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 cranio-caudal progression were defined as Nasally/Paranasally localized (NP), Frontal Base progression (FB), and Brain invasion (BI). Stages of lateral progression were defined as Null (M) or Lateral extension (L), and Midline extension (1). 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 radiation therapy from 2008 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-MU 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.

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 initial symptom was epilepsy in 1 case of glioblastoma and another 3 cases of glioblastoma showed convulsive symptom after convulsion. The initial symptom was epilepsy in 3 cases of glioblastoma. One case showed mutation in ATRX and IDH-1 among 2 cases. 5 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. The median survival of dead 11 glioblastoma cases was 22 months. 4 cases treated with operation, irradiation, chemotherapy during past 62 months. 7 months from the initial diagnosis. MRI demonstrated a rapidly-growing enhancing 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 1p19q codeletion was not evident. Four years later, his 40-year-old woman, had MRI as a routine medical check that found a right frontal tumor at the mirror site of her brother’s tumors, and with identical radiological findings. The tumor was completely removed. The specimen revealed oligodendroglioma, with mutant IDH and 1p19q co-deleted. DNA sequencing showed 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.

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 paratemporal 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 Capricia (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.

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