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Optic Pathway Gliomas (Report of 12 Cases)

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ABSTRACT

Background: Optic pathway gliomas (OPG) represent approximately 3-5% of childhood intracranial tumors. They usually occur in children during the first decade of life and are seen in 11-30% of patients with neurofibromatosis Type 1 (NF1). Although these tumors are typically low-grade gliomas, the functional and vital prognosis can be challenging. The clinical course and natural history are highly variable, making treatment paradigms difficult. Indeed, the appropriate treatment for these tumors remains controversial, some authors prefer surgery, others radiotherapy or chemotherapy, while other offer forbearance. The aim of this work is to evaluate the therapeutic modalities and prognostic factors of glioma of the optical pathways.

Patients and methods: We report 12 cases of OPG, collected in our department between January 2008 and December 2016, whose clinical aspects, imaging data and therapeutic modalities we analysed.

Results: The mean age of patients at the time of diagnosis was 12.67 years (3-27 years) with a slight male predominance (58%). The main functional signs was the decrease of visual acuity (75% of cases), followed by intracranial hypertension syndrome with 58.3%, neurological disorders (hemiplegia and epilepsy) in 33.3% and endocrine signs (amenorrhea) in 16.7% of cases. Tumors were solid in 25% of cases and Solido-cystic in 75% of cases on MRI. Surgical removal was partial in 11 patients (92%) and total in 1 patient whose lesion affected only the optic nerve. Visual improvement was noted in 9 patients (75%), no neurological worsening or operative death was reported. Only one patient had presented 27 months after surgery, epilepsy leading to the death.

Conclusion: Once the diagnosis is made, there are four therapeutic attitudes abstention, surgery, chemotherapy and radiotherapy, whose indications must be studied on a case-by-case. Life expectancy is generally good in the long term, but the visual prognosis remains reserved.

Keywords: Optic pathway gliomas, Astrocytomas, Visual acuity

INTRODUCTION

Optic pathway gliomas are uncommon intracranial tumors and accounts for 3-5% of the children younger than 10 years in 75% [1,2]. The association with neurofibromatosis type 1 is particularly a common clinical entity (in up to 30% of cases) [3,4]. Histologically, most of these gliomas are benign tumors represented by pilocytic astrocytomas [5]. Visual disorders are the most challenging clinical sign. Despite technological progress adequate treatment approaches still controversial, involving surveillance, surgery, radiotherapy and chemotherapy [6]. The aim is to evaluate the therapeutic modalities and prognostic factors of OPG.

PATIENTS AND METHODS

Twelve patients with Optic pathway gliomas were treated in our department between January 2008 and December 2016. The mean age of patients at time of diagnosis was 12.67 years (3-27 years) with male predominance (58%). The mean time to diagnosis was 3 years for patients with optic nerve glioma, 5 months for those with chiasmal glioma and 14.8 months for those with opto-chiasmatic involvement. The main functional sign was the decrease of visual acuity (75% of cases). Four patients had visual acuity greater than 8/10 (33%), 4 patients had visual acuity less than 5/10 (33%), 1 patient had a positive perception of light (8%) and finally 3 patients had blindness (25%). Exophthalmia was found in two cases (17%), one patient had a convergent strabismus and another had a nystagmus. It should be noted

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that fundoscopic examination was abnormal in 75% (9 patients). Exophthalmos was reported in two patients (16.7%), intracranial hypertension syndrome in 58.3%, neurological disorders (hemiplegia and epilepsy) in 33.3% and endocrine signs (amenorrhea) in 16.7% of cases. It should be noted that none of our patients had associated neurofibromatosis. On MRI findings, the composition of the tumor was solid in 25% of cases and Solido-cystic in 75%. The average size of the tumor was 41.9 mm (ranged from 39 to 44.8 mm).We determined tumor sites on isolated lesion of the optic nerve (type 1=1 case/8% **(Figure 1)**), chiasmatic glioma (type 2=2 cases/17% **(Figure 2)**) and opto-chiasmatic glioma (type 3=9 cases/75% **(Figure 3)**).

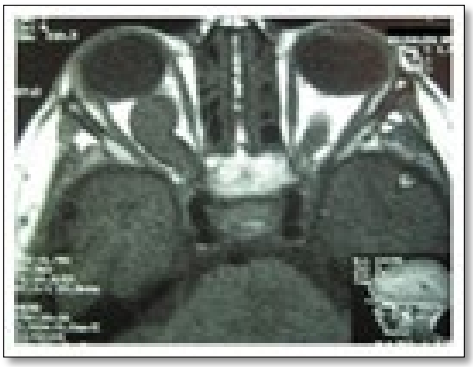


Figure 1. MRI: Right optic nerve glioma.

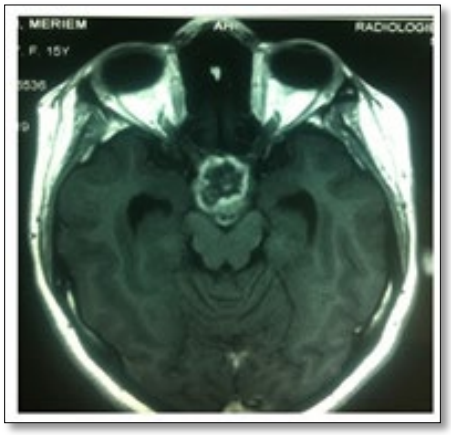
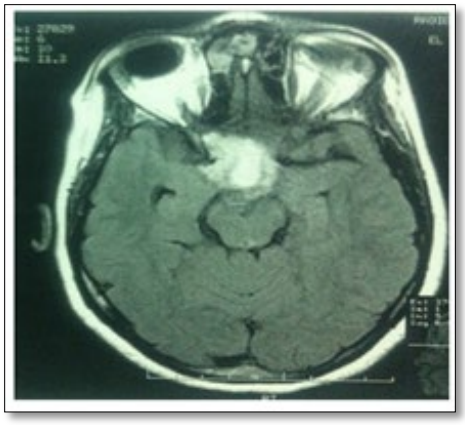


Figure 2. MRI: Chiasmatic glioma.



Figures 3. MRI: Right opto-chiasmatic glioma.

RESULTS

The 12 patients were operated through fronto-pterional or sub-frontal approach. Surgical removal was partial in 11 patients with chiasmatic and opto-chiasmatic glioma (92%). One patient whose lesion affected only the left optic nerve had total resection associated to its sacrifice. All tumors were pilocytic astrocytomas (grade I WHO classification).

No postoperative complications were noted. Two patients received radiotherapy (1 preoperatively and 1 postoperatively). Two patients received chemotherapy postoperatively. Visual improvement was reported in 9 patients (75%). No visual or neurological worsening was described. Only one patient had presented 27 months after surgery, epilepsy leading to the death. The results of the follow-up are illustrated in **Table 1**.

Table 1. The results of the follow-up.

Number 12 cases		Improvement/Stabilisation N (%) 10 cases	Aggravation N (%) 1 case	Death N (%) 1 case
Localization				
Type I	1	1 (8%/100%)	-	-
Type II	2	1 (8%/50%)	1 (8%)	-
Type III	9	8 (67%/89%)	-	1 (8%)
Age				
<3 years	1	1 (8%/100%)	-	-
4-10 years	4	2 (17%/50%)	1 (8%)	1 (8%)
11-15 years	4	4 (34%/100%)	-	-
>16 years	3	3 (25%/100%)	-	-
Surgery				
Total excision	1	1 (8%/100%)	-	-
Partial excision	11	9 (75%/81%)	1 (8%)	1 (8%)
Radiotherapy	2	1 (50%)	1 (50%)	-
Chemotherapy	2	1 (50%)	1 (50%)	-

DISCUSSION

Optic pathway gliomas represents 0.5 to 5.1% of all intracranial tumors [3,7] but 65% of primary tumors in the optic nerve [8]. This frequency is higher in children with neurofibromatosis type 1 (up to 30%) [4]. The average age was 12.67 years. In comparison, one of the most comprehensive studies published by DUTTON about 2297 cases; the average age was 8.8 years of which 90% of cases were under 20 years. The mean time to diagnosis is 3 years for our patients with optic nerve glioma, 5 months for those with chiasmal glioma and 14.8 months for those with opto-chiasmatic involvement [9]. In the BATAINI series [10], the average time between the first symptoms and diagnosis is estimated at 14.6 months. This delay varies according to the nature of the clinical signs and the tumor topography. According to Appeleton [11], only exophthalmia allows early diagnosis because it is easily recognizable. Our series reports a male predominance. This is not corroborated in the various series, which do not find either predominance of sex [12] or on the contrary, a slight female predominance [13]. Exophthalmia, visual loss and strabismus are the three most common symptoms [6]. The decrease of the visual acuity is the first revealing sign. It is usually a slow, gradual decrease that can be uni or bilateral [11,14]. When it is present, it is often very severe. In fact, 80% of Optic pathway gliomas have a low visual acuity less than 1/10 according to RUSH [15] and 67% of patients counted only the fingers, in the JAKOBIEC series [16]. It should be noted that visual acuity is not always quantifiable in very young children because of their lack of “cooperation” and it is a sign that is not constant. Papilledema was present in 2 cases (17%) and optic atrophy was noted in 2 others. Wright [17] notes in his study that papilledema is mainly present in evolutionary tumors. Oculomotor disorders are infrequent and occur in only 17.6% of optic pathway gliomas [18]. However intracranial hypertension (ICH) is the most common tell-tale sign of optic pathway gliomas by hydrocephalus [19]. It was noted in 7 of our patients (58%). ICH is a pejorative sign as reported by a recent study which shows that patients with ICH have a low survival rate compared to patients who do not have this symptom [6]. It may appear in the course of evolution and is related to the posterior chiasmatic extension, blocking the third ventricle and Monro's hole. Other neurological signs (hemiplegia, epilepsy, walking disorder, balance disorder) may sometimes exist [14]. Their presence is a poor prognosis factor. On the other hand, opto-

chiasmatic gliomas may be responsible for endocrine disorders by hypothalamic extension (early puberty, Russel syndrome, diabetes insipidus or anterior pituitary insufficiency) [20]. Lund [21] considers that the use of visual evoked response is more interesting than neuroradiological techniques for the detection of optic pathway gliomas, as they do not require sedation and can play an important role in monitoring in order to evaluate the visual function, residual visualization after surgery or for children for whom no treatment has been recommended. MRI allows a better diagnosisto monitor tumor progression and treatment response. Regarding the tumor localization, it is difficult to obtain reliable statistics. However, it appears that 25% of gliomas remain confined to the optic nerve and 75% reach the chiasm while extension to the optic nerve head and intraocular structures is rare [24]. Despite extensive literature, the treatment of optic pathway gliomas is controversial because of too small series, too short follow-ups, and the heterogeneity of the groups. Similarly, the unpredictable evolution of these tumors justifies the different therapeutic options proposed by different authors, ranging from simple observation to radiotherapy, chemotherapy, surgical resection and/or the combination of these different means [2,6,12,13]. The current approach is to start treatment when visual impairment is evident and/or tumor enlargement on imaging. In case of optic nerve glioma, the surgery must be as radical as possible and offer a definitive cure [23]. The decision to resect the optic nerve should be made in case of blindness and when the tumor is responsible for severe exophthalmos. Otherwise, in case of preserved or partially preserved vision or moderate exophthalmia, the “wait and see” policy should be adopted first [6]. The size and extent of the tumor influences the surgical management.When the tumor affects the optic chiasm, with an anterior extension to the optic nerves or posterior to the hypothalamus, the role of the surgery is not well clarified. These lesions respond better to radiotherapy [22]. To avoid worsening of visual function, a limited biopsy before radiotherapy is the best choice [6,27]. In our series, 11 patients underwent partial excision of the tumor with a visual improvement in 9 patients (75%). In addition, surgery can be indicated in case of differential diagnosis with others sellar tumors as craniopharyngioma. In case of surgery, blindness threatens 66.6% of children treated by partial resection [18,23]. Immediate postoperative complications are poorly documented in the literature but their frequency tends to decrease due to the advent of new neurosurgical techniques [24]. We did not note any of them in our series. Most of optic pathway gliomas are benign pilocytic and pilomyxoid astrocytomas [3].The role of radiotherapy is highly controversial and its usefulness remains to be proven. Some series report an improvement in visual acuity after radiotherapy [15]. Pierce [17], obtained 91% of good visual results after radiotherapy. However, other studies showed no improvement in survival or visual acuity and some researchers have even warned of the dangerous effects of radiation in children [25,26]. It is used at doses ranging from 4500 cGy for tumors limited to chiasma and 5500-6000 cGy for tumors invading the hypothalamus [27]. Chemotherapy, by offering a first-line treatment, may have a therapeutic effect, at least by delaying the indication of radiotherapy. Regarding the role of chemotherapy, Petronio [28] reports that all those for whom the tumor responded to chemotherapy had an improvement or stabilization of visual acuity. But other authors report that chemotherapy has not yet sufficient evidence of efficacy to be proposed as the first therapeutic choice [29].

On the other hand some publications highlight the slow and benign progression of this tumors and even mention cases of spontaneous regression and propose abstention with surveillance [27,30]. With rare exceptions, improvement of visual acuity is not observed with simple monitoring [29]. Tumor-related mortality is estimated at 5% with an average follow-up of 10 years for Dutton [13]. Only one of our patients (8.3%) had presented 27 months after surgery with generalized seizure with disorders of conscience leading to the death.

CONCLUSION

Besides surgery, there are currently three other therapeutic approaches, chemotherapy, radiotherapy and abstention. Tumor localization, age of the patient, clinical and radiological signs progression is important criteria that must be taken into consideration when managing optic pathway gliomas. Life expectancy is generally good in the long term, but the visual prognosis remains reserved.

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