Medulloblastoma in children: a case report and literature review

Diana Marcela Perea Rojas[1]*, Luis Enrique Perea Vásquez[2], Indiana Luz Rojas Torres[1], Carlos Mario Perea Molinares[3], Christian David Seni Hernández[1]

[1]*Faculty of Health Sciences, Universidad Simón Bolívar, Barranquilla, Colombia*
[2]*Quality Department, La Misericordia Clínica Internacional, Barranquilla, Colombia*
[3]*Department of Intensive Care Unit, Clínica General del Norte, Barranquilla, Colombia*

***Correspondence:** Diana Marcela Perea Rojas, Faculty of Health Sciences, Universidad Simón Bolívar, Barranquilla, Colombia, diana.perea@unisimon.edu.co

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ABSTRACT

Most primary nervous system tumors in children are medulloblastomas, this pathology originates in the posterior and lower part of the brain known as the cerebellum. This document aims to describe the case of a 2-year-old who presented to the emergency department with left eye ptosis and deviation of the labial commissure, with a history of headache and vomiting episodes that resulted in asthenic state. Imaging studies revealed a tumor lesion with signs of bleeding located in the right pontocerebellar angle and internal auditory canal. The diagnosis was made as follows: posterior fossa space occupying lesion, right facial paralysis, risk of intracranial hypertension and risk of neurological deterioration, leading to the conclusion: A malignant tumor that responds well to surgery, radiotherapy, and chemotherapy. Although it is rare, it mainly affects children and adolescents, which complicates treatment due to the long term negative effects on the developing brain.

CASE REPORT

CASE REPORT

We present the case of a 2-year-old male patient who was in the emergency room and was referred from a fourth-level complexity clinic in the city of Barranquilla, due to a clinical picture that began on November 5, 2022, characterized by left eye ptosis and deviation of the labial commissure towards the ipsilateral side. The patient had a history of convulsions one year ago, subsequent episodes of headache and vomiting with consequent asthenic state two months ago.

Upon evaluation by pediatrics and pediatric neurology, the patient was found to have a tendency towards drowsiness, with ptosis of the left eyelid and ipsilateral deviation of the labial commissure that did not respond to commands. There were no signs of intracranial hypertension, no signs of meningeal irritation were present, he tolerated ambient oxygen, and was diagnosed with Bell's palsy and brain-occupying mass.

Regarding imaging methods, the patient had underwent a cranial Computed Tomography (CT) scan one year ago due to convulsions, which was within normal limits (Figure 1). However, a Magnetic Resonance Imaging (MRI) revealed a tumor lesion with signs of bleeding located in the right pontocerebellar angle and internal auditory canal (IAC) causing a significant mass effect, displacing the brainstem to the left and collapsing the fourth ventricle with acute supratentorial hydrocephalus (Figure 2).

Given the patient's clinical condition and imaging findings, the treating medical team considered performing a pediatric medium-pressure ventriculoperitoneal shunt due to obstructive hydrocephalus secondary to the posterior fossa space-occupying lesion. At the same time, the patient was being treated in the Intensive Care Unit (ICU) due to the risk of neurological deterioration and secondary ventilatory failure. He remained hospitalized under the following diagnoses:

1. Posterior fossa space-occupying lesion by external MRI
2. Right facial paralysis
3. Risk of intracranial hypertension
4. Risk of neurological deterioration
5. Risk of ventilatory failure

Days later, as the patient showed no improvement, a neurosurgery meeting was held, and it was proposed to perform lesion resection, hence authorization for the procedure was requested. He underwent surgery and underwent craniotomy,

where a tumor lesion was identified; the tumor lesion was removed with the help of an ultrasonic aspirator, samples of the tumor lesion were taken, and pathology studies were ordered. The following imaging studies were performed:

1. Post-surgical cervical spine MRI with contrast, showing nonspecific edema of the soft tissues of the nuchal region and right hemi-neck, with fluid collection in the soft tissues adjacent to the surgical site. Particularly, no secondary neoplastic disease was identified (Figure 3).
2. Thoracic spine MRI with gadolinium within normal limits. Particularly, no lesions suggestive of metastasis were identified (Figure 4).

Furthermore, histological analysis of the tissue was performed, which reported a tumor of small, round cells encircling pale nodular regions, suggesting a nodular architecture, along with the presence of a dense intercellular network, histologically classified as desmoplastic nodular medulloblastoma.

Subsequently, the patient did not evolve satisfactorily, with episodes of desaturation, making weaning from mechanical invasive ventilation impossible. The patient was in poor general condition, looked toxic, in critical condition due to a torpid evolution of respiratory infectious pathology involving acid-base and ventilation-perfusion state, manifesting respiratory failure requiring mechanical support. Chest X-ray showed findings of multilobar pneumonia and a follow up then reported a pneumothorax (Figure 5).

Subsequently, a contrast-enhanced institutional brain MRI was performed, which reported surgical changes of right occipital approach with area of malacia in the lower aspect of the corresponding cerebellar hemisphere and residual extra-axial mass with occupation of right cisterns, internal auditory canal and mass effect especially on the middle cerebellar peduncle (Figure 6). Also, histological findings showed a tumor lesion of small, round, blue cells favoring medulloblastoma.

After discussing the case in the medical board, it was considered that due to the growth rate of the lesion evidenced by current imaging studies compared to pre-surgical ones despite the extensive lesion resection performed on January 25, 2023, urgent evaluation by oncology and radiotherapy for the initiation of adjuvant treatment was necessary.

The pediatric surgery department considered that the patient would not benefit from a new surgical approach, so he continued under neurological surveillance. The patient's prognosis was clinically evolving. Due to medical evolution, fractionated radiotherapy was indicated and performed, despite the treatments instituted, the patient remained with a fixed gaze, unresponsive to stimuli, presenting generalized spasticity, and requiring invasive mechanical ventilation.

DISCUSSION

Etiology & Demographics

Medulloblastoma (MB) is the most common malignant embryonal brain tumor in childhood, accounting for approximately 20% of primary nervous system tumors in children [1]. These tumors can result from various mutations in the genetic material and are associated with multiple hereditary syndromes that predispose to MB development; however, the etiology is unknown. Recent studies suggest that the tumor arises from cerebellar stem cells in the posterior zone of the cerebellar vermis and the neuroepithelial roof of the fourth ventricle in children, rapidly proliferating to form the neoplasm [2].

Tumors occurring in the Central Nervous System (CNS) represent the second leading cause of pediatric cancer diagnosed each year in the United States, which holds significant relevance. Medulloblastoma is primarily diagnosed in developing countries with an incidence ranging from 6.1% to 49.9%, whereas in developed countries, it presents with an incidence of between 20% and 25% of all CNS tumors. This higher incidence in developing countries may be due to factors such as lack of multidisciplinary teams involved in the diagnosis and management of the pathology, which also implies a worse prognosis [3].

The specific cause of medulloblastoma is not well established; however, it has been found to be associated with syndromes such as Gorlin syndrome, ataxia telangiectasia, Li-Fraumeni syndrome, Turcot syndrome, and it is also associated with the presence of extra chromosomes in the 6-12 and 4-5 groups [4].

Classification

They are differentiated either as unipotent or pluripotent, which varies from case to case. This explains the recognized histological variants, ranging from undifferentiated medulloblastoma through various stages to medulloblastoma with glial, neuronal, or even myoblastic components. Rosette formation is observed in half of medulloblastoma cases. They can be classified into four types based on histological findings [5,6].

1. Desmoplastic nodular MB: Consists of tightly packed hyperchromatic cells encircling pale nodular regions exhibiting patterns of neuronal differentiation. Desmoplasia is the word for the dense intercellular network created by these nodular patches.
2. MB with extensive nodularity: This type of MB has a broad lobular architecture and long, reticulin-free regions extending between nodules. Small spherical cells with a neurocytic appearance on a fibrillar background can be found in the internodular zones.
3. Large cell anaplastic MB: Defined by varying quantities of cytoplasm and enlarged, pleomorphic nuclei with conspicuous nucleoli. There are lots of mitotic and apoptotic figures.

4. Classical MB: Consists of patches of neuroblastic rosettes of Homer-Wright and sheets of tiny, poorly differentiated cells organized in parallel rows.

Molecular Genetics

There are subtypes of medulloblastoma, each with specific characteristics and genetic mutations. The Sonic Hedgehog (SHH) subtype, found in approximately 30% of pediatric medulloblastoma cases, is characterized by mutations in genes that regulate this signaling pathway, such as *PTCH1*, *SMO*, and *SUFU*. This subtype may also have mutations in other genes, such as *TP53* and *MLL2* [7,8]. TP53 mutations are present in approximately 20% of all patients, which categorizes them into a very high risk group with poor prognosis, also, around 20% of this patients have metastases at the time of diagnosis, with a 5-year survival rate ranging from 67-89% [9,10].

The WNT subtype (Wingless activated) is less common, present in only about 10% of pediatric medulloblastoma cases. It is characterized by mutations in genes that regulate the WNT pathway, such as *CTNNB1* and *APC* [7, 8]. This subtype rarely metastasizes and has a favorable outcomes compared to the other groups, with a 5-year survival rate ranging from 97-100% [9,10].

The group 3 subtype is the most aggressive of all pediatric medulloblastoma subtypes, found in approximately 25% of cases, and is characterized by a high rate of genetic mutations and amplification of the *MYC* gene. Deletions in chromosomes 8q and 11q may also occur [7,8]. These patients have the poorest prognosis and exhibit leptomeningeal dissemination at the time of diagnosis in 40-50% of cases, with a 5-year survival rate ranging from 42-66% [9,10].

The group 4 subtype is the most common, present in approximately 35% of pediatric medulloblastoma cases. It is characterized by the dysregulation of multiple signaling pathways, including the SSH and WNT pathways. It typically involves changes in the expression of genes that regulate brain tissue formation and cell migration, such as *TSHZ2* and *KCNA1* [7,8]. 35-40% of these patients have metastasis at diagnosis, however, the overall outcome is intermediate with a 5-year-survival rate ranging from 67-83% [9,10].

Physiopathology

The origin of this neoplasm is currently unknown; recent advances suggest that the tumor originates from pluripotent blasts that do not reach complete maturation until they reach the normal state of growth arrest. For this reason, some new classifications from the World's Health Organization (WHO) include the neoplasm among primitive neuroectodermal tumors (PNET) [11].

The body's natural immune defenses are unable to control the abnormally fast and uncontrollable growth and division of immature or incompletely differentiated cells. Neoplasia results from the proliferation of aberrant cells that progresses oncogenously. At the time of diagnosis, it frequently results in an increase in intracranial pressure because of its proximity to the fourth ventricle. According to the WHO classification, they are categorized as grade IV tumors and exhibit aggressive growth both locally and potentially throughout the subarachnoid area [11].

Clinical & Imaging Findings

The clinic depends on the age of the patient and the extent of the disease, whether local or disseminated. Generally, it has a brief duration, less than 3 months, reflecting the tumor's biological aggressiveness. It is usually due to increased intracranial pressure from hydrocephalus secondary to tumor obstruction, leading to headaches, vomiting, papilledema, irritability, diplopia, nystagmus, and an increase in head circumference in younger children. Patients with medulloblastoma present a combination of signs and symptoms of intracranial hypertension and cerebellar dysfunction that evolve over weeks to months. Symptoms include daytime or nighttime headaches, nausea, vomiting, and altered consciousness. Midline tumors may produce ataxic gait or trunk instability, while tumors in the cerebellar hemispheres likely cause limb motor incoordination and intention tremor. Dizziness and diplopia are common symptoms that can be caused by brainstem involvement or compression, or secondary to increased intracranial pressure, for example, sixth nerve involvement due to increased intracranial pressure, as well as papilledema and partial or complete loss of vision [12].

The imaging diagnosis of medulloblastoma, a malignant tumor of the central nervous system, generally involves a combination of imaging studies. The main studies used for the diagnosis and evaluation of medulloblastoma include [13].

- Magnetic Resonance Imaging MRI: MRI is the most commonly used imaging technique for the diagnosis and characterization of medulloblastoma. It provides detailed high-resolution images of the tumor's structure and location, as well as its relationship with surrounding structures. T1- and T2-weighted image sequences, as well as contrast-enhanced sequences, help identify and evaluate the size, extent, and contrast enhancement of the tumor.
- Computed Tomography CT: CT can be used as a complementary study to evaluate medulloblastoma, especially in situations where MRI is not available or suitable. CT can provide information about the location, size, and calcification of the tumor.

It is important to note that the definitive diagnosis of medulloblastoma is made through a biopsy or surgical resection of the tumor, followed by histopathological analysis.

Treatment & Prognosis

Prognosis

- Age greater than three years old: If the tumor is high risk

(disseminated), the 5-year disease-free survival probability is 60–70%; if the tumor is average risk, it is 80% [14].

- Age less than three years old: Because up to 40% of children have widespread disease at the time of diagnosis and because radiation therapy is typically avoided or postponed in this age range, the prognosis is more uncertain. If they survive, children may experience substantial long-term deficiencies in their neurocognitive skills (e.g., language learning, executive function, memory) [14,15].

Treatment

The treatment of medulloblastomas requires a multidisciplinary team, consisting of pediatricians, pediatric oncologists, pediatric neurologists, neurosurgeons, and oncology nurses. Treatment depends fundamentally on the site, size, nature, stage, and progression of the tumor, as well as patient factors such as age, health status, and other factors [16].

Chemotherapy has been added to increase survival and lessen side effects after radiation, even though postoperative radiotherapy plus chemotherapy is thought to be the standard of care for children with medulloblastoma. Eliminating craniospinal radiotherapy in very young children, lowering the dose and field of craniospinal axis, and lowering the boost volume to the tumor bed in older children were the methods used to reduce the negative effects of radiation therapy. Nevertheless, in a different investigation, satisfactory outcomes with medulloblastoma, particularly desmoplastic nodular medulloblastoma, were obtained without the requirement for radiation therapy, ablative bone marrow chemotherapy, or intrathecal methotrexate [17].

Alternatively, by harnessing immune cells, such as T cells, Natural Killer cells, and Dendritic Cells, immunotherapy aims to enhance the body's natural defenses against tumors. This innovative strategy offers hope for more effective and targeted treatments, especially for challenging cancers like medulloblastoma. CAR T cell therapy has shown significant promise in treating hematological malignancies like B cell leukemia, but its efficacy in solid tumors, such as MB, remains limited due to the scarcity of tumor-specific antigens. On the other hand, Natural Killer cells offer a safer alternative, as they do not require specific tumor-antigen recognition and exhibit fewer severe adverse effects. Dendritic cells, known for their antigen-presenting capabilities, have been employed to activate T cells ex vivo, showing potential in increasing survival rates. However, their clinical efficacy in MB remains unclear [18,19].

Complications

Many of these patients present hydrocephalus as a complication after surgery. A study developed a model in 105 patients of multivariate logistic regression and a nomogram to evaluate and predict which of these patients undergoing surgery may develop hydrocephalus, especially when the patient has not undergone ventriculoperitoneal shunt surgery. It was found that in another study of 160 patients operated on for posterior fossa tumor, third ventriculostomy is used as a routine postoperative method for endoscopic treatment of hydrocephalus associated with posterior fossa tumor [20,21].

A systematic review found that risk factors for the development of this complication were brainstem invasion, fourth ventricle invasion, cerebellar peduncle invasion, medulloblastoma diagnosis greater than 50 mm, and the use of imaging during resection decreased the appearance of this complication [22].

It is also important that survivors of medulloblastoma can develop a second neoplasm both inside and outside the central nervous system. In a 10-year follow-up study in 1114 patients after treatment with craniospinal irradiation, it was found that 58% of the second neoplasms were malignant. The highest grade glioma was the second most common malignant neoplasm (45%), and meningioma was the second most common benign tumor (67%). Forty percent of the second neoplasms were outside the central nervous system, and of these, 74% were malignant, with thyroid tumors (45%) and bone and soft tissue tumors (35%) being the most common [23].

Differential Diagnosis

Other brain cancers (ependymoma, glial tumor, atypical teratoid rhabdoid tumor) and other causes of cerebellar abnormalities (hemorrhages, infectious or cystic lesions) are included in the differential diagnosis. The vermis is where MB begins, and the cerebellar hemispheres account for 20% of MB. The dorsal brainstem can give rise to WNT-activated MB. Histologically, MB is defined by small, spherical cells that stain blue when exposed to hematoxylin; these cells can show as anything from big, anaplastic cell features in tumors to tumors with significant nodularity [15].

TEACHING POINT

Medulloblastoma is the most common malignant brain tumor in children, while MRI provides detailed images of the tumor's structure and location, CT offers complementary information on tumor characteristics, both techniques findings typically include an extra-axial mass in the cerebellopontine angle cistern with intracanalicular extension. However, histopathological analysis following biopsy or surgical resection remains essential for definitive diagnosis.

Authors' contributions

Diana Marcela Perea Rojas, Luis Enrique Perea Vásquez and Indiana Luz Rojas Torres: study conception and design, data collection, manuscript writing.

Carlos Mario Perea Molinares and Christian David Seni Hernández: data collection, data analysis and interpretation, manuscript writing.

HUMAN AND ANIMAL RIGHTS

No experiment on human or animal subjects was made.

QUESTIONS

Question 1: Which of the following statements accurately describes a characteristic of medulloblastoma?

A) They typically present with a gradual onset of symptoms over several years.

B) Histopathological analysis is not necessary for definitive diagnosis.

C) Large cell anaplastic medulloblastoma is characterized by small, round, blue cells favoring rosette formation.

D) Posterior fossa space-occupying lesions are typically located in the midline of the brainstem.

E) Medulloblastomas are categorized as grade III tumors according to the World Health Organization (WHO) classification. (applies)

Explanation:

A) Medulloblastomas typically present with a relatively rapid onset of symptoms over weeks to months, reflecting their aggressive nature. [Clinical & imaging findings, para 2]

B) Histopathological analysis following biopsy or surgical resection is essential for definitive diagnosis of medulloblastoma. [Teaching point]

C) Large cell anaplastic medulloblastoma is characterized by enlarged, pleomorphic nuclei with conspicuous nucleoli, not small, round, blue cells favoring rosette formation. [Classification, para 1]

D) Posterior fossa space-occupying lesions associated with medulloblastoma are typically located in the cerebellum, not the midline of the brainstem. [Clinical & imaging findings, para 2]

E) Medulloblastomas are categorized as grade IV tumors according to the World Health Organization (WHO) classification, indicating their aggressive growth both locally and potentially throughout the subarachnoid area. [Physiopathology, para 2]

Question 2: Which imaging modality is considered the most commonly used for the diagnosis and characterization of medulloblastoma?

A) Positron Emission Tomography (PET)

B) Single Photon Emission Computed Tomography (SPECT)

C) Ultrasound

D) Magnetic Resonance Imaging (MRI) (applies)

E) Computed Tomography (CT)

Explanation:

A) PET imaging is not typically used for the diagnosis and characterization of medulloblastoma.

B) SPECT imaging is not typically used for the diagnosis and characterization of medulloblastoma.

C) Ultrasound is not typically used for the diagnosis and characterization of medulloblastoma.

D) MRI is considered the most commonly used imaging modality for the diagnosis and characterization of medulloblastoma, providing detailed high-resolution images of the tumor's structure and location. [Clinical & imaging findings, para 2]

E) While CT can provide complementary information, MRI is the preferred imaging modality for medulloblastoma diagnosis due to its ability to provide detailed images of the tumor's structure and location.

Question 3: Which subtype of medulloblastoma is characterized by mutations in genes that regulate the Sonic Hedgehog (SHH) pathway?

A) Group 3 subtype

B) Group 4 subtype

C) WNT subtype

D) Large cell anaplastic MB

E) SHH subtype (applies)

Explanation:

A) Group 3 subtype of medulloblastoma is characterized by a high rate of genetic mutations and amplification of the MYC gene, not mutations in the SHH pathway.

B) Group 4 subtype of medulloblastoma involves dysregulation of multiple signaling pathways, including Sonic Hedgehog and Wnt pathways, but is not specifically characterized by mutations in the SHH pathway.

C) WNT subtype of medulloblastoma is characterized by mutations in genes that regulate the Wnt signaling pathway, not the SHH pathway.

D) Large cell anaplastic MB is defined by varying quantities of cytoplasm and enlarged, pleomorphic nuclei with conspicuous nucleoli, not mutations in the SHH pathway.

E) The SHH subtype of medulloblastoma is characterized by mutations in genes that regulate the Sonic Hedgehog pathway, such as PTCH1, SMO, and SUFU genes. [Molecular genetics, para 2]

Question 4: What is a distinguishing feature of desmoplastic nodular medulloblastoma?

A) Presence of rosette formation in tumor cells

B) Dense intercellular network encircling pale nodular regions (applies)

C) Enlarged, pleomorphic nuclei with conspicuous nucleoli

D) Sheets of poorly differentiated cells organized in parallel rows

E) Hyperchromatic cells with neurocytic appearance on a fibrillar background

Explanation:

A) Rosette formation is a characteristic of classical medulloblastoma, not desmoplastic nodular medulloblastoma.

B) Desmoplastic nodular medulloblastoma is characterized by a dense intercellular network encircling pale nodular regions, known as desmoplasia. [Classification, para 1]

C) Enlarged, pleomorphic nuclei with conspicuous nucleoli are characteristic of large cell anaplastic medulloblastoma, not desmoplastic nodular medulloblastoma.

D) Sheets of poorly differentiated cells organized in parallel rows are characteristic of classical medulloblastoma, not desmoplastic nodular medulloblastoma.

E) Hyperchromatic cells with neurocytic appearance on a fibrillar background are characteristic of MB with extensive nodularity, not desmoplastic nodular medulloblastoma.

Question 5: What is a common symptom associated with medulloblastoma in children?

A) Chronic cough

B) Vision changes

C) Peripheral neuropathy

D) Intermittent joint pain

E) Headaches and vomiting (applies)

Explanation:

A) Chronic cough is not typically associated with medulloblastoma.

B) Vision changes may occur if the tumor compresses or affects the optic nerves, but it is not a common presenting symptom.

C) Peripheral neuropathy is not typically associated with medulloblastoma.

D) Intermittent joint pain is not typically associated with medulloblastoma.

E) Headaches and vomiting are common symptoms associated with medulloblastoma, reflecting increased intracranial pressure from hydrocephalus secondary to tumor obstruction. [Clinical & imaging findings, para 2]

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FIGURES

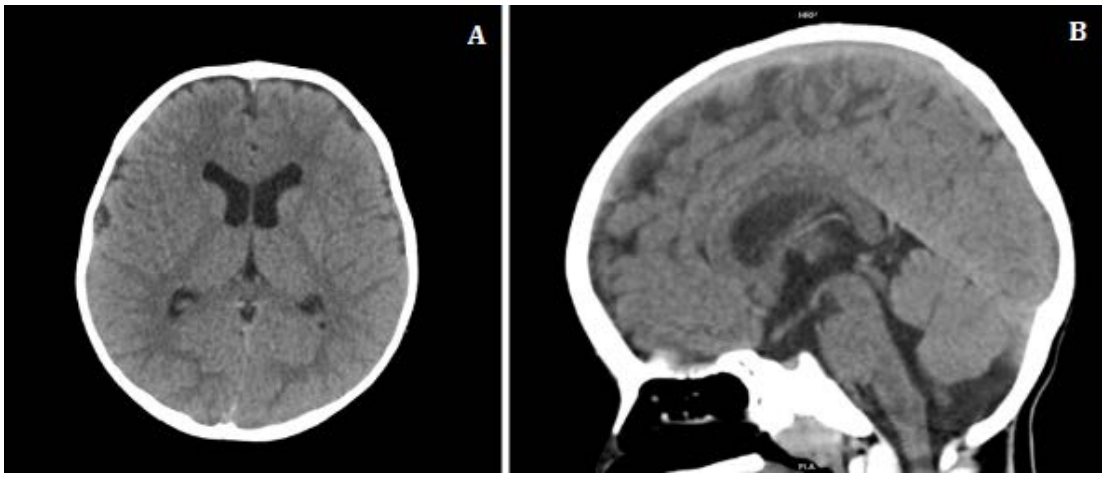

Figure 1: 2-year-old male with convulsions. FINDINGS: Axial (A) and Sagital (B) Cranial Computed Tomography Scan with that shows adequate differentiation between gray and white substance, no supratentorial parenchymal alterations defined, ventricular system of normal morphology, central middle line, no extra-axial collections defined, normal-looking posterior cavity. TECHNIQUE: Multidetector tomograph simple axial acquisitions from the base of the skull to the vertex and subsequent multiplanar reconstructions. **Source:** Patient's medical chart, 2022.

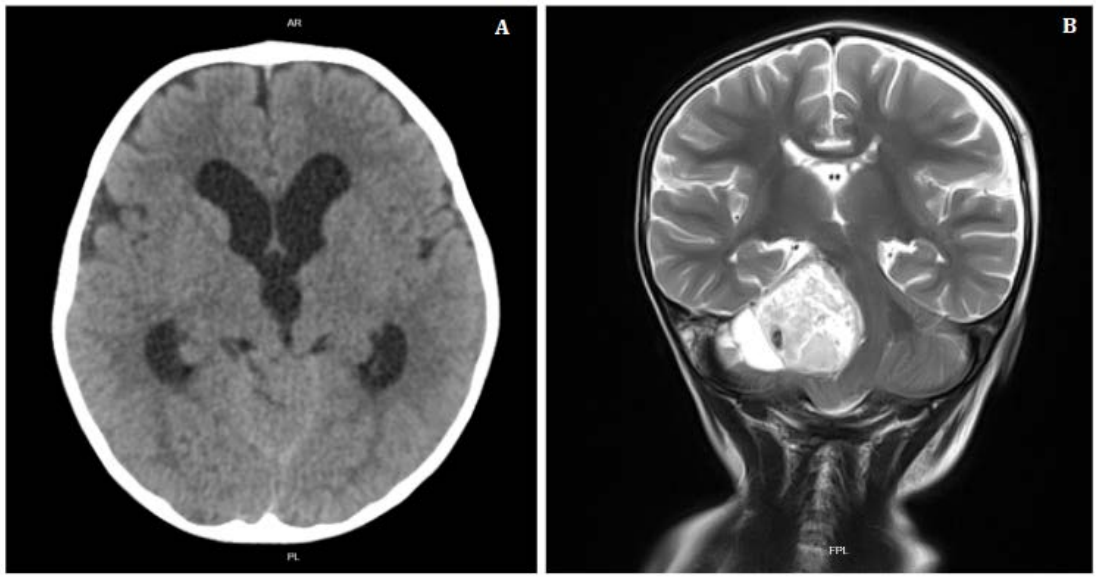

Figure 2: 2-year-old male with medulloblastoma. FINDINGS: Axial (A) and Coronal (B) Brain MRI with tractography (A) and Nuclear MRI (B) that shows an extra-axial mass, well-defined in the cerebellopontine angle cistern, hypointense on T1, hyperintense and heterogeneous on T2 with multiple hypointense foci in the susceptibility sequence, and with an area of intrinsic hyperintensity on T1 towards the periphery of the lesion due to bleeding at different stages of evolution. It occupies and expands the internal auditory canal, presents heterogeneous enhancement, and exerts marked compressive effect on the pons and the right cerebellar hemisphere with almost total obliteration of the 4th ventricle. It measures 40.7 x 56.9 x 33 mm. TECNHIQUE: 1.5T equipment. Radiofrequency pulse sequences with techniques including SE, TSE, FE, or IR. These images evaluate the longitudinal and transverse relaxation times of tissues in a multiplanar manner. **Source:** Patient's medical chart, 2023.

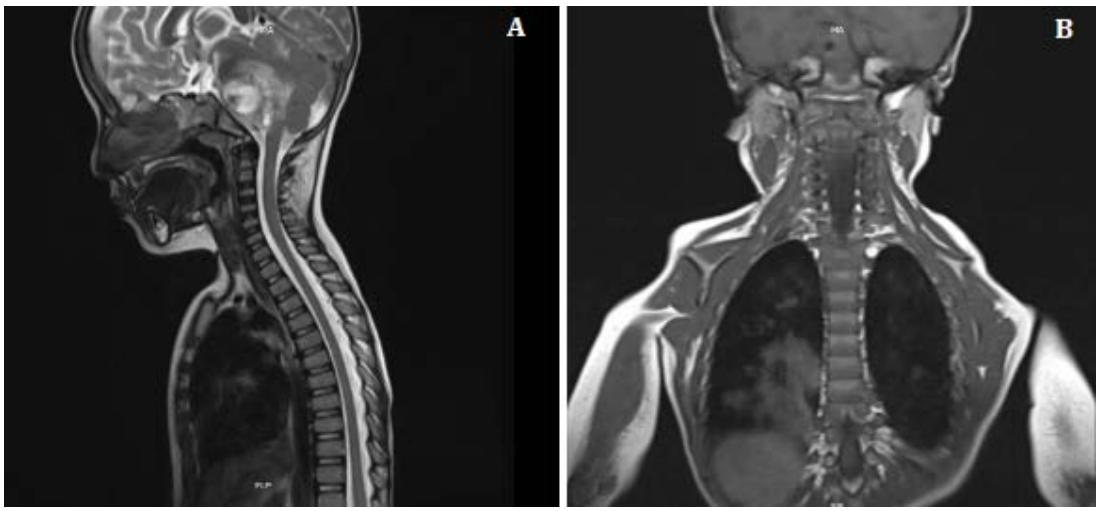

Figure 3: 2-year-old male with medulloblastoma. FINDINGS: Axial (A) and coronal (B) post-surgical contrast-enhaced cervical spine MRI that shows inespecific soft tissue edema in the nuchal region and right side of the neck, with fluid collection in the soft tissues adjacent to the surgical site. Partially visualized mass with cystic and solid component on the right side of the peritroncal cisterns. TECHNIQUE: 1.5T equipment. Radiofrequency pulse sequences with techniques including SE, TSE, FE, or IR. **Source:** Patient's medical chart, 2023.

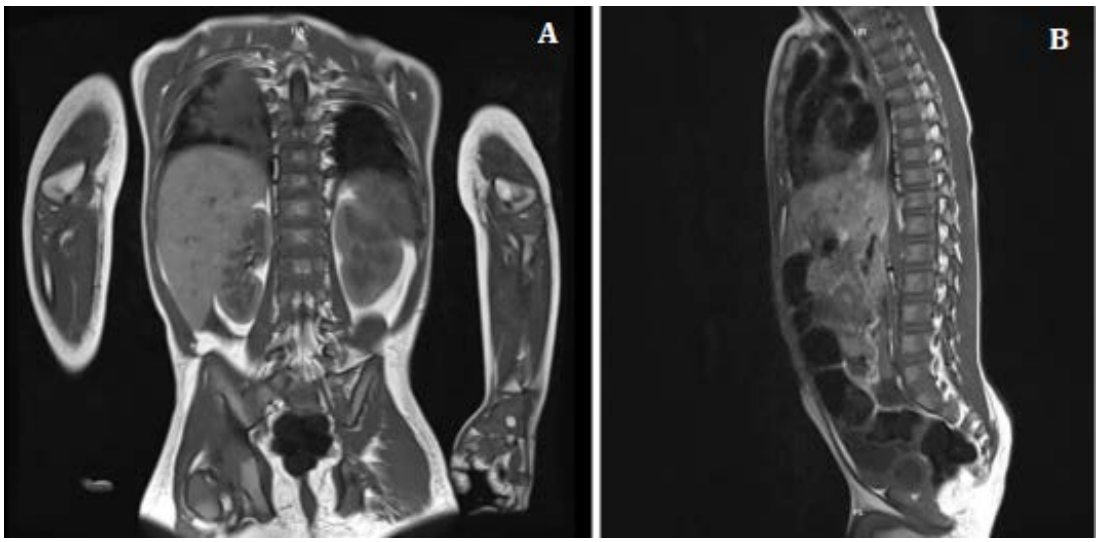

Figure 4: 2-year-old male with medulloblastoma. FINDINGS: Coronal (A) and sagittal (B) thoracic spine MRI with gadolinium that shows that the signal intensity of the bone marrow is normal. The vertebral bodies have normal height, alignment, and configuration. The intervertebral discs have normal height. The posterior elements appear usual. The spinal canal has a normal caliber, and no protrusions into its interior are evident. The neural foramina have a normal caliber. The spinal cord has normal thickness and signal intensity. There are no areas of enhancement after the administration of gadolinium. The paravertebral soft tissues show no abnormalities. There is no mass in the spinal canal, and no areas of pathological dural or leptomeningeal enhancement are visualized. TECHNIQUE: 1.5T equipment. Radiofrequency pulse sequences with techniques including SE, TSE, FE, or IR. **Source:** Patient's medical chart, 2023.

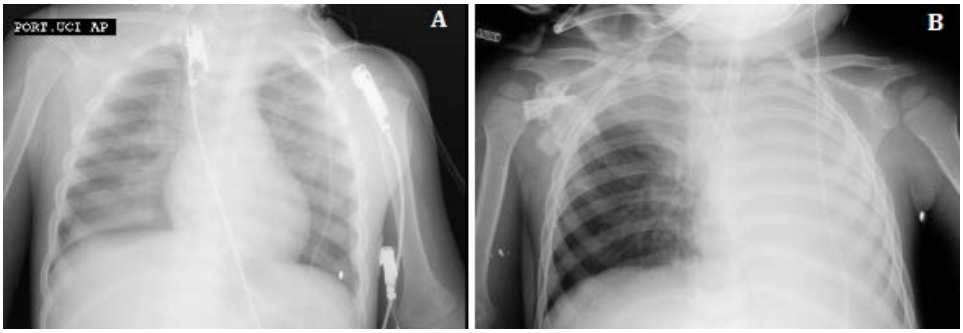

Figure 5: 2-year-old male with acute respiratory infection. FINDINGS: Chest radriography that shows Cardiophrenic silhouette and pulmonary vascularity preserved. Mediastinum without alteration. Thickening of the peribronchovascular axial interstitium. Multilobar consolidation (A). Right pneumothorax (B). Endotracheal tube with distal end at the level of T3. **Source:** Patient's medical chart, 2023.

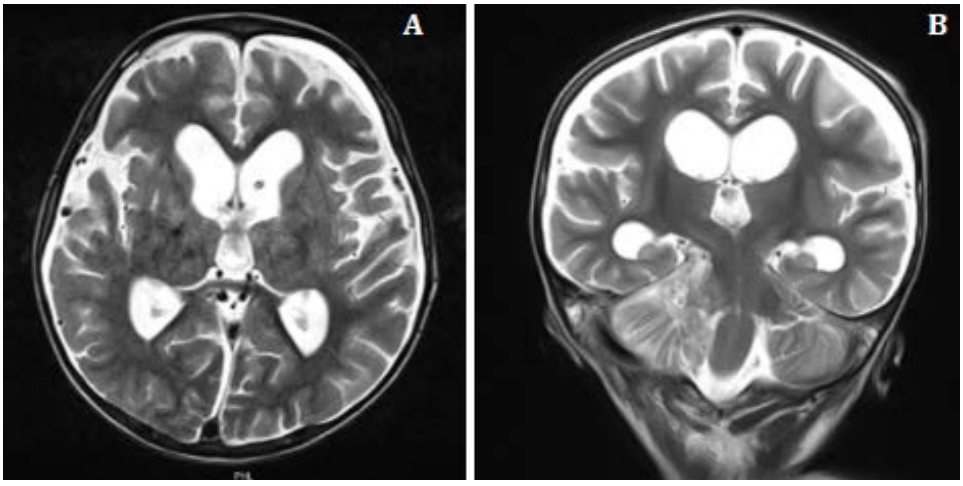

Figure 6: 2-year-old male with medulloblastoma. FINDINGS: Post-surgical contrast-enhaced brain MRI axial (A) and coronal (B) that shows changes consistent with a right occipital craniectomy with an area of encephalomalacia and gliosis at the lower margin of the right cerebellar hemisphere; a lesion with extra-axial characteristics occupying the prepontine, right pontocerebellar, and bulbar cisterns, which is hypointense on T1, heterogeneous on T2, and shows intense enhancement with contrast administration. It presents with an intracanalicular extension component and exerts a moderate mass effect on the middle cerebellar peduncle. Meningeal enhancement is observed supra and infratentorially. Extra-axial collections with fluid signal in subdural morphology are seen in the frontoparietal convexities, without apparent compressive effect. The ventricular system is moderately enlarged in the midline; there are no signs of transependymal migration of cerebrospinal fluid. TECHNIQUE: Study with T1, T2, diffusion, and FLAIR sequences, as well as contrast-enhanced T1 sequences in various plane. **Source:** Patient's medical chart, 2023.

SUMMARY TABLE

Etiology	Unknown.
Incidence	Medulloblastoma accounts for approximately 20% of primary nervous system tumors in children.
Gender ratio	There is no significant gender predilection, it occurs with roughly equal frecuency in both males and females
Age predilection	Primarly affects children and adolescents.
Risk Factors	Associated with Gorlin syndrome, ataxia teleangiectasia, Li-Fraumeni syndrome, Turcot syndrome and the presence of extra chromosomes in the 6-12 and 4-5 groups.
Treatment	Surgical resection, radiotherapy, and chemotherapy.
Prognosis	The 5-year disease-free survival probability is around 60-70% for high risk cases, and 80% for average-risk cases.
Imaging findings	Extra axial mass in the cerebellopointine angle cistern with intracanalicular extension.

KEYWORDS

Neoplasms; Medulloblastoma; Cerebellum; Tomography; Diagnostic Imaging

ABBREVIATIONS

CT = Computed Tomography
MRI = Magnetic Resonance Imaging
IAC = Internal Auditory Canal
ICU = Intensive Care Unit
MB = Medulloblastoma
SHH = Sonic Hedgehog
WNT = Wingless Activated
WHO = World's Health Organization
PNET = Primitive Neuroectodermal Tumor

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