males, ranging in age from 0 to 17 years, median 9.0 years. Three pilocytic astrocytomas (PA) and a polymorphous low-grade neuroepithelial tumor of the young (PLNTY) had BRAF fusion. Four cases were classified as Diffuse midline glioma, H3K27M-altered. Two cases were classified as Diffuse pediatric-type high-grade glioma, H3-wildtype and IDH-wildtype. No genetic alteration was observed in two diffuse astrocytoma cases. An anaplastic oligodendroglioma case with PDGFRA amplification could not be classified as any new entity. All three PA cases with BRAF fusion occurred in cerebellum and all four H3K27M altered cases occurred in midline location such as thalamus, brainstem and cervical spinal cord. Six cases which were classified as pediatric-type diffuse high grade gliomas had poor prognosis. Conclusion:Genetic status is associated to tumor location and patients prognosis. Integrated diagnoses are important in pediatric glioma patients.

Key words: pediatric gliomas | cIMPACT-NOW | integrated diagnoses

PEDT-3

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Background: Germ cell tumors (GCTs) containing a teratoma component is a group of diseases consisting of various pathological conditions such as mature teratoma, immature teratoma, teratoma with malignant transformation, and mixed tumor with other GCTs. There is controversy about the efficacy and safety of radiation and chemotherapy for GCTs with teratoma component other than mature teratomas. Methods: Of 212 cases of GCTs treated at Tohoku University Hospital Neurosurgery from January 1990 to March 2021. In this study, 23 histologically verified GCTs containing teratoma components were included. Pathological findings, recurrence, survival, and late complications were examined. Results: The age of onset was 2 months-21 years (median 10.5 years). Histological diagnosis was mature teratoma alone in 5 cases, mixed GCTs with mature teratoma in 11 cases, immature teratoma in 5 cases, and mixed tumor with mature teratoma and germinoma in 2 cases. Patients except mature teratoma were treated by chemotherapy alone or radiochemotherapy. During follow-up for 7–362 months (median 135 months), 3 patients relapsed. One of these patients was diagnosed with mature teratoma at the time of treatment and did not receive post-treatment, but relapsed as germinoma 21 years later. A review of pathological specimens at the time of initial onset revealed immature teratomas in addition to mature teratomas. Recurrent lesions in 3 cases were controlled by additional treatment, and no deaths due to tumor progression were observed. On the other hand, of the 18 patients who underwent radiochemotherapy, 1 developed primary hypothyroidism and 2 developed thyroid cancer and leukemia. Conclusion: GCTs with teratoma component often contain malignant histological types and require caution when making a pathological diagnosis. In these cases, tumor control can be expected by radiation or chemotherapy, but there is a risk of developing endocrine disorders and secondary tumors, and further studies are needed to optimize treatment.

Key words: radiation | chemotherapy | teratoma

PEDT-5

PROBLEM FOR THE GUIDELINE OF CNS GERM CELL TUMORS

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Primary CNS germ cell tumors (GCTs) are rare neoplasms, therefore, a clinical guideline has not been established so far. While better management has been achieved over recent decades by modifying radiation coverage and selecting appropriate chemotherapy, standardization of treatment remains challenging, partly due to the low volume of cases encountered in each institution. As the incidence is higher in East Asia, including Japan, the Japan Society for Neuro-Oncology established a multidisciplinary task force to create an evidence-based guideline for CNS GCTs. The Medical Information Network Distribution Service (Minds) guideline was referred to and utilized in the course of creating this guideline. We chose 6 topics and 10 clinical questions. This guideline provides recommendations for multiple dimensions of clinical management for CNS GCTs, with particular focus on diagnostic measures including serum markers, treatment algorithms including surgery, radiotherapy and chemotherapy, and under-investigated but important areas such as treatment for recurrent cases, long-term follow-up protocols and long-term sequelae. International collaborations to set standards of clinical management for this rare tumor have proven fruitful, concurrently, many fields continue to show variance in clinical practice, partly due to the rarity of clinical encounters and the absence of documented standards. There still seem to be differences in the treatment concept between Japan and North America or Europe countries. This guideline serves the purpose of helping healthcare professionals keep up to date with current knowledge and standards of management for patients with this rare disease in daily clinical practice, as well as driving future translational and clinical research by recognizing unmet needs concerning this tumor. We discuss about the issues both already clarified and should be cleared in the future.

Key words: CNS germ cell tumor | Clinical Guideline | Tumor Marker

PEDT-9

A STUDY OF NOVOTTF-100A TO EXPAND THE REGULATORY INDICATION FOR CHILDHOOD GLIOBLASTOMA THROUGH A PEDIATRIC CLINICAL TRIAL BASED ON THE ADVANCED MEDICAL CARE SYSTEM

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Background: Tumor-treating fields (TTF) are alternating electric fields applied continuously to the brain by attaching 2-pair arrays on the scalp. Although TTF therapy has demonstrated efficacy against supratentorial glioblastoma (GBM) in adults, its safety and efficacy in children have not been confirmed. In Japan, off-label use of medical devices is almost impossible because the national health insurance system does not cover the cost of off-label use of drugs and medical devices. Therefore, TTF therapy cannot be applied to the treatment of pediatric GBM.

[Objectives] The investigator-initiated clinical trial aims to expand regulatory approval of TTF therapy for pediatric GBM treatment based on safety and exploratory efficacy data. Methods: Patients aging between 5 and 17 years with histopathological diagnosis of GBM (either newly diagnosed or first-recurrence), which located in the supratentorial region would be included. All the patients will receive TTF therapy for 28 days per course for up to 26 courses until the end-of-therapy criteria are met. The primary endpoint is the adverse event rate with causality. The secondary endpoints include various time-to-event measures and QoL. In total ten patients will be enrolled. Current Status: Discussions with the Pharmaceuticals and Medical Devices Agency (PMDA) led to a tentative consensus that the accumulated data on the efficacy of NovoTTF-100A for adult GBM may be extrapolatable to pediatric GBM if the trial is able to demonstrate efficacy equivalent to that found in previous, adult studies. On the other hand, the combination of the pediatric safety data gathered in this trial and the findings of international studies, including clinical trials and post-marketing surveillance studies, may expedite approval of the device for pediatric GBM treatment. The trial started patient enrollment in April, 2021 with the supervision of the Advanced Medical Care administration system and is currently awaiting the first eligible patient.

Key words: tumor treating fields therapy | glioblastoma | children