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CASE REPORT

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A Case of Unexpected Longevity: Glioblastoma Multiforme with Leptomeningeal and Cerebellar Carcinomatosis

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INTRODUCTION/BACKGROUND

Glioblastoma multiforme(GBM) is the most common malignant central nervous system tumor in adults, with a mean survival of 1 year. Despite diagnostic and imaging advances, these lesions are generally poorly delineated, proving complete resection difficult and recurrence common. Utilization of radiotherapy, chemotherapy, and wide surgical resections gives only one tenth of patients an expected survival rate of greater than 2 years[1]. Leptomeningeal carcinomatosis (LC), or neoplastic meningitis, is a complication of cancer that can happen at any time during the course of any type of cancer; it is characterized by the proliferation of cancer cells in the subarachnoid space. LC presents with primary brain tumors at a much lower rate than other malignant tumors[2] (1-2% of primary brain tumors vs 5-15% in leukemias, up to 25% in lung cancer, and 23% in melanomas). Described below are the clinical characteristics and post-surgical outcome of a 60 year old male with a large right frontal lobe GBM, its infiltration of the midline structures, its radiologic presentation and resection, and its subsequent leptomeningeal and cerebellar metastases over one year later.

CASE PRESENTATION

On October 26th, 2008, patient X presented to the Emergency Department of University Medical Center Hospital in El Paso, TX with a 2 week history of right parieto-temporal headaches radiating to the occipital area and both eyes. They were described as 10/10 on a pain scale, pulsatile, dull, and they were accompanied by vomiting and somnolence. The patient was having 2-3 bouts per day, lasting 6-8hrs, without much response to Diclofenac which he had received from a physician in Juarez, Mexico. There was no photophobia, aura, vision changes, memory loss, trauma, personality changes, or nausea associated with the headaches. Prior to his admission, the patient had seen a neurologist in Mexico, and his CT showed a "frontal mass". No records were available, and these were the only results the patient remembered from his prior visits in Mexico. There was no pertinent medical or past surgical history, except for a 20 year history of cigarette use. The patient's physical exam, including neurological evaluation, was within normal limits aside from a blood pressure of 164/73. Laboratory values were significant for a mildly elevated WBC count and hematocrit. EKG showed sinus bradycardia and chest X-ray was within normal limits.

Imaging and Intervention

A CT of the head was ordered and the patient was admitted after an impressive enhancing lesion was observed. The following day an MRI of the head was ordered, showing a heterogeneous intra-axial tumor involving the right internal frontal lobe and extending to the ependyma of the adjacent anterior frontal horn. The lesion measured 51.3 x 31.3 mm (Figure 1a), with perilesional vasogenic edema and an intense heterogeneous enhancement after gadolinium administration. There was marked mass-effect to the adjacent structures with rostral displacement of the anterior cerebral arteries and a 10mm midline shift to the left with compression of the left foramen of Monro producing enlargement of the left lateral ventricle with mild transependymal edema, secondary to incipient hydrocephaly. The following day the patient underwent a right frontoparietal craniotomy and resection of the brain mass with intraoperative microscopy and neuroimaging. The pathology report confirmed a grade 4 Glioblastoma Multiforme, and a day later another MRI was performed which identified diffuse pericranial soft tissue swelling secondary to the craniotomy. Oncology followed the patient with radiation therapy and Temozolamide (Temodar©) chemotherapy. He was discharged home on 03 November, 2008 in stable condition.

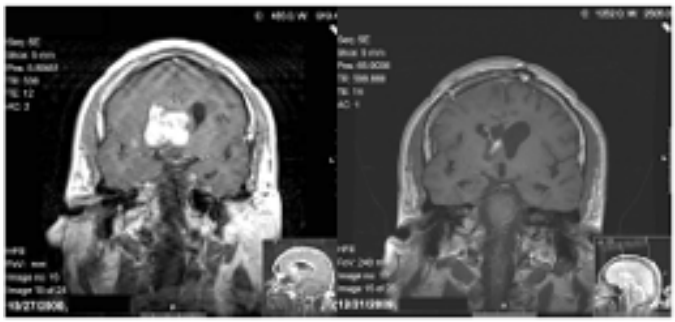

Figure 1a *Left (a): GBM at first presentation in 2008. Right (b). Coronal MRI brain with Gadolinium 14 months after primary tumor resection shows some residual uptake in the primary tumor field. Of concern was the possibility that this lesion could potentially be tumor recurrence.*

Complications

Over one year post craniotomy, the patient returned to the same emergency room with a 5 day history of bilateral lower extremity paralysis, which started as numbness on day one, and shortness of breath. The only pertinent finding on physical exam included paraspinal lumbar tenderness, 0/5 motor strength in the bilateral lower extremities, and no sensation below T10. An MRI showed enhancement of the right frontal cortex and a spinal cord lesion compressing T9-T10. The patient was taken to

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the Operating Room immediately and underwent a posterior thoracic decompression of the spine with a T9-T10 laminectomy and tumor removal. Pathologic analysis revealed a drop metastasis from the glioblastoma with spinal cord infiltration and leptomeningeal gliomatosis.

DISCUSSION

Patients with LC survive an average of 4-6 weeks without therapy due to decline in neurological function. If treated, LC patients usually do not die from LC complications, but rather from complications of the primary tumor[3]. Imaging studies are generally more useful in detecting secondary complications of LC rather than diagnosing LC. In this case, three crucial imaging modalities were implemented to rule out the recurrence of the primary tumor. Perfusion scans were performed to exclude angiogenesis in the primary tumor site post-op (Figure 2a). Since there was a lesion present on MRI on our patient's second visit (Figure 1b), MRI Spectroscopy was performed to differentiate a necrotic lesion from a proliferative lesion. Lactate is produced in significant amounts in necrotic tissue, therefore lactate spikes on cerebral spectroscopy helped identify the lesion in question as radiation-induced necrosis as opposed to tumor re-occurrence. Lastly, since proliferative tumors actively metabolize acetylcholine (ACh), ACh uptake studies were performed (Figure 2b) to ensure that the lesion in the left frontal cortex was not proliferative tissue.

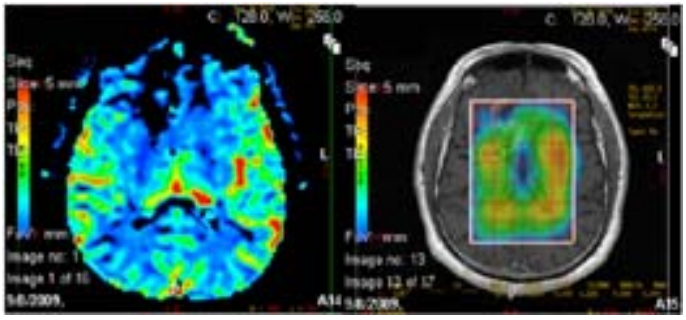

Figure 2b *Left (a): Relative cerebrovascular volume perfusion study. Post-op. Seen here in the right frontal cortex is an area of abnormal enhancement which shows decreased volume as compared to the rest of the brain, which has significantly more vessels per voxel. Right (b): Short-time spectroscopy for ACh and N-acetyl aspartate were utilized to demonstrate the enhancement in the corpus callosum but decreased to no uptake in the right frontal cortex, indicating increased metabolism where appropriate, but not in the area of the suspicious lesion. This, along with lactate uptake spectroscopy studies (not shown), were sufficient to rule out primary tumor recurrence and confirm the suspicion that the lesion was in fact radiation-induced necrosis.*

This proved to be a challenging case of a multicentric/multifocal glioblastoma multiforme with somewhat unexpected outcomes. Surviving this long after being diagnosed with such an aggressive tumor is remarkable, especially with evident metastases. The neuroimaging spectroscopy and perfusion studies perhaps saved this patient a second craniotomy. The lesion evident in his frontal cortex in the spot where his primary tumor was before it was resected was worrisome initially. With conventional MRI and CT, it is impossible to tell necrosis from neoplasia. The value of University Medical Center Neuroradiology department was evidenced by this amazing ability to tell what certain regions of the brain are metabolizing and where the blood flow is most pronounced, via MRI Spectroscopy, via MRI Spectroscopy. In summary, the team of neurosurgeons, oncologists, and radiologists at UMC were able to help this gentleman attain precious time with his family and friends for longer than anyone ever expected.

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