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Case report

Rare incidence of parietal lobe metastasis in an adult with desmoplastic/nodular medulloblastoma: A case report and review of the literature

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ABSTRACT

Introduction and importance: Medulloblastoma in adults is a rare and highly aggressive central nervous system (CNS) tumor, representing less than 1 % of all brain tumors. Supratentorial metastasis is uncommon, and extra-neural metastasis occurs in approximately 5 % of cases, primarily in frontal and temporal lobes. Here, we present an exceptional case of parietal lobe metastasis in an adult with desmoplastic/nodular medulloblastoma. To explore prior cases and establish the uniqueness of our case, we conducted a thorough search on the PubMed database.

Case presentation: A 46-year-old male, who was previously treated for medulloblastoma with surgery and adjuvant chemoradiotherapy seven years ago, presented with clinical symptoms suggestive of potential tumor recurrence. Despite two years of dedicated adjuvant chemoradiotherapy, the patient exhibited progressive right hemiparesis, ataxia, and gait disturbances. Subsequent brain magnetic resonance imaging (MRI) revealed a distinct 6 × 4 × 2 cm lesion in the left parietal lobe, which, upon post-operative histopathological examination, was identified as a supratentorial metastasis originating from desmoplastic/nodular medulloblastoma.

Clinical discussion: Medulloblastomas, once categorized as primitive neuroectodermal tumors (PNET), are now distinctly classified as high-grade embryonal tumors, mainly characterized by their histological features and cellular origin. Common clinical presentations include hydrocephalus, headache, unsteady gait, and truncal ataxia. Surgical intervention aims for radical excision, complemented by vital adjuvant chemoradiotherapy to minimize recurrence risk.

Conclusion: Considering the possibility of tumor recurrence or intracranial metastasis in patients with medulloblastoma is crucial. Therefore, regular follow-ups are strongly recommended to promptly detect any signs of reoccurrence in these atypical presentations.

1. Introduction

Medulloblastoma (MB) is a highly aggressive central nervous system (CNS) neoplasm consisting of undifferentiated primitive small round cells. It predominantly affects children, constituting approximately 25 % of all pediatric brain tumors. However, its occurrence in adults is rare, accounting for less than 1 % of all brain tumors [1,2]. Adults diagnosed with MB experience a more unfavorable prognosis in comparison to children; hence, early detection is vital, as metastatic disease at diagnosis is associated with a poor outcome. Additionally, long-term complications have a considerable impact on the quality of life and survival of these patients [3].

Patients with medulloblastoma may present with various initial symptoms, such as nocturnal or morning headaches, nausea, vomiting, altered mental status, ataxia, and other nonspecific indications of increased intracranial pressure. Due to inconsistent findings on imaging, histopathological examination is essential to confirm the diagnosis [2,4]. Adult medulloblastoma metastasizing to supratentorial regions is rare. When it occurs, it often involves the sub-frontal region [5]. Furthermore, extra-neural metastasis is infrequent, affecting

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approximately 5 % of cases, with the bone marrow, lungs, and liver commonly involved sites. Therefore, for adult patients with medulloblastoma, adjuvant radiation therapy is the standard of care and should be initiated promptly after surgery. Any delays or interruptions in treatment have been associated with worse outcomes [6,7].

This study aims to emphasize the importance of a rare incidence of supratentorial parietal lobe metastasis of medulloblastoma in a 46-year-old man, highlighting the need for vigilant surveillance and personalized treatment approaches for these exceptional occurrences. Additionally, this work has been reported in line with the SCARE criteria [8].

2. Case presentation

In September 2016, a 39-year-old man presented with complaints of dizziness, loss of balance, difficulty walking, nausea, and vomiting at the hospital. Upon physical examination, the patient exhibited a tandem gait and tested positive for loss of proprioception and cerebellar function (finger-to-nose and heel-to-shin test). Magnetic Resonance Imaging (MRI) scans confirmed the presence of a posterior fossa tumor accompanied by hydrocephalus. Consequently, the patient underwent an urgent endoscopic third ventriculostomy (ETV) procedure, which was performed through a right-sided burr hole to address the hydrocephalus.

Ten days after the ETV, an elective surgical procedure was carried out, resulting in the complete removal of the cerebellar mass measuring 8 × 5 × 4 cm. Later on, hemostasis was effectively achieved, and duraplasty was performed using a pericranium patch to reduce the risk of complications. Subsequently, the patient was closely observed in the intensive care unit (ICU), and the postoperative period proceeded without any notable incidents. The tumor's histopathological examination confirmed the diagnosis of medulloblastoma, prompting the patient's referral to an oncologist for further management. The patient underwent adjuvant concurrent chemoradiotherapy, followed by a monthly course of adjuvant intravenous chemotherapy.

Five years later, a surveillance brain MRI showed mild hyperintensity (T2-weighted imaging) in the left cerebellum with mild edema, suggesting a possible tumor recurrence. Additionally, a whole-body bone scan (WBBS) revealed widespread bone marrow metastases in various locations, including the spine, ribs, sternum, proximal bilateral humeri, pelvic bones, and proximal bilateral femora. In the past two years, despite receiving adjuvant chemoradiotherapy, the patient has experienced a gradual, progressive weakness on the right side, which has led to right hemiparesis, ataxia, and gait disturbances. Consequently, a brain MRI with and without contrast revealed a well-defined 6 × 4 × 2 cm supra-tentorial mass [Fig. 1]. Initially, the tumor raised suspicion of a parasagittal meningioma, primarily due to its location and consistent contrast uptake pattern. As a result, the patient underwent a complete surgical removal of the tumor.

Histopathological examination revealed distinctive characteristics of desmoplastic/nodular medulloblastoma, an uncommon finding in this particular location. The tumor consisted of desmoplastic/nodular architecture infiltrated by rich reticulin fibers around the pale nodules. Pale reticulin-poor nodules comprise tumor cells with variable neurocytic maturation set in a fibrillary matrix. The reticulin-rich area comprises densely packed cellular sheets of undifferentiated small round blue cells with brisk mitotic activity [Fig. 2]. Immunohistochemistry (IHC) study confirmed the mentioned diagnosis with the positive results for GFAP, Synaptophysin, D2–40, ATRX, B-Catenin (cytoplasmic), Neurofilament 70/200 kDa (scattered positive) and Ki-67 (50 %). The other markers of the intended IHC panel were negative (CK, EMA, NKX-2, SOX-10, S-100a, ALK-1, IDH-1, OLIG-2, PHOX2B, P-53).

The patient was discharged from the hospital in stable condition following the surgery. Currently, the patient is undergoing further assessment to determine the most appropriate action for adjuvant therapy. An overview of the timeline can be found in Fig. 3.

3. Discussion

This report outlines the case of a 46-year-old male diagnosed with desmoplastic/nodular medulloblastoma, initially manifesting as a posterior fossa tumor, and despite receiving appropriate adjuvant concurrent chemoradiotherapy, a supratentorial recurrence occurred five years later. The initial non-specific symptoms and the subsequent clinical progression [2,4] emphasize the need for vigilant monitoring and awareness of unusual metastatic patterns.

In our study, we conducted a comprehensive literature search in the PubMed database using the following search strategy: “(((parietal) OR

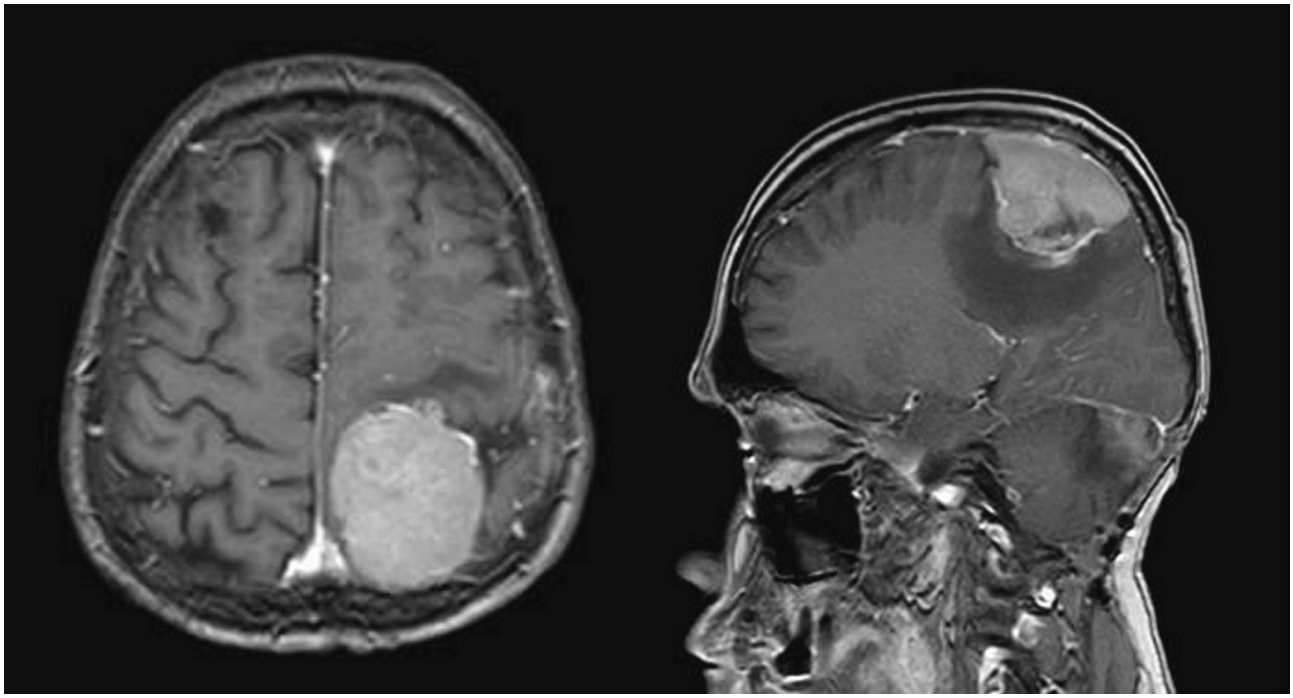

Fig. 1. A well-defined enhancing oval mass in the left parietal lobe measuring about 6 × 4 × 2 cm in T1-weighted MRI with Gadolinium contrast.

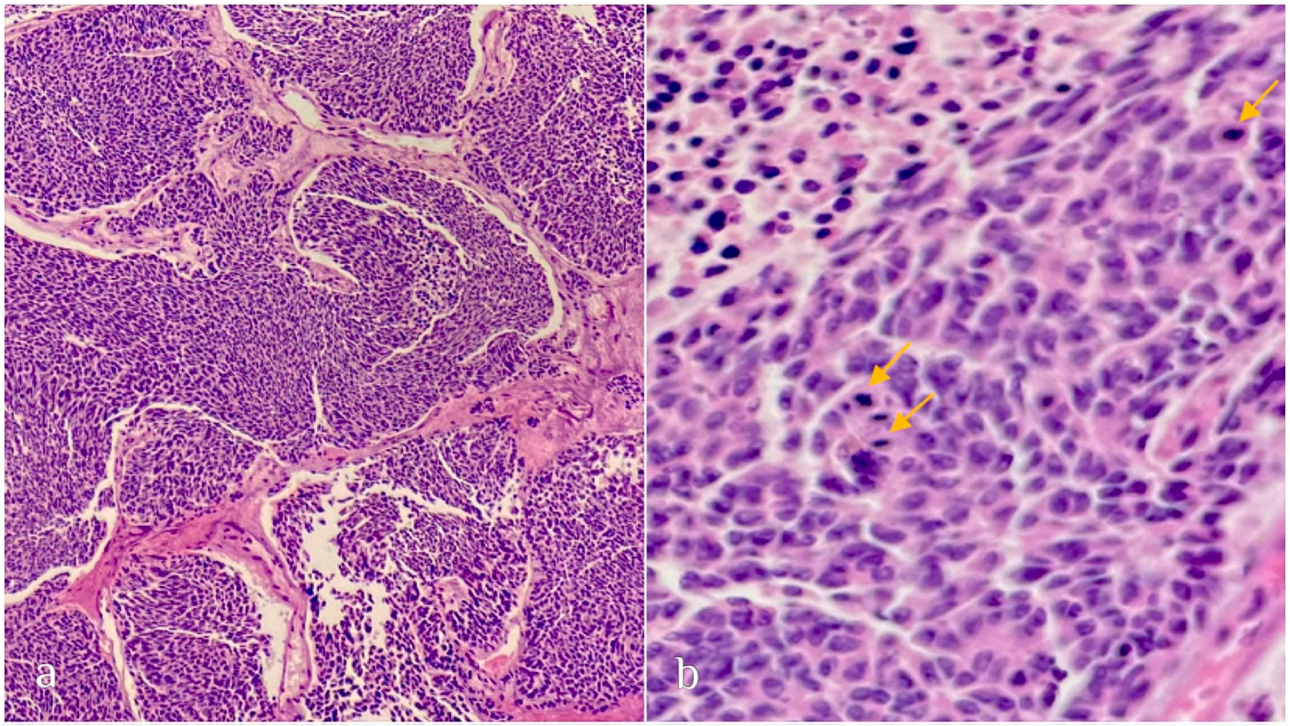

Fig. 2. a. Desmoplastic/Nodular growth pattern (×10, H&E) b. Brisk mitotic activity (arrows) in the highly proliferative undifferentiated area surrounding the central pale nodular islands with a neurocytic differentiation (×40, H&E).

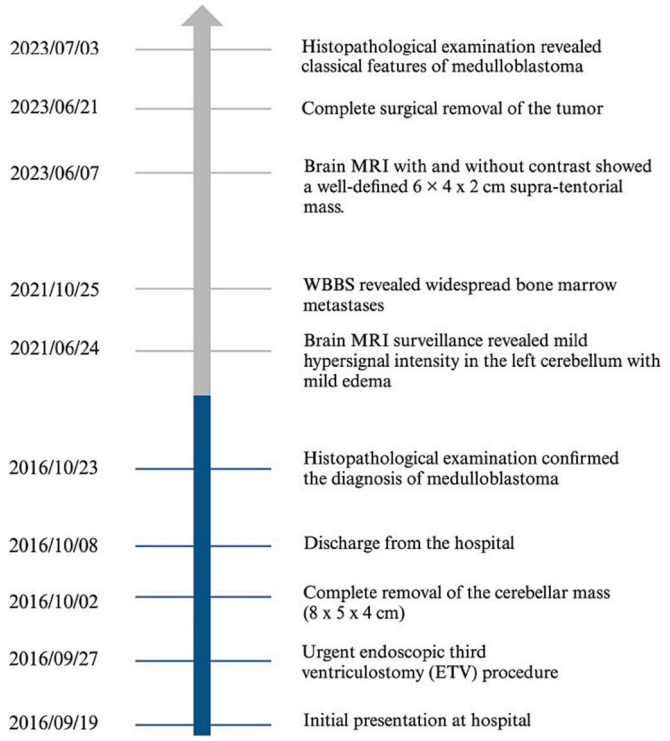

Fig. 3. A chronological overview of notable events.

((supratentorial) OR (supra tentorial)))) AND (((medulloblastoma) OR (PNET)) OR (primitive neuroectodermal tumor) AND (adult)).” Our most recent search update was performed on September 7, 2023. After excluding irrelevant article types and pediatric cases mentioned solely in the title and abstract, we meticulously examined the full text of the remaining articles. To the best of our knowledge, and considering the available pathological and imaging data in the literature, our case represents a unique instance of parietal lobe metastasis in an adult with desmoplastic/nodular medulloblastoma.

Medulloblastomas are rare among adults, with an annual incidence varying between 0.5 and 20 cases per 1 million individuals [9]. These neoplasms are categorized as high-grade embryonal tumors based on their histological features and cellular origin. Historically, medulloblastomas were grouped with other embryonal tumors under the primitive neuroectodermal tumors (PNET) classification. However, according to the WHO classification, they are now recognized as a distinct entity, encompassing classic medulloblastoma, desmoplastic/nodular medulloblastoma, medulloblastoma with extensive nodularity, and large-cell/anaplastic medulloblastoma. Additionally, medulloblastomas are further divided into four subcategories: wingless (WNT) activated, sonic hedgehog (SHH) activated, group 3, and group 4, according to the molecular pathways responsible for their development [10].

In addition to physical examinations, diagnosing medulloblastomas relies on brain Computed Tomography (CT) and MRI scans. On CT imaging, medulloblastomas appear hyperdense, while MRI shows isointensity or hypo-intensity in T1-weighted imaging and hyperintensity in T2/FLAIR (fluid-attenuated inversion recovery) imaging [11].

They commonly present with hydrocephalus and cerebellar dysfunction symptoms, such as nausea/vomiting, headache, unsteady gait, and truncal ataxia. Metastases typically occur in the posterior fossa, spine, and bones [3,5,12]. Supratentorial metastasis of adult medulloblastoma is scarce, primarily involving the frontal lobe, sub-frontal region, or cribriform plate. Kumar et al. reported two cases, aged 31 and 20 years, with supratentorial metastases occurring 3½ years and 11 months after the complete removal of posterior fossa medulloblastoma. Both patients underwent craniospinal irradiation; notably, the first case also developed spinal metastasis [13].

Surgery is the primary treatment for all MB patients, aiming to achieve the most radical excision possible. The surgical goals include relieving the mass effect, restoring CSF circulation, obtaining diagnostic tissue, and reducing tumor burden. While complete resection is

considered the standard approach upon diagnosis, the extent of survival advantage between this and near-total resection (removal of over 90 % of the tumor) remains uncertain. Given the substantial risk of recurrence associated with surgery alone, adjuvant radiotherapy plays a crucial role in MB treatment [14,15].

Radiation therapy usually commences approximately 3 to 4 weeks after surgery. The treatment involves irradiating the entire craniospinal axis, called craniospinal irradiation (CSI) [15]. Adult medulloblastoma patients classified as high-risk, which includes individuals with large cell or anaplastic medulloblastoma, supratentorial primitive neuroectodermal tumors (PNETs), disseminated disease, tumors that cannot be surgically removed, or residual tumors exceeding 1.5 cm after surgical intervention, are subjected to the standard dose of craniospinal irradiation (CSI) and radiation targeted at the posterior fossa. Additionally, chemotherapy is an integral component of the therapeutic regimen in pediatric cases. Standard adjuvant chemotherapeutic regimens include cisplatin, carboplatin, and etoposide with or without cyclophosphamide [16]. However, routine chemotherapy in adult patients remains controversial.

In cases where hydrocephalus is present, the consideration of ventricular shunting may be required either prior to or following surgery. Nevertheless, certain patients may be able to avoid the need for shunting by effectively managing hydrocephalus through primary decompression and the restoration of cerebrospinal fluid (CSF) pathways [17].

4. Conclusion

This report details the case of a 46-year-old male with desmoplastic/nodular medulloblastoma, presenting initially as a posterior fossa tumor. Despite receiving appropriate adjuvant chemoradiotherapy, a supratentorial recurrence occurred after five years. Emphasizing the importance of routine examination and MR imaging in post-treatment medulloblastoma patients, regular follow-up imaging can be exceedingly advantageous in identifying any instances of local recurrence or distant metastasis, as the availability of multiple imaging datasets enhances the accuracy of diagnosis in such rare cases.

Ethics approval and consent to participate

This study received approval from the Institutional Review Board at Guilan University of Medical Sciences, with the ethics approval code IR.GUMS.REC.1402.298.

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Author contribution

N.Z. and E.A. co-authored in writing and collecting data. E.A. and S.K. designed figures; N.Z. and S.K. conceived the idea. S.K. revised the manuscript, aided by input from S.Y.C., Z.R., and P.K., who provided valuable feedback and contributed to the manuscript's refinement.

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Consent

Written informed consent was obtained from the patient to publish this case report and accompanying images. A copy of written consent is available for review by the editor-in-chief of this journal on request.

Conflict of interest statement

There are no conflicts of interest.

Data availability

The data used to support the findings of this case report are available from the corresponding author upon request. Anonymized and aggregated data that do not compromise patient confidentiality can be made available to researchers for further analysis upon request and appropriate ethical approvals.

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