LONG-TERM SURVIVAL IN A PATIENT WITH BUTTERFLY GLIOBLASTOMA: A CASE REPORT

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• THE AUTHORS HAVE NO CONFLICTS TO DISCLOSE.
INTRODUCTION

Glioblastoma is an aggressive 1º CNS malignancy\(^1\).

Out of 3/100,000 Americans are diagnosed yearly.

LIFE EXPECTANCY

- 15 months: Glioblastoma\(^1\) (GBM)
- 3 months: Butterfly Variant\(^2\) (bGBM)

3% invade midline structures symmetrically\(^2\) resembling the wings of a butterfly.
CASE PRESENTATION

Low impact motor vehicle collision prompts evaluation

CT head without contrast shows hydrocephalus with remarkable dilation of the temporal horns (Figure 1A, left). Post-contrast T1-weighted MRI of the brain show an enhancing mass involving the septum pellucidum and corpus callosum with a necrotic component (Figures 1B, center; 1C, right).
Hematoxylin and eosin stain from brain biopsy performed the day after initial presentation show hypercellular glial tissue with pseudopalisading necrosis (Figure 2A, left) and microvascular proliferation (Figure 2B, center). The compilation of nuclear pleomorphism, nuclear atypia, microvascular proliferation, and necrosis (Figure 2C, right) are consistent with glioblastoma, grade IV.
TREATMENT

• Post-contrast axial T1-weighted MRI brain shows significant improvement of lesion two years after diagnosis, one year after completion of twelve rounds of temozolomide and six weeks of radiation (Figure 3A, left).

• Brain MRI the following year, three years after diagnosis, shows a bilobed focus of enhancement is within the right ventricle (Figure 3B, center). She underwent two rounds of temozolomide, a repeat biopsy (BRAF+), and two months of lomustine but developed thrombocytopenia.

• Eighteen months later, brain MRI showed progression of the lesion (Figure 3C, right). At that time the patient declined further treatment. She died six months later, more than five years from the original diagnosis.
DISCUSSION

AGE AT DIAGNOSIS
Indirectly related to:
- Capacity to withstand disease
- Likelihood of aggressive tumor

Median Age
- of all GBM diagnoses\(^3\) is 64
- of long-term GBM survivors\(^4\) is 31

FUNCTIONAL STATUS
Low Karnofsky Performance Scores:
- Represent a critically ill cohort
- Fare worse than increased age alone

PREDICTORS of survival\(^3\)

TUMOR PROFILE
- Isocitrate Dehydrogenase (IDH1/IDH2)\(^5\)
  - Depletes ROS seeking bi-products
  - Potentiates efficacy of modern chemotherapy
- O6-methylguanine-DNA methyltransferase (MGMT)\(^6\)
  - DNA repair protein – acts as dealkylating agent
  - Methylation (silencing): more sensitive to temozolomide
  - 95% of long-term survivors have MGMT methylation

TREATMENT
- Surgical resection improves survival by 3.5 months\(^7\)
- Infiltrative behavior of bGBM makes it nearly impossible to eradicate
- Preferred treatment is needle biopsy with adjuvant therapy

PSYCHOSOCIAL STRESS
- The link between depression and terminal illness is well-established\(^8\)
- Depression may affect outcomes through release of stress hormones\(^9\)
- Chronic release of stress hormones may inhibit anti-tumor responses
SUMMARY POINTS

• Butterfly glioblastoma has poor survival outcomes
• Many factors contribute to prognosis, all of which are not fully understood
• Our patient’s survival or more than five years with bGBM is unparalleled in the literature
REFERENCES