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Review Paper

Optic Pathway Glioma Treatment: A Mini-review of the Current Literature

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ABSTRACT

Background and Aim: Optic pathway glioma (OPG) is a chronic condition that needs a multi-disciplinary management strategy. Most of these tumors are observed in the pediatric population and the tumor tends to stabilize after the child's growth. This benign course can be observed mostly in neurofibromatosis 1 (NF1) patients, which are about half of the pediatric patients.

Methods and Materials/Patients: The current literature in PubMed and Scopus databases was searched. The recent data regarding OPG and treatment options were reviewed to design this narrative mini-review.

Results: The brief data extracted from 17 articles, cited in the reference list, were included in the study.

Conclusion: Chemotherapy is the first and best treatment modality for patients with OPG. It is more useful at younger ages because it has lower rates of complications and cancer in the future compared with radiotherapy, the treatment of choice in previous decades for these patients. However, in recent practice, it has been substituted by chemotherapy because of its serious adverse effects on the pediatric population. Neurosurgical treatments for OPG are used for three main purposes, third ventricle obstruction-related hydrocephalus, biopsy, and tissue diagnosis for cases with an uncertain diagnosis, and tumor decompression due to mass effect on vital structures. Surgical decompression is not considered the first-line treatment in OPG. It can be used for patients with progressive exophthalmos with ipsilateral blindness or patients with refractory pain after adjuvant treatment. This short review discusses the main aspects of OPG treatment modalities.

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Highlights

- Optic pathway gliomas (OPG) is a chronic condition that needs a multi-disciplinary management strategy.
- Chemotherapy is the first and best treatment modality for OPG patients.
- Previously, radiotherapy was preferred for treating these patients but recently, it has been replaced by chemotherapy.
- In OPG patients, radiosurgery can be a novel option.
- Neurosurgical treatments for OPG are used for third ventricle obstruction-related hydrocephalus, biopsy, and tissue diagnosis for cases with an uncertain diagnosis, and tumor decompression due to mass effect on vital structures.

Plain Language Summary

Optic pathway gliomas (OPG) are benign tumors. These tumors are often found in the pediatric population. According to the current experience to treat these lesions, chemotherapy is the best option. Surgery should be reserved for patients with special conditions. Radiotherapy was used as the main treatment modality in past decades but now it has been substituted by chemotherapy. Radiosurgery is another novel option for the treatment of these lesions which should be investigated in future research projects.

1. Introduction

Optic pathway glioma (OPG) is a chronic condition that needs a multi-disciplinary management strategy. Most of these tumors are observed in the pediatric population and the tumor tends to stabilize after the child's growth. This benign course can be observed mostly in neurofibromatosis 1 (NF1) patients, which are about half of the pediatric patients. With multi-disciplinary management, long-term survival is favorable for pediatric patients. For patients with small tumors without hydrocephalus and patients without visual impairment, observational management can be the best plan. Indications for medical or surgical interventions include raised (ICP) (intracranial pressure), progressive visual impairment, and tumor growth during follow-up imaging. Treatment options for these include chemotherapy, surgical management, and radiotherapy [1, 2].

2. Methods and Materials/Patients

The current literature in PubMed and Scopus databases was searched and all of the recent data on OPG and its treatment options were used in writing this review.

3. Results

The brief data extracted from 17 articles were entered into the study. They are cited in the reference list.

4. Discussion

Chemotherapy

Chemotherapy is the first and best treatment modality for patients suffering OPG. It is more useful at lower ages because it has fewer complications and less likelihood of future cancer development compared with radiotherapy. Chemotherapy can stabilize the growth of the tumor and prevent future neurosurgical procedures and radiotherapy. Various chemotherapy agents have been used to treat this condition but the most popular regimen contains vincristine and carboplatin. Chemotherapy can be used as a first-line therapy in patients with visual disturbances. 6-Thioguanine, procarbazine, and lomustine are also used as chemotherapeutic agents in these patients [1, 3]. After chemotherapy, visual acuity (VA) improved or remained stable on 59% and 61% of children with NF1 and sporadic OPGs, respectively. Sporadic OPGs present more often than NF1-OPG with progressive disease and associated vision loss [4]. Following treatment, 72% of children with NF1-OPG have improved or stable VA, while 74% of children with sporadic OPG have evidence of visual progression or tu-

mor [4-6]. Patients should be followed-up with clinical examinations, visual examination, and neuroimaging. The interval between follow-ups can be 3 months to 2 years, which can be chosen in a case-by-case manner. The vital visual examination is VA. For follow-up neuroimaging, MRI with contrast-enhanced T1 or T2 can be obtained [4, 7]. Optic atrophy, various visual symptoms in primary clinical manifestation, squint, proptosis, and posterior optic pathway are risk factors for visual deterioration after observation or treatment. NF-1 diagnosis is a positive predictor factor for visual outcome [8].

Radiotherapy

Radiotherapy was the treatment of choice in past decades for these patients, but in recent practice, chemotherapy preferred, due to the serious and unfavorable effects of radiotherapy on children. Also, a considerable number of patients with OPG are NF1 cases, who are more prone to cancers of the central nervous system (CNS). Radiotherapy should be saved as the last treatment modality for patients in whom previous treatment modalities have failed. Proton-beam therapy showed fewer late effects in the pediatric population in recent studies [3, 9].

Radiosurgery

Radiosurgery is another option that has been used also in hypothalamic low-grade gliomas after histological confirmation in recent case studies [10]. In OPG cases, radiosurgery can be a novel option. In a recent retrospective cohort, single fraction gamma knife radiosurgery was used for patients older than 10 years old, with a tumor located in the orbit, and VA<0.2. In this study, fractioned gamma knife surgery was used in 2-4 sessions in patients younger than 10 years old, VA>0.2, or tumors located in the hypothalamic or chiasmatic region [11]. In another study, 19 patients with OPG with clinical symptoms of vision loss were treated with gamma knife radiosurgery (GKRS) in three sessions at 12-hour intervals [12]. Single-session gamma knife surgery (GKS) was also reported as a safe treatment modality in another study [13]. Further research is needed on the safe use of radiosurgery in OPGs.

Surgery

Neurosurgical treatments for OPG are used for three main purposes, third ventricle obstruction-related hydrocephalus, biopsy, and tissue diagnosis for cases with an uncertain diagnosis, and decompression of tumor due to mass effect on vital structures [2, 14]. Endoscopic third ventriculostomy is a popular treatment modality in OPG that can be used for dual purposes, biopsy, and non-communicating hydrocephalus management. Septostomy should also be considered in all these patients, which can be useful in case of obstruction of Monro foramen in future tumor growth. Also, in patients who intend to undergo ventriculoperitoneal shunting, septostomy can be helpful for cases with Monro foramen obstruction and management of the patient with a ventriculoperitoneal shunt. Biopsy and tissue diagnosis are not mandatory in NF1 cases with typical imaging findings but should be considered in patients with NF1 with atypical imaging findings and non-NF1 patients, before adjuvant treatment. Various surgical biopsy strategies exist but endoscopic trans ventricular biopsy is the best modality due to the high rates of hydrocephalus in these patients [3, 14]. Chemotherapy is commonly selected as the first treatment when a tumor progresses, to avoid adverse effects on cognitive function caused by radiation or other therapies. If progression is associated with proptosis, and vision is poor/absent in that eye, debulking surgery should be considered. However, surgery invariably results in complete loss of vision in the involved eye [15-17]. Safe debulking of the exophytic part of the tumor is the main goal in the surgical management of these tumors and complete gross resection should be avoided [3, 14]. Aggressive resection could cause disastrous conditions associated with hypothalamic damage and endocrine and visual impairments. Depending on the tumor growth pattern, surgical decompression can be performed with different surgical routes. If the tumor has a major component in the third ventricle, the interhemispheric trans-callosal or trans-cortical F2 approach can be used. It is important to leave a peripheral rim of the tumor to avoid possible hypothalamic damage [1, 18, 19]. In tumors with a considerable lateral exophytic component, a subfrontal or pterional approach can be more useful. The subfrontal approach can be more useful in younger patients due to the lower relative volume of the frontal lobe compared to adults [18, 19]. Endoscopic endonasal approaches to sellar and parasellar pathologies have become more popular in recent years [20, 21], and OPG management with this approach has been reported in the literature [22].

5. Conclusion

This short review sought the fundamental aspects of treatment modalities for OPG. Chemotherapy should be the first and the main treatment modality in most patients, surgical decompression should be reserved for certain situations. Radiosurgery could be an appropriate

option for these patients but due to limited evidence, further investigations are needed.

Ethical Considerations

Compliance with ethical guidelines

There were no ethical considerations to be considered in this research.

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Conflict of interest

The author declared no conflict of interest.

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