eccentric, or lobulated.

see Optic pathway hypothalamic pilocytic astrocytoma.

Clinical features

Optic pathway gliomas (OPG) represent an important cause of visual loss in pediatric population. Visual acuity loss is the most important symptom of disease progression, but children with OPG are frequently unable to complete the testing of visual function.

Clinical features may include:

Decreased visual acuity / optic nerve atrophy ≈50%

Diencephalic syndrome [5) 6) 7)].

Obesity; sexual precocity; endocrine dysfunction (short stature)

Diabetes insipidus.

Diagnosis

Optic pathway glioma Diagnosis.

Differential diagnosis

The main differential is that of optic nerve sheath meningioma, however, the potential list is much longer including most causes of optic nerve enlargement.

The absence of calcification can be used to differentiate optic nerve glioma from optic nerve sheath meningioma.

Additionally, when the bulk of the tumour is located at the chiasm, the differential should include pituitary region masses.

They usually present earlier (first 5 years of age) than craniopharyngioma. Hypothalamic glioma poses a diagnostic dilemma with craniopharyngioma and other hypothalamic region tumors, when they present with atypical clinical or imaging patterns. Neuroimaging modalities especially MRI plays a very important role in scrutinizing the lesions in the hypothalamic region.

The main differentiating features between craniopharyngioma and hypothalamic glioma are the presence of mixed intensity cysts on T1 and calcification in craniopharyngioma and the relative young age of presentation in hypothalamic gliomas. Yet, it is not always possible to differentiate craniopharyngiomas from hypothalamic glioma. Thus, obtaining a tissue diagnosis via biopsy may be the right course of action in planning further management, whenever diagnosis is in doubt. [8)].

Treatment

Optic pathway glioma treatment.

Outcome

see Optic pathway glioma outcome.

Case series

2017

A retrospective follow-up study of 40 children with NF1 and OPG evaluated between August 1996 and May 2015 was undertaken. Patients who underwent radiotherapy or surgical resection were excluded and 36 patients were studied. Tumour location was classified according to the Dodge criteria: stage I, optic nerve alone; stage II, optic chiasm with or without optic nerve involvement; and stage III, involvement of the hypothalamus or other adjacent structures.

Endocrinopathies were diagnosed in 20/36 (55.6%) children during a mean follow-up of 9.1 (0.2-13.6) years: 0/4 OPGs were Dodge stage I, 12/21 (57.1%) stage II, and 8/11 (72.7%) stage III. The first endocrinopathy was found at a mean age of 7.4 (5.0-13.2) years, 2.4 (0-6.7) years after tumour diagnosis. We found growth hormone deficiency (GHD; 36.1%), central precocious puberty (33.3%), obesity with insulin resistance/impaired glucose tolerance (11.1%), early puberty (5.5%), GH excess (5.5%), ACTH deficiency (5.5%), hypogonadotropic hypogonadism (2.7%), and thyrotropin deficiency (2.7%). GHD was transient in all of those who were retested.

This population is at high risk of endocrinopathies due to tumour location. Lifelong endocrine follow-up is recommended. [9)].

2016

A retrospective case series study was led for 3 patients diagnosed with primary optic nerve and chiasm glioblastoma (Glioblastoma), coming from two referral neurosurgical centers. 2) An electronic search was conducted on MEDLINE via PUBMED, COCHRANE, from October 1973 to April 2016. Cohort, case reports, and case series were screened for investigating treatment and overall survivals of malignant optic nerve gliomas. Pooled means and 95% confidence intervals of overall survival for each treatment were generated.

1) From our retrospective case series, all patients had initial visual impairment (2 women and 1 man). The histological diagnosis was done by biopsy. The patients' mean age of was 67.3 years (SD 18.5). The disease was rapidly lethal for all patients: median overall survival (OS) was 5 months (SD: 15.1). Two patients underwent chemotherapy by single cure of temozolomide, while the third one was treated with a radio-chemotherapy protocol. 2) Due to the fact that there is no gold standard treatment as first choice treatment, a large heterogeneity in first-choice oncological treatment is observed. However, we did not find any significant differences for overall survival between WHO grade III and grade IV optic gliomas.

Malignant optic glioma is a rare and fatal disease in adults. Despite the modalities of treatment, the treatment outcomes remain unsatisfactory. There is no significant difference in the median overall survival of patients with malignant optic nerve, as compared to those diagnosed with other supratentorial glioblastoma. Chemoradiotherapy with temozolomide currently remains the best

treatment in terms of overall survival. Advances in the understanding of tumor biology have yet failed to translate into effective treatment regimens [10].

2015

Ten patients managed surgically utilising ioMRI at Alder Hey Children's Hospital between 2010 and 2013 were retrospectively identified. Demographic and relevant clinical data were obtained. MRI was used to estimate tumour volume pre-operatively and post-resection. If ioMRI demonstrated that further resection was possible, second-look surgery, at the discretion of the operating surgeon, was performed, followed by post-operative imaging to establish the final status of resection. Tumour volume was estimated for each MR image using the MRIcron software package.

Control of tumour progression was achieved in all patients. Seven patients had, on table, second-look surgery with significant further tumour resection following ioMRI without any surgically related mortality or morbidity. The median additional quantity of tumour removed following second-look surgery, as a percentage of the initial total volume, was 27.79 % (range 11.2-59.2 %). The final tumour volume remaining with second-look surgery was 23.96 vs. 33.21 % without ($p = 0.1$).

OPHGs are technically difficult to resect due to their eloquent location, making them suitable for debulking resection only. IoMRI allows surgical goals to be reassessed intra-operatively following primary resection. Second-look surgery can be performed if possible and necessary and allows significant quantities of extra tumour to be resected safely. Although the clinical significance of additional tumour resection is not yet clear, we suggest that ioMRI is a safe and useful additional tool, to be combined with advanced neuronavigation techniques for partial tumor resection [11].

2014

Forty-two patients were treated between 1998 and 2011. Their median age at diagnosis was 5 years 7 months. Nineteen patients were positive for neurofibromatosis Type 1 (NF1) and 23 patients were negative for NF1. The median duration of follow-up was 77 months (range 21.8-142.3 months). Presenting symptoms included visual impairment (in 50% of cases), headache (in 24%), and hypothalamic/pituitary dysfunction (in 29%). Twenty-two debulking procedures were performed in 21 patients. Four biopsies (3 open, 1 endoscopic) were also performed. The histological diagnosis was pilocytic astrocytoma in 21 patients and pilomyxoid astrocytoma in 2 patients. Ten patients (Group 1) had primary surgical debulking alone and were then observed. Four patients (Group 2) had surgical debulking, plus planned chemotherapy within 3 months. Seven patients (Group 3) required surgical debulking for progressive disease following a variety of treatments. Patient age had the greatest impact on subsequent tumor progression. In total, 13 patients received chemotherapy, 4 on initial presentation, 4 in combination with surgery, and 5 for further tumor progression. Five patients were treated with radiotherapy, 3 prior to referral to Alder Hey. Eleven patients required shunt insertion for hydrocephalus. Vision was stabilized for 74% of patients. The number of patients with hypothalamic/pituitary dysfunction increased from 12 at presentation to 16 by the end of treatment. The overall survival rate was 93%. Three patients died-1 from tumor progression, 1 from infective complications from tumor biopsy, and 1 from a spontaneous posterior fossa hemorrhage. NF1 was associated with improved outcome-fewer patients required active intervention and rates of visual impairment and/or or hypothalamic/pituitary dysfunction were lower.

Good long-term survival and functional outcomes can be achieved in children with OPHG. Tumor control was achieved through an individualized approach using surgery, chemotherapy, or

radiotherapy in varied combinations. The authors aim to limit radiotherapy to cases involving older children in whom other therapies have failed, due to the well-described and often devastating late effects associated with midline cranial irradiation. Surgery has a clear role for diagnosis, tumor control, and relief of mass effect. In particular, primary surgical debulking of tumor (without adjuvant therapy) is safe and effective. Recent advances in intraoperative MRI may add value and need further assessment [12)].

Zoli et al. analyze their experience with hypothalamic gliomas treated via the endoscopic endonasal approach. Methods Consecutive cases of hypothalamic gliomas treated since 2007 via an endoscopic endonasal approach were reviewed. Preoperative and postoperative neuroimaging as well as endocrinological, neurological, and visual symptoms were analyzed to assess the surgical outcome. Signs and symptoms of hypothalamic dysfunction including body mass index (BMI), memory, sleep-wake rhythm, and polyphagia were prospectively collected pre- and postoperatively to assess hypothalamic function. Quality of life was evaluated using the Katz scale. Results In the initial phase the endoscopic endonasal approach was adopted in 3 cases with a palliative intent, to obtain a biopsy sample or for debulking of the mass followed by radio- or chemotherapy. In 2 later cases it was successfully adopted to achieve gross-total tumor resection. Complications consisted of 2 postoperative Cerebrospinal fluid fistulas, which required an endoscopic endonasal reintervention. Visual deficit improved in 3 cases and normalized in the other 2. Four patients developed diabetes insipidus, and 3 an anterior panhypopituitarism. All patients had a moderate increase in BMI. No patients presented with any other signs of hypothalamic damage, and their quality of life at follow-up is normal. Conclusions Despite the limitations of a short follow-up and small sample, the authors' early experience with the endoscopic endonasal approach has revealed it to be a direct, straightforward, and safe approach to third ventricle astrocytomas. It allowed the authors to perform tumor resection with the same microsurgical technique: dissecting the tumor with 2 hands, performing a central debulking, and controlling the bleeding with bipolar coagulation. The main limitations were represented by some anatomical conditions, such as the position of the chiasm and the anterior communicating artery complex and, finally, by the challenge of Water-tight plastic repair. To definitively evaluate the role of this approach in hypothalamic gliomas, a comparison with transcranial series would be necessary, but due to the rarity of these cases such a study is still lacking. The authors observed that more aggressive surgery is associated with a worse endocrinological outcome; thus they consider it to be an open question (in particular in prepubertal patients) whether radical removal is an advisable goal for hypothalamic gliomas. [13)].

A retrospective review of patients diagnosed with suprasellar glioma between 2000-October 2012, included patients diagnosed with optic pathway glioma based on radiological features (with or without biopsy) and those who had a biopsy confirming pilocytic astrocytoma.

Fifty-three patients included (sporadic tumours 24 and NF1 related 29). Fifteen sporadic and four NF1 patients were biopsied. Twelve sporadic and 13 NF1 patients were initially treated with chemotherapy while only 1 patient had radiotherapy initially. Progression was noted in 58 % of the sporadic group and 24 % of the NF1 group. The only significant factor for progression was NF1 status ($p = 0.026$).

Management should be guided by individual patient circumstance. In our cohort, chemotherapy did not significantly improve progression free survival; however, NF1 status significantly correlated with the decreased progression [14)].

2013

A total of 101 patients with optic glioma newly diagnosed between 1975 and 2008 were evaluated retrospectively. COPP (cyclophosphamide, vincristine, procarbazine, prednisolone) and cisplatin plus etoposide were the most commonly used chemotherapy regimens. Radiotherapy was administered in patients with progressive or unresponsive disease.

The median age at the time of diagnosis was 6 years, and the male/female ratio was 1.15. The most common referral complaint was strabismus. The most common site of optic glioma was the hypothalamic-chiasmatic region (31.7%). Fifty-three patients (52.5%) had neurofibromatosis type 1 (NF-1). Treatment consisted of surgery, radiotherapy, and chemotherapy. Forty-nine patients (48.5%) underwent surgery, which was predominantly subtotal resection, radiotherapy was administered to 39.4%, and 30 patients received chemotherapy. The 5-year progression-free survival (PFS) and overall survival (OS) rates were 65.8% and 88.4%, respectively, and the 10-year PFS and OS were 54.2% and 83.4%, respectively, with an 8-year median follow-up. OS was significantly lower in patients with hypothalamo-chiasmatic involvement and significantly higher in patients with NF-1. The 5- and 10-year PFS rates were significantly higher in patients 10 years or older at diagnosis (P=0.0001) and in patients with intraorbital involvement (P=0.032). Eighteen patients (17.8%) died of disease.

Patients with NF-l and those older than 10 years have a better prognosis, whereas patients younger than 3 years and those with hypothalamic-chiasmatic optic glioma have a worse outcome. Further studies are needed to find appropriate treatment strategies [15].

1979

Diencephalic syndrome and its relation to opticochiasmatic glioma: review of twelve cases [16].

1974

Twenty children with hypothalamic gliomas from the Hospital for Sick Children, Great Ormond Street, were reviewed in an attempt to determine the appropriate management. If the child's condition at the time of diagnosis is such that survival for several months is likely, the long-term prognosis for good survival is excellent and is enhanced by treatment. It is concluded that radiotherapy has a definite beneficial effect and should be given to every child not presenting in poor or critical condition. These are best left untreated. An exception is a child with the diencephalic syndrome whose general condition is poor but there is little or no impairment of consciousness; such a child should be treated by radiotherapy sometimes preceded by a shunt operation. If there is increased intracranial pressure with radiological evidence to suggest that the obstruction may be relieved by operation, then partial removal should be carried out. If it is judged that obstruction cannot be relieved, a shunting procedure is required. Craniotomy is also indicated if there is anything in the clinical or radiological examination to suggest that the lesion may be extracerebral. Otherwise, biopsy through a burr hole may be adequate for confirmation of the diagnosis [17].

Case reports

Hidalgo et al., present the case of an adult patient with a benign OPG who underwent subtotal resection without adjuvant therapy and has had no tumor progression for more than 20 years. A 50-year-old woman presented with a 2-year history of personality changes, weight gain, and a few months of visual disturbances. Ophthalmological evaluation showed incomplete right homonymous hemianopsia. MRI demonstrated a 2.5 × 2.5 × 2.5-cm enhancing left-sided lesion involving the hypothalamus with extension into the suprasellar cistern, extending along the left optic tract and anterior to the level of the optic chiasm. A biopsy procedure revealed a juvenile pilocytic astrocytoma. A subtotal resection of approximately 80% of the tumor was performed. Postoperatively, the patient experienced complete resolution of her personality changes, and her weight decreased back to baseline. Ophthalmological examination showed increased right homonymous hemianopsia. In the years following her surgery, there was a spontaneous decrease in tumor size without adjuvant therapy. The patient continues to have an excellent quality of life despite a visual field defect, and no further tumor growth has been observed [18].

2015

Loh et al. present here the case of a 4-year-old boy with exophthalmos and near blindness due to an intraorbital OPG. Despite chemotherapy he showed progressive exophthalmos and vision loss. Bony orbital decompression with ON transection temporally reduced his exophthalmos. OPG resection was required later for recurrence of his exophthalmos secondary to tumor progression. Post operatively, he had preserved oculomotor nerve functions but developed globe ischemia. Unusually, his ischemic globe caused him to have pain and severe photophobia, which later lead to enucleation. Photophobia has been reported in blind patients. Animal models and MRI functional imaging showed activation of trigeminal pathway during photophobia in completely transected ON. However, the exact neuro-ophthalmology pathway requires further study.

This is the first described case of photophobia after excision of OPG with ON denervation. Photophobia can be a serious side effect that significantly lowers the patient's quality of life [19].

2013

Cavicchiolo et al. describe the case of a 3-year-old child, diagnosed with familial neurofibromatosis type 1 (NF1) and asymptomatic optic pathway tumor at the age of two, who developed diencephalic syndrome (DS) due to tumor progression 1 year after diagnosis. Magnetic resonance imaging disclosed an enlarging hypothalamic contrast-enhanced mass. Because of the tumor progression, in terms of tumor volume and DS, chemotherapy (CT) treatment was started according to the international protocol for progressive low-grade glioma, with rapid clinical improvement in terms of gain weight and DS resolution. Interestingly, tumor volume was unchanged after CT.

This case report highlights the following facts: (1) optic pathway glioma (OPG) in young children with NF1 may have definitive growth potentials and thus, they are worth an accurate clinical follow-up; (2) also, OPG occurring in NF1 patients can be responsible for DS in case of hypothalamus involvement; (3) consequently, the child's growth pattern must be included among the clinical parameters, which must be specifically evaluated during the follow-up of children, with or without NF1, bearing an OPG; and, finally, (4) that DS can improve after CT, even in face of a stable tumor volume [20].

Vyas et al. report a case of a hypothalamic glioma masquerading as a craniopharyngioma on imaging along with brief review of both the tumors [21].

Diffusion tensor imaging localization of the pyramidal tract and spectroscopy in diencephalic pilocytic astrocytoma: a case report [22].

Case report from the HGUA

A 5-year-old male with neurofibromatosis type 1 (NF1) who underwent biopsy of a left optic nerve glioma and port-a-cath placement. We discuss the multidisciplinary approach to management, surgical intervention, and postoperative care, highlighting both positive aspects and areas of concern in the patient's treatment course.

Case Presentation: A 5-year-old boy with a known diagnosis of NF1 presented with a left optic nerve glioma. The patient had been under multidisciplinary follow-up, including ophthalmology, neuropediatrics, and dermatology. Serial magnetic resonance imaging (MRI) scans showed significant tumor progression with increased infiltration and invasion of adjacent structures. The patient's left eye visual acuity decreased from 0.8 to 0.3 over the past few months.

Given the tumor's growth, a decision was made to perform a biopsy and place a port-a-cath for potential chemotherapy administration. The patient also has attention deficit hyperactivity disorder (ADHD), currently unmedicated, and an asymptomatic paravertebral plexiform neurofibroma.

Surgical Procedure: The patient underwent a left optic nerve glioma biopsy and port-a-cath placement under general anesthesia. The procedure was performed using standard microsurgical techniques to minimize trauma to surrounding structures.

Postoperative Course: The patient was admitted to the pediatric intensive care unit for close monitoring. Pain management was adequate, and no major complications were observed. A postoperative CT scan revealed a small hemoventricle, apparently without clinical repercussions. The patient required school support, indicating possible cognitive impact of the disease or treatments.

Discussion: This case highlights several important aspects in the management of optic pathway gliomas in NF1 patients:

1. The importance of regular multidisciplinary follow-up and serial imaging to monitor tumor progression. 2. The challenge of determining the optimal timing for surgical intervention in pediatric brain tumors. 3. The need for careful consideration of the risks and benefits of biopsy in optic pathway gliomas, given the potential for visual deterioration. 4. The complexity of managing concurrent conditions (such as ADHD) in the perioperative period.

The presence of a small hemoventricle postoperatively, while not clinically significant at present, warrants close follow-up. Urgent histopathological evaluation of the biopsy is crucial for guiding further treatment decisions, including the potential need for chemotherapy or radiotherapy.

Conclusion: This case demonstrates the complexities involved in managing optic nerve gliomas in children with NF1. While the immediate postoperative course was without major complications, the tumor's progression and visual deterioration are concerning. Moving forward, a balanced approach

considering both tumor control and preservation of function will be crucial. Close monitoring, potential oncological treatment based on biopsy results, and ongoing multidisciplinary care will be essential for optimizing this patient's outcome.

The management of optic nerve gliomas, particularly in the context of NF1, presents unique challenges and requires a tailored approach. In this case, several factors influenced the management strategy:

Observation vs. Intervention: Initial management often involves close observation with serial imaging and ophthalmological examinations. In this case, the decision to intervene surgically was based on:

Significant tumor progression on serial MRI scans Deterioration of visual acuity (from 0.8 to 0.3) The need for tissue diagnosis to guide further treatment

Surgical Approach:

The primary goal of surgery in this case was to obtain a tissue diagnosis rather than attempt complete resection. Biopsy of optic pathway gliomas carries risks, including potential further visual deterioration. The decision to proceed was based on the tumor's progression and the need for histological confirmation to guide therapy.

Port-a-cath Placement:

Concurrent placement of a port-a-cath demonstrates proactive planning for potential chemotherapy, reducing the need for future procedures.

Post-surgical Considerations:

Close monitoring in the pediatric ICU allowed for early detection of complications such as the small hemoventricle seen on postoperative CT. Pain management is crucial in pediatric neurosurgical cases and was reported as adequate in this patient.

Future Treatment Planning:

Histopathological results will guide the next steps in management. Options may include: a) Chemotherapy: Often the first-line treatment for progressive optic pathway gliomas in NF1 patients. b) Radiotherapy: Generally avoided in young NF1 patients due to the risk of secondary malignancies, but may be considered in certain cases. c) Targeted therapies: Emerging options such as MEK inhibitors show promise in NF1-associated tumors.

Visual Rehabilitation:

Given the visual deterioration, early involvement of visual rehabilitation services should be considered.

Neurocognitive Considerations:

The patient's ADHD and need for school support highlight the importance of neurocognitive assessment and support in the overall management plan.

Long-term Follow-up:

Regular MRI scans and ophthalmological examinations will be crucial for monitoring tumor response to treatment and visual function. The asymptomatic paravertebral plexiform neurofibroma also

requires periodic assessment.

This comprehensive approach to managing optic nerve gliomas in NF1 patients emphasizes the need for a multidisciplinary team, including neurosurgeons, neuro-oncologists, ophthalmologists, and rehabilitation specialists. The balance between tumor control and preservation of function remains a key consideration throughout the treatment course.

[1)](#), [2)](#)
https://radiopaedia.org/articles/optic-pathway-glioma
[3)](#)
A retrospective, single-center, cohort study of 176 patients (93 boys), aged 6 years (range, 0.2-18 years), with hypothalamic-pituitary lesions was performed. The lesions were craniopharyngioma (n = 56), optic pathway glioma (n = 54), suprasellar arachnoid cyst (n = 25), hamartoma (n = 22), germ cell tumor (n = 12), and hypothalamic-pituitary astrocytoma (n = 7). The most common presenting symptoms were neurologic (50%) and/or visual complaints (38%), followed by solitary endocrine symptoms (28%). Precocious puberty led to diagnosis in 19% of prepubertal patients (n = 131), occurring earlier in patients with hamartoma than in patients with optic-pathway glioma (P < .02). Isolated diabetes insipidus led to diagnosis for all germ-cell tumors. For 122 patients with neuro-ophthalmic presenting symptoms, the mean symptom interval was 0.5 year (95% CI, 0.4-0.6 year), although 66% of patients had abnormal body mass index or growth velocity, which preceded the presenting symptom interval onset by 1.9 years (95% CI, 1.5-2.4 years) (P < .0001) and 1.4 years (95% CI, 1-1.8 years) (P < .0001), respectively. Among them, 41 patients were obese before diagnosis (median 2.2 years [IQR, 1-3 years] prior to diagnosis) and 35 of them had normal growth velocity at the onset of obesity. The sensitivity of current guidelines for management of childhood obesity failed to identify 61%-85% of obese children with an underlying hypothalamic-pituitary lesion in this series.
[4)](#)
Raelson C, Chiang G. Chiasmatic-hypothalamic masses in adults: a case series and review of the literature. J Neuroimaging. 2015 May;25(3):361-4. doi: 10.1111/jon.12132. Epub 2014 Jul 4. PubMed PMID: 25039315.
[5)](#)
Pelc S. The diencephalic syndrome in infants. A review in relation to optic nerve glioma. Eur Neurol. 1972;7(6):321-34. PubMed PMID: 4659145.
[6)](#)
Pelc S, Flament-Durand J. Histological evidence of optic chiasma glioma in the “diencephalic syndrome”. Arch Neurol. 1973 Feb;28(2):139-40. PubMed PMID:4683151.
[7)](#)
Khan AA, El-Borai AK. Pilomyxoid astrocytoma presenting as diencephalic syndrome. J Ayub Med Coll Abbottabad. 2014 Oct-Dec;26(4):611-5. PubMed PMID: 25672198.
[8)](#), [21)](#)
Vyas S, Prabhakar N, Tewari MK, Radotra BD, Khandelwal N. Hypothalamic glioma masquerading as Craniopharyngioma. J Neurosci Rural Pract. 2013 Jul;4(3):323-5. doi: 10.4103/0976-3147.118790. PubMed PMID: 24250172; PubMed Central PMCID:PMC3821425.
[9)](#)
Sani I, Albanese A. Endocrine Long-Term Follow-Up of Children with Neurofibromatosis Type 1 and Optic Pathway Glioma. Horm Res Paediatr. 2017;87(3):179-188. doi: 10.1159/000458525. Epub 2017 Mar 27. PubMed PMID: 28346917.
[10)](#)
Alireza M, Amelot A, Chauvet D, Terrier LM, Lot G, Bekaert O. Poor prognosis and challenging in treatment of optic nerve malignant gliomas: A literature review and case report series. World Neurosurg. 2016 Oct 25. pii: S1878-8750(16)31072-5. doi: 10.1016/j.wneu.2016.10.083. [Epub ahead of print] PubMed PMID: 27793766.
[11)](#)
Millward CP, Da Rosa SP, Avula S, Ellenbogen JR, Spiteri M, Lewis E, Didi M, Mallucci C. The role of

early intra-operative MRI in partial resection of optic pathway/hypothalamic gliomas in children. Childs Nerv Syst. 2015 Nov;31(11):2055-62. doi: 10.1007/s00381-015-2830-3. Epub 2015 Jul 28. PubMed PMID: 26216059.
12)

Goodden J, Pizer B, Pettorini B, Williams D, Blair J, Didi M, Thorp N, Mallucci C. The role of surgery in optic pathway/hypothalamic gliomas in children. J Neurosurg Pediatr. 2014 Jan;13(1):1-12. doi: 10.3171/2013.8.PEDS12546. Epub 2013 Oct 18. PubMed PMID: 24138145.
13)

Zoli M, Mazzatenta D, Valluzzi A, Marucci G, Acciarri N, Pasquini E, Frank G. Expanding indications for the extended endoscopic endonasal approach to hypothalamic gliomas: preliminary report. Neurosurg Focus. 2014;37(4):E11. doi: 10.3171/2014.7.FOCUS14317. PubMed PMID: 25270130.
14)

Mandiwanza T, Kaliaperumal C, Khalil A, Sattar M, Crimmins D, Caird J. Suprasellar pilocytic astrocytoma: one national centre's experience. Childs Nerv Syst. 2014 Feb 25. [Epub ahead of print] PubMed PMID: 24566674.
15)

Varan A, Batu A, Cila A, Soylemezoğlu F, Balcı S, Akalan N, Zorlu F, Akyüz C, Kutluk T, Büyükpamukçu M. Optic glioma in children: a retrospective analysis of 101 cases. Am J Clin Oncol. 2013 Jun;36(3):287-92. doi: 10.1097/COC.0b013e3182467efa. PubMed PMID: 22547006.
16)

DeSousa AL, Kalsbeck JE, Mealey J Jr, Fitzgerald J. Diencephalic syndrome and its relation to opticochiasmatic glioma: review of twelve cases. Neurosurgery. 1979 Mar;4(3):207-9. PubMed PMID: 460550.
17)

http://jnnp.bmj.com/content/37/9/1047.full.pdf
18)

Hidalgo ET, McQuinn MW, Wisoff JH. Regression after subtotal resection of an optic pathway glioma in an adult without adjuvant therapy: case report. J Neurosurg. 2018 Jun 1:1-4. doi: 10.3171/2017.12.JNS172188. [Epub ahead of print] PubMed PMID: 29999469.
19)

Loh CK, Weis B, van Velthoven V, Reiff C, Rössler J. Photophobia in a blind eye after removal of a progressive orbital optic glioma with denervation. J Neurol Sci. 2015 Nov 15;358(1-2):522-4. doi: 10.1016/j.jns.2015.09.375. Epub 2015 Oct 3. PubMed PMID: 26474792.
20)

Cavicchiolo ME, Opocher E, Daverio M, Bendini M, Viscardi E, Bisogno G, Perilongo G, Da Dalt L. Diencephalic syndrome as sign of tumor progression in a child with neurofibromatosis type 1 and optic pathway glioma: a case report. Childs Nerv Syst. 2013 Oct;29(10):1941-5. doi: 10.1007/s00381-013-2109-5. Epub 2013 Apr 25. PubMed PMID: 23615855.
22)

Knorr Z, Leblond P, Baroncini M, Pruvo JP, Jissendi Tchofo P. Diffusion tensor imaging localization of the pyramidal tract and spectroscopy in diencephalic pilocytic astrocytoma: a case report. J Neuroradiol. 2013 Mar;40(1):68-70. doi: 10.1016/j.neurad.2011.12.002. Epub 2012 Feb 1. PubMed PMID: 22300962.