Case Report

Anaplastic Meningioma: A Rare Case Report

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Abstract

Meningiomas are tumors arising from the leptomeninges. The malignant counterpart of them is known as anaplastic meningioma, which are WHO grade 3 tumors. Histopathology and immunohistochemistry are confirmatory for subtyping of meningioma. We present a case of a 45-year-old male who presented with usual symptoms of headache and projectile vomiting. After surgery, the clinical diagnosis of meningioma was made, and the tissue was sent to the pathology department. Definitive diagnosis revealed a high-grade spindle cell neoplasm disposed in sheets with markedly elevated and atypical mitoses. Large cells with eccentric cytoplasm were also seen. IHC was positive for vimentin and EMA and negative for GFAP. A final diagnosis of anaplastic meningioma was made. During follow-up, the patient also received adjuvant radiotherapy post-surgery. Anaplastic meningioma is very rare and generally known to have a poorer prognosis. The extent of resection is associated with prognosis of patients with all WHO grades of meningioma.

Keywords:

Anaplastic meningioma, Meningioma, Immunohistochemical

Introduction

Meningiomas are primary CNS tumours originating from arachnoid cells located on the inner surface of the dura mater. These are the most common primary CNS malignancy, accounting for 36% of all intracranial tumours, and are mostly benign (CNS WHO grade 1). By contrast, anaplastic meningiomas are rare tumours, representing 1% of all meningiomas. Meningiomas comprise 37.6% of all primary CNS tumors and 53.3% of all benign CNS tumors [1]. The incidence of meningiomas increases with age, with the median age at diagnosis being 66 years old [1]. The incidence rate in patients aged 40+ years is 18.69/100,000, and in ages 0–19 years it is 0.16/100,000 [1]. In patients aged 40+ years, 15–39 years, and 0–14 years, meningiomas make up 43.6%, 15.6%, and 1.7% of all CNS tumors, respectively [1]. Benign and malignant meningiomas are more common in females, with incidence rate ratios of 2.33 and 1.12, respectively [1]. Females and males in the 0–19 year age range had similar incidence ratios of meningiomas [1]. Different from low-grade meningiomas that occur mostly in women and are associated with a relatively good

outcome, anaplastic meningiomas are more frequent in men and have poor prognosis [2]. The symptoms and clinical presentation of anaplastic meningiomas depend on the location of the primary tumor. They can cause symptoms due to mass effect, such as headaches, seizures, or focal neurological deficits resulting from compression of nearby structures or increased intracranial pressure. Anaplastic meningiomas are often found on the convex surfaces of the cerebellum and parietal region, with rare occurrences in the spinal cord [3, 4, 5]. Distant metastases are possible but rare and have been described in about 3% of cases. The lung is a frequent site of distant spread; however, liver and lymph node metastases have also been reported. Anaplastic meningiomas are characterized by markedly elevated mitotic activity, typically 20 or more mitoses in 10 high-power fields without papillary architecture or rhabdoid cytology, and >1 per high-power field [6, 7]. Immunohistochemical stains, including EMA, progesterone receptor, and vimentin, are commonly used to confirm the diagnosis of meningioma. However, it's important to note that focal or absent staining, particularly with EMA and progesterone receptor, can occur, especially in high-grade subtypes. The assessment of proliferative activity is aided by the Ki-67 stain, which highlights the most proliferative foci and facilitates mitotic count. On computed tomography (CT), meningiomas typically appear as slightly hyperdense, well-circumscribed, extraxial masses attached to the dura. They may exhibit intratumoral calcifications, especially in slow-growing subtypes often discovered incidentally. Adjacent skull remodeling or hyperostosis can also be observed [8, 9]. Gadolinium-enhanced magnetic resonance imaging (MRI) is the preferred method for further characterizing these lesions and assessing response in meningioma patients, providing detailed information about tumor location, size, and enhancement pattern [10, 11].

Case Report

Here we report a case of a 45-year-old male who presented to the neurosurgery department with the chief complaint of projectile vomiting with headache for the past one month, along with dizziness and decreased appetite. Imaging studies (CT: computed tomography) scan showed a heterogeneous hypodense mass lesion in the left frontal lobe with perilesional edema, causing a mass effect with midline shift. Frontal craniectomy and excision were performed. Grossly, we received grey-white to grey-brown soft tissue pieces measuring together 10 × 8 × 2 cm. Microscopic examination revealed an anaplastic meningioma; WHO Grade 3.

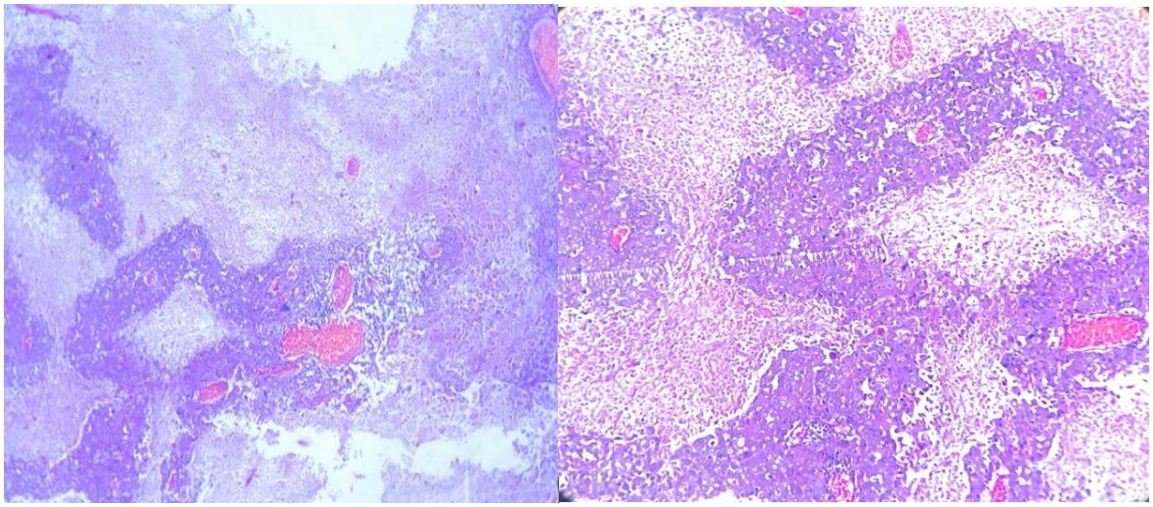

Figure 1: Tumor cells dispersed in sheets with markedly elevated activity of mitosis (H&E; 100X 400X)

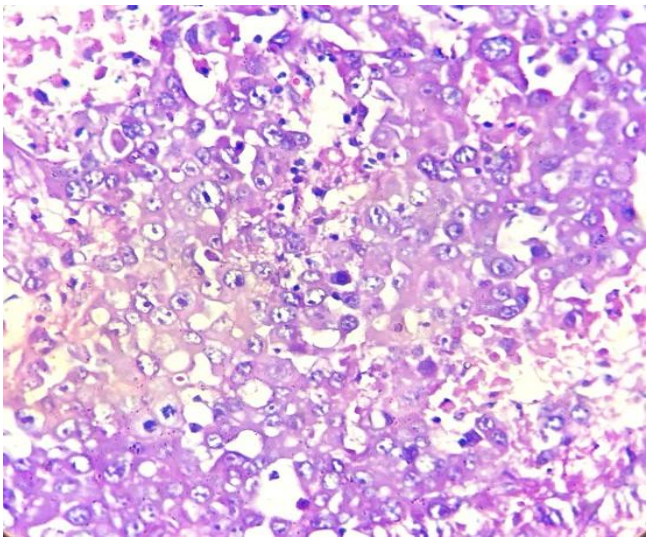

Figure 2: Tumor cells with pleomorphism and mitosis(H&E; 400X)

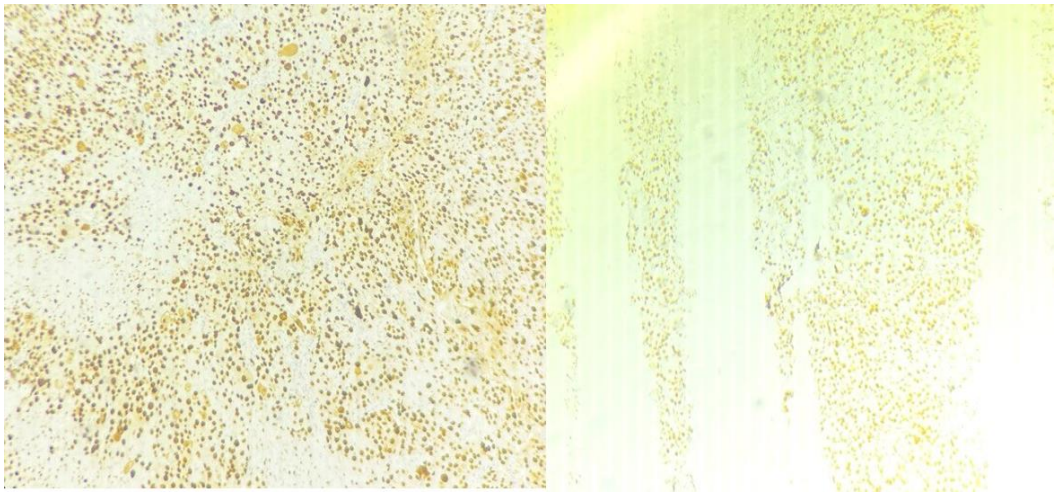

Figure 3: IHC Stained section showing P53-Strong and diffuse positive, EMA- Positive

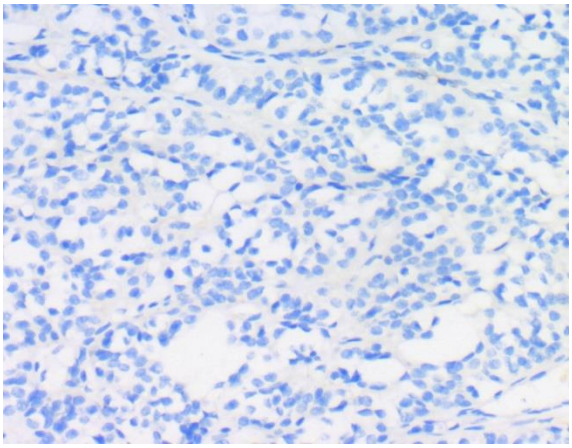

Figure 4: IHC Stained section showing GFAP-Negative

Discussion

Meningioma is the most common intracranial brain tumor, accounting for over one-third of primary brain neoplasms. Meningioma is divided into 15 subtypes and 3 grades. Grade 3 has 3 variants, namely Anaplastic, Rhabdoid, and Papillary. Anaplastic Meningioma is the most common grade 3 type, which has a high degree of malignancy and a high recurrence rate.

The age at onset of Anaplastic Meningioma ranges from 18 to 70 years old. Variable reports of median survival are found in the literature, with some series reporting a survival of 1.5–3.5 years, a 5-year survival rate of 35%–61%, which may be due to variations in the times of distant metastasis [12]. Female predominance is noted only in Benign Meningioma, and male predominance is found in Atypical and Malignant variants. The clinical presentation of Anaplastic Meningioma can indeed vary widely, often reflecting the location and size of the tumor. Typical symptoms such as headaches, seizures, and focal neurological deficits are common due to mass effect. However, some patients may remain asymptomatic until the tumor reaches a certain size or location.

The diagnostic approach for meningiomas typically involves radiological imaging, such as CT and MRI, to assess the characteristics and location of the tumor. Contrast-enhanced imaging is particularly useful, showing hyperdensity and strong enhancement on CT and hyperintensity on T2-weighted MRI. The presence of calcifications, necrotic areas, hemorrhage, or cystic changes can add further insight into the tumor's nature. Confirmation through histopathological examination remains essential for definitive diagnosis.

Distinguishing between spindle cell intracranial lesions can indeed be challenging due to overlapping morphological features. Immunohistochemical staining plays a crucial role in narrowing down the differential diagnosis. While both ependymomas and gliomas typically show GFAP expression, meningiomas are GFAP-negative but express EMA and vimentin. EMA expression in ependymomas tends to be localized to the Golgi zone, presenting as dot-like staining, contrasting with the cytoplasmic expression seen in meningiomas.

Surgery remains the mainstay treatment for intracranial tumors, including meningiomas, with the goal of achieving maximal safe resection. The role of adjuvant therapies such as radiotherapy is still debated, particularly regarding its efficacy in preventing progression of primary or recurrent meningiomas. While radiotherapy can be effective in certain cases, its use as an adjuvant treatment remains controversial due to concerns about potential side effects and long-term outcomes. Chemotherapy is generally reserved for recurrent tumors after surgical and radiation treatments, although its effectiveness in this context is limited [13].

Conclusion

Anaplastic meningioma is prone to metastasis and recurrence. It represents a major challenge in terms of treatment, making accurate diagnosis essential. Anaplastic meningioma exhibits the general imaging characteristics of a malignant tumor, but when it causes bone restriction, observing whether symmetry exists between bone restriction and mass may provide clues to diagnosis. Intraventricular anaplastic meningiomas are rare and aggressive tumors with a high recurrence rate. The presence of rhabdoid differentiation and a young age of presentation further add to their rarity. Despite their uncommon occurrence, meningiomas should be considered in the clinical and radiological differentials of intraventricular tumors, alongside the more typical ependymomas. This highlights the importance of comprehensive evaluation and consideration of less common possibilities in clinical practice.

Abbreviations:

WHO – World Health Organization

IHC – Immunohistochemical

CNS – Central Nervous System

GFAP – Glial Fibrillary Acidic Protein

EMA – Epithelial Membrane Antigen

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Conflict of Interest: The authors declare no conflict of interest.

Patient Consent Status: The consent for images or other clinical information relating to the case to be reported in a medical publication had been taken by the patient.

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