HGG-45. HIGH GR ADE G LIOMA PRESENTING AS A SYNCHRONOUS CANCER IN A PATIENT WITH DIFFUSE LARGE B CELL LYMPHOMA: A UNIQUE TREATMENT DILEMMA
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BACKGROUND: Patients with synchronous tumors pose significant treatment dilemmas. We present a pediatric patient found to have concomitant Diffuse Large B-cell Lymphoma (DLBCL) and a High Grade Glioma (HGG). His unique presentation and family history uncovered a familial diagnosis of Lynch Syndrome. CASE: A 7-year-old male presented with acute appendiceal rupture. Surgical resection identified an appendiceal mass consistent with localized DLBCL. While undergoing staging evaluations, the patient complained of new onset blurry vision with normal neurologic and ophthalmologic exams. Subsequent imaging was concerning for a high-grade lesion. Biopsy confirmed HGG and he underwent gross total resection. Family history included a brother with relapsed leukemia status post bone marrow transplant and multiple first cousins with lymphoma and gastrointestinal cancers. RESULTS: While awaiting genetics and mutational analysis, we initiated treatment. The HGG was treated with focal radiation to the tumor bed starting 2 weeks after resection. Lymphoma therapy was initiated concurrently per the REBOOT regimen with the goal of decreasing doxorubicin dosing during radiation. Lumbar punctures and intrathecal chemotherapy were deferred until completion of radiation. Biopsy of residual abdominal masses following consolidation 1 showed <5% viable cells and therapy was intensified to ANHL1331, Group C1. Evaluation of both lesions identified hypermutation (mutational burden > 150mutations/Mb). Two unique germline mutations of PMS2 were also identified, consistent with Lynch syndrome. After completion of above therapy, the patient received maintenance therapy with nivolumab checkpoint inhibition. The patient developed severe neutropenia leading to cessation of nivolumab and quick progression of his HGG. CONCLUSION: Synchronous tumors pose significant treatment dilemmas given the need to treat multiple lesions while balancing side effects. While checkpoint inhibitors may provide a way to address multiple lesions, our patient had unacceptable side effects limiting the utility of this therapy.