

## Pediatric Meningiomatosis: A Rare Radiological Case Report

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### ABSTRACT

Meningiomatosis is defined as the presence of multiple synchronous meningiomas arising at separate intracranial or spinal locations in the absence of prior cranial irradiation. While meningiomas represent one of the most common extra-axial intracranial tumors in adults, they are rare in children, accounting for less than 3% of pediatric central nervous system tumors. Pediatric meningiomatosis is exceedingly uncommon, with only sporadic cases reported, and its true incidence remains unknown.

Diagnosis relies primarily on neuroimaging, with magnetic resonance imaging (MRI) serving as the modality of choice. Typical MRI features include multiple well-defined extra-axial masses that are isointense to gray matter on T1-weighted images, iso- to hyperintense on T2-weighted and fluid-attenuated inversion recovery sequences, and demonstrate strong, usually homogeneous contrast enhancement with broad-based dural attachment and dural tail signs. Diffusion-weighted imaging generally shows no significant diffusion restriction, supporting a benign etiology.

We report a rare case of pediatric meningiomatosis in which MRI revealed multiple characteristic extra-axial enhancing lesions, subsequently confirmed by histopathological examination as World Health Organization grade I meningioma.

**KEYWORDS:** Pediatric meningiomatosis; Magnetic resonance imaging.

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### INTRODUCTION

Meningiomas are extra-axial neoplasms originating from arachnoid cap cells and constitute approximately 20–30% of all primary intracranial tumors in adults [1,2,5]. In the pediatric population, however, meningiomas are rare, accounting for less than 3% of childhood central nervous system tumors, and they often demonstrate imaging and biological characteristics distinct from those seen in adults [1,2,10]. Pediatric meningiomas are more frequently associated with atypical locations, higher histological grades, and genetic conditions such as neurofibromatosis type 2 [2,3,11].

Meningiomatosis refers to the presence of multiple meningiomas occurring synchronously at different intracranial or spinal sites in the absence of prior cranial irradiation [1,5]. While multiple meningiomas are reported in up to 10% of adult cases, meningiomatosis in children is exceedingly rare, with only a limited number of cases described in the literature [1,3]. From a radiological perspective, the occurrence of multiple extra-axial lesions in pediatric patients presents a diagnostic challenge and requires careful differentiation from metastatic disease, lymphoma, inflammatory meningeal processes, and other neurocutaneous disorders [3,4,8,14].

Magnetic resonance imaging (MRI) is the modality of choice for the evaluation of meningiomas due to its superior soft-tissue contrast and multiplanar capabilities [1,6]. Typical MRI features include well-circumscribed extra-axial masses that are isointense to gray matter on T1-weighted images, iso- to hyperintense on T2-weighted images, and demonstrate strong, usually homogeneous enhancement following gadolinium administration, frequently accompanied by a dural tail sign [1,3,4]. MRI is particularly valuable in meningiomatosis for accurately depicting the number, distribution, and extent of lesions, as well as associated findings such as peritumoral edema, mass effect, and involvement of adjacent bone or neurovascular structures [2]. In pediatric meningiomatosis, imaging characteristics may be variable, and lesions may show heterogeneous signal intensity or enhancement patterns, making diagnosis more complex. Advanced MRI techniques, including diffusion-weighted imaging and perfusion imaging, can provide additional information regarding tumor cellularity and vascularity, which may assist in characterization and management planning [2,4,5,12].

Given the rarity of meningiomatosis in children and the limited radiological descriptions available in standard literature, reporting such cases is important to expand the understanding of its MRI spectrum and to support accurate diagnosis. We present a rare case of pediatric meningiomatosis, with emphasis on the MRI findings and key radiological considerations.

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## CASE REPORT

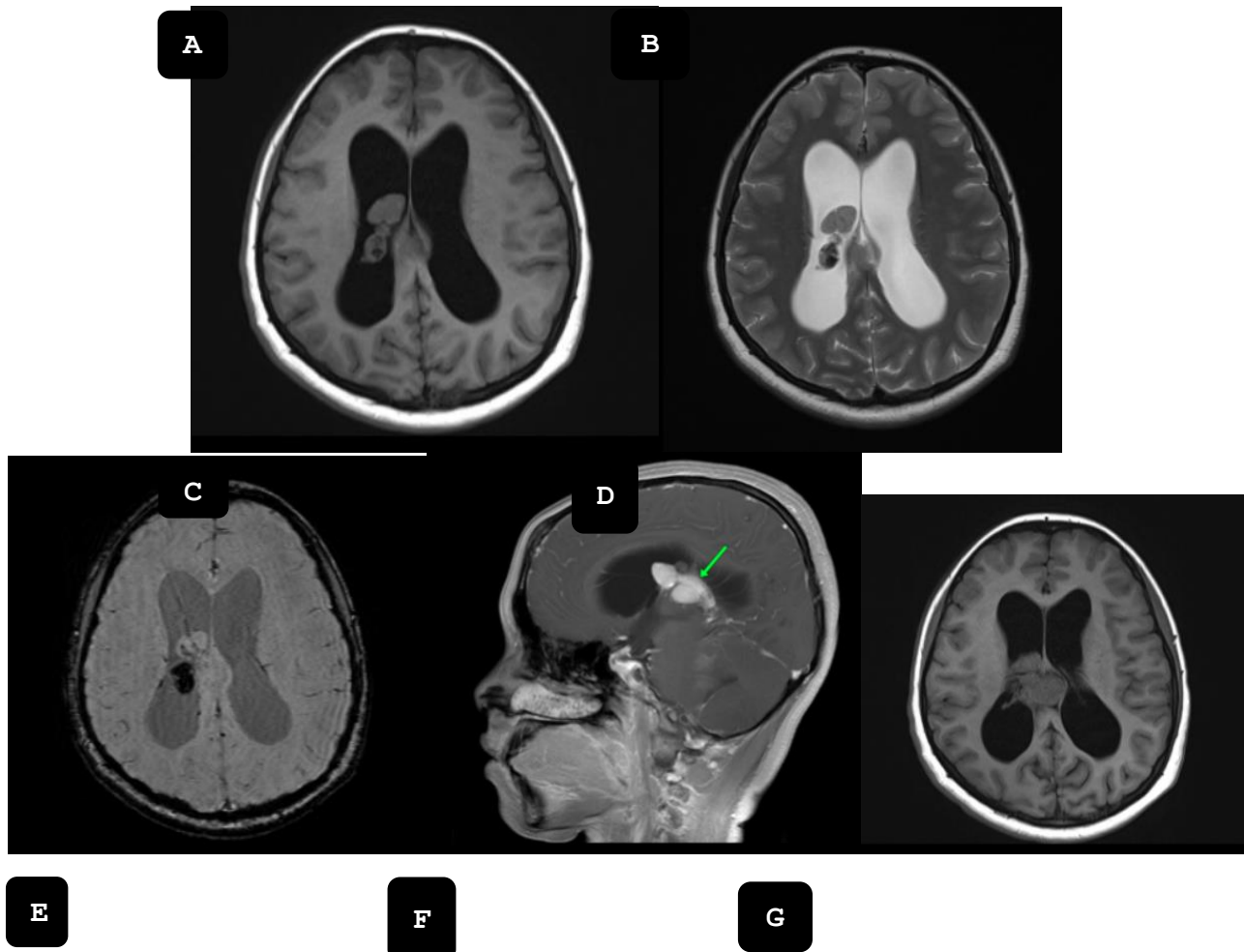
### Case Presentation

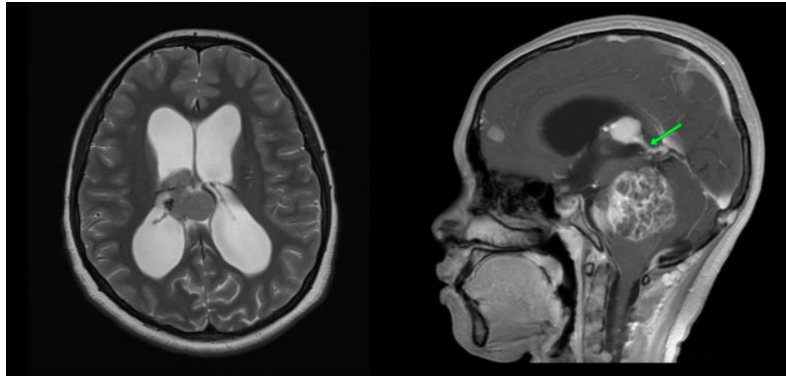
A 16-year-old female presented with bilateral strabismus, impaired voluntary eye movements, accompanied by hearing impairment and balance disturbances for approximately 10 years, with worsening of symptoms during the month prior to hospital admission. Patient had neither experienced seizures nor reported any weakness in her limbs. Throughout this period, the patient remained conscious, could understand speech, and could follow commands. Physical examination, including assessment of vital signs, presented normal results. The neurological examination was largely unremarkable, except for a slight weakness in the motor function of her left extremity. Her laboratory tests showed an elevated leukocyte count.

### Diagnostic Evaluation

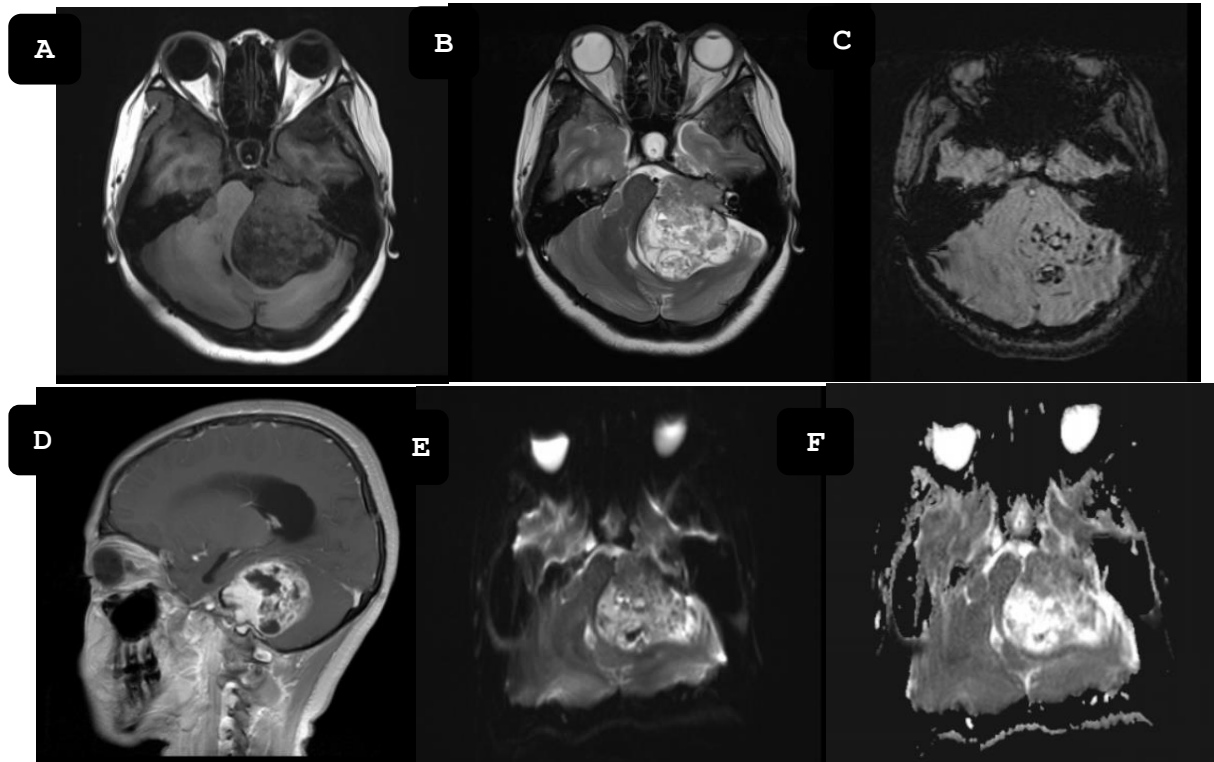
Brain magnetic resonance imaging was obtained using standard sequences, including T1-weighted, T2-weighted, fluid-attenuated inversion recovery (FLAIR), diffusion-weighted imaging, and post-contrast T1-weighted imaging [6,18]. MRI demonstrated multiple well-circumscribed extra-axial masses located at different intracranial sites. On T1-weighted images, the lesions were predominantly isointense to gray matter. T2-weighted and FLAIR images showed iso- to mildly hyperintense signal intensity with surrounding vasogenic edema [1,3]. Following gadolinium administration, all lesions exhibited intense, relatively homogeneous enhancement with associated dural tail signs, a classic imaging feature of meningiomas [1,4,6]. No significant diffusion restriction was observed, consistent with typical benign meningioma characteristics [3,15]. Based on the imaging findings, a radiological diagnosis of meningiomatosis was considered [1,5].

In this case, Magnetic resonance imaging (MRI) examination of the head revealed multiple masses with a “dural tail” sign filling the right lateral ventricle and third ventricle with associated dilatation of the lateral ventricles and the third ventricle. (Fig. 1). A cerebellopontine angle mass causing uncus herniation and bilateral compression of the oculomotor, abducens, and vestibulocochlear nerves. No restricted areas were observed in diffusion weighted imaging with an apparent diffusion coefficient value of  $1.2 \times 10^{-3} \text{ mm}^2/\text{s}$  and blooming artifacts in susceptibility weighted imaging. (Fig. 2).





**Fig. 1.** MRI showed hypointense mass in T1WI (A and E), isointense mass in T2WI (B and F), blooming artifacts in SWI (C), T1+ Contrast showed enhanced mass, *dural tail sign* (green arrow) (D and G).



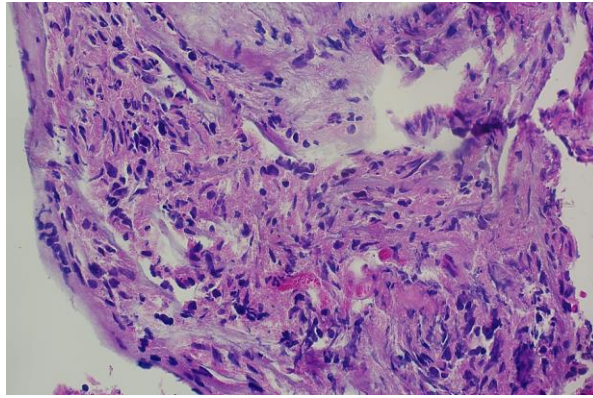
**Fig. 2.** MRI showed hypointense cerebellopontine angle mass in T1WI (A), isointense mass in T2WI (B), blooming artifacts in SWI (C), T1+ Contrast showed enhanced mass (D), No restricted areas diffusion weighted imaging (E and F).

### Histopathological Findings

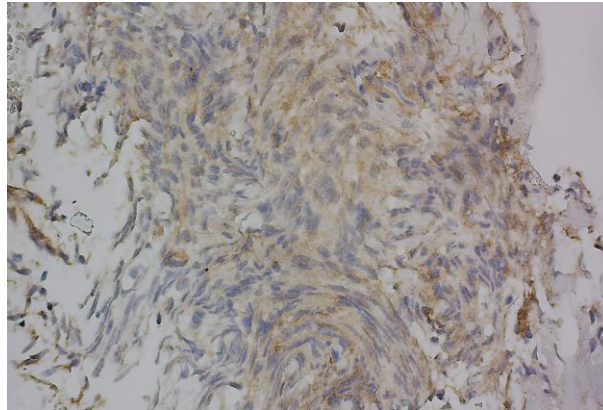
Surgical biopsy and histopathological examination were performed to confirm the radiological diagnosis. Gross examination revealed firm, well-circumscribed dural-based tumor tissue. Microscopic analysis demonstrated features consistent with meningioma, including whorled arrangements of meningothelial cells and oval nuclei with fine chromatin. Psammoma bodies were focally identified.

There was no evidence of significant nuclear atypia, increased mitotic activity, necrosis, or brain invasion. Immunohistochemical staining showed tumor cell positivity for epithelial membrane antigen (EMA) and vimentin, supporting the diagnosis of meningioma. The Ki-67 (MIB-1) labeling index was low, consistent with a World Health Organization (WHO) grade I meningioma [1,4]. These pathological findings correlated well with the benign imaging characteristics observed on MRI.

In this case, a histopathological examination of the tissue biopsy confirmed the diagnosis of meningiomas, was composed of round to oval cells with focal spindle-shaped hyperplastic cells, displaying dense cellularity and monomorphic features. The stroma showed lymphocytic inflammatory infiltration with vascular dilatation (Fig. 3). Immunohistochemical staining showed tumor cell positivity for epithelial membrane antigen (EMA), supporting the diagnosis of meningioma (Fig. 4).



**Fig. 3. Tumor masses composed of hyperplastic round to oval cells, confirmed the diagnosis of meningiomas**



**Fig. 4. (EMA) immunohistochemical staining, 400x**

## DISCUSSION

Meningiomatosis in children is an exceptionally rare condition, and its true incidence remains unknown due to the limited number of reported cases [1,2,10]. Pediatric meningiomas, including those occurring as part of meningiomatosis, are more likely to present at atypical locations and may show variable imaging characteristics compared with adult cases [2,11,16].

MRI is essential for the diagnosis of meningiomatosis, allowing accurate assessment of lesion number, size, location, and relationship to adjacent structures [1,3,6]. Typical imaging features, such as extra-axial location, broad-based dural attachment, and avid contrast enhancement with dural tail signs, are key to suggesting the diagnosis [1,4]. However, in children, the presence of multiple enhancing intracranial lesions necessitates careful consideration of alternative diagnoses, including metastatic disease, lymphoma, inflammatory conditions, and meningeal infections [3,8,14].

Early and accurate radiological diagnosis of pediatric meningiomatosis is important for guiding clinical management, surgical planning, and follow-up strategies [12,15]. Reporting rare cases contributes to a broader understanding of the imaging spectrum and assists radiologists in recognizing this uncommon entity [1,3].

## CONCLUSION

Pediatric meningiomatosis is an exceptionally rare clinical entity, and its diagnosis requires a high index of suspicion, particularly when evaluating multiple intracranial extra-axial lesions in children. This case highlights the pivotal role of magnetic resonance imaging as the best diagnostic modality, enabling accurate identification of lesion multiplicity, distribution, and characteristic imaging features such as homogeneous enhancement, broad-based dural attachment, and dural tail signs.

Comprehensive MRI assessment, combined with histopathological confirmation, is essential for establishing a definitive diagnosis and excluding other potential causes of multifocal intracranial disease, including metastatic, inflammatory, and infectious conditions. Early and accurate recognition of pediatric meningiomatosis has important implications for clinical decision-making, surgical planning, and long-term follow-up strategies. Increased awareness of this rare entity among radiologists and clinicians may facilitate timely diagnosis, appropriate management, and improved understanding of its imaging spectrum in the pediatric population.

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