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BACKGROUND: Primary central nervous system(CNS) choriocarcinoma(CC) is very rare and has the poorest prognosis among germ cell tumor (GCT). CC usually has extremely high level (HL) of serum beta-human chorionic gonadotropin (bhCG) over than 1,000 mIU/ml. Some studies assign HL bhCG cases to poor prognosis group even without biopsy. The purpose of this study was to find out if there was a good prognosis subset in the HL bh group. MATERIALS AND METHODS: We analyzed 103 cases diagnosed with GCT from 1998 to 2019 in Hokkaido University Hospital and reviewed the literature of CNS CC and bhCG. Survival was assessed using Kaplan-Meier method and log-rank statistics between the group with CC component and that with no CC component but HL bhCG. RESULTS: One out of 103 our cases was diagnosed as a mixed GCT with CC component and did not respond to treatment and died 9 months later. Two cases were treated as CC because of HL bhCG (1,226 and 2,739 mIU/ml) despite that the biopsy showed only germinomas and survived(105 and 37 months), that is, no CC component. Combining our cases with 69 cases in the literature, all 7 cases with no CC component but HL bhCG survived but the median survival of the other 65 cases with CC component was 38.2 months (P=0.02). CONCLUSION: This study has a limitation of selection bias, however, it suggests that patients with no CC component but HL bhCG may have a better prognosis.

GCT-12. INTRACRANIAL GROWING TERATOMA SYNDROME: CLINICAL IMPLICATION FROM SINGLE UNIVERSITY EXPERIENCES

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In general, intracranial germ cell tumors (GCT) are sensitive to chemotherapy, radiation therapy, and have favorable outcomes. However, a rare chemotherapeutic retro conversion phenomenon, known as intracranial growing teratoma syndrome (iGTS), shown a poorer prognosis. We analyze the diagnostic characteristics and the result of treatment response for the patients with iGTS treated in our institutes (SNUH and SNUBH, from 1997 to 2019). The electronic medical records and PACS were used for reviewing the clinical information, follow-up MRI images, tumor markers (alpha-fetoprotein, human chorionic gonadotropin, in serum or cerebrospinal fluids), and pathological findings. Out of 328 intracranial GCT patients, seventeen were finally identified as iGTS. Sixteen out of 17 patients were non-germinomatous GCTs, and 1 were germinomas. Initial pathology was common in order of immature teratoma (26.7%), other than immature teratoma (11.5%), and germinoma (0.5%). All of the tumors showed typical 'honeycomb appearance' in their follow-up MRI images. Sixteen out of 17 tumors were surgically resected as 2nd look surgery. Among them, 13 tumors were gross totally resected. Twelve were alive without evidence of recurrences during follow-up periods, and the other was dead from the progression of the disease. Among the other than the gross total resection group (n=4), two patients were dead, one recurred the tumor, and the other is following up with stable disease after adjuvant radiation therapy. Early detection and total resection of the tumor as possible might be meaningful for favor prognosis, especially in non-germinomatous GCTs patients.

GCT-13. THE TREATMENT OUTCOMES OF INTRACRANIAL GERM CELL TUMORS WITH KSPNO PROTOCOL: SINGLE CENTER RETROSPECTIVE ANALYSIS

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Dho et al. (BTRT, 2017) reported that 1.1% (127/11,827) of primary brain tumors are intracranial germ cell tumors (iGCT) in Korea. We analyzed the epidemiology and treatment results of germ cell tumors in our institution. From 2004 to 2019, among 6494 patients with intracranial neoplasms the 61 (0.9%) patients with iGCTs were enrolled: histologically diagnosed in 50 patients and clinically in 11 respectively. Pediatric patients underwent treatment according to the KSPNO protocol, and adult patients were treated with bleomycin, etoposide, and cisplatin regimens. The median age was 20 years (range: 1–42) and the follow-up period was 7.7 months (range: 10.0–203.4 months), respectively. The tumors arise most frequently in the pineal area (n=30, 49.2%). There were no significant differences in outcomes between protocols, but in KSPNO protocol group showed lower tumor recurrence rate (11.5% vs. 20%, p=0.494) and mortality (0% vs. 5.2%, p=0.503). According to the pathological subtype, the outcomes showed statistically significant differences between germinoma and non-germinomatous germ cell tumor (NGGCT) groups. The 10-year progression-free survival was 93.2% and 67.1% in the germinoma and the NGGCT group, respectively (p=0.009). The NGGCT pathological type (p=0.021) was a significant recurrence associated factor in multivariate analysis. Significant adverse events (CTCAE version 5.0 grade≥3) were showed in 14 patients (7 patients in both KSPNO and other treatment protocol groups). Pure germinoma has a higher survival rate and a lower recurrence rate than NGGCT. And KSPNO protocol might be safe and effective. For appropriate treatment for iGTCs, a multidisciplinary approach might be needed.

GCT-14. SECOND-LOOK SURGERY FOR INTRACRANIAL GERM CELL TUMORS

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OBJECTIVE: The authors present their experiences of second-look surgery in patients with intracranial GCTs who showed less than complete response despite normalizing or decreasing tumor markers after chemotherapy. METHODS: Retrospective review of 14 patients who underwent second-look surgery for an intracranial GCT was performed. RESULTS: Of 40 consecutive patients with newly diagnosed intracranial GCTs treated between August 2003 and 2019, 14 patients (35%) underwent second-look surgery. The mean age was 9.2 years. The initial diagnoses were mixed germ cell tumor in 6, immature teratoma in 4, yolk sac tumor in 2, and germinoma 2. Second-look surgery was performed after 1–3 courses of chemotherapy. Magnetic resonance imaging (MRI) at the surgery demonstrated increasing residual tumor in 8 and stable residual tumor in 6. Tumor markers were normalized in 10 and nearly-normalized in 4. Gross total resection was achieved in 12 patients and near-total resection in 2. Histopatholgy at second-look surgery revealed mature teratoma in 6, immature teratoma in 3, fibrosis with atypical cells in 2, and fibrosis in 3. Eleven patients subsequently underwent additional chemo-radiation therapy according to the initial diagnosis. All patients are alive with no evidence of recurrence with a mean follow-up of 69 months. CONCLUSIONS: Second-look surgery plays an important role in the treatment of intracranial GCTs. Surgery may be encouraged at a relatively early phase after chemotherapy when the residual tumor increases or does not change the size despite normalized or nearly-normalized tumor markers in order to achieve complete resection and improve the outcome.

GCT-15. INTEGRATED CLINICAL, HISTOPATHOLOGICAL, AND MOLECULAR DATA ANALYSIS OF 190 CENTRAL NERVOUS SYSTEM GERM CELL TUMORS FROM THE IGCT CONSORTIUM

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BACKGROUND: We integrated clinical, histopathological, and molecular data of central nervous system germ cell tumors to provide insights into their management. METHODS: Data from the Intracranial Germ Cell Tumor Genome Analysis Consortium were reviewed. A total of 190 cases were classified as primary GCTs based on central pathological reviews. RESULTS: All but one of the cases that were bifocal (neurohypophysis and pineal glands) and cases with multiple lesions including neurohypophysis or pineal gland were germinomas (34 of 35). Age was significantly higher in patients with germinoma than other histologies. Comparison between tumor marker and histopathological diagnoses showed that 18.2% of histopathologically diagnosed germinomas were marker-positive and 6.1% of non-germinomatous GCTs were marker-negative, suggesting a limitation in the utility of markers or histopathology alone using small specimens for diagnosis. Comparison between local and central histopathological diagnoses revealed a discordance of 12.7%. Discordance was significantly less frequent in biopsy cases, implying difficulty in detecting all histopathological components of heterogeneous GCTs. Germinomas at the typical sites (neurohypophysis or pineal gland) showed a better PFS than those at atyp-