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Posterior fossa medulloblastoma in an atypical extra-axial location: A case report

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ABSTRACT

Medulloblastoma is the most common posterior fossa tumor of childhood typically within the fourth ventricle. However, extra-axial medulloblastoma in posterior fossa is an uncommon diagnosis. We report a case in a 33-month-old male who presented with repeated complaints of abdominal pain, intermittent emesis, and diarrhea, and diagnosed with right cerebellar extra-axial medulloblastoma, which was surgically resected. Majority of the reported extra-axial medulloblastoma in posterior fossa in the United States are located in the cerebellopontine angle. However, to the best of our knowledge, our case is the first to document medulloblastoma occurring exclusively in the cerebellar hemispheric extra-axial space rather than the cerebellopontine angle. Although the diagnosis can present as a radiological dilemma, a systematic multimodality imaging approach can aid in narrowing the differential diagnosis and timely management. In this case report, we will discuss the imaging characteristics, differential diagnosis, and management strategies, alongside a brief review of the world literature of extra-axial medulloblastoma.

Introduction

Medulloblastoma is the most common malignant, invasive, childhood embryonal tumor that occurs typically in midline posterior fossa. Approximately 500 children are diagnosed with medulloblastoma each year in the United States. It accounts for 20% of all pediatric tumors, with a peak age distribution of 5-9 years old and a preponderance for males [1]. Medulloblastoma typically presents midline or paramedian in the cerebellum, often compressing and invading the fourth ventricle. Diagnosis requires histopathologic confirmation at time of resection [2]. Children diagnosed at younger than 5 years of age, and especially those younger than 3, have been shown to have poorer prognosis with an estimated 5-year survival of 32% [3]. That said, histopathologic and molecular subtype have

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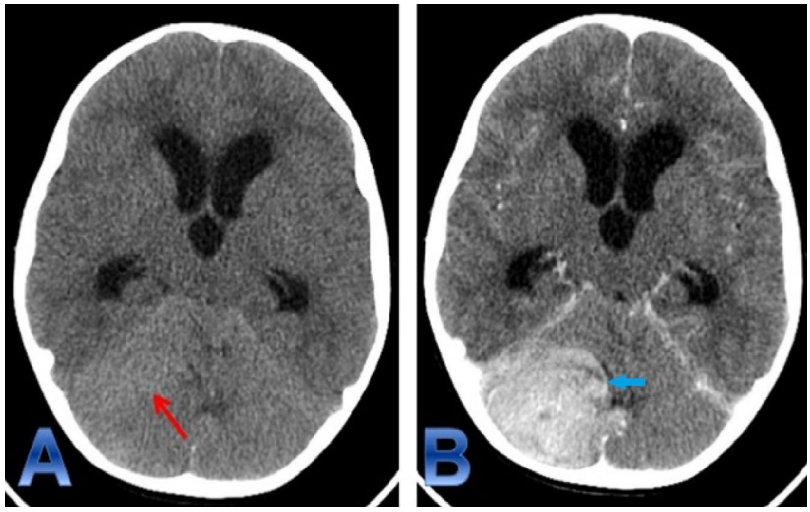

Fig. 1 – Noncontrast CT head (A) demonstrates a mildly hyperattenuating focus (red arrow) in the region of right cerebellar hemisphere. The lesion demonstrates homogenous enhancement (blue arrow) seen on the contrast-enhanced CT (B). In addition, mass effect of the right cerebellar hemisphere and brainstem, along with near complete effacement of the fourth ventricle results in obstructive hydrocephalus. CT, computed tomography.

been used to stratify patients into risk categories and determine appropriate therapy [4]. Herein, we present the atypical case of a 33-month-old patient diagnosed with medulloblastoma presenting in a rare location overlying the cerebellar hemisphere posterolateral to the cerebellopontine angle.

Case report

A 33-month-old white male presented with 1-week history of increased sleepiness, abdominal pain, emesis, and diarrhea previously diagnosed as constipation and dehydration. Initial contrast-enhanced (CE) head computed tomography (CT) scan demonstrated an enhancing mass overlying the right cerebellar hemisphere with secondary mass effect on adjacent structures including effacement of the fourth ventricle, resulting in hydrocephalus (Fig. 1). Subsequent contrast-enhanced magnetic resonance imaging (CEMRI) brain depicted a well-circumscribed, extra-axial T2 hyperintense mass in the right posterior fossa measuring 4.3 × 4.7 × 3.1 cm, which exerted mass effect on the right cerebellar hemisphere, causing slight inferior herniation of the right cerebellar tonsil as well as effacement of the fourth ventricle and resulting in obstructive hydrocephalus. The mass demonstrated restricted diffusion on diffusion-weighted images (Figs. 2-8). Given the infratentorial location in the posterior fossa, mild heterogeneous enhancement, and age of the patient, extra-axial medulloblastoma was

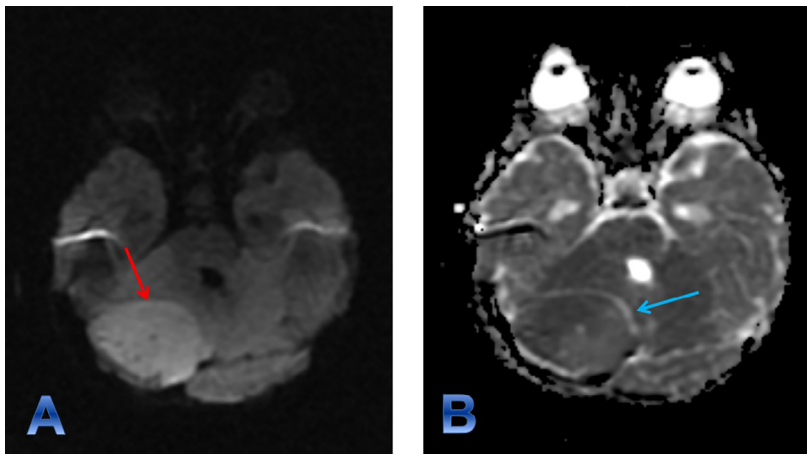

Fig. 2 – Diffusion-weighted imaging (DWI) (A) and apparent diffusion coefficient (ADC) sequences (B) clearly depict a large well-circumscribed focus of diffusion restriction (red arrow) within the right cerebellar hemisphere with corresponding signal loss on ADC maps. The hypercellularity of the lesion can be attributed to its low ADC value. A small rim of CSF (T2 signal intensity) well outlines the mass anteromedially (blue arrow). This finding along with the nature of the mass effect on the adjacent cerebellum and fourth ventricle guides the diagnosis toward an extra-axial tumor. CSF, cerebrospinal fluid.