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Treatment of Recurrent Anaplastic Pleomorphic Xanthoastrocytoma with BRAF V600E mutation in a 33- Year- Old Female Previously Diagnosed with WHO Grade I Ganglioglioma

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Abstract
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Abstract

Introduction:

Anaplastic Pleomorphic Xanthoastrocytoma are rare neoplasms accounting for approximately 1% of primary brain tumors and generally present in the 2nd and 3rd decades of life in the temporal areas of the brain. Grade 3 lesions have a poor prognosis with 40-50% 5-year survival. We present the case of a woman ganglioglioma who presented with a high-grade neuroepithelial tumor thought to represent anaplastic pleomorphic xanthoastrocytoma.

Case:

The patient initially presented at 25 years of age with new onset seizures. Magnetic resonance imaging conducted at the time identified the right anterior temporal mass. She was placed on seizure medication and the mass was completely resected. Pathology revealing WHO grade 1 ganglioglioma. She continued antiseizure medication with two failed attempts to wean off medical treatment. 8 years after her initial resection, surveillance MRI showed new growth in the right temporal lobe. She underwent craniotomy and resection with pathology revealing high grade neuroepithelial tumor with BRAF V600E mutation. This was initially thought to represent WHO Grade IV gangliosarcoma but after second review with John Hopkins University, the consensus was WHO Grade III neuroepithelial tumor that is more suggestive of anaplastic pleomorphic xanthoastrocytoma. MGMT status was negative, IDH-1 and IDH-2 mutations were found to be absent, and Ki-67 was initially 32.8%. She was treated postoperatively 60 Gy in 30 fractions following EORTC guidelines for glioblastoma with adjuvant dabrafenib and trametinib for BRAF/MEK inhibition. 7 weeks after therapy initiation, she had a dose reduction due to cutaneous toxicities. 5 months after therapy initiation, she developed right-sided hearing loss attributed to CSF leak and presented to the OR for craniotomy and correction. Intraoperative tumor recurrence was noted and resected with pathology confirming high-grade neuroepithelial lesion with BRAF V600E and BCOR mutations and new mutation in NF-1 with elevated Ki-67 to 46%. She was treated with re-irradiation to the site of her recurrence with 35 Gy in 10 fractions and concomitant temozolomide 75 mg per mg/m2 in conjunction with the initiation of Nivolumab. She developed worsening radiation necrosis for 8 months after her radiation course, which warranted bevacizumab treatment and led to a perforation of a duodenal ulcer. She underwent encorafenib and binimetinib therapy shortly after, which showed response to therapy for almost 6 months after there was a new finding of leptomeningeal disease in the left frontal lobe. Unfortunately, the patient continued to have progression following treatment and treatment-related complications, passing away after one year since the re-irradiation. In total, she survived 2 years and 8 months after the diagnosis of high grade anaplastic pleomorphic xanthoastrocytoma.

Discussion:

High grade pleomorphic xanthoastrocytoma is a primary glial tumor with poor prognosis. It often manifests as transformation from low grade disease, which can bear a close histological resemblance to ganglioglioma. In this particular case, treatment was attempted with TKI to target the notable BRAF V600E mutation and chemoradiation following the regimen published by Stupp et al. with limited impact on prognosis and increased aggressive transformation of tumor. Additional research into treatment regimens needs to be conducted to more effectively treat these rare primary glial neoplasms.