CS-09

PROPOSAL OF A NEW CLASSIFICATION FOR DETERMINING THERAPEUTIC STRATEGY ACCORDING TO THE PROGRESSION OF OLFACTORY NEUROBLASTOMA

Hiroaki Motegi[1], Shigeru Yamaguchi[1], Yukitomo Ishi[1], Shogo Endo[1], Hiroyuki Kobayashi[1], Shunsuke Terasaka[1], Masanobu Suzuki, Yuji Nakamaru, Akihiro Homma, Kiyohiro Houkin[1]; [1]Department of Neurosurgery, Graduate School of Medicine, Hokkaido University, Sapporo, Japan

INTRODUCTION: Olfactory neuroblastoma (ONB) is a rare type of malignancy that infiltrates and propagates from the nasal cavity to the anterior skull base and into the cranium. Various treatment strategies have been used at different institutions and time of treatment. Although the staging system proposed by Kadish is commonly adopted has not proven useful for predicting prognosis or choosing among treatment strategies. Factors to be considered have increased accordingly, for example, whether to perform ESS alone or in combination with craniotomy, whether to try preserving the olfactory sense and whether to use neoadjuvant chemotherapy. In this study, we reviewed ONB cases treated at our institution to propose a new classification system to help determine treatment strategies. METHODS: Thirty-four patients treated at Hokkaido University were included. Stages of craniocaudal progression were defined as Nasally/Paranasally localized (NP), Frontal Base progression (FB), and Brain invasion (BI). Stages of lateral progression were defined as Midline (M) or Lateral extension (L), and Unilateral (U) or Bilateral (B). RESULTS: Between 2008 and 2016, patients at the BI stage were proactively treated with neoadjuvant chemotherapy and achieved long-term survival (mean overall survival, 64.2 months). However, no standard way of choosing among treatment options was established. M-stage patients underwent concurrent craniotomy. From 2017 onwards, 5 patients were treated according to the new classification system. All were FB-M cases, including 4 cases of B disease, in which ESS alone followed by radiotherapy was used. One patient in the FB-M-U category underwent unilateral resection and the olfactory sense was preserved. In general, the treatment with ESS alone appeared to be preferred for M disease, and surgery after neoadjuvant chemotherapy was advisable for BI cases. CONCLUSION: The result suggests that the new classification system is helpful to decide the treatment strategy according to the progression of ONB.

CS-11

PITUITARY EPENDYMOMA: A CASE REPORT

Tadateru Fukami[1], Yayoi Yoshimura[1], Ryoko Fujikawa, Kazuhiko Nozaki[1]; [1]Department of Neurosurgery, Shiga University of Medical Science

INTRODUCTION: Neoplasms of the sellar region generally includes pituitary adenoma, craniopharyngioma, meningioma. We report a case of pituitary ependymoma. CASE: A 39 years-old man. He experienced the sense of discomfort of the inside upper part field of vision of the left eye for a few months since May, 201X. Ophthalmological examination showed right homonymous hemianopia of right upper 1/4. He was introduced to the department of neurosurgery of nearby hospital. MRI showed intrasellar tumor and the lesion was partially removed because of solidness by endoscopic transsphenoidal surgery on July, 201X. Postoperative pathological diagnosis was pituitary adenoma. The residual tumor was followed up, but the compression to the left optic nerve was not resolved. So he was introduced to our hospital in January, 201X+1 and endoscopic transsphenoidal surgery was performed on May, 201X+1. OPERATION: Supposing the change to extended transsphenoidal surgery, we prepared rescue flap. Enlarging the window of sellar floor and removing the tuberculum sellae, the tumor was totally removed. The boundary between the tumor and the normal pituitary gland was obscure. We inserted fat piece to the intrasellar space, and reconstructed the sellar floor with the absorbable plate following fixation with a polyglycolic acid sheet, fibrin glue, and sinus balloon. PATHOLOGY: Fusiform cells having an oval or a short spindle shape nucleus multiplied in strand and palisading pattern through capillary vessels were the main findings, and ependymal rosettes were confirmed. Immunohistchemical study showed chromograninA(focally+), synaptophysin (-), EMA (+, dot and ring pattern), CAM5.2(+), bcl-2(+), TTF-1(-), S100(focally+), GFAP(-). Final diagnosis was pituitary ependymoma. Mild diabetes insipidus was occurred postoperatively but it was controlled medically. Now he is followed up in outpatient department. DISCUSSION: Pituitary ependymoma was reported only eight cases in the past literatures. Though it is extremely rare, pituitary ependymoma should be included as a differential diagnosis of the sellar tumors.

CS-12

IDH-1 MUTANT GLIOMA IN BROTHER AND SISTER, ONSET AT AGE OF 30s.

Ryuhei Kitai[1], Takahiro Yamauchi[1], Hiroyuki Neishi[1], Makoto Isozaki[1], Kenzo Tsunetoshi [1], Ken Matsuda [1], Hidetaka Arishima[1], Toshiaki Kodera[1], Ken-ichiro Kikuta[1]; [1]Department of Neurosurgery, University of Fukui, Japan

A 38-year-old man consulted with our neurosurgery group at University of Fukui hospital, due to tonic-clonic seizures. An MRI revealed a non-enhanced intra-axial tumor at the left frontal lobe. CT showed no calcification in the tumor. The tumor was removed by awake brain surgery. The pathological specimen was diagnosed as a diffuse astrocytoma with IDH-mutant. Immunohistochemical staining and DNA sequencing confirmed a R132H mutation at IDH-1. Telomerase Reverse Transcriptase (TERT) promoter mutation and 1p 19q codeletion was not evident. Four years later, his sister, a 40-year-old woman, had an MRI as a routine medical check that found a right frontal tumor at the mirror site of her brother's tumor, and with identical radiological findings. The tumor was completely removed. The specimen revealed oligodendrocytoma, with mutant IDH and 1p/19q co-deleted. DNA sequencing showed also R132H at IDH-1. TERT promoter mutation was evident at C228T, which is a surrogate marker for oligodendroglioma. IDH-mutant astrocytoma and oligodendroglioma in siblings; and germline mutation of IDH have not been reported. However, the respective incidences of astrocytoma and oligodendroglioma are 0.55/100,000/year and 0.26/100,000/year according to United State statistics, which indicates that merely coincidental occurrence of these tumors is extremely unlikely. A trigger for IDH mutation that runs in rare families could warrant whole-genome sequencing.

CS-14

A CASE OF CIC-REARRANGED INTRACRANIAL SARCOMA

Shota Tanaka[1], Daisuke Sato[1], Masako Ikemura, Shunsaku Takayanagi[1], Kazuo Tsutsumi, Nobuhito Saito[1]; [1]Department of Neurosurgery, The University of Tokyo

INTRODUCTION: Intracranial sarcoma is extremely rare among primary brain tumors and often misdiagnosed. Its standard treatment is yet to be established, and treatment options are discussed on a case-by-case basis. Here we report our recent case of intracranial sarcoma review the relevant literature. CASE ILLUSTRATION: A 57-year-old right-handed man presented with headache and was found to have a 5cm mass in the right frontal lobe. Gross total resection was achieved without complications. Given the local pathological diagnosis being glioblastoma, adjuvant radiotherapy with concurrent temozolomide was administered. Further pathological examination revealed Capicua (CIC) rearrangement on FISH, which lead to the diagnosis of sarcoma. No further treatment was pursued at that time. However, he noticed rapid decline in the right visual acuity 7 months from the initial diagnosis. MRI demonstrated a rapidly-growing mass in the right optic nerve sized 1.5cm, which was depicted as a high uptake area on FDG-PET, suggestive of recurrence. Two cycles of chemotherapy with vincristine, ifosfamide, doxorubicin, and etoposide as well as GammaKnife stereotactic radiosurgery were performed with partial response. Sustained myelosuppression and debilitating constitutional symptoms precluded additional chemotherapy. No further recurrence was noted 1 year after diagnosis. CONCLUSION: We have recently experienced a case of CIC-rearranged intracranial sarcoma. FISH was useful in detecting CIC rearrangement and reaching the correct pathological diagnosis. Rapid recurrence of the tumor was noted, but well controlled with radiochemotherapy.

CLINICAL OTHERS (COT)

COT-01

SYMPTOMATIC EPILEPSY INDUCED BY MALIGNANT BRAIN TUMOR

Hirofumi Oyama[1]; [1]Department of Neurosurgery, Toyohashi Municipal Hospital, Toyohashi, Japan

CASES: We report about 41 cases of malignant brain tumor which were treated with operation, irradiation, chemotherapy during past 62 months. 34 cases of glioblastoma, 2 cases of malignant glioma(WHO grade III), 2 cases of medulloblastoma, 1 case of germ cell tumor, 1 case of malignant neurocytoma, and 1 case of malignant ependymoma were included in 41 cases. Two cases of glioblastoma survived over than 25 years, but the median survival of dead 11 glioblastoma cases was 22 months. 4 cases were expired among another 7 cases. RESULTS: 6 cases showed mutation in ATRX and one case had mutation of IDH-1 among 34 cases of glioblastoma. One case showed mutation in ATRX and IDH-1 among 2 cases of malignant glioma. But the mutation in ATRX and IDH-1 had no correlation with convulsion. The initial symptom was epilepsy in 3 cases of glioblastoma and another 3 cases of glioblastoma showed convulsive seizure thereafter. One case of glioblastoma showed rapid aggravation of symptom after convulsion. The initial symptom was epilepsy in 1 case of