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(CASE REPORT)

THE ROLE OF RADIOTHERAPY IN THE MANAGEMENT OF OPTIC CHIASM GLIOMA: A CASE REPORT AND LITERATURE REVIEW

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ABSTRACT

Optic pathway gliomas are rare brain tumors that mainly affect children. They commonly present with progressive visual impairment. Radiotherapy remains an effective treatment option in selected cases, although its use in young children is limited by the risk of long-term complications. We report the case of a 6-year-old boy presenting with a one-year history of progressive bilateral visual impairment. Ophthalmological examination revealed severe visual loss, left convergent strabismus, bilateral nystagmus, and optic disc pallor. Brain magnetic resonance imaging (MRI) showed a 21 × 22 × 21 mm mass centered on the optic chiasm, predominantly extending to the right, with heterogeneous contrast enhancement. Multiple additional T2/FLAIR hyperintense lesions were suggestive of unidentified bright objects (UBOs), raising suspicion of neurofibromatosis type 1 (NF1), although no clinical features of NF1 were identified. No biopsy was performed. Following multidisciplinary discussion, the patient received intensity-modulated radiation therapy (IMRT), with a total dose of 54 Gy in 27 fractions. No severe treatment-related toxicity was reported. Follow-up MRI showed stable disease, with no significant change in the optic chiasm lesion or associated UBOs.

Key words: Optic Glioma; Optic Chiasm; Radiotherapy; IMRT.

1 INTRODUCTION

Optic pathway gliomas (OPGs) are rare, usually low-grade brain tumors that mainly affect children during the first decade of life. They can involve the optic nerves, optic chiasm, or optic tracts, sometimes extending to the hypothalamus [1,2].

Clinical presentation mainly includes progressive visual impairment, proptosis, strabismus, and nystagmus. Endocrine or neurological abnormalities may also occur, particularly in cases of hypothalamic involvement [1,2].

Diagnosis is based on ophthalmological examination and magnetic resonance imaging (MRI), which is the preferred imaging technique for assessing tumor location and extension. Biopsy is not always necessary, especially in patients with neurofibromatosis type 1 (NF1), when clinical and radiological findings are typical [1,2].

Treatment depends on visual function, tumor progression, age, and NF1 status. Stable tumors can be monitored, while chemotherapy is generally the first-line treatment for progressive or symptomatic disease. Surgery has a limited role because of the risk of visual damage [2,3].

Radiotherapy can provide effective tumor control but is generally reserved for selected cases because of the risk of long-term complications. Modern techniques, such as intensity-modulated radiation therapy (IMRT) and proton therapy, aim to reduce radiation exposure to surrounding healthy tissues [3,4].

2 CASE PRESENTATION

We report the case of a 6-year-old boy with no significant medical history who presented with a one-year history of progressive bilateral visual impairment.

On clinical examination, the patient was in good general condition. Ophthalmological examination showed severe visual impairment in both eyes, with a visual acuity of 0.5/10 in the right eye and no light perception in the left eye. Left convergent strabismus, bilateral nystagmus, optic disc pallor, and diffuse chorioretinal atrophy were also noted. No other cranial nerve abnormalities or sensorimotor deficits were found.

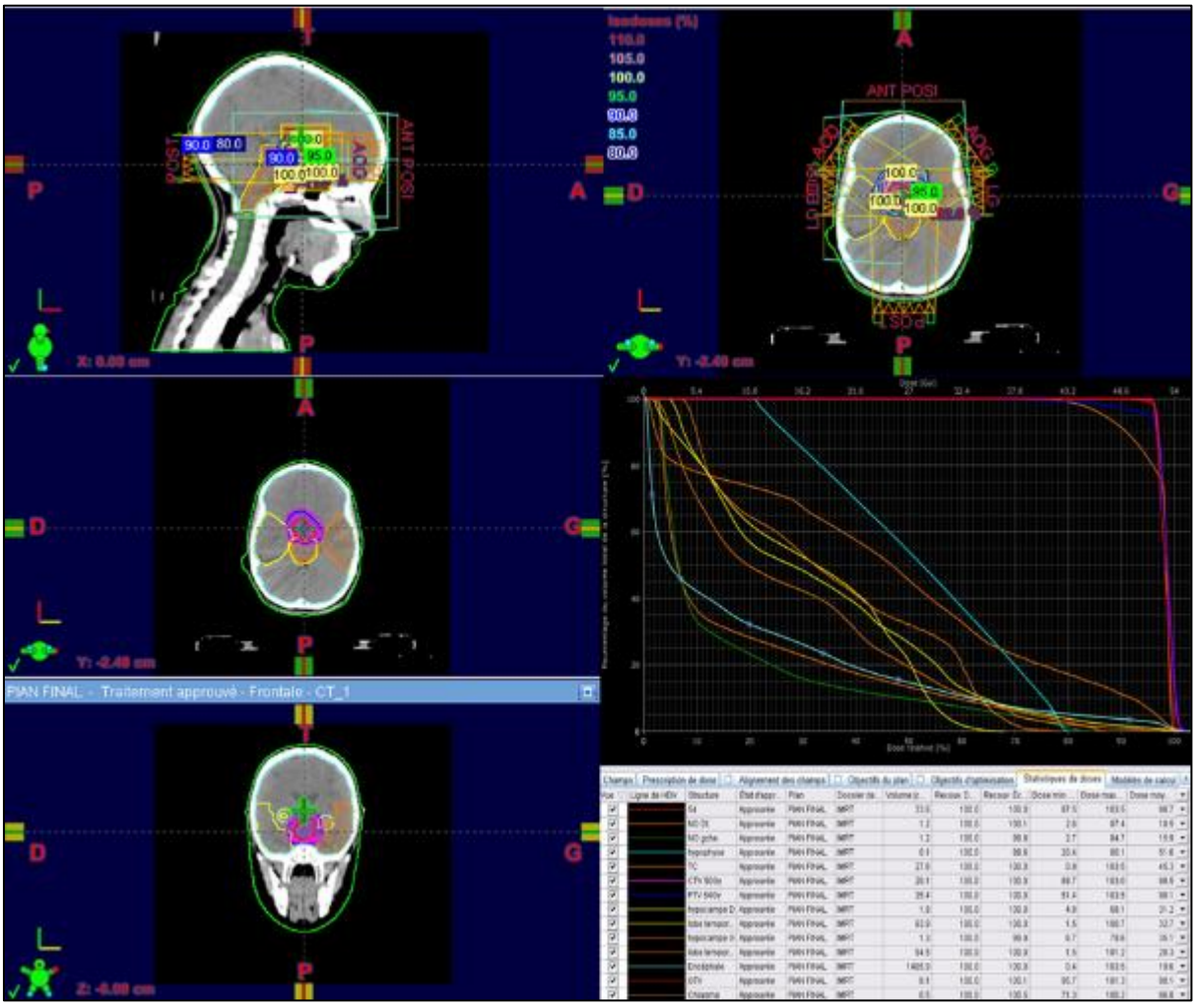


Figure 1: Radiotherapy treatment planning and dose evaluation.

Brain MRI revealed a 21 × 22 × 21 mm mass centered on the optic chiasm, predominantly extending to the right, with superior extension towards the mammillary bodies. The lesion showed an isointense signal on T1-weighted images, a hyperintense signal on T2/FLAIR sequences, and intense heterogeneous enhancement after gadolinium injection. Multiple T2/FLAIR hyperintense lesions were also seen in the bilateral hippocampal, internal capsular, midbrain, and cerebellar regions, as well as in the right parahippocampal region. These lesions were suggestive of unidentified bright objects (UBOs). Unfortunately, the original MRI images were not available for inclusion in this report.

The radiological findings were consistent with an optic chiasm glioma associated with multiple UBOs, raising suspicion of NF1. However, no clinical features of NF1 were found. No biopsy was performed, and the diagnosis was based on clinical and radiological findings.

The case was discussed at a multidisciplinary team meeting, and definitive radiotherapy was recommended.

A planning CT scan was performed with the patient in the treatment position, using a thermoplastic mask for immobilization. The patient received IMRT, with a total dose of 54 Gy delivered in 27 fractions (Fig. 1).

No severe acute or late toxicities were reported following radiotherapy. The latest follow-up MRI showed no significant change in the perichiasmatic tumor involving the optic chiasm. The previously described bilateral hippocampal, internal capsular, midbrain, and cerebellar lesions, as well as the right parahippocampal lesions, also remained stable and were still consistent with UBOs (Fig. 2).

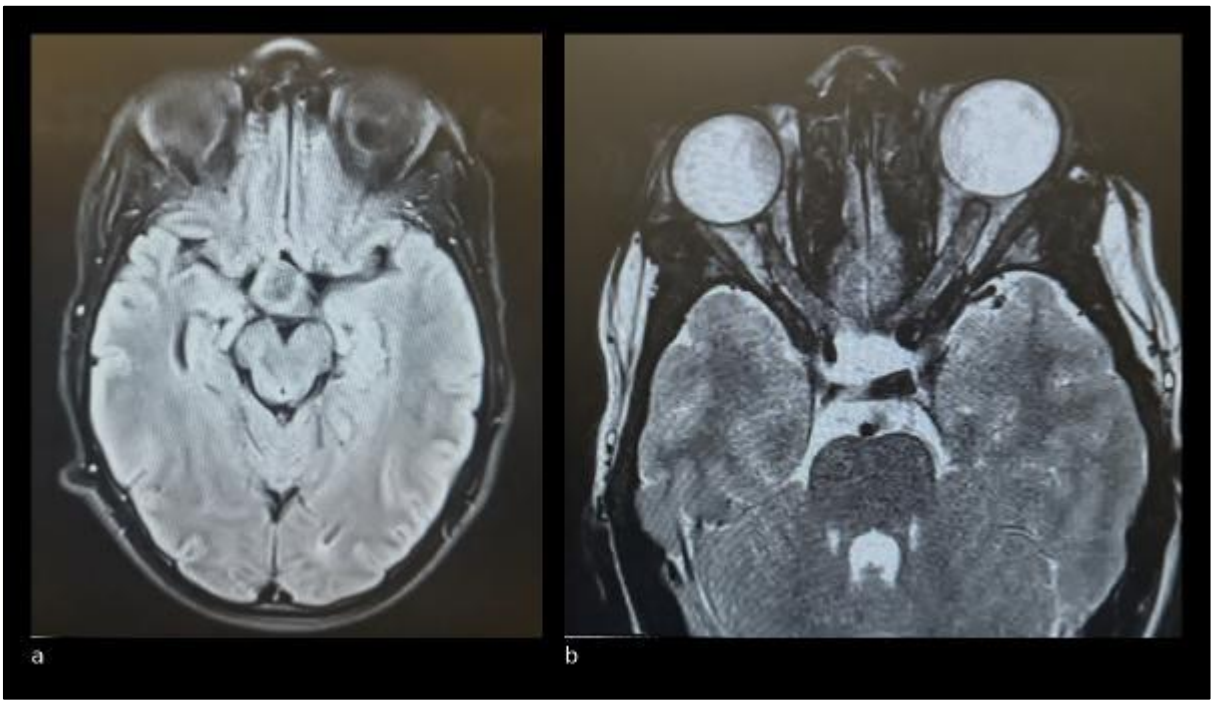


Figure 2: Follow-up brain MRI performed five years after radiotherapy. (a) Axial T2-weighted dark-fluid (FLAIR) sequence. (b) Axial T2-weighted TSE Dixon opposed-phase sequence, showing the perichiasmatic region.

3 DISCUSSION

3.1 Epidemiology and clinical presentation

OPGs are rare tumors, accounting for approximately 3–5% of pediatric central nervous system tumors. They mainly affect children during the first decade of life and are usually low-grade gliomas, most commonly pilocytic astrocytomas. They may involve the optic nerves, optic chiasm, or optic tracts, sometimes extending to the hypothalamus [1,3].

OPGs can occur sporadically or in association with neurofibromatosis type 1 (NF1). Approximately 15–20% of children with NF1 develop an OPG, although most do not require treatment. NF1-associated tumors generally have a more favorable course than sporadic tumors [1,3].

Clinical presentation depends on tumor location and extension. Visual impairment is the most common symptom and may be associated with strabismus, nystagmus, proptosis, and optic disc pallor. Hypothalamic involvement may cause endocrine disorders and growth disturbances. Other neurological manifestations are less frequent [1,3]. In their study of 95 children, Kebudi et al. also identified visual abnormalities, proptosis, and nystagmus as frequent presenting signs [3].

Our patient was a 6-year-old boy presenting with progressive bilateral visual impairment. Ophthalmological examination revealed severe visual loss, left convergent strabismus, bilateral nystagmus, and optic disc pallor, consistent with optic pathway involvement.

3.2 Diagnosis and imaging findings

The diagnosis of OPG is based mainly on clinical examination and MRI, which is the preferred imaging technique for assessing tumor location and extension. Typical findings include enlargement of the optic pathway, T2 hyperintensity, and variable contrast enhancement [1,2].

Ophthalmological evaluation includes visual acuity, visual fields, ocular motility, pupillary responses, and fundoscopic examination. Optical coherence tomography (OCT) can also help assess and monitor optic pathway damage [1].

Biopsy is usually avoided when clinical and MRI findings are typical, particularly in patients with NF1, because of the risk of visual damage. However, it may be considered in atypical cases or when the diagnosis is uncertain [1,2].

In our patient, MRI revealed a 21 × 22 × 21 mm mass centered on the optic chiasm, predominantly extending to the right, with heterogeneous gadolinium enhancement. No biopsy was performed, and the diagnosis was based on clinical and radiological findings. Therefore, the histological subtype could not be established.

MRI also showed multiple T2/FLAIR hyperintense lesions in different brain regions, interpreted as UBOs. Although these lesions are frequently associated with NF1, they are not specific to the disease. As our patient had no other clinical features of NF1, the diagnosis could not be confirmed, and further clinical or genetic assessment may be considered.

3.3 Therapeutic strategies

Treatment decisions depend on visual function, tumor progression, age, tumor location, and NF1 status. Patients with stable tumors and preserved visual function can be managed with regular clinical, ophthalmological, and radiological follow-up. Treatment is generally indicated in cases of significant visual deterioration or documented tumor progression [1,2].

Chemotherapy is currently the preferred first-line treatment for progressive or symptomatic OPGs in children. Carboplatin and vincristine are commonly used, while vinblastine, bevacizumab, and targeted therapies such as MEK inhibitors are alternative options in selected patients [1,3].

In their retrospective series of 95 children, Kebudi et al. reported that chemotherapy was the main treatment approach, while radiotherapy was used in only one patient following disease progression. The 10-year progression-free survival rate was 85.7% in patients with NF1 and 54.2% in those without NF1 [3]. These findings support the preference for chemotherapy and the need to limit radiation exposure in children whenever possible.

Surgery has a limited role because of the risk of permanent visual and neurological damage. It is mainly reserved for selected situations, such as severe proptosis, a blind painful eye, hydrocephalus, or when tissue diagnosis is necessary [1,2].

3.4 Role of radiotherapy

Radiotherapy has demonstrated effective long-term tumor control in pediatric OPGs. However, its use has decreased because of the risk of late complications, particularly in young children and patients with NF1. It is now mainly considered when other treatment options are unsuitable or have failed to control progressive disease [3,4].

Tsang et al. reported long-term outcomes in 89 patients treated with photon radiotherapy, generally using a total dose of 54 Gy. The reported 10-year progression-free survival rates ranged from approximately 62% to 68%, with overall survival rates between 92% and 98% [4]. These results demonstrate the potential of radiotherapy to provide durable tumor control.

Proton therapy has also shown encouraging results. Indelicato et al. evaluated 174 children with low-grade gliomas treated with proton therapy. At a median follow-up of 4.4 years, the 5-year local control, progression-free survival, and overall survival rates were 85%, 84%, and 92%, respectively [7]. However, this study included different tumor locations and was not limited to OPGs.

Conventional fractionated radiotherapy generally delivers 50.4–54 Gy, usually in fractions of 1.8 Gy [4]. In our patient, a total dose of 54 Gy was delivered in 27 fractions, corresponding to 2 Gy per fraction.

Modern techniques such as IMRT and proton therapy allow better dose distribution and reduce unnecessary irradiation of surrounding healthy tissues. Proton therapy offers a dosimetric advantage through the Bragg peak effect, which limits the exit dose beyond the target. However, neither technique completely eliminates the risk of toxicity, particularly when critical structures are close to the treatment volume [4,7]. Moreover, there are no strong randomized studies demonstrating the superiority of proton therapy over modern photon techniques specifically for OPGs.

In our patient, IMRT was selected following multidisciplinary discussion. The treatment delivered 54 Gy in 27 fractions and allowed conformal irradiation of the perichiasmatic lesion while limiting exposure to adjacent organs at risk.

3.5 Treatment-related toxicity

Radiotherapy in children may cause late complications, including visual deterioration, endocrine disorders, cerebrovascular events, neurocognitive impairment, and secondary malignancies. These risks depend on age at treatment, radiation dose, irradiated volume, tumor location, and NF1 status [4,6,7].

Endocrine disorders are mainly related to irradiation of the hypothalamic-pituitary region, while cerebrovascular complications, such as arterial stenosis and moyamoya syndrome, are particularly concerning in patients with NF1 [4,6].

In a long-term study of 182 childhood OPG survivors, Morin et al. reported that nearly 95% developed at least one late health condition after a median follow-up of 17.2 years. These included visual impairment, pituitary dysfunction, and neurocognitive difficulties [5].

In our patient, no severe treatment-related toxicity was reported. However, longer follow-up is needed to monitor for possible late complications.

3.6 Prognosis and follow-up

The overall prognosis of pediatric OPG is generally favorable, but visual outcomes remain variable. Prognosis depends on tumor location, hypothalamic involvement, NF1 status, age at diagnosis, and initial visual function [3,5,6].

In a 2025 nationwide study of 117 children, Bennebroek et al. reported severe visual impairment or blindness in both eyes in 18.8% of patients after a median follow-up of 8.3 years. Diagnosis before the age of two and hypothalamic involvement were associated with a higher risk of visual impairment [6]. Thus, tumor stability does not necessarily mean visual recovery.

Follow-up should include regular ophthalmological examinations and brain MRI. Visual acuity, visual fields, ocular motility, fundoscopic findings, and, when available, OCT should be assessed [1,2]. Long-term monitoring of endocrine function, growth, neurocognitive development, and cerebrovascular complications is also important, particularly after radiotherapy [5,6].

In our patient, the latest MRI showed a stable perichiasmatic tumor and unchanged UBOs, without significant regression. Continued ophthalmological, neurological, endocrine, and radiological follow-up is therefore required.

4 CONCLUSION

Pediatric OPGs require individualized management based on visual function and tumor progression. Although chemotherapy remains the preferred first-line treatment in children, radiotherapy can provide durable tumor control in selected cases. Modern techniques such as IMRT help limit radiation exposure to surrounding organs at risk, but long-term complications remain a concern. Our case illustrates radiological stability following IMRT and highlights the importance of careful treatment selection and prolonged follow-up.

STATEMENTS ON COMPLIANCE WITH ETHICAL STANDARDS

Disclosure of conflict of interest

All authors have no conflict of interest to declare.

Statement of ethical approval

This case report was conducted in accordance with ethical guidelines.

Statement of informed consent

The patient provided informed consent for the publication of this case report.

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