Recurrent Atypical Teratoid/Rhabdoid Tumor: Case Report of a Recurrence Involving the Internal Auditory Canal

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INTRODUCTION

- Atypical teratoid/rhabdoid tumor (AT/RT) is a rare, aggressive, embryonal malignant tumor of the central nervous system that most often affects young children and was first described in 1987.[1-2]
- Represents1.6% of all CNS neoplasms in patients < 19 y/o, but up to 10% of CNS tumors in children < 1 y/o.[1]
- In 2016, WHO defined AT/RT by alterations of either INI1 protein (SMARCB1 gene), or rarely, BRG1 protein (SMARCA4 gene) in the presence of rhabdoid cells.[3]
- Presenting symptoms: headaches, increased intracranial pressure, ataxia, cranial nerve deficits, vomiting, and visual disturbances. [4-5]
- Posterior fossa is most common tumor location in children.[6]
- Survival time after diagnosis ranges from 0.5 months to 11 months and is significantly shorter for infants.[7]

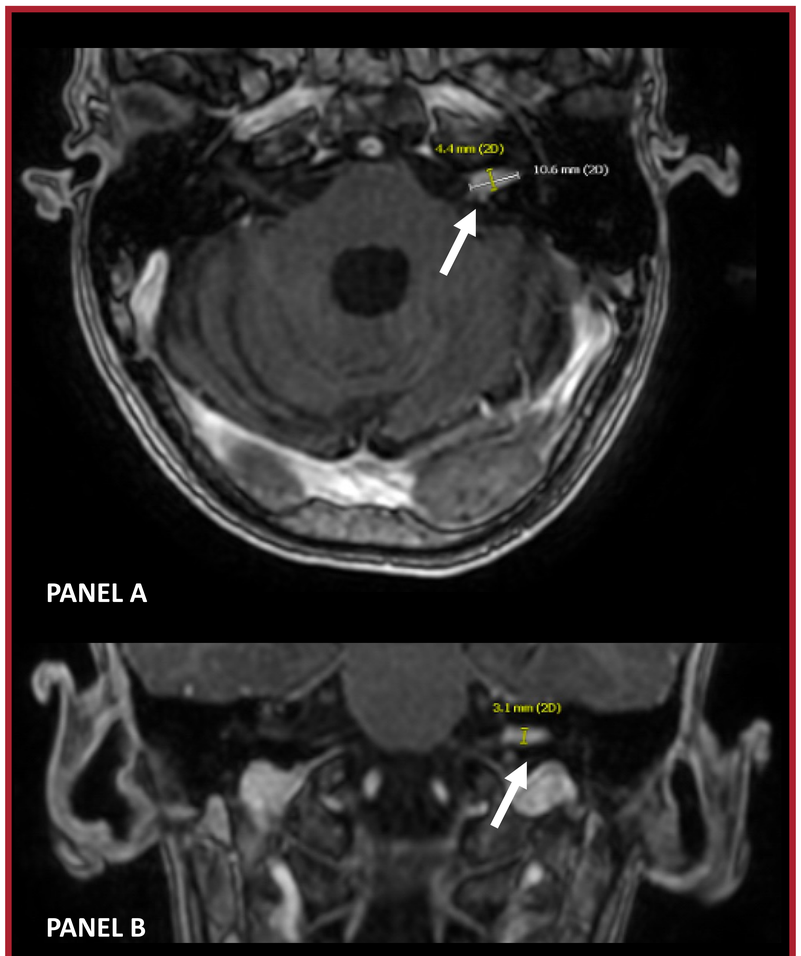

Figure 1
Axial and coronal images of T1 post gadolinium MRI demonstrating enhancing left IAC lesion. Lesion measures 1cm x 3mm x 4.4mm.

CASE DESCRIPTION

- 3 y/o presented with 4-6 weeks of 5 or more episodes of emesis each morning.
- Work-up included a CT head which showed a posterior fossa tumor causing hydrocephalus.
- Patient underwent gross total resection of the posterior fossa tumor followed by adjuvant chemoradiation.
- Two siblings diagnosed with AT/RT and SMARCB1 gene mutation identified.
- Disease free until age 20 when a recurrence was found in the spinal canal. Patient underwent gross total resection and adjuvant chemoradiation.

NEUROTOLOGY CONSULT

- At age 21, surveillance imaging showed a new enhancing mass within the left internal auditory canal. (Figure 1)
- Symptoms included left ear fullness, mild autophony and bilateral high frequency profound SNHL. No balance disturbances.
- Patient underwent gross total resection of the left IAC mass via a middle fossa approach and adjuvant chemoradiation.
- Surgical pathology consistent with recurrent AT/RT.
- 1 year after surgery, patient was found to have a large recurrent tumor in posterior fossa (Figure 2) and passed away from cerebellar herniation.

DISCUSSION

- To our knowledge, this is the 3rd report of an AT/RT involving the internal auditory canal, and the first known report of a tumor recurrence in this area.[8-9]
- Given the rarity of AT/RT and its potential involvement of the IAC, this case report fills a significant knowledge gap related to AT/RT and the potential need for neurotologist intervention.

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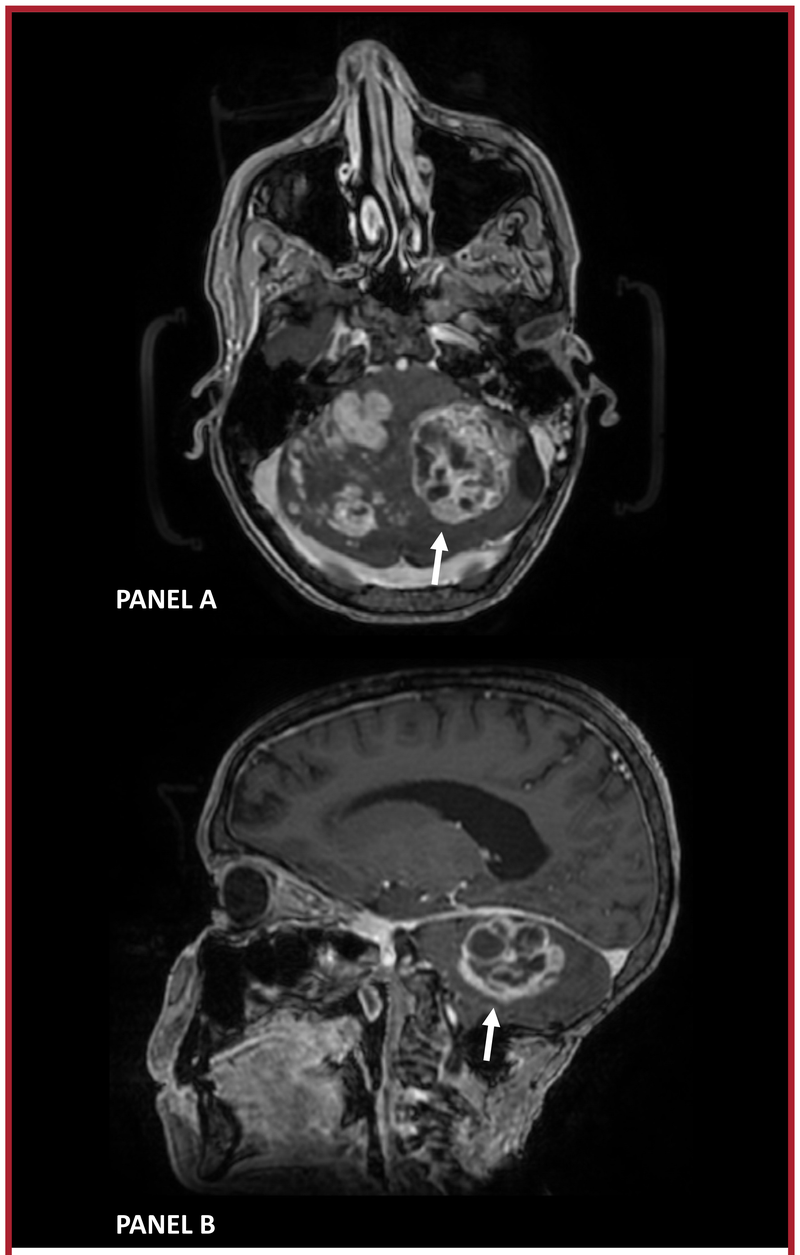

Figure 2
Axial and sagittal MRI images showing extensive AT/RT recurrence.

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