INTRODUCTION: 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. From 2013 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 neo-adjuvant chemotherapy was advisable for B cases.

CONCLUSION: The result suggests that the new classification system is helpful to decide the treatment strategy according to the progress of ONB.

CS-11
PITUITARY EPENDYOMA: A CASE REPORT
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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, 2018. In July, 2019, he was admitted to our hospital. MRI demonstrated intrasellar tumor and the lesion was partially removed because of solidness by endoscopic transnasal surgery. The specimen revealed ependymoma, with mutant IDH and 1p 19q co-deletion. No further recurrence was noted as GammaKnife stereotactic radiosurgery were performed with partial resection.

CS-14
A CASE OF CIC-REARRANGED INTRACRANIAL SARCOMA
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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. The lesion was a CIC-rearranged intracranial sarcoma. FISH was useful in detecting CIC-rearrangement.

CS-12
IDH-1 MUTANT GLIOMA IN BROTHER AND SISTER, ONSET AT AGE OF 30s.
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A 38-year-old man consulted with our neurosurgery group at University of Fukuoka 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-1 mutant. Immunohistochemical staining and DNA sequencing confirmed a R132H mutation at IDH1-1. Telomerase Reverse Transcriptase (TERT) promoter mutation and 1p 19q co-deletion were noted. Four years later, his 40-year-old younger brother, who 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 1p 19q co-deleted. DNA sequencing showed also R132H at IDH1-1. TERT promoter mutation was evident at C228T, which is a surrogate marker for oligodendroglioma. IDH1-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.