A 31-year-old male with a seizure

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A 31-year-old male patient without specific medical history was admitted with a seizure. Brain computed tomography revealed 7cm-sized contrast enhancing cystic mass with internal necrosis and calcification, in the right frontal lobe. With suspicion of Glial tumor, gross total removal of the tumor was performed. Patient is alive well after subsequent radiotherapy.



Material Submitted

- 1. Preoperative MRI Image
- 2. Representative H-E slides (two slides No. 1 and No. 2)

- 1. Histological characteristics
- 2. Correlation of molecular findings
- 3. Differential Diagnosis

9 year-old boy suffering rapidly developing multicystic hemorrhage

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Clinical History:

9 year-old boy developed a sudden onset of generalized seizure and admitted to local hospital. MRI demonstrated blurring of corticomedullary junction at left frontal cortex and spotty T2/ FLAIR high intensity at left basal ganglia. As he had an episode of infection 1 month prior to disease onset, acute disseminated encephalomyelitis was suspected. Half an year later, he developed headache, anorexia, and vomiting. MRI obtained at this point revealed multicystic hemorrhagic mass with perifocal edema at his left frontal lobe. CT scan revealed calcification. Opthalmological examination revealed no abnormalities. Genetic examinations of *SNORD118* turned out to be negative. Anti-Echinococcus Ab was also negative. After then, the lesion enlarged with multi-stage hemorrhage and was subjected to surgical removal.



FLAIR image at onset

Preoperative GdT1WI(Lt) and T2WI(Rt)



Materials Submitted:

- 1. Preoperative MRI image
- 2. Representative H&E slide

- 1. Histological characteristics
- 2. Differential diagnosis

A 13-year-old female with diminution of vision in both the eyes and headache

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Clinical history:

A 13-year-old female presented with insidious onset, gradually progressive loss of vision over 1-year duration, headache for 8 months, weakness in bilateral lower limbs for 1 month, squint in eyes since 2 weeks, generalised tonic-clonic seizures since 8 days, and bladder and bowel incontinence for 3 days. Four months prior to presentation, she had been empirically given antitubercular therapy and steroids with no symptomatic improvement. On examination, she was conscious and oriented. Perception to light was absent in bilateral pupils. There was sensory loss at D2 level and power was reduced in bilateral upper and lower limbs. MRI revealed leptomeningeal thickening along with spinal lesions at D1-D4 and L5-S1 levels, and multiple lesions in brain stem, thalamus and cerebellum. The patient underwent L5-S1 laminectomy with an open biopsy of the L5-S1 dural lesion.



Material Submitted:

- 1. Preoperative MRI image
- 2. Representative H&E slide

- 1. Histological characteristics
- 2. Molecular findings
- 3. Differential diagnosis

A 38-year-old female with right parietal lobe mass lesion

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Clinical history: A 38-year-old female began to show paroxysmal left-sided numbness more than 5 months ago. Her symptom spontaneously relieved after 1-2 minutes each time, without physical weakness and hallucinations. No disturbance of consciousness, nausea and vomiting. The patient felt uncomfortable before each attack, but it cannot be clearly stated. The patient visited orthopedics over 3 months ago. Cervical MRI showed no obvious abnormality. Then, the examination of head CT taken 11 days ago indicated a low density in the right basal ganglia, parietal lobe and temporal lobe, which was considered as space-occupying lesion. A cranial MRI showed a right parietal lobe mass lesion, being likely low-grade glioma.

The patient was admitted to the department of neurosurgery and the tumor was completely removed under the microscope (5cm×4cm×3cm).



Material Submitted: 1.Preoperative MRI image 2.Representative H&E slide and immunohistochemistry of IDH1R132H

Points for Discussion: Molecular findings and diagnosis

A 7-year-old girl presenting gradually worsening of headache and nausea

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Clinical history : A 7-year-old girl presented with 4 months history of gradually worsening of headache and nausea. On admission, she presented with left hemiparesis. CT and MRI demonstrated a well demarcated large right temporo-parietal cystic tumor with mural nodule which was strongly enhanced by Gd-DTPA, accompanying intratumoral hemorrhage. 3D-CT image of skull demonstrated calvarial erosion or lytic change. Our pre-operative diagnosis was pilocytic astrocytoma or pleomorphic xanthoastrocytoma, and underwent 2-stage total resection of the nodule and cyst wall of the tumor.



Material Submitted:

- 1. Preoperative neuro-images (CT scan, MRI, Angiography etc.)
- 2. Intraoperative findings
- 3. Representative H&E slide (and immunohistochemistry, electron microscopy, if necessary)

- 1. Histological characteristics
- 2. Molecular findings
- 3. Differential diagnosis

A 38-year-old female of a tumor of the lateral ventricle

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[Clinical History] A 38-year-old female presented with headache caused by right temporal tumor 60 mm in the major axis. She underwent surgical resection of it. Although she didn't receive postoperative treatment, the tumor recurred at the removal cavity wall 2 months after the surgery. She underwent repeated surgery. Then, she was treated with radiochemotherapy including temozolomide. After 4cycle of maintenance chemotherapy, thoracic spinal cord metastasis caused her paraplegia. She received irradiation and second line chemotherapy with bevacizumab. However, her intrathecal seeding of the tumor was progressing and her treatment was terminated.



[Submitted Material] 1. Preoperative MRI image: The lesion was heterogeneous gadoliniumenhancement. 2. Representative H&E slide

A 27-years-old female patient of right temporal lobe tumor which recurred many times

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Clinical History:

A 27-year-old female patient with left upper quadrantanopia and headache demonstrated a cystic mass lesion in the right temporal lobe on MRI. The tumor showed a well-circumscribed cyst with a mural nodule that was slightly low on T1WI and homogeneously enhanced by gadolinium (Gd). The tumor was subtotally removed. No adjuvant therapy was provided. Eighteen months after the initial resection, a small-mass lesion was observed on Gd-enhanced MR images (1st recurrence). Although the patient underwent stereotactic radiosurgery (SRS), the lesion continued to increase in size, and a second tumor resection was performed 13 months after SRS. The pathological diagnosis of the first recurrent tumor was the same as the first time with BRAFV600E mutation, but the tumor had gained a TERT promoter mutation. An enhanced lesion appeared 5 months after the second surgery (2nd recurrence); the tumor was removed 11 months after the second surgery. Pathological examination confirmed that the tumor was highly malignant and the diagnosis had been changed to highgrade glioma (malignant transformation). After the third resection, chemotherapy with ACNU was administered for three courses. However, the tumor recurred 8 months after the third resection (3rd recurrence). The fourth tumor removal was performed 9 months after the third resection. After the fourth resection, ACNU chemotherapy was administered again, but the tumor recurred 4 months later (4th recurrence). For the fifth surgery, we performed extended tumor removal and provided intraoperative photodynamic therapy (PDT). The pathological diagnosis and genetic information of the fourth and the fifth resected specimens were the same as the third resected specimen.



Initial MRI: T1WI / T2WI / Gd(+)T1WI

Material Submitted:

- 1. Preoperative MRI images
- 2. Representative H&E slides

- 1. Pathological diagnosis after the malignant transformation
- 2. Differential diagnosis
- 3. The significance of an acquired *TERT* promoter mutation