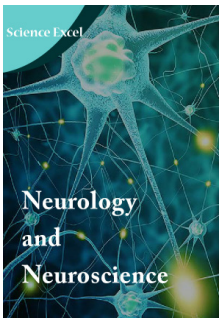
Neurology & Neuroscience

Case Report



Intracranial Meningeal Hemangiopericytoma: Case Report and Literature Review

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Abstract

Hemangiopericytoma (HPC) is a rare mesenchymal tumor, often located in extracranial areas, and when found in the meninges, it can be mistaken for meningiomas. Intracranial HPC accounts for approximately 0.4% of all brain tumors. These tumors are classified into three types based on biological behavior: benign (Type I), atypical (Type II), and malignant (Type III). Malignant HPCs are characterized by high recurrence rates and metastatic potential. Differential diagnosis between HPC and meningioma is challenging due to similar clinical and radiological features; however, STAT6 immunohistochemical expression is a key marker for differentiation. Imaging techniques such as contrast-enhanced MRI and positron emission tomography (PET) are valuable in preoperative tumor characterization. This report presents a 29-year-old female with recurrent severe headaches and a temporoparietal extra-axial tumor, initially diagnosed as meningioma via imaging. Postoperative histological analysis confirmed the diagnosis of HPC. Despite total surgical resection, the patient experienced local recurrence, requiring further surgical intervention and adjuvant radiotherapy and chemotherapy. HPC is more aggressive than meningioma, with a recurrence rate of 91% and metastasis to various organs. Preoperative embolization can minimize intraoperative bleeding risks due to the tumor's vascularity. Early diagnosis and vigilant postoperative follow-up are critical for improving patient outcomes and managing recurrence and metastasis.

Introduction

Hemangiopericytoma (HPC) is a rare mesenchymal neoplasm originating from pericytes of venules and capillaries, typically observed in extracranial areas. When located in the meninges, it forms a solid tumor that adheres to the dura mater, often being confused with meningiomas. Intracranial HPC accounts for approximately 0.4% of all brain tumors [1,2]. Depending on biological behavior and histopathological grade, HPCs can be classified into three types: Type I (benign, solitary fibrous), Type II (atypical), and Type III (malignant, highly mitotic, and aggressive). Malignant HPCs have a high recurrence rate and the potential for metastases, making the management of these tumors challenging and frequently complex [3-5].

The differential diagnosis between HPC and meningioma is often difficult, as both tumor types share similar clinical and radiological characteristics. However, immunohistochemical evaluation, including the expression of the STAT6 marker, has proven to be a crucial tool for differentiating these two neoplasms [6-8]. Furthermore, due to the vascular nature of the tumor, imaging exams such as contrast-enhanced magnetic resonance imaging (MRI) and positron emission tomography (PET) scans have been helpful in better characterizing the tumor before surgical intervention [9,10].

Case Report

A 29-year-old female patient sought medical attention due to recurrent episodes of severe headache, with no previous history of significant comorbidities. A brain MRI revealed a solid extra-axial tumor located in the right temporoparietal region, measuring 35 mm in diameter, showing characteristics suggestive of meningioma, such as dural attachment and increased vascularity (Figure 1).

The patient underwent microsurgery with total resection of the lesion, which appeared hypervascular and atypical, inconsistent with typical meningiomas. The surgery was performed with the patient in right lateral decubitus, with the superior sagittal sinus at the top of the surgical opening, aiming to allow the tumor to separate from the superior sagittal sinus with the assistance of gravity. During the

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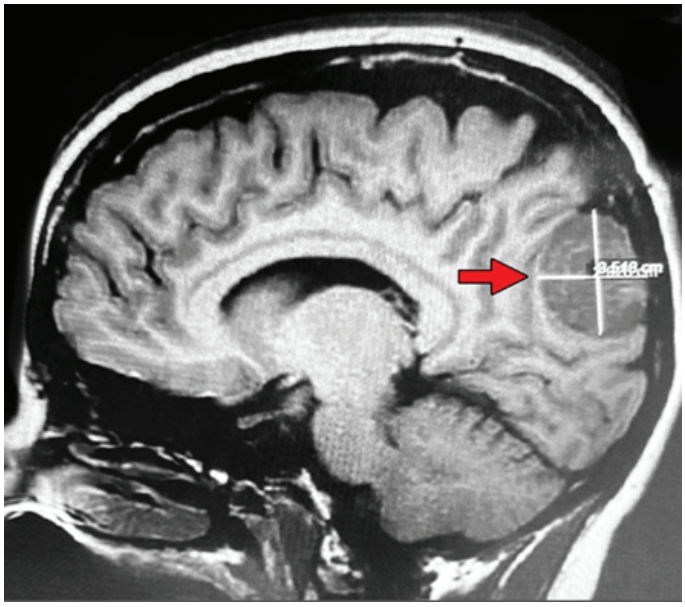

***Figure 1**. Brain MRI (sagittal section) showing a solid extra-axial lesion in the right temporoparietal region, with a diameter of 35 mm (red arrow).*

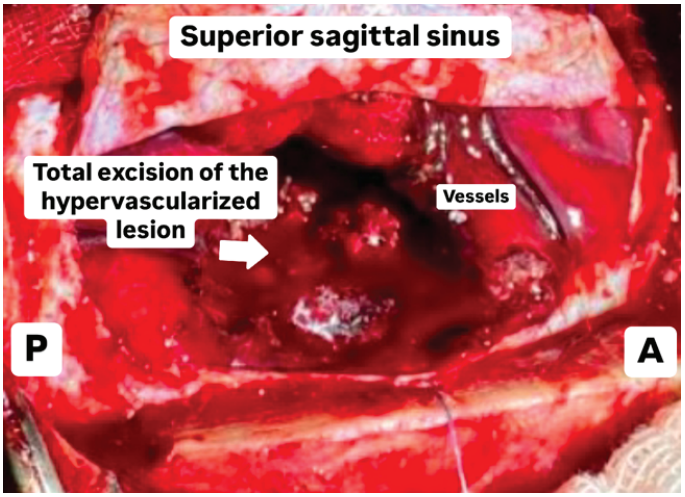

***Figure 2**. Intraoperative image, with the patient positioned in right lateral decubitus (A - anterior and P - posterior), with the superior sagittal sinus located at the top of the surgical opening, highlighting large vessels around the lesion, and total resection of the hypervascularized lesion with a significant amount of blood.*

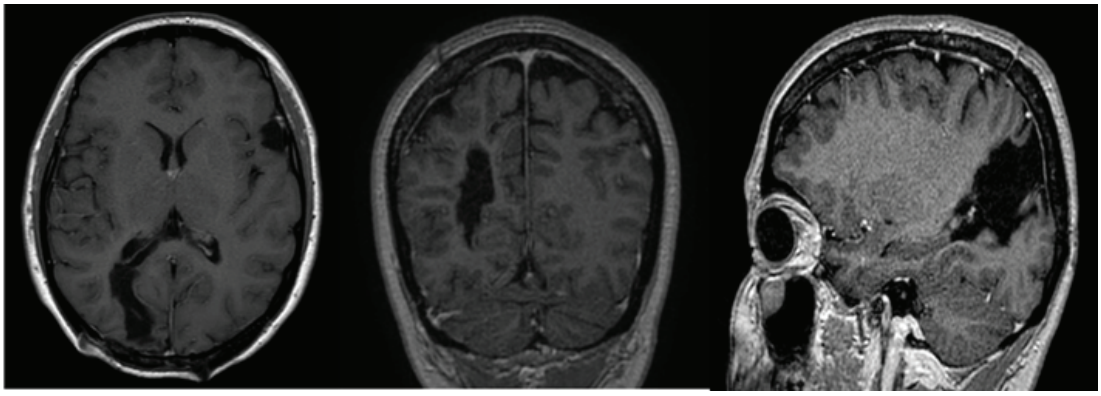

***Figure 3**. Post-operative brain MRI in axial, coronal, and sagittal sections showing complete resection of the lesion.*

surgical procedure, there was significant bleeding, but no blood transfusion was required (Figure 2).

A post-operative brain MRI showed complete resection of the lesion (Figure 3).

Histological analysis revealed a fusocellular neoplasm with diffuse positivity for the STAT6 marker, confirming the diagnosis of hemangiopericytoma/solitary fibrous tumor. Eighteen months after surgery, the patient developed local recurrence, requiring another surgical intervention. Additionally, adjuvant chemotherapy and radiotherapy were initiated due to the high recurrence rate and the risk of metastases.

Discussion

Intracranial meningeal hemangiopericytoma is a more aggressive tumor than meningioma, with a recurrence rate of 91% and up to a 70% chance of metastases, even after complete resection of the lesion. Studies suggest that after 15 years of follow-up, more than 50% of patients may experience recurrences or metastatic spread [2,3]. The most common sites of metastasis include the lungs, liver, bones, subcutaneous tissues, breast, pleura, and even the thyroid [4,5,11].

Total surgical resection of the tumor is considered the standard treatment, but the risk of intraoperative bleeding due to the tumor's intense vascularity can complicate the procedure. Preoperative embolization has been recommended to reduce these risks and improve surgical outcomes [12,13]. Additionally, adjuvant radiotherapy has proven effective in reducing recurrence rates and controlling metastases [14].

The differential diagnosis between HPC and meningioma is crucial, as treatment strategies may differ significantly. Histological analysis with immunohistochemistry is fundamental for diagnosis, with STAT6 expression showing high sensitivity (100%) and specificity for HPC. Other markers, such as CD34 (present in about 60% of cases), may also assist in diagnosis [2,3,6,15].

Resection of meningeal HPCs is complex due to their high vascularity, which increases the risk of intraoperative bleeding. Preoperative embolization is recommended to reduce this

risk and improve surgical outcomes [5,13]. Advanced MRI techniques, including diffusion, perfusion, and spectroscopy, can be useful in characterizing the tumor's microstructure, vascularity, and metabolism, aiding in surgical planning and post-operative follow-up [9,16].

Conclusion

Meningeal hemangiopericytoma is a rare but aggressive neoplasm that can present similarly to meningioma on radiological images. However, differentiating between these two entities is critical for proper management, as HPC requires special considerations such as preoperative embolization and adjuvant treatments (radiotherapy and chemotherapy) due to its high recurrence rate and metastatic potential. Rigorous follow-up after resection is essential for early detection of recurrences and metastases, improving long-term prognosis.

Conflict of Interests

The authors have no conflict of interests to declare

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