respectively. Median PFS: 6.28 years (4.9–11.2); median OS: 8.8 (7.8–12.4) years. Median time to first recurrence: 8.2 (5.1-not reached); 5-year recurrence-free rate: 71% in SR and 45% in HR patients. Survival after first recurrence: 2 years (1.64–2.4). Documented recurrence and risk group were associated with survival (HR 30.4, p< 0.0001 and 2.33, p< 0.001), but metastatic status and local vs. distant recurrence were not. The effect of upfront chemotherapy on survival did not reach statistical significance in SR patients. The use of chemotherapy regimens with or without vincristine or containing cisplatin vs. carboplatin, did not alter the survival outcome. In a subgroup of 57 SR patients who were seen at initial diagnosis and for whom complete staging and treatment information were available 3 and 5-year PFS were 94% (88%, 100%) and 76% (66%, 86%); OS at 5 years, 94% (87%, 100%). CONCLUSION: No statistically significant difference was noted in outcome of patients treated without vincristine or with carboplatin instead of cisplatin, suggesting attenuated upfront regimens could be considered to reduce toxicity. Adult MB patients recur late and have a poor post-recurrence survival, therefore long-term follow-up, development of molecular risk-adjusted upfront treatment, and optimization of rescue treatments are needed.

RARE-48. GROWING KNOWLEDGE AND EMERGING QUESTIONS RELATED TO POT-1 GERMLINE MUTATIONS AND ASSOCIATED MALIGNANCIES: A CASE REPORT

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Protection of telomeres 1 (POT1) is a member of a family of genes responsible for maintenance of telomere integrity. Germline mutations of the POT1 gene have been associated with tumorigenesis of multiple tissue types, conferring an increased risk of development of malignancies such as melanoma, chronic lymphocytic leukemia, gliomas, and cardiac sarcomas. We present the case of a 28 year old male who first presented at the age of 15 with personality changes. Several years later, he was found to have a left frontal, IDH mutated, WHO grade III anaplastic astrocytoma. Following successful treatment with gross total resection, radiation therapy, and adjuvant chemotherapy, the patient represented with an MRI showing enhancement concerning for osteomyelitis and possible tumor recurrence. Pathology of the dura was consistent with myxofibrosarcoma. Staging PET scan at that time revealed multiple hypermetabolic lung lesions found to be Lanagerhans cell histiocytosis. Given the patient's complex oncological history, genetic testing was performed. He was found to carry a heterozygous mutation of the POT1 gene. This case raises the suspicion for a broader POT1-associated cancer predisposition than previously described in the literature. Moreover, this prompts further questioning of an association between POT1 mutations and IDH status in related gliomas. Patients with POT1 mutations might also be at increased risk of secondary malignancies after radiation exposure, which may require closer observation after treatment.

RARE-49. SEX-SPECIFIC SURVIVAL ANALYSIS IDENTIFIES DIFFERENTIAL CLUSTERS OF PROGNOSTIC RELEVANCE IN PATIENTS WITH PRIMARY CNS LYMPHOMA

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Sex-specific differences are increasingly recognized in various diseases but have not yet been investigated for patients with primary CNS lymphoma (PCNSL). We collected clinical information and MR-images from 130 patients with PCNSL (66 f/ 64 m). We segmented MR-images into contrast-enhanced tumor, necrosis and edema for volumetric analysis and additionally quantified anti-PCNSL immune response using anti-CD3 staining in 67 cases. Median overall survival was 10 months for females and 8 months for males (p=0.979). Sex-specific Cox-regression analyses implicated age, performance and immunodeficiency as significant prognostic markers in females, whereas only age remained as significant marker in males. We then performed cluster analyses for females (fC) and males (mC) with complete sets of clinical, imaging, and tissue phenotypes (n=55). In females, two main clusters emerged that were mainly driven by clinical performance with fC1 (n=14) featuring patients with better performance (most of whom received MTX) compared to fC2 (n=18; mOS 21 vs 3 months, p< 0.01). fC2 resolved into two subclusters with better outcome for fC2a based on larger enhancing tumor volume and high immune response (mOS 12 vs. 1 months, p< 0.01). In males, two major clusters emerged (16 vs 7 patients, mOS 5 vs 23 months, p=0.414).), which differed mainly according to treatment approach with higher prevalence of MTX-chemotherapy in mC1. Each cluster could be subdivided into 2 subclusters based on differences in clinical performance in mC1, or according to treatment strategy, i.e., combined chemo-radiotherapy vs radiotherapy-only or best supportive care (mOS 49 vs 2 months, p=0.12) in mC2. In summary, we find prognostically relevant sex-specific clusters in patients with PCNSL that implicate differential roles of tumor-related contrast enhancement and immune response in female versus treatment modality in male patients. Initial differences in cluster-defining factors need further validation in independent cohorts but might have implications for differential patient management.

RARE-50. PRIMARY CENTRAL NERVOUS SYSTEM LYMPHOMA – OUTCOMES IN THE 'HAEMATOLOGY ERA'

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BACKGROUND: Primary CNS lymphoma (PCNSL) requires a biopsy for diagnosis. Administration of corticosteroids can lead to inconclusive results and delay diagnosis. The aim of this study was to compare outcomes of patients treated under haematology compared to radiation oncologists. METHODS: Retrospective case review of patients treated under radiation oncology (2006–2010) and haematology (2011–2016). RESULTS: 121 cases were identified (median age 63 years; range 19–84). Median WHO performance status (PS) was 1. Fourteen patients (11.6%) required repeat biopsy. 10 patients were managed palliatively due to poor PS. 67 cases were managed under haematology. Median symptom duration was 28 days (range 2–540). Median time from MRI to diagnosis was 18 days (range 6–232). 66 patients received chemotherapy, 1 received radiotherapy. Median overall survival (OS) was 8 months (95%CI:0.7–15.3), 5-year OS was 22.4%. 44 cases were managed under radiation oncology. Median symptom duration was 28 days (range 2–365). Median time from MRI to diagnosis was 16 days (range 6–309). 34 patients received radiotherapy first-line, 10 received chemotherapy. Median OS was 7 months (95%CI:0–21.5), 5-year OS was 15.9%. Multivariate analysis demonstrated PS (HR 2.02 (95%CI: 1.08–3.76)) and symptom duration (HR 0.63 (95%CI: 0.41–0.96)) to be significant prognostic indicators for OS. CONCLUSION: The outcomes from the 'haematology era' are similar to those achieved by radiation oncologists. Delay in diagnosis leads to worse outcomes and highlights the ongoing need to streamline the patient pathway to improve outcomes.

RARE-51. RITUXIMAB, METHOTREXATE, BCNU, ETOPOSIDE, AND PREDNISONE (RMBVP) FOR THE TREATMENT OF RELAPSED/RECURRENT PRIMARY CENTRAL NERVOUS SYSTEM LYMPHOMA: A RETROSPECTIVE SINGLE CENTER STUDY

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BACKGROUND: Primary Central Nervous System Lymphoma (PCNSL) is an aggressive rare non-Hodgkin lymphoma. Prognosis is particularly poor for relapsed/recurrent (R/R) patients with no established treatment modality. Methotrexate/BCNU/Teniposide/Prednisone with or without rituximab has efficacy in newly diagnosed PCNSL. Here, we report efficacy and toxicity of RMBVP for the treatment of R/R PCNSL. METHODS: This retrospective study at MSKCC included PCNSLs treated with MBVP for R/R disease between 5/2009 and 5/2019. Methotrexate (3.5 g/m2; day 1 and 15), etoposide (100 mg/m2; day 2), BCNU (100 mg/m2; day 3), prednisone (60 mg/m2/day; day 1–5) and rituximab (500 mg/m2; day 0 and 14) were given in 28-day cycles. Granulocyte colony-stimulating factor support was given for 5 days in between doses of chemotherapy. RESULTS: Thirty patients received MBVP; 27 (90%) received RMBVP with a median of 2 cycles given (0.5–5). Median age was 66 years (23–81); median KPS was 70 (30–90); 14 (46.7%) were women. Median prior lines of therapy were 2 (1–5); all received prior methotrexate. Median time from last treatment was 1 month (0–88.3; 21 (70%) < 12 months). Of twenty-nine evaluable patients for response, 23 (76.7%) had a response (9 complete response (CR) (30%), 5 CR-unconfirmed (16.7%), 8 partial response (26.7%), 1 stable disease (3%)). At a median follow up of 14.2 months, median progression free survival (PFS) was 15.6 months and median overall survival was not reached. Median PFS was poorer for patients with < 12 months since last chemotherapy, 10.1 vs 60.6 months. Rates of serious (grade 3 or 4) neutropenia, anemia, thrombocytopenia and transaminitis were 20%, 20%, 16.7%, and 16.7%, respectively. One patient experienced grade 3 nephrotoxicity. No treatment related mortalities occurred. CONCLUSIONS: RMBVP is a tolerable treatment option for R/R PCNSL, particularly in those with recurrent disease.