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Case Report

Pilocytic Astrocytoma of the Brainstem in a Child Followed for Hereditary Spherocytosis: A Case Report

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Abstract

The majority of pediatric brain tumors originate within the posterior fossa, most commonly including juvenile pilocytic astrocytomas, medulloblastomas (MB), ependymomas, and brainstem gliomas. This report describes the case of a 10-year-old patient with underlying hereditary microspherocytosis presenting with headaches, vomiting, and a cerebellar syndrome. Brain computed tomography (CT) and brain magnetic resonance imaging (MRI) with magnetic resonance spectroscopy (MRS), displaying classic features, revealed a well-circumscribed dual cystic-solid lesion of the medulla oblongata without obstructive hydrocephalus. Pilocytic astrocytoma, ganglioglioma, ependymoma, and hemangioblastoma are among the main differential diagnoses.

Keywords: *Pilocytic Astrocytoma; Brainstem; Cerebellar Syndrome; MRI; Magnetic Resonance Spectroscopy; Child*

Introduction

Brain tumors represent the most common solid neoplasms in the pediatric population, with approximately 50% to 60% originating within the posterior fossa [1,2]. The most frequent histological subtypes in this anatomical compartment include juvenile pilocytic astrocytomas (PAs), medulloblastomas, ependymomas, and brainstem gliomas [2]. Among these, pilocytic astrocytoma is classified as a World Health Organization (WHO) Grade 1 tumor, typically demonstrating a benign biological behavior and favorable long-term outcomes when complete surgical resection is achievable [1].

Although pilocytic astrocytomas predominantly arise within the cerebellar hemispheres, vermis, and optic pathway, primary focal involvement of the brainstem—specifically isolated to the medulla oblongata—remains relatively uncommon [3]. Clinical presentation often mimics non-specific pediatric conditions or post-traumatic symptoms, leading to diagnostic challenges [4].

Initial imaging with non-contrast brain Computed Tomography (CT) is frequently performed in emergency settings to rule out acute hemorrhage, post-traumatic lesions, or acute hydrocephalus [4]. However, multi-parametric Magnetic Resonance Imaging (MRI) combined with Proton Magnetic Resonance Spectroscopy represents the gold standard for accurate lesion characterization, anatomical localization, and preoperative differential diagnosis [5].

Here, we report the case of a 10-year-old child with underlying hereditary microspherocytosis presenting with posterior fossa symptoms, where brain CT and MRI objectified a focal, well-circumscribed medullary lesion highly suggestive of a pilocytic astrocytoma in the first place.

Case Report

A 10-year-old child followed in the pediatric hematology department for hereditary spherocytosis, with no other notable medical history, presented recently, with no history of trauma, with a clinical picture combining vomiting and headaches. Neurological examination revealed a cerebellar syndrome.

Given this presentation, a non-contrast brain computed tomography (CT) scan was performed first (Figure 1). It showed no parenchymal density abnormality of evolving appearance at the supratentorial level. At the brainstem level, it identified a roughly rounded, well-defined medullary (bulbar) mass of tissue density, spontaneously hyperdense with a few scattered hypodense areas, without associated tonsillar herniation. This CT pattern was primarily suggestive of an acute/subacute brainstem hematoma, with a bleeding astrocytoma also considered, prompting correlation with prior imaging and further work-up with brain MRI.

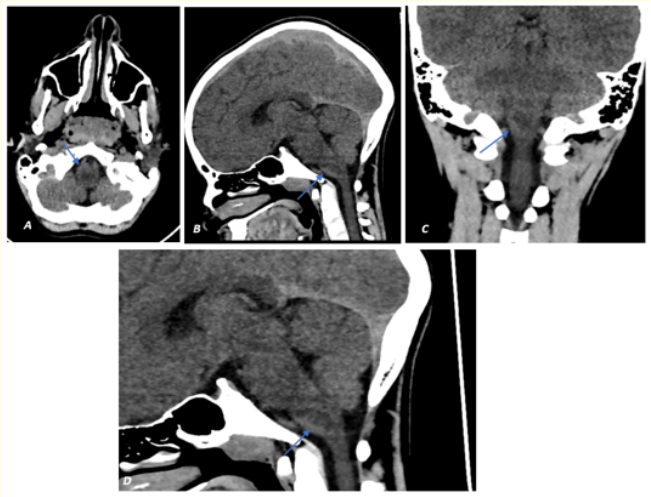

Figure 1: *Non-contrast brain CT scan showing: A) Axial view demonstrating a rounded, well-defined medullary (Bulbar) mass of tissue density, spontaneously hyperdense with scattered hypodense areas (Blue arrow); B, D) Sagittal reformations highlighting the focal swelling of the medulla oblongata (Blue arrow); C) Coronal reformation confirming the well-circumscribed nature of the medullary lesion (Blue arrow).*

A brain MRI was subsequently performed, including axial and coronal T2-weighted sequences, axial FLAIR, sagittal T1, Coronal TIR (Turbo Inversion Recovery), diffusion-weighted imaging, gradient-echo (GE) sequences, with contrast injection (Figure 2).

At the infratentorial level, the medulla (bulb) was the site of an oval, well-defined lesion with regular contours, showing two components: a predominantly cystic component, hypointense on T1 and hyperintense on T2/FLAIR, and a solid (tissue) component, isointense on T1 and T2, without diffusion restriction, showing heterogeneous enhancement after contrast injection, measuring 20 x 13 x 10 mm. The cerebellum and the rest of the brainstem were otherwise of normal morphology, without other signal abnormality. Spectroscopy performed on the lesion showed an inverted choline/creatine ratio as well as a lactate peak.

Based on this workup, the diagnosis considered in the first instance was a brainstem (bulbar) pilocytic astrocytoma, with other diagnoses still to be discussed.

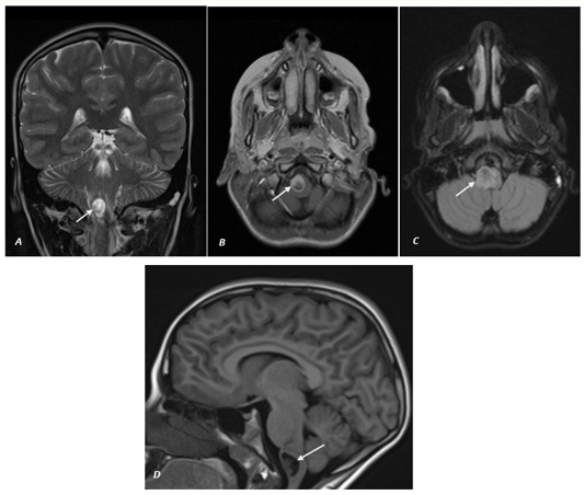

Figure 2: *Brain MRI showing the focal medullary lesion: A) Coronal T2-weighted image showing a well-defined, oval-shaped lesion within the medulla oblongata with high T2 signal intensity (Arrow); B) Axial FLAIR image highlighting the hyperintense cystic-solid nature of the mass (Arrow); C) Axial post-contrast T1-weighted image demonstrating heterogeneous enhancement of the solid mural component (Arrow); D) Sagittal T1-weighted image displaying the low T1 signal intensity of the predominantly cystic lesion within the medulla (Arrow).*

Clinical course: Three days after the MRI, the child developed acute respiratory deterioration, rapidly progressing to death.

Discussion

Pediatric brainstem gliomas represent a clinically and biologically heterogeneous group of tumors. They are broadly categorized into two main entities: diffuse intrinsic pontine gliomas (DIPGs), which carry a dismal prognosis, and focal brainstem gliomas, which typically exhibit low-grade histology and a more indolent clinical course [5]. Focal medullary lesions are particularly rare, representing a small subset of pediatric infratentorial neoplasms [5,6].

In our patient, non-contrast brain CT initially demonstrated a well-defined medullary mass of tissue density with high attenuation (spontaneously hyperdense) containing scattered hypodense areas. Spontaneous high attenuation on non-contrast CT in pilocytic astrocytomas is relatively atypical but can be attributed to high cell density, compact neuroglial architecture, or proteinaceous contents within micro-cystic changes. This underscores the inherent limitations of non-contrast CT in definitively characterizing brainstem tumors and highlights the vital role of multi-parametric MRI [6,7].

Subsequent MRI accurately delineated the classic dual-component structure of the lesion, featuring a predominant cystic component and a solid mural portion [8]. The absence of diffusion restriction on DWI supported a low-grade histology rather than a highly cellular, high-grade embryonal tumor like medulloblastoma or an acute ischemic lesion [9]. Proton MR spectroscopy further contributed to characterization by demonstrating an inverted Choline/Creatine ratio and a prominent lactate peak [9,10]. Although lactate accumulation typically reflects anaerobic metabolism and can raise suspicion of high-grade malignancy or necrosis, in juvenile pilocytic astrocytomas, lactate and lipid peaks are frequently identified within benign cystic cavities without denoting high-grade transformation [10].

The differential diagnosis for a focal cystic-solid medullary mass in a child includes [6]:

1. Pilocytic astrocytoma (WHO Grade 1): The leading diagnosis given the distinct margins, dual architecture, lack of diffusion restriction, and classic heterogeneous enhancement [1,8].
2. Ganglioglioma: A well-delineated mixed solid-cystic tumor, though calcifications are more frequently observed on CT [6].
3. Ependymoma: Typically arises from the floor of the fourth ventricle, often expanding through the foramina of Luschka or Magendie, with higher cellularity and restriction on DWI [6].
4. Hemangioblastoma: Rare in early childhood unless linked to Von Hippel-Lindau (VHL) disease, usually demonstrating marked hypervascularity and prominent T2 flow voids [11].

Despite the histologically benign appearance expected in pilocytic astrocytomas, their anatomical location within the brainstem poses critical risks [12]. The medulla oblongata houses vital autonomic reflex centers governing cardiorespiratory control and cranial nerve nuclei (IX-XII), including the nucleus tractus solitarius, whose disruption is specifically implicated in the generation of acute pulmonary edema [13]. Rapid neurological deterioration, as observed in our case, can occur secondary to focal mass effect, perilesional edema, intratumoral micro-hemorrhage, or acute brainstem compression. Such acute brainstem compromise is a recognized trigger of neurogenic pulmonary edema (NPE), a centrally mediated, sympathetically driven form of acute lung injury with a reported mortality of 60 - 100% regardless of the underlying etiology [14]. This fatal outcome illustrates that even low-grade, benign neoplasms can have a catastrophic clinical course when situated in eloquent neural structures.

Conclusion

This case highlights the diagnostic challenge of focal medullary lesions in pediatric patients presenting with non-specific posterior fossa symptoms. Although pilocytic astrocytoma represents the most likely diagnosis for a focal, well-circumscribed cystic-solid brainstem mass, its critical location within the medulla oblongata carries a grave risk of sudden, life-threatening cardiorespiratory failure, including neurogenic pulmonary edema, regardless of its benign histopathological grade.

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