OTHER BRAIN TUMORS (BT)

BT-1

AN AUTOPSY CASE OF MULTICENTRIC MALIGNANT GLIOMA, H3K27M MUTANT

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Introduction: In 2016, the World Health Organization (WHO) defined diffuse midline glioma, H3K27M mutant (WHO Grade 4) as a tumor with K27M mutation in histone H3.3 (H3F3A) or H3.1 (HIST1H3B/C) that develops mainly in the midline regions of the central nervous system. Here, we report a rare case of the abovementioned disease with remote multiple lesions in addition to the midline regions that was diagnosed on the initial visit. Case: The 52-year-old man, suffered from dysarthria, dysphagia, gait disturbance, and headache that gradually worsened over several months. Non-contrast-enhanced lesions were noted in the pons(swelling and involvement of the basilar artery trunk), cerebellum, thalamus, fornix, periventricular area, hippocampus, medial aspect of bilateral frontal lobes, and distally in the right frontal cortex and apical region of the left temporal lobe. The open biopsy was performed for left cerebellar surface lesion, and the pathological and genetic diagnosis was diffuse midline glioma, H3K27M mutant. Extended focal radiation at 50Gy/25fr and corpus callosal/cerebellar boost at 10Gy/5fr were performed. The lesions were markedly reduced, and neurological symptoms were also alleviated. However, 20 months after the initial visit, neurological symptoms had worsened and cerebrospinal fluid dissemination occurred, after that died at 29 months. An autopsy revealed tumor invasion mainly in the midline regions of the cerebrum and in the cerebellum, brain stem, pituitary gland, entire spinal cord, and cauda equina. Immunostaining of the distally cerebral cortex lesions showed that with a negative result for H3G34V. Discussion/Conclusion: It was suggested that caution is required for primary differential diagnosis may be presented at multiple lesions such as remote cerebral cortex of diffuse midline glioma, H3K27M-mutant.

Key words: Diffuse midline glioma | H3K27M-mutant | Remote infiltration

BT-4

THE TREATMENT HISTORY OF LONG-TERM SURVIVORS OF OPTIC PATHWAY GLIOMA IN KOBE CHILDREN'S HOSPITAL

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Optic pathway glioma(OPG) is almost recognized in childhood and about 0.01–0.02% of whole brain tumor in Japan. Because of the rare tumor, there are few reports about results of its treatment. In 2021, Guideline of Optic pathway/hypothalamic glioma is indicated from The japan Society for Neuro-Oncology. We retrospectively study 9 cases history of OPG who have treated for more than 5 years from January 2005 to March 2021 in Kobe Children's Hospital. Cases are 4 boys and 5 girls. Average age at diagnosis is 3.8 years old and average follow up term is 11 years 10 months. They are 4 Pilocytic astrocytoma, 3 Pilomyxoid astrocytoma, and 1 Fibrillary astrocytoma. Al l cases have survived. There are only 2 cases who could be controlled with single series of surgical treatment and chemotherapy. Most cases need several times of resection and chemotherapy and uncontrollable 5 cases required radiological treatment. 2 cases are still under treatment for over 10 years. For OPG, partial resection to control hydrocephalus is recommended and several trial of chemotherapy must be carried out. There exist a few cases who need continuous treatment for long term. The other side, a few cases are uncontrollable those need radiotherapy to manage tumor volume. Because the history of OPG would be long term, we should adjust the treatment plan to environment of patients.

Key words: Optic Pathway Glioma | Long-term Prognosis | Multimodal Treatment

BT-7

LONG-TERM OUTCOME AND LATE ADVERSE EFFECTS OF INTRACRANIAL PRIMARY GERM CELL TUMOR

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Background: Current standard of care for intracranial germ cell tumor (IGCT) have favorable cure rates. However, long-term treatment-related adverse event data are limited. The present study examined the long-term outcome of IGCT.Methods: The data from 27 patients with IGCT treated at our institutes from 1993 to 2020 were retrospectively analyzed. The patients were divided into two groups: group A; who received whole-ventricle RT (30 Gy) or focal RT (40-50Gy) from 1993 to 2012, group B; who received whole-ventricle RT (23.4 Gy) from 2013 to 2020. Complications and physical-activity level after treatment were retrospectively analyzed.Results: Pathological diagnosis was germinoma in all cases, and chemotherapy was CARE in 19 cases and ICE in 2 cases. Radiation therapy was performed in 15 cases in group A and 6 cases in group B. The follow-up period was 8–19 years (mean 11.3 years) in group A and 0.4–7 years (mean 3.6 years) in group B. Radiological cure was obtained in all cases, there was no recurrence. Hypopituitarism requiring hormone replacement therapy was observed in 53% of patients in group A and 50% of patients in group B. Late complications were cerebral hemorrhage from venous malformation (4 years after treatment), symptomatic cerebral atrophy in 2 cases (3 years / 6 years after treatment), radiation induced malignant glioma (19 years after treatment) in group A. The rate of good physical-activity was 71% of group A and 100% of group B.Discussion/Conclusions: CARE + whole-ventricle radiation therapy is appropriate as a standard treatment for ICGT. Late complications are directly linked to poor quality of life and may be radiation dose dependent. Optimize radiation therapy to further improve outcomes is required.

Key words: germinoma | outcome | radiotherapy

RARE CASE SERIES (CS)

CS-1

CEREBELLAR LIPONEUROCYTOMA; REPORT OF TWO CASES WITH DETAILED METABOLIC, IMMUNOHISTOCHEMICAL, AND GENETIC EVALUATIONS

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Cerebellar liponeurocytoma (cLNC), World Health Organization grade II neoplasm, is a rare brain tumor characterized by advanced neuronal/neurocytic differentiation and focal lipid accumulation in neuroepithelial tumor cells. However, the expression and genetic profiling of cLNC, as well as metabolic imaging characteristics, have been poorly studied. Two patients with lower vermian tumors were operated on with telovelar approach. Moderate methionine uptake in positron emission tomography was observed in both cases. Histologically, the tumor was composed of small, uniform cells with round nuclei in a sheet-like fashion. Vacuolate cells with displacement of nuclei suggested the lipid accumulation, which was further supported by immunohistochemical staining of S-100. Although the extent of lipidization was relatively low compared with the reported cLNC cases, the immunohistochemical findings confirmed the diagnosis of cLNC. Next-generation sequencing of tumoral DNA in one case detected a splice site mutation of the ATRX gene, which is the first observation in the literature. Neither chemotherapy nor radiotherapy were administered postoperatively in both cases. In one case with spinal dissemination, residual tumor demonstrated progression 7 months after the resection. Long term follow-up data of cLNC cases with detailed expression and genetic profiles are essential for precise diagnosis and better understanding of the oncogenic pathway as well as the natural history of cLNC.

Key words: cerebellar liponeurocytoma | S-100 | ATRX

CS-5

A CASE OF ANAPLASTIC PLEOMORPHIC XANTHOASTROCYTOMA IN THE PINEAL REGION

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Tumors in the pineal gland are rare tumors that account for about 0.3% of all brain tumors and have various histological types of tumors develop with germinoma, pineocytoma, and pineoblastoma in that order. On the other hand, pleomorphic xanthoastrocytoma (PXA) is a rare tumor of less than 0.2% and frequently occurs in supratentorial cerebral surface of children and young adults.A case was a 61-year-old man whose pineal tumor was found due to visual disturbance. MRI showed a 23 mm-sized lesion with cysts and inhomogeneous enhancement in the pineal gland. Partial calcification was observed, but there was no non-communicating hydrocephalus, and no increase in HCG-β and AFP with blood sampling. A midline