First Incidence of Parietal Lobe Metastasis in an Adult with Desmoplastic/Nodular Medulloblastoma: A Case Report and Review of the Literature

Nooshin Zaresharifi
1- Department of Pathology, Faculty of Medicine, Guilan University of Medical Sciences

Elahe Abbaspour
2- Clinical Research Development Unit of Poursina Hospital, Guilan University of Medical Sciences

Shahrokh Yousefzade-Chabok
3- Guilan Road Trauma Research Center, Guilan University of Medical Sciences

Zoheir Reihanian
3- Guilan Road Trauma Research Center, Guilan University of Medical Sciences

Paridokht Karimian
1- Department of Pathology, Faculty of Medicine, Guilan University of Medical Sciences

Sahand Karimzadhagh (✉ Sahand.karimzad.md@gmail.com)
2- Clinical Research Development Unit of Poursina Hospital, Guilan University of Medical Sciences

Case Report

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Abstract

**Background:** 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 extraneural metastasis occurs in approximately 5% of cases, primarily in frontal and temporal lobes. Here, we present a unique case of parietal lobe metastasis in an adult with desmoplastic/nodular medulloblastoma, adding a novel dimension to the existing knowledge base. We conducted a thorough search on the PubMed database to explore prior cases and establish the uniqueness of our case.

**Case presentation:** A 46-year-old male with a posterior fossa tumor and hydrocephalus was diagnosed with medulloblastoma seven years ago. Adjuvant concurrent chemoradiotherapy was administered after surgery. After five years, a surveillance brain MRI suggested possible tumor recurrence. A whole-body bone scan revealed widespread bone marrow metastases. Despite receiving adjuvant therapy in the past two years, the patient has experienced progressive right hemiparesis, ataxia, and gait disturbances. A brain MRI showed a distinct 6 x 4 x 2 cm in the left parietal lobe. The patient's tumor was entirely removed, initially suspected to be a parasagittal meningioma. However, it was identified as a supratentorial metastasis of desmoplastic/nodular medulloblastoma after a thorough histopathological examination.

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

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 medulloblastoma experience a more unfavorable prognosis in comparison to children; hence, early detection is vital, as disseminated or metastatic disease at diagnosis is linked to 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, and when it occurs, it often involves the sub-frontal region.(5) Furthermore, extraneural metastasis is infrequent, affecting 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 novel 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.

**Case Presentation**

In September 2016, a 39-year-old man presented with complaints of dizziness, loss of balance, difficulty walking, nausea, and vomiting. 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 x 5 x 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 hypersignal intensity (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 therapy, 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 was conducted, revealing a well-defined 6 x 4 x 2 cm supra-tentorial mass. [Figure 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 classical features of desmoplastic/nodular medulloblastoma, which is a rarity in this 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. [Figure 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 released from the hospital after the surgery in stable condition. 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 Figure 3.

Discussion

In our study, we conducted a comprehensive literature search in the PubMed database using the following search strategy: "(((parietal) OR ((supratentorial) OR (supra tentorial))) AND (((medulloblastoma) OR (PNET)) OR (primitive neuroectodermal tumor) AND (adult)))". Our most recent search update, performed on September 7, 2023, yielded a total of 279 articles. After excluding review articles and pediatric cases mentioned solely in the title and abstract, we meticulously examined the full text of the remaining articles. Based on the available pathological and imaging data in the literature, our case represents a unique instance—the first reported occurrence 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.(8) 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, they are now acknowledged as a separate entity. 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.(9)

In addition to physical examinations, the diagnosis of medulloblastomas relies on Brain CT and MRI scans. On CT imaging, medulloblastomas appear hyperdense, while MRI shows iso-intensity or hypointensity in T1-weighted imaging and hyperintensity in T2/FLAIR (fluid-attenuated inversion recovery) imaging.(10)

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.(11) Supratentorial metastasis of adult medulloblastoma is scarce, primarily involving the frontal lobe, sub-frontal region, or cribiform 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.(12)

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. As surgery alone carries a high recurrence risk, adjuvant radiotherapy is crucial in MB treatment.(13,14)

Radiation therapy usually commences approximately 3 to 4 weeks after surgery. The treatment involves irradiating the entire craniospinal axis, called craniospinal irradiation (CSI).(14) 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.(15) 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.(16)

**Conclusion**

Our report recommends routine examination and MR imaging for 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.

**Abbreviations**

CNS
Central nervous system
CSI
Craniospinal irradiation
CSF
Cerebrospinal fluid
ETV
Endoscopic third ventriculostomy

FLAIR
Fluid-attenuated inversion recovery

IHC
Immunohistochemistry

ICU
Intensive care unit

MB
Medulloblastoma

MRI
Magnetic Resonance Imaging

PNETs
primitive neuroectodermal tumors

SHH
Sonic Hedgehog

WBBS
Whole-body bone scan

WNT
Wingless INT-1

Declarations

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.

Consent for publication

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.

Availability of data

The data used to support the findings of this case report are available from the corresponding author upon request.

Competing Interest

The authors declare that they have no competing interests.

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Authors Contributions

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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None.

References


**Figures**

**Figure 1**

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

a. Desmoplastic/Nodular growth pattern (x10, H&E) b. Brisk mitotic activity (arrows) in the highly proliferative undifferentiated area surrounding the central pale nodular islands with a neurocytic differentiation (x40, H&E).
Figure 3

Timeline of notable events (a). Schematic illustration portraying the tumor’s location within the parietal lobe (b).