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MALIGNANT OPTIC GLIOMA OF CHILDHOOD

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SUMMARY

Optic glioma is known as a tumor of childhood. However, malignant glial tumors of the anterior visual pathway are rarely encountered in children, but in the elderly. The tumor presents with visual deterioration. Malignancy may occur primarily, or a preexisting low-grade glioma may become malignant following factors such as radiation therapy.

A 6-year-old girl presented with visual deterioration for about two months. Radiological evaluation revealed an intra, para and suprasellar lesion. The patient was operated on through a right pterional craniotomy and a subtotal tumor resection performed. The histopathologic diagnosis was malignant glioma of the optic chiasma.

Although considered to be a pathology of adults, malign optic glioma may occur in childhood. Presentation may occur with visual deterioration or endocrinological abnormality. Therapeutic intervention is similar to that of the management combination for a malignant glial tumor of the cerebral cortex and optic glioma.

Key Words: *Malignant Optic Glioma, Optic Glioblastoma Multiforme*

ÖZET

MALİNG OPTİK GLİOMA

Optik glioma genellikle çocukluk çağı tümörü olmasina rağmen anterior vizual yolun malign glial tümörleri az rastlanan bir patolojidir ve genellikle ileri yaşlarda ortaya çikarlar. Görmede bozulmayla kendini gösterir. Malignensi primer olarak ortaya çıkabilir veya düşük grade'li gliomada radyoterapi gibi bazi faktörleri takiben malign tümöre dönüsebilir.

6 yaşında kız çocuğu 2 aylık görme bozukluğu şikayetiyle başvurdu. Hastanın radyolojik değerlendirmesinde sellar, parasellar ve suprasellar lezyon tespit edildi. Hasta sağ pterional kraniotomiyle opere edildi. Subtotal tümör rezeksiyonu yapıldı. Histopatolojik tanı optik kiazmanın malign gliomu olarak geldi. Bu patoloji ileri yaşlarda seyrekde olsa görülebilir, fakat çocukluk çağında oldukça nadirdir. Hastalık görme bozukluğu veya endokrinolojik anormalliklerle kendini gösterir.

Anahtar Kelimeler: *Malign Optik Glioma, Optik Glioblastome Multiforme*

A malign optic glioma is a rather rare tumor that usually develops in adults. The tumor is located predominantly in the optic chiasm and presents with progressive loss of vision. In a verbal pre-school child, typical presentation is visual impairment with optic canal enlargement and optic atrophy. The clinical course is extremely fatal. In a review article Taphorn et al. reported 30 cases of malign optic glioma (1). Diagnosis depends on neuroradiological and opthalmological findings (2). We present a child with histologically confirmed malign optic glioma.

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Case Report

A 6-year-old girl complained of progressive deterioration of vision in both eyes for two months. On examination, she was cooperative and alert. Opthalmological examination revealed ambylopia of the right eye. Visual acuity was decreased in both eyes to 5/20, and a visual field examination showed bitemporal hemianopsia, indicating the involvement of the chiasm. Optic discs were normal. On T1-weighted Magnetic Resonance Imaging (T1W-MRI), enhanced with gadolinium, a lesion was observed in the chiasmatic region, extending both upward and downward and heterogenously. In the center of the lesion there was a hypointense region, indicating necrosis (Figures 1a, 1b). A malignant glioma of the optic tract was suspected, and a right pterional craniotomy was performed with the partial resection of the tumor. As expected, the tumor arose from the chiasma and infiltrated both temporal lobes and the hypothalamus. The part infiltrating the hypothalamus and temporal lobes was dissected and resected. There were necrotic and hemorrhagic areas in the center of the tumor. Histological evaluation revealed a glioblastome multiforme with anaplastic hyperchromatic cells, atypical mitoses, vascular proliferation and areas of necrosis. In the postoperative course, the patient developed abundant metabolic disorders, including resistant hypernatremia (up to 175mg/dl) and diabetes insipidus, and her general condition worsened. She died on the postoperative sixth day.

Discussion

Gliomas of the anterior visual pathway are rare orbital lesions occurring principally among children in the first decade of life. They appear to be true neoplasms that characteristically show early growth, followed by stability in many patients. Visual prognosis is fair, and the outlook for life depends on tumor location. When initially confined to the optic nerve alone, overall mortality is about 5%. Once the hypothalamus becomes involved, mortality rises sharply to 50%. With involvement of the chiasm or hypothalamus, no form of therapy significantly alters the final outcome. Because of their indolent course, gliomas may be conservatively followed when confined to the optic nerve. In these cases, surgery is indicated only when blindness and pain or severe proptosis intervene. However, all such cases should be followed radiologically for evidence of posterior extension. When the chiasm is threatened, surgical excision is warranted to prevent subsequent hypothalamic or third ventricle involvement.

Malignant optic glioma is a distinct disease primarily affecting middle-aged adults. The chiasm is always involved, and rapid progression to bilateral blindness is usual. The disease is uniformly fatal (3,4). When occurring in a verbal pre-school child, the typical presentation is visual impairment with optic canal enlargement and optic atrophy. An intraorbital location leads to axial irreducible, nonpulsatile proptosis, whereas an intracranial location may disturb hypothalamic and pituitary function and produce hydrocephalus. Ocular findings may include limited motility, a pupillary-related afferent defect, nystagmus and nonspecific field defects. Roentgenographic studies may show concentric unilateral enlargement of the optic canal with preservation of a well-corticated margin, a fossa under the anterior clinoid process in continuity with the optic canal (J-shaped sella) and increased intracranial pressure (3,5).

Malignant gliomas of the optic nerve and chiasm are extremely rare, but may develop after radiotherapy for a preexisting malignancy (6). Although malignant optic glioma patients receive radiotherapy and chemotherapy, these measures do not significantly improve survival rates. The infiltrative nature of the tumor into the normal brain and the presence of tumor foci in regions far from the main tumor burden make cure using current therapies virtually impossible. Management therefore consists of tumor control and maintaining the patient's quality of life. Craniotomy decreases the overall tumor burden and provides room for normal brain, edema and recurrent tumors (3,6,7,8,9).

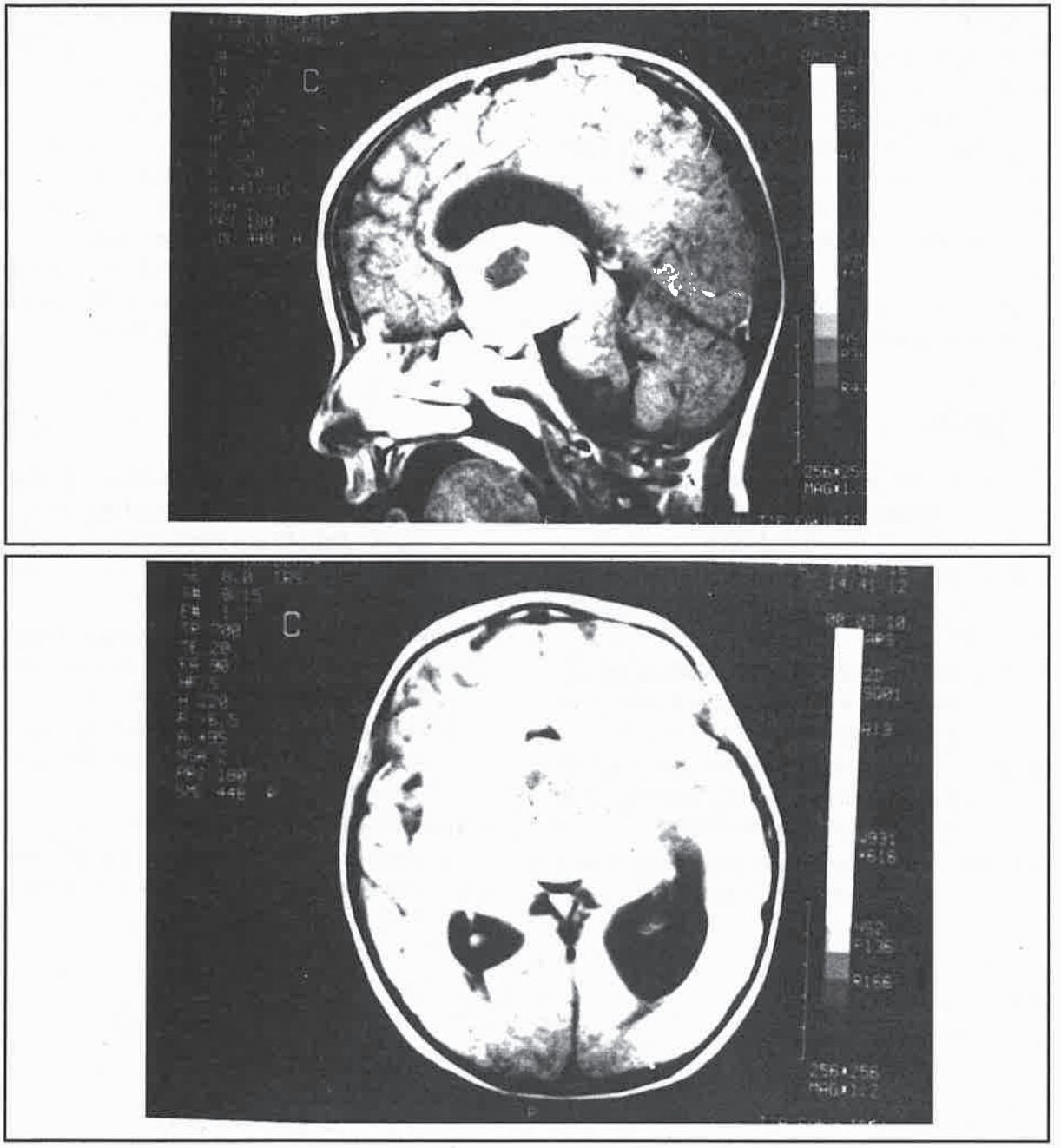

Figure 1: TIW MRI (a: sagital, b: coronal) sections of the tumor enhanced after GDTPA administration; the central hypointensity, indicating necrosis, should be noted.

Our case is unique in that the malignant form of the tumor is extremely rare in childhood. Presentation and radiological evaluation is no different than in adult cases, but the postoperative course is worse in children than in adults. Progression is similar to that of a craniopharyngioma infiltrating the hypothalamic region; therefore, a surgeon who opts for a radical resection of the lesion must keep in mind the presence of postoperative metabolic changes and may require the assistance of neuroendocrinological expertise.

Careful pre- and postoperative neuroendocrinological and opthalmological evaluation is necessary. Since our patient did not survive the early postoperative course, we cannot evaluate how radiotherapy and chemotherapy might have affected the outcome. However, the literature on adult cases of malign optic glioma indicate such measures have little effect on increasing survival rates.

In conclusion, childhood malign optic gliomas are rather rare and present with a variety of visual problems. Once the lesion infiltrates the hypothalamus, the course is usually fatal. The postoperative course may be complicated by endocrinological abnormalities that increase morbidity and mortality. Postoperative radiochemotherapy has not played a significant role in affecting the general outcome. However, surgical planning may affect the postoperative course. Attempts at total resection may increase morbidity and mortality by decreasing vision and causing postoperative endocrinological problems.

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