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Case Report

A rare case of multifocal atypical teratoid rhabdoid tumor ☆,☆☆

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ABSTRACT

Atypical rhabdoid teratoid tumor of the primary central nervous system is an uncommon and highly aggressive tumor that often affects infants and young children. Approximately two-thirds of tumors start in the posterior fossa. The best treatment for atypical rhabdoid teratoid tumor is yet unknown. Chemotherapy, radiation, and surgery are available as treatment options. Their respective roles are yet not each well defined. The prognosis for atypical rhabdoid teratoid tumor is absolutely poor. In this article, we intended to introduce a very rare case of multifocal atypical rhabdoid teratoid tumor in a 7-year-old boy.

Introduction

Atypical teratoid rhabdoid tumor of the central nervous system is a relatively uncommon, rapidly expanding tumor that starts in the brain and spinal cord. The majority of atypical teratoid rhabdoid tumors, which are rare WHO grade 4 tumors, affect young children under the age of 2 with a little male preponderance [1–5]. The brain stem or cerebellum are where around half of these cancers develop. The pineal region, the spine, and other regions have also been documented to be impacted. Imaging and even H&E microscopy frequently show similarities between atypical teratoid rhabdoid tumor and medulloblastoma, making cytogenetic study of the tissue necessary for the diagnosis [1–5].

Case presentation

A 7-year-old male, suffering from 3-month headache, nausea, and vomiting, was admitted to department of neurosurgery,

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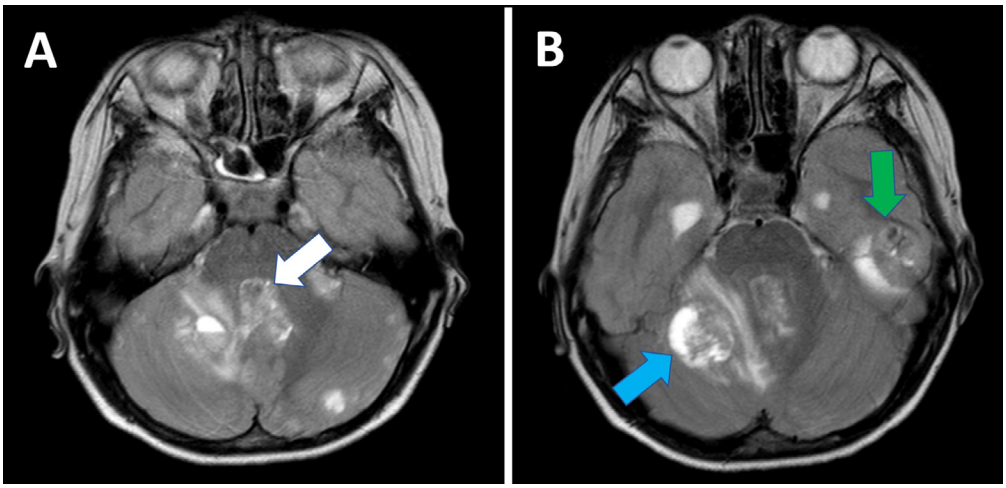

Fig. 1 – Axial T2-weighted images revealed the solid tumor inside the fourth ventricle (white arrow) (A) and mixed cyst and solid tumors at right cerebellar hemisphere (blue arrow) and left temporal lobe (green arrow).

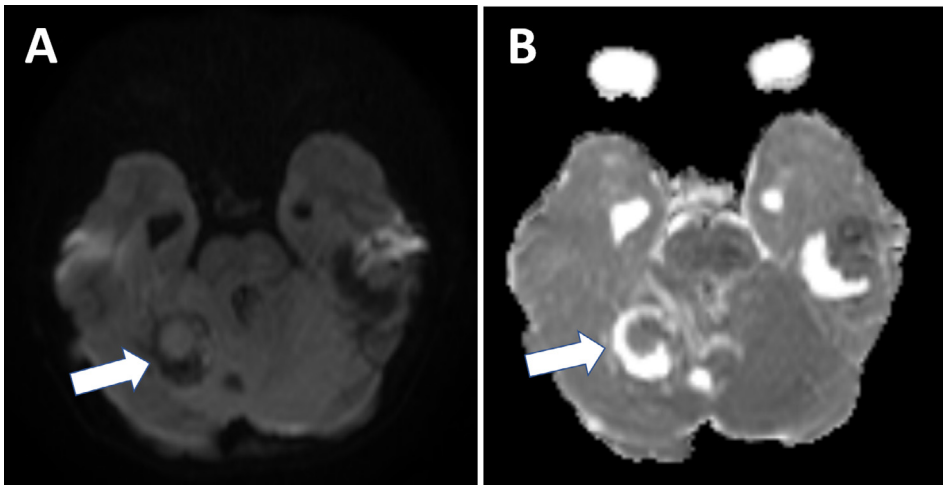

Fig. 2 – (A) Diffusion-weighted image and (B) apparent diffusion coefficient map of tumor (arrow).

Children's Hospital 2. His medical profile manifested no abnormalities. Neurological deficits were not detected during clinical evaluation. The routine laboratory tests and tumor markers were within normal ranges. Brain magnetic resonance imaging (MRI) with contrast agent was immediately indicated for him. There were multifocal masses observed on MRI scan. Among these masses, there were 3 biggest lesions (intra-fourth-ventricle solid tumor with size of 40 mm × 23 mm; right cerebellar hemisphere heterogeneous mass with size of 23 mm × 27 mm; and left temporal lobe heterogeneous mass with size of 31 mm × 35 mm) (Fig. 1). There was no hemorrhage or calcification observed inside the lesion. The mean apparent diffusion coefficient (ADC) values for the solid component of mass was 0.6×10^{-3} mm^2/s (Fig. 2). The fractional anisotropy value of the tumor was 0.3 (Fig. 3). The tumors were enhanced heterogeneously on T1-weighted image with contrast agent (Fig. 4). On MRI spectroscopy, the choline/N-acetyl aspartate ratio of the solid part of mass was 2.24 (Fig. 5). The initial diagnosis, relied on the clinical and subclinical informa-

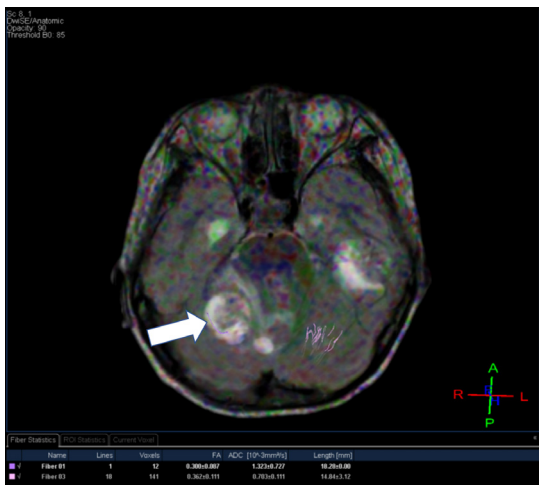

Fig. 3 – Diffusion tensor imaging of the lesion.

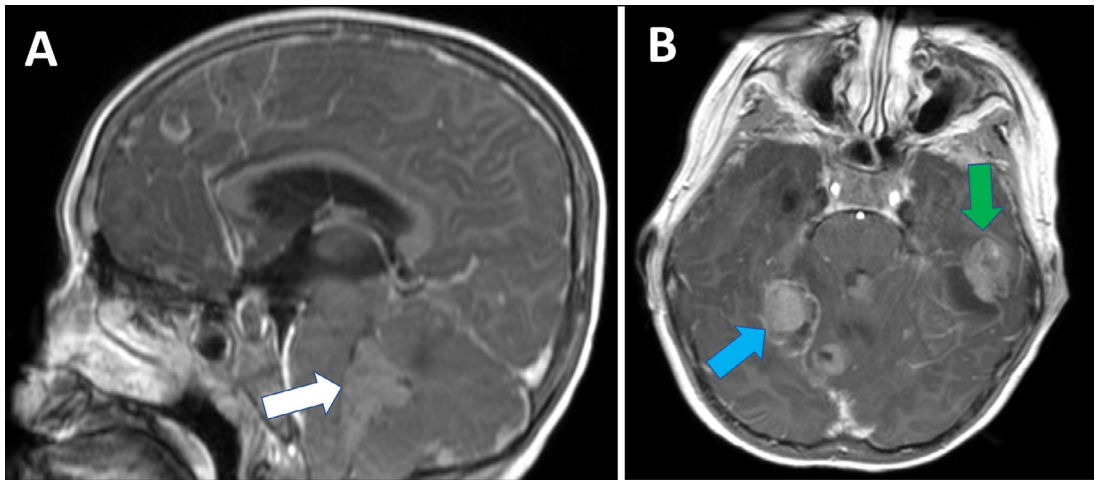

Fig. 4 – Sagittal (A) and axial (B) T1-weighted images with contrast enhancement revealed the solid tumor inside the fourth ventricle (white arrow) and mixed cyst and solid tumors at right cerebellar hemisphere (blue arrow) and left temporal lobe (green arrow).

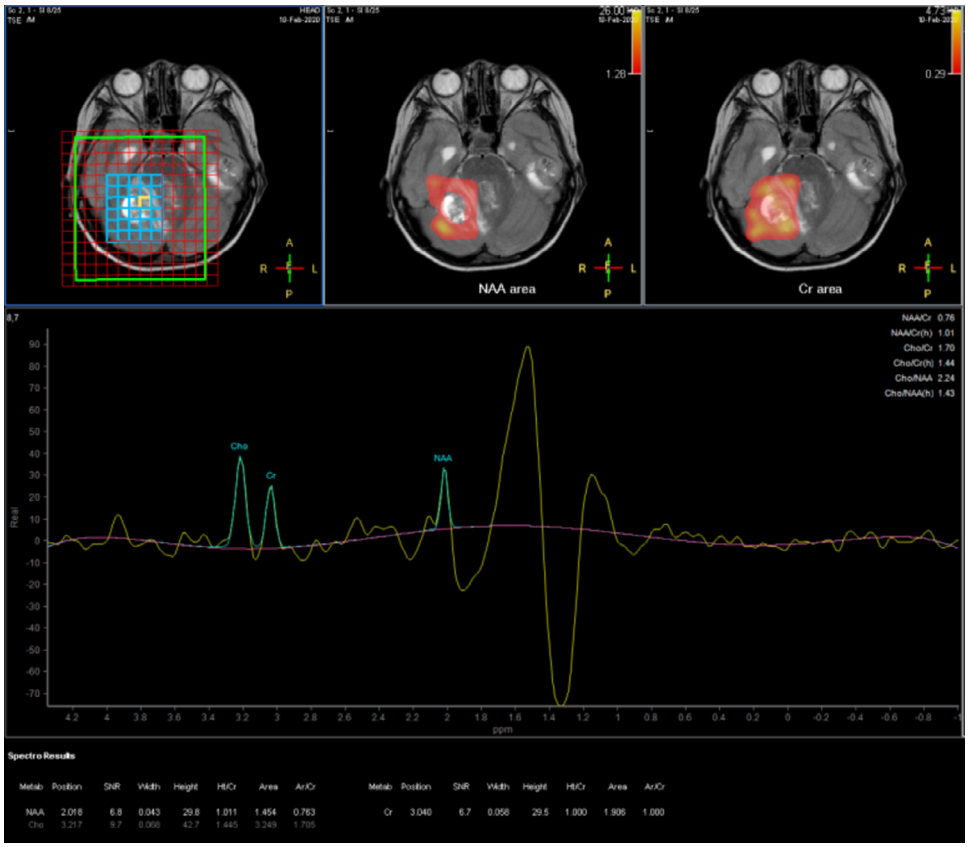

Fig. 5 – Magnetic resonance spectroscopy of the lesion.

tion, was a medulloblastoma. The patient underwent a surgical biopsy. The histopathological evaluation of the excised tumor tissues displayed atypical teratoid rhabdoid tumor. Even though, the patient was transferred to a distinct oncological center to adopt adjuvant chemo- and radiotherapy, he died 3 months later.

Discussion

The central nervous system atypical rhabdoid teratoid tumor is an uncommon, malignant tumor that often manifests in infancy. This form of tumor was initially identified as an ex-

tremely bad prognosis histology variation of Wilm's tumor, which primarily affects newborns. Rhabdoid cell tumors in the central nervous system were initially identified in 1985, but it wasn't until 1995-1996 that their clinical and pathological characteristics were clearly established [2].

Typically, the age at presentation is under 2 years old with a male to female ratio of 1.6:1 [6]. However, reports suggest that older children and adults might also experience it. Twenty to 30% of the time, distributed forms are present [7]. The median time to death for patients with atypical rhabdoid teratoid tumor is only a few months following the diagnosis, and their prognoses are often grim. When diagnosed, up to one-third of individuals have CSF dispersion [1].

The radiological results are vague. Some physical characteristics of atypical rhabdoid teratoid tumor are similar to those of medulloblastoma/primitive neuroectodermal tumors, and it can very closely resemble choroid plexus papilloma or choroid plexus carcinoma [8]. Rhabdoid cells, which may or may not include fibrillary globoid inclusions and strongly eosinophilic cytoplasm, big and eccentrically positioned nuclei, and a single conspicuous nucleolus, are the histological sign of atypical rhabdoid teratoid tumor. Additionally, rosette development was visible. Atypical rhabdoid teratoid tumor exhibit positive vimentin, GFAP, EMA, cytokeratins, synaptophysin, chromogranin, and SMA immunohistochemistry staining [2,5,9].

There is no agreement on how best to treat this tumor because of its rarity and fast progression. However, a combination of chemotherapy, radiation treatment, and surgery may be employed to enhance a bleak prognosis [1–4].

Conclusion

Multifocal atypical rhabdoid teratoid tumor in a child is exceptionally rare without previous reports. The best course of therapy for this extremely aggressive tumor is yet unknown. Noted that this disease had a catastrophic prognosis. Further study needs to be carried out to achieve better diagnostic and therapeutic solutions for atypical rhabdoid teratoid tumor.

Author contributions

NMD performed all steps to complete this article.

Ethics approval

This study has been approved by the hospital ethics committee (Ref: 352/NĐ2-CĐT).

Data sharing statement

Not applicable.

Patient consent

Informed consent has been obtained from the family members of the patient included in this study.

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REFERENCES

[1] Hilden JM, Meerbaum S, Burger P, Finlay J, Janss A, Scheithauer BW, et al. Central nervous system atypical teratoid/rhabdoid tumor: results of therapy in children enrolled in a registry. J Clin Oncol 2004;22(14):2877–84.
[2] Rorke LB, Packer RJ, Biegel JA. Central nervous system atypical teratoid/rhabdoid tumors of infancy and childhood: definition of an entity. J Neurosurg 1996;85(1):56–65.
[3] Bouvier C, De Paula AM, Fernandez C, Quilichini B, Scarvada D, Gentet JC, et al. Atypical teratoid/rhabdoid tumour: 7-year event-free survival with gross total resection and radiotherapy in a 7-year-old boy. Childs Nerv Syst 2008;24(1):143–7.
[4] Lee IH, Yoo SY, Kim JH, Eo H, Kim OH, Kim IO, et al. Atypical teratoid/rhabdoid tumors of the central nervous system: imaging and clinical findings in 16 children. Clin Radiol 2009;64(3):256–64.
[5] Louis DN, Perry A, Wesseling P, Brat DJ, Cree IA, Figarella-Branger D, et al. The 2021 WHO classification of tumors of the central nervous system: a summary. Neuro Oncol 2021;23(8):1231–51.
[6] Lee YK, Choi CG, Lee JH. Atypical teratoid/rhabdoid tumor of the cerebellum: report of two infantile cases. ANJR Am J Neuroradiol 2004;25(3):481–3.
[7] Tinsa F, Jallouli M, Douira W, Boubaker A, Kchir N, Hassine DB, et al. Atypical teratoid/rhabdoid tumor of the spine in a 4-year-old girl. J Child Neurol 2008;23(12):1439–44.
[8] Minh Thong P, Minh Duc N. The role of apparent diffusion coefficient in the differentiation between cerebellar medulloblastoma and brainstem glioma. Neurol Int 2020;12(3):34–40.
[9] Reedy AT. Atypical teratoid/rhabdoid tumors of the central nervous system. J Neurooncol 2005;75(3):309–13.