

## **Radiotherapy in the management of optic pathway gliomas**

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**A retrospective study was performed in 28 patients with optic pathway gliomas treated at our institution between 1975-1995. The median age was 8 (range 1-55) years. The female-to-male ratio was 16:12. Seven patients (25%) had a history of neurofibromatosis. Twenty-two patients had histologic confirmation of the diagnosis and six patients were clinically diagnosed. Of the 22 surgically treated patients 12 (55%) had biopsy only and 10 (45%) underwent subtotal excision. Four patients showed tumor limited to optic nerve. Three patients had tumor located on chiasma only and nine patients were with tumors involving the chiasma and one or two optic nerves. More extensive involvement including involvement of hypothalamus and the third ventricle was recorded in 12 patients. Radiotherapy was applied with Co-60 unit or 6 MV photon beams and all patients received irradiation to local fields. The median dose was 5040 cGy in conventional fractions. The median follow-up time was 72 months. Ten-year overall survival and progression free survival were 93% and 84% respectively. The vision improved in 42% and remained stable in 58% of patients. We conclude that postoperative radiotherapy is beneficial in patients with chiasmal involvement and with incomplete resections. [Turk J Cancer 2000;30(1):31-35]**

**Key words: Optic gliomas, radiotherapy, childhood, adult**

Optic gliomas are rare tumors comprising 1-2% of all gliomas and their estimated incidence is 1 in 100.000 hospital patients (1-3). The disease usually occurs in childhood presenting with 75% in the first decade and 90% in the first 2 decades of life (2). Optic gliomas are often associated with type I neurofibromatosis (4).

Although most of these neoplasms are low grade astrocytomas with a long natural history, the untreated clinical course is that of progressive blindness or death from local tumor progression in a significant number of patients (5). Lesions that extent outside of the optic nerve to involve the chiasm and

neighboring structures can not be totally resected without substantial morbidity and in these patients radiotherapy is most useful (3,6).

### Materials and Methods

Twenty-eight patients were treated with a diagnosis of optic glioma at Hacettepe University, Faculty of Medicine, Department of Radiation Oncology between 1975-1995. The median age was 8 with a range from 1 to 55 years. The female-to-male ratio was 16:12. Seven patients (25%) had a history of neurofibromatosis.

Presenting signs and symptoms are shown in table 1. The duration of symptoms prior to presentation varied between 1 month to 8 years.

**Table 1**  
**Signs and symptoms at presentation**

	n	%
Optic atrophy	20	71
Strabismus	10	36
Blindness	24	86
Proptosis	5	18
Visual field defect	5	18
Headache	14	50
Nystagmus	4	14
Papilledema	4	14
Endocrine disorders	4	14

Twenty-two patients had histologic confirmation of the diagnosis and six patients were clinically diagnosed. Of the 22 surgically treated patients 12 (55%) had biopsy only and 10 (45%) underwent subtotal excision.

The extent of tumor showed a wide range. Involvement limited to optic nerve was seen in four (13%) patients. Three patients (11%) showed a tumor located on chiasma only and 9 (32%) patients were with tumors involving the chiasma and one or two optic nerves. More extensive involvement including involvement of hypothalamus and the third ventricle was recorded in 12 (44 %) patients.

Radiotherapy was administered from Co-60 teletherapy unit or 6 MV photon beams. All patients had local fields only to encompass the known disease with a 1-2 cm margin. A total dose of 4500-6000 cGy (median 5040 cGy) with conventional daily fractions was applied.

Statistical analysis: Statistical analysis was made using 'Life-table' method. Only 21 patients with at least 5 year follow-up time were included in the survival analyses.

## Results

Median follow-up time in 21 patients included in statistical analysis was 72 months. The overall and progression free survival at 10 years were 93% and %84 respectively (Figure 1). Two patients developed local recurrence; one at the eighth month and the other at the 60<sup>th</sup> month after radiotherapy. Both patients with local recurrence were with tumors involving hypothalamus or the third ventricle.

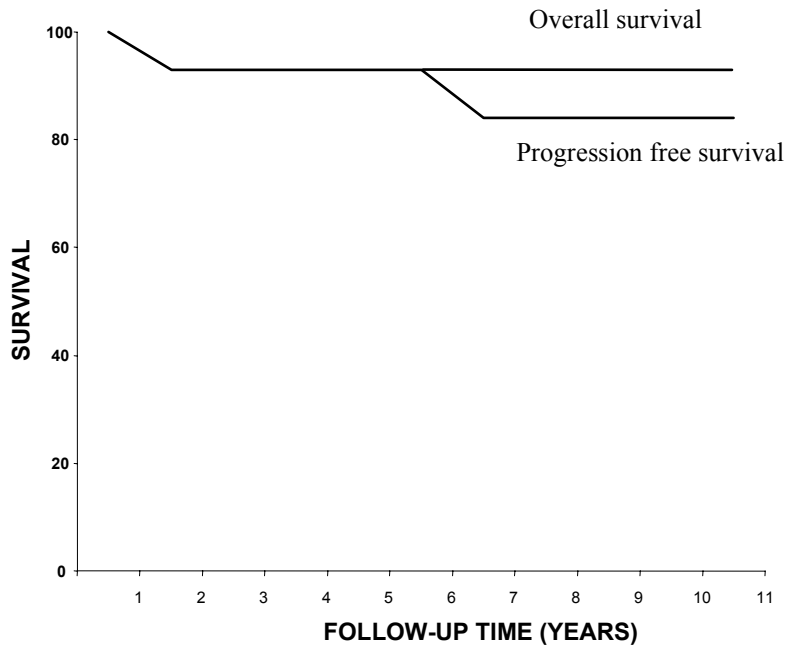


Fig 1. Overall and progression free survival

The vision following radiotherapy was assessed in 19 patients. Eight (42%) patients showed improved visual acuity while 11 (58%) patients were considered as stable after radiotherapy.

Four patients had endocrine disorders at initial presentation: two with diabetes insipidus and two with precocious puberty. All the four patients with endocrine disorders were without any sign of endocrine abnormality after radiotherapy till the time of this analysis.

## Discussion

Although first defined as benign hamartomatous growths in 1969 by Hoyt and Baghdassarian (8), optic gliomas are low grade tumors that progress locally or cause death in 75% of patients if left untreated (5). The most important factor in patients with optic glioma is the extent of disease at the time of diagnosis

(3,8). In large series from the Mayo Clinic (9) with a median follow-up time of 10 years, patients with glioma confined to the optic nerve survived almost twice as long as those with involvement of optic chiasm. Total surgical resection of the tumors limited to optic nerve only, leads to high long term survival up to 100% of 15 year progression free survival (3). Radiotherapy in the management of chiasmal lesions produce local control and survival advantage when compared to biopsy only (5,10-12). Tenny et al. (10) reported that only 3/14 (21%) patients with chiasmal gliomas survived following biopsy only when compared to 28/44 (64%) survival after radiotherapy. The majority of our patients included in the statistical analysis (18/21) had tumors involving chiasma and radiotherapy produced 93% 10 year overall and 84% progression free survival which is consistent with Kovalic et al. (3). The presence of neurofibromatosis in optic glioma patients has been suggested as a favorable prognostic sign (3,13). Only 4/16 patients in our series had neurofibromatosis and these patients were without any sign of progression at the time of analysis. Since only four patients were with neurofibromatosis, we could not estimate a prognostic value in this regard.

Radiation doses lower than 5000 cGy are reported to be associated with significantly lower progression free and overall survival rates (8). Only two patients who are younger than three years at the time of irradiation in our series received 4500 cGy. The remaining nineteen patients were treated with doses equal or more than 5000 cGy. Two patients in our series developed local recurrences. These patients were with tumors extending hypothalamus or the third ventricle and 5000 cGy external irradiation was applied. Since most of our patients received more than 5000 cGy, it is difficult to make a conclusion about the dose effect.

Radiotherapy leads to 9-44% of improved vision and 77% stable vision (1,14). Eight patients (42%) in our series were reported to have improved vision while 11 patients (58%) showed stable vision after radiotherapy which is consistent with the literature.

In conclusion, optic pathway gliomas involving chiasma and contiguous structures should be referred for radiotherapy either after debulking surgery or biopsy or clinical diagnosis. Our treatment policy is to treat these tumors with doses above 5000 cGy.

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