

Medulloblastoma: Intraventricular Recurrences

Hendy Setiawan¹, Celia², and Ridha Dharmajaya³

^{1,2}Resident, Department of Neurosurgery, Faculty of Medicine, Sumatera Utara University/RSUP. H. Adam Malik Medan

³Head of Departement, Department of Neurosurgery, Faculty of Medicine, Sumatera Utara University/RSUP. H. Adam Malik Medan

Abstract: Medulloblastoma is the most common malignant pediatric brain tumor. Incidence in adult population is less than 1 %. Medulloblastoma in adult usually between 20 and 40 years old, which is lateral hemispheric and not midline vermian. Survival rate in adult is 67% lower than adolescents is 69% and children is 72%. Location of recurrences most commonly presents at posterior fossa, spinal, supratentorial, and bone metastases. Recurrences of medulloblastoma in supratentorial ventricular is uncommon. The outcome for patients with recurrent medulloblastoma has historically been poor. 37 years old male who presented with severe headache gradually. Patient has history of headache 2 months ago and gait ataxia 1,5 months ago. Patient had surgery 2 years ago with result of hystopathology medulloblastoma. He had irradiation completely 22 months ago. Physical examination showed GCS 15 with gait ataxia. Head CT scan show multiple isodense lesion intraventricular. Before patient had surgery, Head CT scan showed hiperdense lesion at cerebellopontine angle. Seeding medulloblastoma to intraventricular is recurrences which have poor prognosis. Patient had poor condition in few days. Patient died before got reradiation

Keyword: Intraventricular, Medulloblastoma, Reirradiation, Recurrences, Seeding

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1 Introduction

Medulloblastoma is the most common malignant pediatric brain tumor. Incidence in adult population is less than 1 %.[1] Medulloblastoma in adult usually between 20 and 40 years old, which is lateral hemispheric and not midline vermian.[2] Survival rate in adult is 67% lower than children is 72%. Location of recurrences most commonly presents at posterior fossa, spinal, supratentorial, and bone metastases.[3] Late recurrences of medulloblastoma in supratentorial ventricular is uncommon. Recurrent medulloblastoma has poor outcome, although gets radiotherapy and chemotherapy.[4] Recurrence of this tumor is well recognized and may require salvage therapy. Time to recurrence typically occurs within two years of initial diagnosis

*Corresponding author at: hsns987@gmail.com

E-mail address: hsns987@gmail.com

in the pediatric population. Tumors that follow Collin's Law (tumor free period of 9 months plus the age at diagnosis) are considered to be cured. However, late recurrence of medulloblastoma has been documented in the pediatric population.[5]

2 Case Report

37 years old male patient who presented with severe headache gradually. Patient has history of headache 2 months ago and gait ataxia 1,5 months ago. Patient had been operated ventriculoperitoneal shunt (VP shunt) and resection tumor 2 years ago with result of histopathology desmoplastic medulloblastoma. He had cranio-spinal irradiation completely 22 months ago. He had not complaint after irradiation. Physical examination showed GCS 15 with gait ataxia. CT scan shows multiple isodense lesion intraventricular.



Figure 1. Non contrast CT Scan of brain before surgery shows right cerebellopontine angle lesion isodense

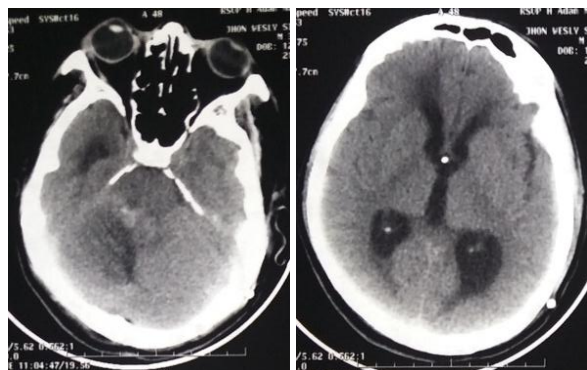


Figure 2. Non Contrast CT Scan of brain after he had been tumor removed and VP Shunt

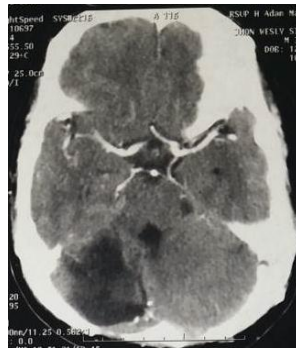


Figure 3. Contrast CT Scan of brain after irradiation shows mixed density at right cerebellum

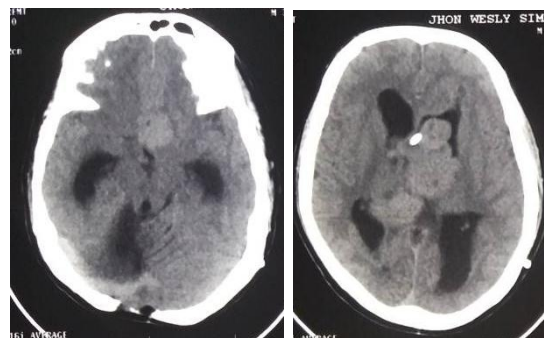


Figure 4. Non contrast CT Scan of brain 2 years later shows multiple isodense lesion intraventricular

3 Discussion

Medulloblastoma, a malignant tumor typically arising from the cerebellar vermis in young children and lobes in older children, represents 4–8% of all intracranial tumors, and is the most common malignant central nervous system tumor of childhood,⁵ with approximately 80% occurring in patients under 15 years of age.⁶ It accounts for 15–25% of all childhood brain tumors in comparison with only 1% of adult intracranial neoplasms, with a slight male predominance in both groups.[7]

Differences in childhood and adult tumors can also be observed in terms of histologic variant. Classical histologic presentation of medulloblastoma includes densely-packed primitive cells with hyperchromatic nuclei, scant cytoplasm, and nuclear molding.[8] This classical tumor histology is more common in children.⁷ Homer-Wright rosettes may be seen.[9] Adults are more likely to have laterally-located tumors, which more often desmoplastic.[9] Both tumor types include markers of neuronal lineage, such as class III beta tubulin and MAP-2 neurofilaments, whereas more adult tumors will express GFAP.[10] The anaplastic/large cell variant is more frequently seen in older children and adults.

Medulloblastoma tends to seed the cerebrospinal fluid resulting in extensive leptomeningeal involvement, and treatment therefore requires surgical resection followed by neuro-axis radiotherapy and chemotherapy.[11] Recurrence in the pediatric population differs from that seen in adults. Age at diagnosis has been shown to be a significant predictor of time until relapse.[12] The majorities of pediatric recurrences are within two years of initial diagnosis, and are in the posterior fossa.[12] Those with tumor-free period equal to the age at diagnosis plus nine months may be considered cured (Collin's Law).[13] While a good predictor of many childhood tumors, there are known exceptions to Collins law for medulloblastoma. However, a recent review of 125 patients by Massimino et al., gender, age at diagnosis, metastases, and therapeutic protocol were shown to have no prognostic impact.[13]

In the event of recurrence, relapse at a single site and further out from initial time of diagnosis is considered a more favorable prognostic indicator.[14] Recurrences are most common in the posterior fossa, followed by spinal, supratentorial and boney metastases.[7] Supratentorial dissemination is often found in the subfrontal area and may be due to overly generous radiation protection of the orbital roof to prevent irradiation of the cribriform plate, allowing for a nidus of recurrence.[14] Late recurrence of medulloblastoma in the supratentorial ventricular compartment is uncommon. We present a case of late recurrent medulloblastoma 13 years after initial diagnosis, again validating the need for long term follow-up in the pediatric population following diagnosis of medulloblastoma

4 Conclusion

Seeding medulloblastoma to intraventricular is recurrences which have poor prognosis. Patient died before got reirradiation. Although he had not complaint after surgery and first irradiation.

CONFLICT OF INTEREST

The authors declare no conflict of interest regarding this manuscript.

FUNDING

The authors declare no financial interest in any of the materials discussed in this work.

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