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Case Report

Adult tentorial medulloblastoma mimicking meningioma: A case report and systematic review

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ABSTRACT

Background: Tentorial medulloblastomas in adults are exceedingly rare and may clinically and radiologically mimic meningiomas. This case report, with a systematic review, aims to outline the clinical, radiological, pathological, and management strategies for adult tentorial medulloblastoma.

Case Description: A 37-year-old male patient presented with headaches, vertigo, and vomiting. Imaging investigations revealed a tentorial extra-axial mass, initially considered a meningioma. The patient subsequently underwent surgical resection followed by chemoradiation. Histopathological examination ultimately identified the mass as an eccentrically located adult medulloblastoma. We conducted a systematic review of the literature, analyzing four studies that reported similar cases. This analysis included clinical and demographic information, diagnosis through imaging and histopathology, treatment methods, and outcomes for seven cases, including our own.

Conclusion: Adult tentorial medulloblastomas are extremely rare tumors that may mimic meningiomas, posing significant clinical challenges. Accurate diagnosis necessitates advanced imaging techniques and histopathological confirmation. The primary treatment strategy involves maximal surgical resection, supplemented by chemoradiotherapy.

Keywords: Adult, Histopathology, Medulloblastoma, Meningioma, Posterior fossa, Tentorial

INTRODUCTION

Medulloblastomas are classified as the World Health Organization (WHO) grade IV tumors, recognized as highly aggressive embryonal neoplasms with a strong preference for the cerebellum.[6,9] These tumors are primarily diagnosed in pediatric patients, making up approximately 15% of all central nervous system (CNS) tumors in children and over 40% of tumors found in the posterior fossa.[26] While medulloblastomas have traditionally been viewed as malignancies primarily affecting children, cases in adults, though rare, do occur, representing <1% of all CNS neoplasms.[1,26] In pediatric cases, medulloblastomas typically develop in the midline, involving the cerebellar vermis, and are often linked with symptoms of increased intracranial pressure, such as headaches, vomiting, and ataxia. In contrast, in adults,

medulloblastomas are more likely to occur in the lateral cerebellar hemispheres and often manifest with more insidious or atypical symptoms.[10] The observed differences in anatomical localization and clinical presentation correspond with specific molecular and genetic variations that affect tumor phenotype, prognosis, and treatment responses.[10,17]

Understanding adult medulloblastomas is complicated by rare variants, especially in extra-axial locations such as the tentorium cerebelli, which are primarily associated with meningiomas. Tentorial medulloblastomas are extremely rare, and their radiological features may closely resemble those of meningiomas, making accurate diagnosis more challenging.[14]

This report outlines an unusual case of tentorial medulloblastoma in an adult patient, thus contributing to the limited literature on this atypical manifestation. Through a systematic review of previous cases, our objective is to clarify the clinical, radiological, and pathological features associated with this case in detail, ultimately addressing the challenges and complexities related to the diagnosis and treatment of such rare neoplasms. In addition, this case emphasizes the necessity of employing advanced imaging modalities and histopathological assessments alongside molecular diagnostic methods in adult medulloblastomas, especially in atypical presentations.

CASE DESCRIPTION

A 37-year-old male presented to the neurology outpatient clinic 3 months ago, complaining of a persistent holocranial headache, imbalance, and fear of falling for 3 weeks. Upon clinical examination, the patient was alert and showed some imbalance while walking. Fundoscopy revealed bilateral hyperemia with blurred margins of the right optic disc.

The patient was referred to the hospital's emergency department (ED) due to complaints of a progressively worsening headache, persistent vertigo, and episodes of vomiting. Upon arrival at the ED, a neurological examination indicated no motor deficits except a broad-based gait. A head computed tomography scan revealed a large, tentorial, extra-axial hyperdense mass lesion, which exhibited signs of fourth ventricular compression, leading to supratentorial hydrocephalus [Figure 1]. Magnetic resonance imaging (MRI) findings demonstrated a right tentorial, bi-lobulated extra-axial lesion extending both infratentorially and supratentorially, characterized by diffusion restriction and substantial contrast enhancement [Figures 2 and 3]. The MR spectroscopic perfusion study revealed a notable increase in choline levels and a high choline-to-N-acetylaspartate (NAA) ratio. The lesion was well-circumscribed but was associated with perilesional parenchymal cerebellar edema involving the midline structures, with left-sided extension, resulting in considerable mass effect and partial effacement of the fourth ventricle. In addition, cerebellar tonsillar herniation was noted, measuring 14 mm, along with signs of hydrocephalus. These MR findings were consistent with a diagnosis of tentorial lesion meningioma.

The patient underwent a right-sided suboccipital craniotomy for the resection of a tentorial-based lesion. During the operation, there was a well-defined interface between the tumor and the cortex. It appeared grayish-white in color and had a firm consistency. A per-operative frozen section revealed the lesion as a blue round cell tumor, raising suspicion for neuroblastoma while making a diagnosis of meningioma unlikely. Complete resection was achieved, including the supratentorial portion that was resected from the infratentorial surgical exposure.

The microscopic examination reveals a hypercellular tumor composed of small, round blue cells with uniform, round nuclei. These cells are arranged in a trabecular pattern, separated by fibrous bands, and show numerous mitotic figures [Figure 4a]. Immunohistochemical studies indicate

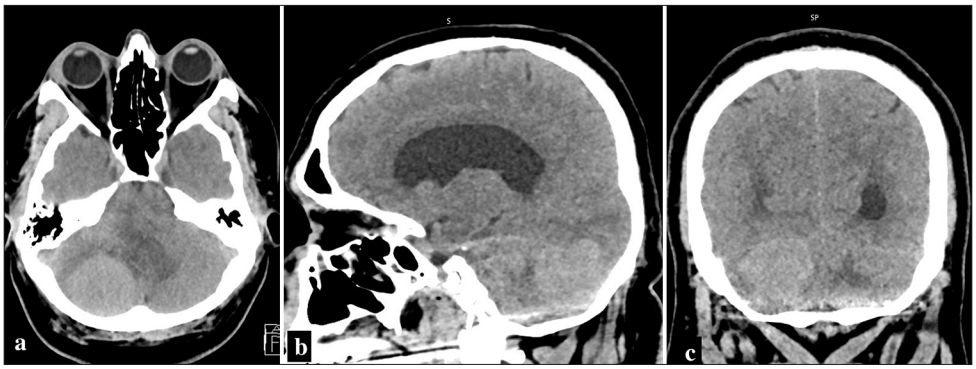

Figure 1: An unenhanced computed tomography (CT) scan of the brain shows (a) axial, (b) sagittal, and (c) coronal sections revealing a hyperdense mass lesion situated in the right posterior fossa adjacent to the tentorium, accompanied by significant perifocal edema that causes effacement of the fourth ventricle.

that the tumor cells are diffusely positive for synaptophysin [Figure 4b] and Somatostatin receptor 2a (SSTR2A). In addition, the tumor exhibits focal positivity for glial fibrillary acidic protein. Notably, the expression of INI-1 and BRG1 is retained. The tumor is negative for keratin (CK AE1/AE3) [Figure 4c], chromogranin, NKX2.2, SS18-SSX, smooth muscle markers, and vascular markers. Molecular studies further confirm the diagnosis, revealing mutations in the telomerase reverse transcriptase promoter, smoothened (SMO), and isocitrate dehydrogenase 1 (IDH1) (p.R132C). These mutations are characteristic of Sonic Hedgehog (SHH)--activated medulloblastomas, driven by aberrations in the SHH signaling pathway. The presence of triploidy and a gain of chromosome 3q, identified by chromosomal microarray, further support this molecular subtype. This classification places the tumor in the CNS WHO grade 4 category, highlighting its malignant and aggressive nature.

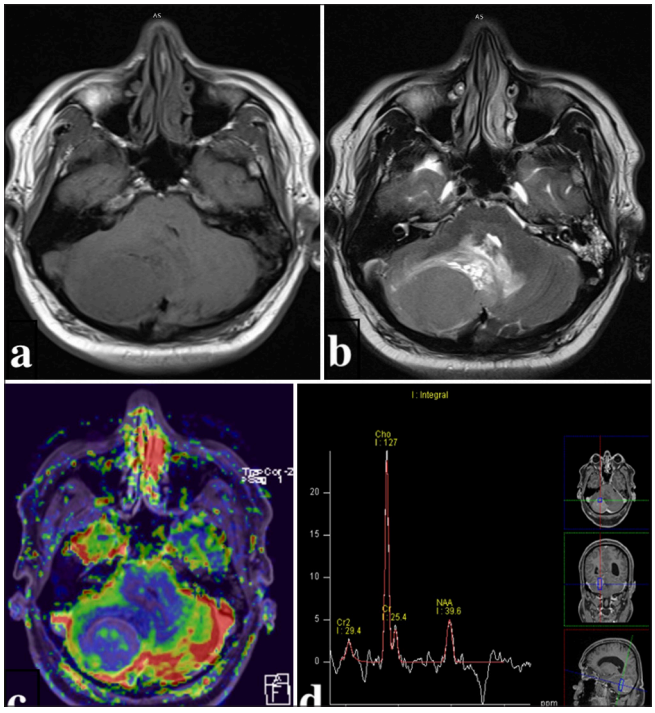

Figure 2: (a) Axial sections from magnetic resonance (MR) imaging using T1 weighted, (b) T2 weighted, and (c) perfusion cerebral blood flow scans of the brain illustrate a hypointense mass lesion on both T1 and T2-weighted imaging, along with perifocal edema, while the perfusion scan indicates no significant vascularity, while (d) shows the MR spectroscopic study revealed a notable increase in choline levels and a high choline-to-N-acetylaspartate ratio.

In light of the final diagnosis of eccentric medulloblastoma, the screening MRI of the spine and cerebrospinal fluid showed no spinal metastatic spread. The patient was discussed in our weekly neuro-oncology multidisciplinary meeting, and the consensus was to administer chemotherapy (vincristine and cisplatin), followed by radiotherapy of the craniospinal axis and tumor bed in the posterior fossa. At the 6-month follow-up, the patient showed a complete recovery without any neurological deficits.

MATERIALS AND METHODS

Literature search

This systematic review was conducted according to the Joanna Briggs Institute (JBI) methodology for systematic reviews of observational studies. The research protocol adhered to Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) 2020 guidelines to ensure comprehensive reporting.

Search strategy

An extensive search strategy was developed to identify relevant studies addressing rare adult tentorial medulloblastoma cases. The databases searched included Medline, Embase, Scopus, Web of Science, ScienceDirect,

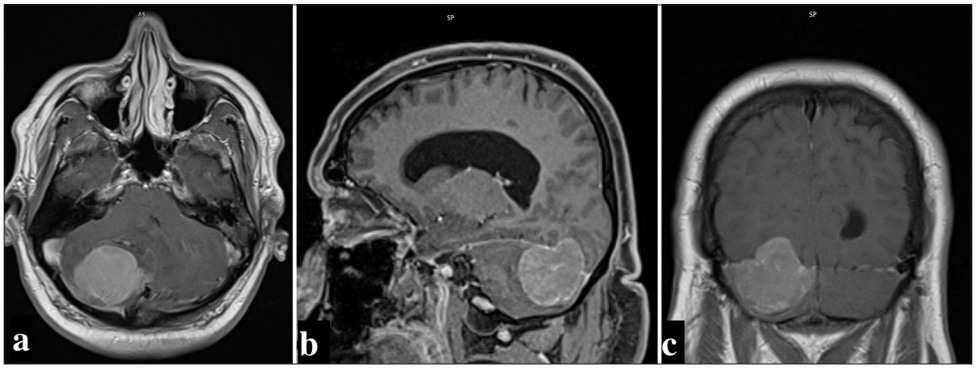

Figure 3: Gadolinium-enhanced magnetic resonance images depict (a) axial, (b) sagittal, and (c) coronal sections indicating a prominently enhancing mass lesion attached to the right tentorial cerebelli, with slight extension above the tentorium. The perifocal edema exerts a mass effect, causing displacement and effacement of the fourth ventricle.

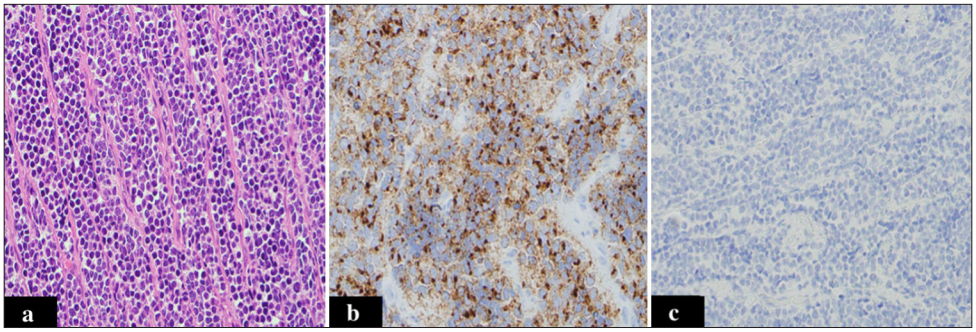

Figure 4: (a) A histological image demonstrates a hypercellular malignant tumor composed of small round cells arranged in trabecular formations and separated by fibrous bands (hematoxylin and eosin ×200); (b) Immunohistochemical staining of the tumor cells indicates perinuclear positivity for synaptophysin (×200); (c) Immunohistochemical staining reveals negativity for keratin (CK AE1/AE3) in the tumor cells (×200).

Cochrane, Clinical Key, and Access Medicine. The key terms included tentorial medulloblastoma, posterior fossa tumor, adult medulloblastoma, atypical medulloblastoma presentation, mimicking meningioma, radiological meningioma look-alikes, and meningioma differential diagnosis. The search strategy utilized a combination of Medical Subject Headings (MeSH) terms: ((tentorial medulloblastoma) OR (posterior fossa tumor) OR (adult medulloblastoma) OR (atypical medulloblastoma presentation)) AND ((mimicking meningioma) OR (meningioma misdiagnosis) OR (radiological meningioma look-alikes) OR (meningioma differential diagnosis)).

Selection and screening process

After the initial search, duplicate entries were removed using web-based reference management software (Rayyan). The remaining records were screened based on their titles and abstracts, with two independent reviewers assessing eligibility according to predefined inclusion and exclusion criteria. Studies were considered suitable for inclusion if they reported on adult patients diagnosed with medulloblastoma located in the tentorium, provided sufficient clinical and radiological data, and were published in English.

Exclusion criteria included studies that focused on pediatric populations, cases involving alternative tumor locations, such as the cerebellopontine angle, nonprimary research (e.g., reviews), and publications in languages other than English. Full-text articles of potentially eligible studies were obtained and assessed for inclusion. Any disagreements with the reviewer were resolved through consensus or consulting a third reviewer. Figure 5, the PRISMA flowchart, illustrates the systematic process used for study identification.

Data extraction

Data extraction was performed using a standardized JBI data extraction method in Excel sheets, which systematically captured information regarding study characteristics, patient demographics, clinical presentations, radiological findings, and outcomes of treatment modalities [Table 1].

Quality assessment

The quality appraisal of the studies included in this review was conducted using the JBI Critical Appraisal Checklist for Case Reports. This method ensured a comprehensive evaluation of methodological quality while minimizing potential biases identified in the reviewed literature. Due to the limited number of eligible studies and the heterogeneity in the reported outcomes, the extracted data were synthesized narratively. The findings were summarized to provide insights into the rare clinical entity of adult tentorial medulloblastoma, emphasizing diagnostic challenges, treatment strategies, and outcomes [Table 2].

RESULTS

This review included six cases of medulloblastoma that mimicked tentorial meningioma from four previously published articles, which were analyzed in full and summarized alongside our cases [Table 1]. The patients' ages ranged from 18 to 37 years (mean 28.6, standard deviation ±7.03), with a slight male predominance (4:3). Clinical presentations varied but frequently included headaches in four out of the seven cases, as well as other symptoms associated with increased intracranial pressure, such as vomiting and vertigo – two cases presented with cerebellar signs, which included gait imbalance and truncal instability ataxia. MRI findings consistently demonstrated heterogeneous contrast enhancement, with varying intensity noted among the cases. Diffusion restriction was observed in two cases, including ours. Obstructive hydrocephalus was recorded in three cases at presentation, as seen in our patient. Tonsillar herniation was observed in two cases, including ours, indicating a significant mass effect. Histopathological

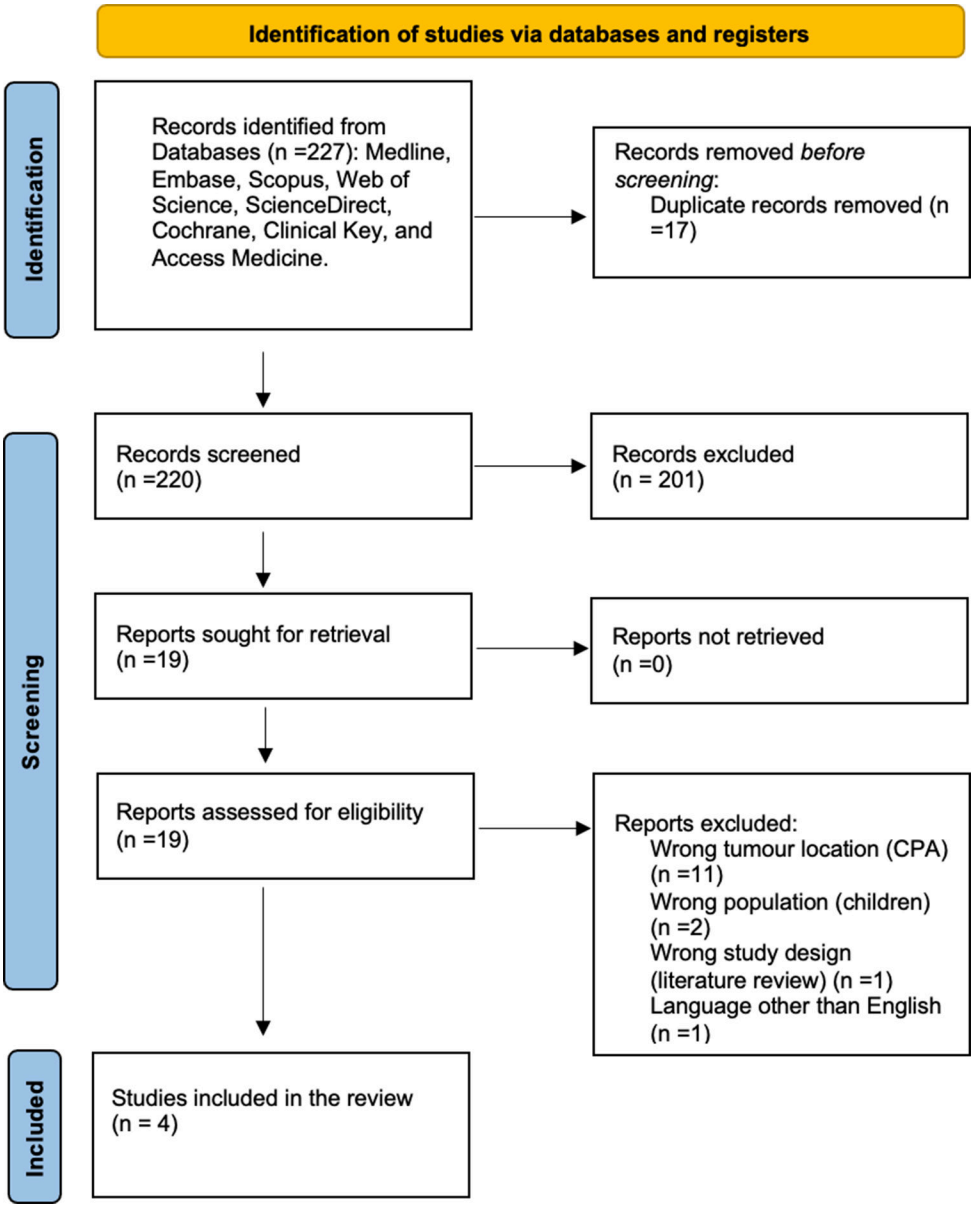

Figure 5: A flowchart based on Preferred Reporting Items for Systematic Reviews and Meta-Analyses guidelines illustrates the flow of information through various phases of a systematic review.

analysis confirmed the presence of medulloblastoma in all cases. Classification of three cases identified SHH-activated subtypes, including those found in our case. Management strategies primarily involved surgical intervention, with two patients receiving additional adjuvant chemoradiation. Our patient successfully underwent tumor resection, followed by chemoradiation and adjuvant chemotherapy.

DISCUSSION

Medulloblastomas account for 15% of CNS tumors in pediatric populations, 40% of all tumors found in the posterior fossa, and 90% of all embryonal tumors, making them one of the most prevalent forms of embryonal tumors originating from the posterior fossa.[11,14] The peak incidence of medulloblastomas typically occurs in children aged 5–7 years, with a significantly higher prevalence in males, indicated by a male-to-female ratio of 1.7:1.[26] These tumors primarily appear in the midline; however, a few rare, documented cases of extra-axial medulloblastomas have been reported, mainly located in the cerebellopontine angle and the lateral cerebellar hemisphere.[3,10,30] In adults, medulloblastomas are relatively rare, comprising only 0.4–1% of all CNS tumors in this demographic, with an annual incidence rate of about 0.6–1 case per million individuals.[1,4,13,24,25] Unlike pediatric cases, adult

Table 1: Summary of studies included for full-text analysis.

Study ID	Age\ Sex	Clinical presentation	Physical examination	Tumor size (cm)	MRI characteristics	Hydrocephalus	Histopathology	Management
Becker *et al*. 1995	34\F	N\A	N\A	3.5 × 6	• Moderate contrast enhancement, heterogeneous	Marked	N\A	N\A
	35\F	N\A	N\A	3 × 4	• Mild-to-moderate contrast enhancement	Marked	N\A	N\A
	28\M	N\A	N\A	2.5 × 3.5	• Slight contrast enhancement	No	N\A	N\A
Meshkini *et al*. 2014	19\F	Headache and vomiting and mild truncal ataxia	N\A	N\A	• An extra-axial on the left side of the tentorium. • Infra-and supratentorial necrotic areas. • Severe vasogenic at the cerebellum and tonsillar herniation	N\A	Classic medulloblastoma	N\A
Doan *et al*. 2018	29\M	Syncopal episode and headache	Unremarkable	N\A	• Enhanced extra-axial midline tentorial mass with the tentorial dural-tail sign	N\A	Desmoplastic medulloblastoma (SHH subgroup)	Surgery, then radiation and adjuvant chemotherapies (vincristine, cisplatin, and Cyclophosphamide)
Singh *et al*. 2021	18\M	Dull, aching left-sided headache and gait imbalance	Cerebellar signs were positive	N\A	• Iso to hypointense on T1W, hypointense on T2W and intermediate on T2-FLAIR • Significant restriction on diffusion-weighted imaging • Mild and heterogeneous contrast enhancement	N\A	Medulloblastoma, SHH-activated, and TP-53 wild type	Surgery then chemoradiation with etoposide and cisplatin, and CSI
Our report	37\M	Headache, vomiting, and persistent vertigo	Unremarkable	2.8 × 3.5 × 3	• Right tentorial bi-lobulated extra-axial lesion extending significantly infratentorial and slightly supratentorial • Showing diffusion restriction and significant contrast enhancement	Supratentorial hydrocephalus	Medulloblastoma, SHH-activated and TP-53 wild type.	Surgery

SHH: Sonic hedgehog, FLAIR: Fluid-attenuated inversion recovery, CSI: Craniospinal irradiation, NA: Not available

Table 2: JBI critical appraisal checklist for case studies.

Study ID	A	B	C	D	E	F	G	H
Becker *et al*. 1995	No	No	No	Yes	No	No	No	Yes
Meshkini *et al*. 2014	No	Yes	Yes	Yes	No	No	No	Yes
Doan *et al*. 2018	Yes	No	Yes	Yes	Yes	No	No	Yes
Singh *et al*. 2021	Yes	Yes	Yes	Yes	Yes	Yes	No	Yes

JBI: Joanna Briggs Institute. Criteria for Quality Assessment: A: Were the patient's demographic characteristics clearly described? B: Was the patient's history clearly described and presented as a timeline? C: Was the current clinical condition of the patient on presentation clearly described? D: Were diagnostic tests or assessment methods and the results clearly described? E: Was the intervention (s) or treatment procedure (s) clearly described? F: Was the postintervention clinical condition clearly described? G: Were adverse events (harms) or unanticipated events identified and described? H: Does the case report provide takeaway lessons?

medulloblastomas are more frequently found eccentrically. To the best of our knowledge, this report represents the fifth documented study and the seventh case of tentorial medulloblastoma in the adult population.

The commonly reported symptoms include morning headaches, nausea, vomiting, lethargy, and ataxia, primarily attributed to increased intracranial pressure – either due to the tumor mass itself or associated obstructive hydrocephalus – and cerebellar dysfunction.[22] Previous studies have documented similar clinical presentations. In the case described by Doan *et al*.,[4] there was a 2-week history of headaches accompanied by a syncopal episode. The report by Singh *et al*.[23] indicated that the patient experienced a 3-month history of chronic dull, aching headaches on the left side, along with 2.5 months of gait instability. In addition, the study by Meshkini *et al*.[13] reported a patient with headache, vomiting, and mild truncal ataxia. Our case also illustrates this classic presentation of a posterior fossa lesion linked to elevated intracranial pressure and mild cerebellar dysfunction, resulting in gait ataxia.

In the current case, the MRI characteristics of the lesion showed significant enhancement and diffusion restrictions on the MR Spectroscopy, indicating increased neoplastic activity. This finding is further supported by an elevated choline level and a high choline/NAA ratio. Notably, these imaging features can sometimes mimic those typically associated with meningiomas. Becker *et al*.[1] observed three patients, each presenting different degrees of contrast enhancement categorized as mild, moderate, and slightly moderate, with heterogeneous features, respectively. In addition, Singh *et al*.[23] reported mild and heterogeneous enhancement of the tumor, an increased signal for choline and creatine, and the absence of ascending NAA or lipid-lactate signals significantly complicating the differentiation between these two entities. Furthermore, Doan *et al*.[4] emphasized the presence of a tentorial dural-tail sign, similar to what is observed in meningiomas; however, this sign can also appear in medulloblastomas as a manifestation of neoplastic infiltration within the dura mater, extending beyond the primary tumor mass.[6] These observations underscore that eccentric medulloblastomas, particularly in adults, may pose a diagnostic challenge due to their neuroradiological resemblance to meningiomas.

According to the 2021 classification by the WHO regarding CNS tumors, medulloblastomas are molecularly classified into four distinct subgroups: wingless integration(WNT)-activated, sonic hedgehog (SHH)-activated, tumor protein 53 (TP53) wild type, SHH-activated TP53 mutant, and non-WNT non-SHH.[2,9,12] In our case, the patient is classified as group 2, identified as SHH-activated and TP53 wild type, as reported by Singh *et al*.[23,24] However, Doan *et al*.[4] classified their patient solely within the SHH group. It is noteworthy that the SHH subgroup of medulloblastoma is particularly associated with tumors occurring in the cerebellar hemispheres, unlike the other subgroups.[14,16] Nevertheless, our case represents a unique instance of SHH-activated medulloblastoma located outside the cerebellar hemispheres in an extra-axial location.

The SHH-activated subtype of medulloblastoma, as identified in our case, has significant therapeutic implications. Unlike other subtypes, SHH-driven tumors show potential responsiveness to targeted therapies, particularly SMO inhibitors such as vismodegib and sonidegib, which have shown promise in clinical trials.[7,21] This subtype also demonstrates distinct patterns of spread and prognosis, with TP53-wild-type cases generally exhibiting better outcomes compared to TP53-mutant variants.[19,29] These findings highlight the necessity of molecular profiling in medulloblastoma cases, as it can guide treatment decisions, potentially reducing the need for extensive craniospinal irradiation (CSI) in selected patients.[15]

The evolving landscape of precision medicine in neuro-oncology suggests that future strategies may involve tailored therapeutic approaches based on molecular and genetic markers, improving survival while minimizing long-term treatment-related morbidity.[18-20]

Current treatment regimens are based on pediatric protocols, as specific guidelines for adult medulloblastoma are lacking due to its relative rarity.[8,24] The standard of care currently involves maximal safe resection, adjuvant chemotherapy, and CSI.[25] Historically, craniospinal radiation was administered postoperatively at a dose of 36 Gy, with an additional boost of 54–55.8 Gy to the posterior fossa tumor bed, owing to the high radiosensitivity of medulloblastomas. Since the 1990s, numerous researchers have demonstrated that incorporating adjuvant chemotherapy significantly enhances the absolute survival rate.[5,28] In the documented case by Singh *et al*.,[23] the patient underwent surgical intervention followed by chemoradiation,

which included etoposide, cisplatin, and CSI. Similarly, Doan *et al.*[4] described a strategy combining surgery, radiotherapy, and adjuvant chemotherapy with vincristine, cisplatin, and cyclophosphamide. These cases illustrate that chemotherapy protocols may vary. In our case, the same standard management approach was utilized: maximal safe resection combined with chemoradiation, followed by radiotherapy.

The prognosis and clinical outcomes of medulloblastomas vary significantly depending on the molecular subtype and the age of the patient. As in our case, patients with SHH-activated medulloblastomas typically have an intermediate prognosis, falling between the excellent outcomes seen in WNT-activated tumors and the poorer prognosis associated with non-WNT/non-SHH-activated (group 3) medulloblastomas. However, prognosis can be highly variable based on specific clinicopathological features.[20,27] In infants with SHH-activated wild-type medulloblastomas, the outcomes are generally favorable. For young children and adolescents, survival rates exceed 80% unless high-risk factors such as metastatic disease or MYCN amplification are present, which are linked to a worse prognosis.[21] In adults, SHH-activated medulloblastomas tend to have relatively favorable outcomes, although studies on this age group are limited.[29]

CONCLUSION

Reports regarding tentorial medulloblastomas, especially among the adult population, are notably scarce. These tumors pose considerable diagnostic challenges due to their morphological similarities to meningiomas. Precise identification requires the use of advanced imaging techniques along with detailed histopathological evaluation, which also includes molecular and genetic analyses. The main treatment options involve surgical intervention followed by chemotherapy and/or radiation therapy, highlighting the need for comprehensive evaluation and personalized management strategies for these rare cases.

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