Introduction
Malignant Rhabdoid Tumor (MRT) has been reported as a variant of the Wilms tumor in 1978. MRTs most often arise from the central nervous system and the kidney. And one of the most lethal tumors in childhood. Occur in six out of 100 million children. Reported survival rates in the first year were 17%, and those with the onset of infancy survival in the first one year, were 0%. In most tumors, the hSNF5/INI1 (SMARCB1) tumor suppressor gene is inactivated.

Case Report
3 months girl. Birth at 39w4d 2910g. The patient was transferred at age 3 months for management of large extracranial tumor diagnosed with hemangioma. Although propranolol therapy was initiated, it was not effective, resulted in growth in size. Pathological examination of head tumor revealed a diagnosis of MRT suggested by eosinophilic perinuclear cytoplasmic inclusions and loss of INI1-protein expression, and by no stain of BaF47, immunohistochemically.
We performed chemotherapy (VDC (vincristine+cyclophosphamide+dorubicin) 3 course and ICE (ifosfamide+etoposide+carboplatin 2 course). The pediatric oncologist, we pediatric otolaryngologist and parents hold conference and decided to resect tumor by operation.

After Chemo-Therapy (4 month)

Operation
The left radical neck resection (Level I –V) was performed. Surgery completion time was 3 hours 45 minute. The parotid gland, left facial nerve, accessory nerve and sternocleidomastoid muscle were resected. And hemorrhage volume was 60ml. The lymph node metastasis were 11/20 Pieces. The tumor was completely resected.

2 weeks after operation
multiple intracranial tumor was found at the postoperative follow up CT.

And she was managed palliatively and succumbed at the age of 8 month despite surgical resection and initial good response to chemotherapy.

The therapy for MRT is multidisciplinary treatment but no effective therapy has been established.
SMRCB1 gene mutation was found in MRT. It is hoped that the treatment method, for example gene therapy, will be established by piling up the cases.

The conclusion
(headlines: A case report of MRT of head and neck of infant.)

• We report a case of Malignant Rhabdoid Tumor (MRT) in head and neck.
• MRTs are uncommon and aggressive tumors that typically present in childhood, and that are associated with extremely poor prognosis.
• After chemotherapy (VDC (vincristine+cyclophosphamide+dorubicin) 3 course and ICE (ifosfamide+etoposide+carboplatin 2 course), we operated for total tumor removal with facial nerve sacrifice. However, intracranial MRT was found after two weeks postoperatively. The patient was died at the age of 8 months.