CASE REPORT

Visual Epilepsy in Glioblastoma Multiforme

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SUMMARY

We report a 33 year old Chinese gentleman who presented with visual epilepsy and symptoms of raised intracranial pressure in which clinical examination revealed normal visual fields and acuity despite Magnetic Resonance Imaging (MRI) brain showing large contrast enhancing mass at the right occipital lobe. Craniotomy and excision of tumour was done and the histology confirmed glioblastoma multiforme (GBM). He completed radiotherapy and recovered well except developing left inferior homonymous quadrantropia post operatively which improved with time.

KEY WORDS:
Occipital glioma, Glioblastoma multiforme, Visual epilepsy

INTRODUCTION

Glioblastoma multiforme (WHO Grade IV) is the most devastating form of intrinsic brain cancer. It is invasive and associated with severe disability and high mortality. The commonest sites are in the frontal, parietal and temporal lobes. Here, we report a case of GBM in an uncommon site in the occipital lobe with visual symptoms.

CASE REPORT

A 33 year old Chinese gentleman presented with gradual onset of right sided occipital headache associated with seeing flashes of light for the past two months. The flashes were sudden, brief and episodic, with no definitive form. He had no history of epilepsy, psychiatric disorder or history of psychotropic substance abuse.

Neurological examination was unremarkable with perfect visual acuity. Visual fields, pupillary light reflexes, cranial nerve functions and fundoscopy were normal.

MRI brain showed intrinsic right occipital lobe peripheral contrast enhancing mass measuring 3.3cm x 3.4cm x 3.52cm with perilesional white matter vasogenic edema (Figure 1). Initial working diagnosis was GBM with differential diagnosis of brain abscess and metastasis in mind.

After initial treatment with steroids and analgesics, his symptoms improved. Elective right occipital craniotomy and debulking of tumour were done to confirm diagnosis and to alleviate mass effect. Intraoperatively, it was a soft to firm tumour, heterogenous in consistency with moderate vascularity. Presence of thrombosed intratumoural vessel and copious necrosis has reaffirmed our suspicion. Histopathological examination confirmed our diagnosis with the presence of both endothelial cells proliferation and necrosis.

Conscious level, speech and limbs neurology were normal post operatively. However, detailed ophthalmological examination showed a left homonymous inferior quadrantropia (Figure 2), which gradually improved with time.

As maximum cytoreduction would only contribute to 2 log reduction of tumour cells, a total of 60Gy of radiotherapy was fractionated over six weeks to complement the treatment. MRI brain three months and one year later showed minimal residual tumour in right occipital lobe.

DISCUSSION

Glioblastoma multiforme is a devastating disease, accounting to 27.7% of all brain tumours in USA. Occipital lobe is an uncommon site of GBM, occurring as low as 5%[1]. Sixty-four percent of space occupying lesions originating in the occipital lobe comprise of primary brain tumour of which 49% of them are malignant gliomas. The incidence of glioblastoma per 100,000 population rises from 0.2 in the under 14 age group to 4.5 after the age of 45. 90% of the tumours are supratentorial in location above the age of 25 .

This patient presented with classical symptoms of visual epilepsy as evidenced by seeing flashes of light. Visual hallucinations arising in cases of occipital lobe tumours are typically unformed, e.g. flashing lights, colours or zig zag lines. They may appear before a field defect is evident[1].

Interestingly enough, he did not suffer from any visual field defect as one would expect in a tumour of that size in the occipital lobe to produce congruous hemianopia. Parkinson and Craig studied a series of 50 patients with verified primary tumours without extra-occipital extension and found the following symptoms, in order of frequency: visual field defect 38%, failing visual acuity 32%, and unformed hallucinations 12%[2].

During the surgery, the abnormal tumour areas were carefully excised with the help of an ultrasonic aspirator. Most reports suggest the use of T2WI MRI abnormalities as a guide for tumour margin is better than using gadolinium enhancing margin in T1WI MRI. A careful balance is to be made between inflicting additional neurodeficit and leaving too much tumour behind. Most study indicates gross total

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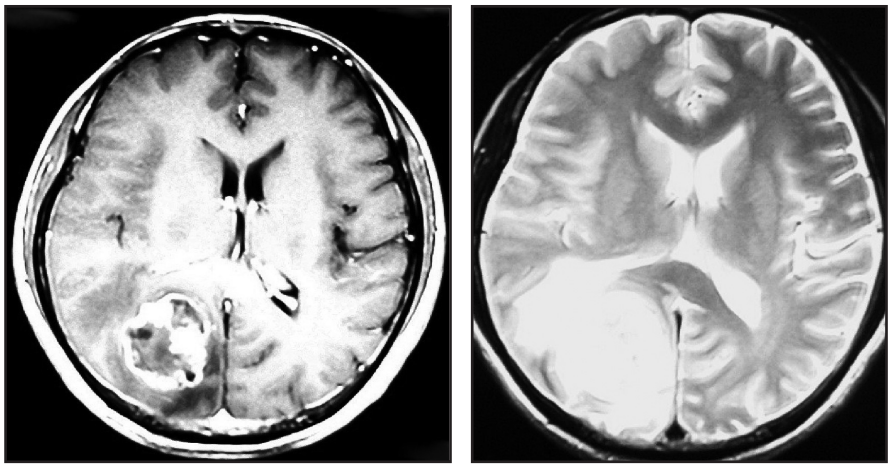

Fig. 1: T1WI showing contrast enhancing mass at right occipital lobe (left) and T2WI showing white matter perilesional edema (right)

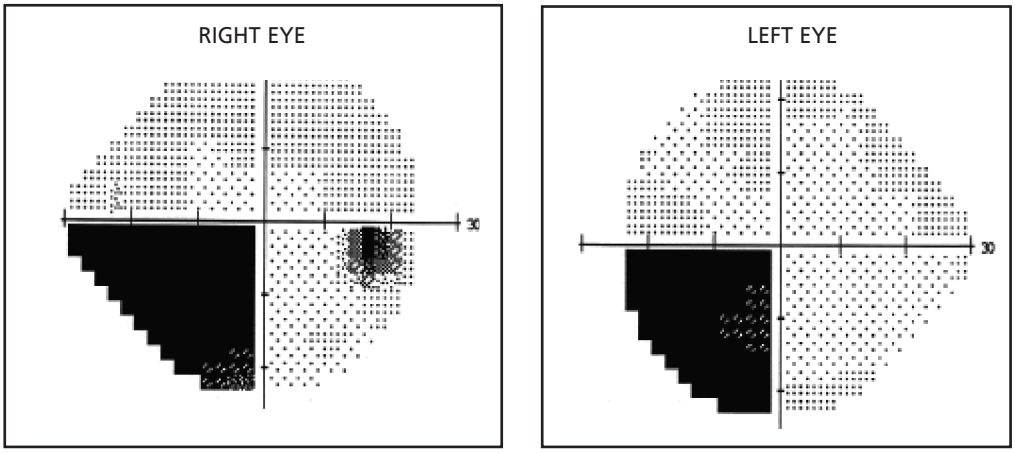

Fig. 2: Automated Humphrey Visual Field analyzer showing (L) homonymous inferior quadrantropia

debulking of tumour is associated with longer symptoms free survival.

Though we are aware of the presence of optic radiation in the vicinity of the tumour, we did not have the benefit of visual evoked potential to monitor and map out this eloquent fibre intraoperatively. As a result, he developed visual field deficit post operatively.

The principal hallmarks of GBM are endothelial hyperplasia and intercellular areas of necrosis[1], which forms the basis of diagnosis by Daumas-Duport criteria.

As demonstrated by numerous trials, radiation remains the most effective treatment for GBM. Patients whose tumours remained unchanged in size or actually became smaller on comparison of the pre- and post-irradiation scans had much better survival than those patients whose tumours grew during treatment[1].

GBM are highly vascularized and have over express angiogenic factors. New chemotherapy agents such as thalidomide and temozolomide are particularly effective in young patients with tumour that have stayed the same size or have been reduced in size during the course of radiation. Baumann F *et al* quoted the combination of thalidomide and temozolomide appears to be more effective than that of temozolomide alone in the treatment of GBM[3].

Other therapy under study includes hyperthermia as adjunctive modality to interstitial brachytherapy, immunotherapy and gene therapy. As over activation of epidermoid growth factor receptor (EGFR) signaling has been recognized as an important step in pathogenesis and progression of malignant gliomas, EGFR-targeted therapies, in future will enhance the effectiveness of existing therapies.

CONCLUSION

With aggressive surgical cytoreduction and radiotherapy with or without chemotherapy, median survival of GBM is 62 weeks compared to 17 weeks for untreated patients. Therefore, in young patients with resectable tumour, a meticulous, yet aggressive resection followed by complete radiotherapy should be advocated to achieve a good outcome.

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