CASE REPORT

Intracranial Glioblastoma Multiforme (Stage IV Glioma) Presenting as Papilledema: Two Case Reports

Basavashree Chimkode, Bharathi Megur, Deepak Megur, Geeta S Bandi

ABSTRACT

Glioblastoma multiforme (GBM) is fast growing invasive gliomas often referred to as Grade IV glioma, arising from astrocyte spreading rapidly into nearby brain tissue and occur most commonly in temporal and frontal lobes. Here, we present case reports of two young female patients who presented to an eye hospital with predominant complaints of a headache and blurred vision, without any other neurological deficits. Comprehensive evaluation revealed intracranial frontal lobe mass lesion (GBM) causing severe papilledema and secondary optic atrophy.

KEY WORDS: Glioblastoma multiforme, headache, loss of vision, papilledema

INTRODUCTION

Glioblastoma multiforme (GBM) is a type of malignant brain tumor. It is the most common primary brain neoplasm in adults and has a frequency of 50%.[1-3] They arise from astrocytes - the star-shaped cells that make up supportive tissue of the brain. These tumors are often aggressive and infiltrate the surrounding brain tissue. They are more common in the United States, Scandinavia, and Israel than in Asia, accounting for approximately 12-15% of all intracranial neoplasms and 50-60% of all astrocytic tumors. The incidence increases with age (45-70 years)[4] and affects more men than women (2:1 ratio).[5]

CASE REPORTS

Case 1

An 18-year-old girl presented with severe headache of 3 months duration which was associated with episodes of vomiting and drowsiness. During the onset of headache, the patient also had blurring of vision and diplopia. She presented to us 15 days after the onset of symptoms. On examination, there was 15° of esotropia and relative afferent pupillary defect in the left eye which was associated with bilateral lateral rectus palsy. Her best corrected visual acuity in the right eye was less than 20/200 and no perception of light in the left eye. Fundus examination established papilledema in both the eyes [Figure 1].

Magnetic resonance imaging (MRI) brain revealed a well-defined hypointense mass lesion measuring 4.5 cm × 3.4 cm × 4.5 cm in left frontal region. Mass lesion was hyperintense on T2W images, the images with contrast showed eccentric peripheral thick wall enhancement, suggestive of left frontal region GBM [Figure 2].

Case 2

A 35-year-old female presented to us with complaints of double vision since 1 month which was associated with severe headache and vomiting with progressive loss of vision in both the eyes over a period of 4-month. Her best corrected visual acuity was less than 20/200 in the right eye and counting fingers close to face in the left eye. Anterior segment examination revealed sluggishly reacting pupils on both sides with exotropia of the left eye with normal ocular movements in both the eyes. Fundus examination revealed bilateral chronic (vintage) papilledema with pale discs, optociliary shunts, and dilated tortuous vessels [Figure 3].

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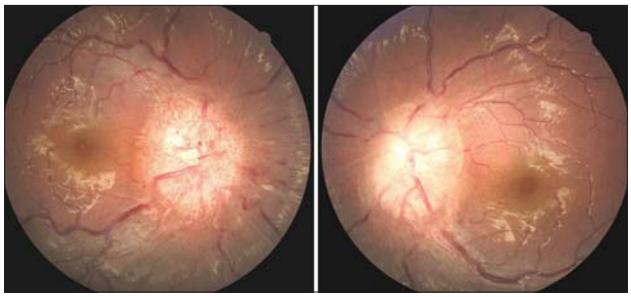

Figure 1: Fundus photograph showing established papilledema

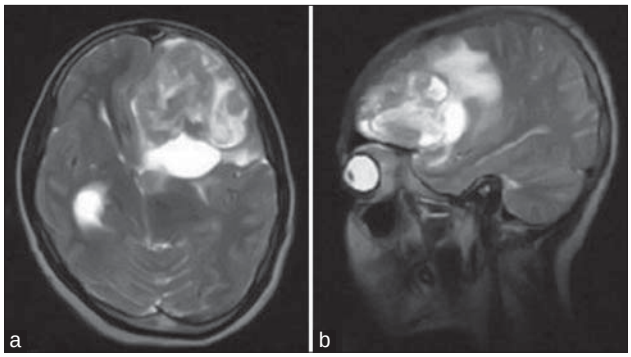

Figure 2: (a and b) Magnetic resonance T2-weighted images showing well-defined hyperintense mass lesion measuring 4.5 cm × 3.4 cm × 4.5 cm in left frontal region

MRI brain plain and contrast showed large ill-defining heterogeneous enhancing lesion in the left frontal lobe abutting callosum with perilesional edema suggestive of GBM [Figure 4].

DISCUSSION

GBM is by far the most common and most malignant of the glial tumors. GBMs are mainly composed of star-shaped glial cells known as astrocytes, which are cells that form the tissue that surrounds and protects other nerve cells found within the brain and spinal cord. Gliomas are classified according to a grading system developed by the World Health Organization. Grade IV gliomas are known as GBM.

The exact etiology of GBM has not been fully elucidated. The familial form of this tumor is described for 1% of cases. Ionizing radiation is one of the factors, which increases the risk.

About 90% of GBM cases develop *de novo* (primary glioblastoma)[6] from normal glial cells by multistep tumorigenesis. They are very aggressive and rapid growth of the lesion is seen sometimes within

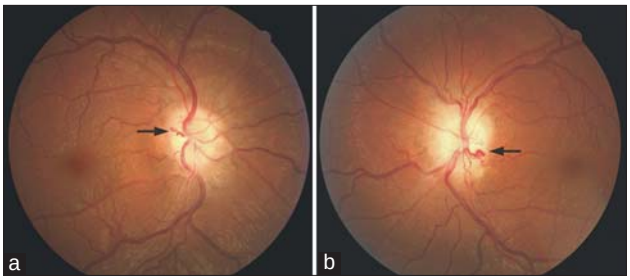

Figure 3: Fundus photograph showing chronic papilledema with optociliary shunts (black arrow)

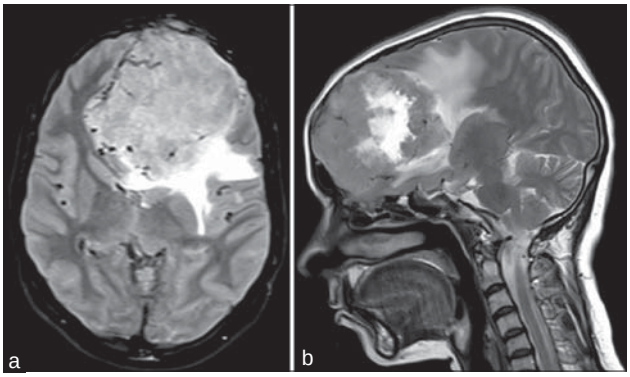

Figure 4: (a and b) Magnetic resonance imaging T2-weighted images showing large, ill-defined heterogeneous, hyperintense lesion measuring 75 mm × 85 mm in left frontal lobe abutting callosum with perilesional edema and midline shift to right side

3 months. The remaining 10% of gliomas are cases of secondary neoplasm, developing through the progression from low-grade tumors, diagnosed mostly in persons with mean age of 39 years and grow more slowly and have a better prognosis.

Clinical findings depend on the location, size, and rate of growth of the tumor, as with any other central nervous system tumor. Tumors in less critical areas (e.g., anterior frontal or temporal lobe) may present with subtle personality changes and memory problems which were not observed in our both patients. The clinical history is usually short (<3 months in >50% of patients). Common presenting symptoms are usually caused by raised intracranial pressure causing headache, nausea, vomiting, drowsiness, and frequent syncope. Depending on the location of the tumor, patients can develop weakness on one side of the body, memory, and/or speech difficulties and visual changes such as loss of vision and diplopia. In contrast, both our patients presented mainly with chronic severe headache and visual problems without any neurological deficits. One of our patients had

bilateral optociliary shunts, which have been reported with other conditions such as central retinal vein occlusions,[7] optic nerve meningioma,[8] and optic nerve glioma.[9]

MRI is the primary diagnostic tool for GBM. The tumor involving the corpus callosum and growing bilaterally into occipital and temporal lobes results in a butterfly pattern on MRI. Definitive diagnosis is based on histopathological examination of the intraoperatively removed tumor parts.

Glioblastoma can be difficult to treat because the tumors contain so many different types of cells. This is why the treatment plan for glioblastoma may combine several approaches. The standard treatment option is surgical resection to the extent feasible followed by radiotherapy and chemotherapy. Although current therapies remain palliative, they have been shown to prolong quality survival. Mean survival is inversely correlated with age. Without therapy, patients with GBM uniformly die within 3 months. Patients treated with optimal therapy, including surgical resection, radiation therapy, and chemotherapy, have a median survival of about 12-19 months[3,5] and 2 years survival is 30%.

Recurrent gliomas bear an even more dismal outcome than primary lesions. Newer agents such as intravenous bevacizumab (anti-vascular endothelial growth factor) and irinotecan have been tested in recurrent gliomas, in a Phase II setting. Based on the promise shown by bevacizumab in the western data, it is soon going to be tested in a Phase III trial in GBM.[10]

REFERENCES

1. Demir MK, Hakan T, Akinci O, Berkman Z. Primary cerebellar glioblastoma multiforme. Diagn Interv Radiol 2005;11:83-6.
2. Mattos JP, Marenco HA, Campos JM, Faria AV, Queiroz LS, Borges G, *et al.* Cerebellar glioblastoma multiforme in an adult. Arq Neuropsiquiatr 2006;64:132-5.
3. Stark AM, Nabavi A, Mehdorn HM, Blömer U. Glioblastoma multiforme-report of 267 cases treated at a single institution. Surg Neurol 2005;63:162-9.
4. Levin VA, Leibel SA, Gutin PH. In: DeVita VT Jr, Hellman S, Rosenberg SA, editors. Neoplasms of the Central Nervous System. Cancer: Principles and Practice of Oncology. 6th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2001. p. 2100-60.
5. Dohrmann GJ, Dunsmore RH. Gliobastoma multiforme of the cerebellum. Surg Neurol 1975;3:219-23.
6. Ohgaki H, Kleihues P. Genetic pathways to primary and secondary glioblastoma. Am J Pathol 2007;170:1445-53.
7. Giuffrè G, Palumbo C, Randazzo-Papa G. Optociliary veins and central retinal vein occlusion. Br J Ophthalmol 1993;77:774-7.
8. Hollenhorst RW Jr, Hollenhorst RW Sr, MacCarty CS. Visual prognosis of optic nerve sheath meningiomas producing shunt vessels on the optic disk: The Hoyt-Spencer syndrome. Trans Am Ophthalmol Soc 1977;75:141-63.
9. Spencer WH. Primary neoplasms of the optic nerve and its sheaths: Clinical features and current concepts of pathogenetic mechanisms. Trans Am Ophthalmol Soc 1972;70:490-528.
10. Munshi A, Jalali R. Therapy for glioma: Indian perspective. Indian J Cancer 2009;46:127-31.

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