GC-05. MANAGEMENT OF PRIMARY INTRASELLAR/CAVERNOUS SINUS PURE EMBRYONAL CARCINOMA IN THE SETTING OF DOWN SYNDROME
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BACKGROUND: Primary central nervous system (CNS) germ cell tumors (GCT) are the most common solid tumors in children with Down Syndrome (DS); such children are especially susceptible to Moyamoya disease, enhanced by irradiation-associated cerebrovascular damage. Pure embryonal carcinoma (EC) is rare among CNSGCT; however, EC is recognized as being more radio-resistant than other GCT components, and outcome for patients less favorable. PATIENT/METHODS: A 17 years-old girl with DS and severe developmental delay presented with headaches and visual changes; she was found on MRI to harbor an intrasellar/cavernous sinus tumor without radiographic evidence of neuraxis dissemination. Lumbar cerebrospinal fluid (CSF) cytology and serum/CSF tumor markers were normal. Partial tumor resection revealed tumor entirely characteristic of “pure” EC: 100% positive for CD30, pancytokeratin and Ki-67; generally OCT4 and Sall4 positive; entirely negative for beta-HCG, AFP and CD117 (cKIT). RESULTS: She received 3 cycles of standard chemotherapy (carboplatin/etoposide x 2 cycles alternating with cyclophosphamide/etoposide) with achievement of radiographic complete response. She then received three cycles of ifosfamide (300mg/M2/day x 2 days) and high-dose carboplatin (1500mg/m2/day x 2 days, adjusted by the Calvert formula) each followed by autologous hematopoietic cell rescue. Subsequent treatment with intravenously-administered brentuximab-vedotin (anti-CD30 monoclonal antibody (moab) therapy) will be presented. CONCLUSIONS: Although follow-up is short, our experience with this patient suggests that in patients with substantial components of CD30 + ve EC within a CNSGCT, especially those already pre-morbidly at additional risk from CNS irradiation, avoidance of high-dose irradiation might be achieved through early consolidation with myeloablative chemotherapy and anti-CD30 moab therapy.