MALIGNANT OPTIC GLIOMA OF ADULTHOOD

Pandey MK¹*, Mittra P², Kumar A¹, Yadav AK¹

1. Department of Medicine, Rohilkhand Medical College, Bareilly (U.P.) INDIA
2. Department of Pathology, Rohilkhand Medical College, Bareilly (U.P.) INDIA

Correspondence: Dr. Manmohan Krishna Pandey. Department of Medicine, Rohilkhand Medical College, Bareilly (U.P.) INDIA
Email: drmkp12@yahoo.com


ABSTRACT

Optic pathway glioma is a slow growing neoplasm. Malignant optic glioma of adulthood is a very rare entity. We are describing a case of malignant optic glioma in a female presented with eight weeks history of headache and blurred vision with characteristic radiological features in MR imaging of brain.

Key words: Optic glioma, optic nerve

INTRODUCTION

Optic pathway gliomas are slow growing neoplasms that may cause visual or neurological complications and death. Gliomas of the optic pathway are two types: (1) the relatively benign optic nerve glioma - occurs in pediatric population and (2) the malignant optic glioma of adulthood ¹. The more common benign optic nerve glioma is considered a low grade astrocytoma ². ³ and is frequently associated with neurofibromatosis type 1.

CASE REPORT

A 26 year female was presented with a eight week history of blurred vision in both eyes with off and on headache. She was unable to sit properly on bed and maintain erect posture as she had tendency to fall. Physical examination was with in normal limit. Pupillary reflex (both direct and consensual) was normal in both eyes. Exact visual acuity could not be tested as patient was unable to sit properly. MR imaging of brain revealed a large heterogeneous necrotic lesion with mixed cystic and solid component in the midline suprasellar region with the optic chiasma, nerves were not seen separately causing marked splaying of the cerebral peduncle and medial temporal lobes with edema in the ganglio-capsular region on both sides suggestive of optic glioma (Figure 1 and 2). Patient was treated symptomatically with steroid and mannitol and referred to advanced neurology center.
Figure 1. MR brain scan(sagittal view).
Figure 2. MR scan brain (Cross-sectional View).
DISCUSSION

Malignant optic glioma of adulthood is an extremely rare optic pathway tumor and are not associated with neurofibromatosis. Malignant optic glioma of adulthood is classified pathologically as either an anaplastic astrocytoma or a glioblastoma multiforme. Differential diagnosis of optic glioma are conditions which enlarge the optic nerve without usually affecting the optic nerve sheath (optic neuritis, optic nerve ischemia) and conditions which enlarge the optic nerve sheath without usually affecting the optic nerve are meningioma and pseudotumor. Conditions which affect optic nerve and sheath both are optic glioma, perioptic neuritis from infectious and granulomatous causes, leukemia, lymphoma, and metastatic disease.

Exact origin of malignant optic glioma of adulthood is controversial because of its rarity and rapid progression. Malignant optic glioma of adulthood involving the optic chiasm may directly invade the hypothalamus, basal ganglia, and internal capsule. Leptomeningeal and sub-pial spread of malignant optic glioma of adulthood to the medial temporal lobes and brain stem has been reported. The diagnosis of optic glioma depends on clinical recognition of an optic nerve or chiasmal lesion followed by appropriate neuroimaging. Orbital and cranial computed tomography scans of optic gliomas may show fusiform enlargement of the optic nerve, kinking of the optic nerve, or enlargement or enhancement of the optic nerve, chiasm, or retrochiasmal visual pathways. Magnetic resonance imaging is superior for demonstrating these findings in optic glioma and documenting intracranial involvement (hypothalamic and retrochiasmal involvement and hydrocephalus). In some cases, there is a hemorrhagic, cystic, or exophytic (extrinsic) component to the lesion. Magnetic resonance images are superior to computed tomography scans for defining the extent of intracanalicular or intracranial involvement (in the hypothalamus and along the optic tract for example) and also may be useful in the identification of other intracranial lesions suggestive of NF1. Typical neuroimaging in the appropriate clinical presentation is sufficient for diagnosis to obviate the need for tissue confirmation via biopsy sampling, but some atypical cases may require pathological confirmation.

Consideration of malignant optic glioma of adulthood in the differential diagnosis of optic neuritis, particularly in older patients, may alter the course of rapidly fatal malignancy. Because early physical signs are usually absent in the syndrome of malignant optic glioma of adulthood, the role of MR imaging in the early diagnosis of this disease assumes greater importance. Major goal in managing cases of optic glioma is to preserve vision for as long as possible, initial observation is recommended for all cases of optic glioma. Continued observation is generally recommended for optic glioma that do not progress clinically or radiographically. Sometimes optic glioma may spontaneously regress. The tumor-related mortality rate for optic nerve glioma (ONG) is low (probably < 6%) and presumably occurs from intracranial extension. Patients with ONGs treated with either complete or partial excision followed by radiotherapy suffered vision loss as a complication. Thus, surgery alone is probably counterproductive in ONG if the goal is to preserve vision. Patients with no useful vision, severe eye-threatening proptosis, or a blind, painful eye might benefit from gross-total resection.
As compared to optic nerve gliomas (ONGs), optic chiasmal gliomas (OCG) are not generally amenable to complete excision without having bilateral visual loss. Debulking of the exophytic or cystic component of the OCG can be considered in individual cases (especially in those with rapid progression), but the indications for surgery are not well defined. Patients with OCGs or hypothalamic gliomas who develop hydrocephalus may benefit from shunt placement. As in ONGs, the general recommendation for both OCGs and hypothalamic gliomas is initial clinical and radiographic observation for evidence of progression.

Chemotherapy is useful for delaying the need for radiotherapy in progressive OPGs. Carboplatin-based regimens such as carboplatin and vincristine have been the most commonly used. The carboplatin regimens also appear to be relatively well tolerated, but neutropenia, thrombocytopenia, and allergic reactions may occur. Although chemotherapy may delay progression or stabilize visual function in patients with progressive OPGs, there remains a significant rate of progression or recurrence after chemotherapy.

CONSENT

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

COMPETING INTERESTS

The authors declare that they have no competing interests.

REFERENCES