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Intracranial Solitary Fibrous Tumour / Hemangiopericytoma: A Rare Meningeal Neoplasm with Aggressive Clinical Behaviour

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Abstract: ***Solitary fibrous tumor/hemangiopericytoma (SFT/HPC) is a rare, aggressive central nervous system tumor with significant diagnostic and therapeutic challenges. This article presents a case of a 65-year-old female with intracranial HPC, initially misdiagnosed as meningioma, and discusses its clinical presentation, imaging features, histopathology, and immunohistochemical findings. The case underscores the tumor's aggressive nature, potential for recurrence and metastasis, and the necessity for long-term surveillance. Through imaging and pathological correlations, the article highlights the evolving classification and diagnostic strategies for such rare meningeal tumors.***

Keywords: hemangiopericytoma, solitary fibrous tumor, meningeal tumor, CNS metastasis, intracranial neoplasm

1.Introduction

Intracranial hemangiopericytoma (HPC) is a rare malignant tumor, accounting for less than 1% of all intracranial neoplasms and approximately 0.4–0.5% of central nervous system (CNS) tumors [1]. These tumors were initially believed to arise from Zimmermann's pericytes of the meninges[2] and were previously classified as angioblastic meningiomas due to their overlapping clinical and radiological features with meningiomas[3]. However, subsequent studies demonstrated that they share genetic alterations with solitary fibrous tumors (SFTs) of the dura, leading to the 2021 World Health Organization (WHO) classification that unified the two entities under the term solitary fibrous tumor/hemangiopericytoma (SFT/HPC)[4].

HPC originates from meningeal mesenchyme and often presents as an intracranial mass mimicking meningioma on neuroimaging [5]. Despite this similarity, the biological behavior of HPC is significantly more aggressive, with reported recurrence rates of up to 60% following surgical resection and extracranial metastases occurring in approximately 20% of cases [6]. Common metastatic sites include the long bones, liver, lungs, CNS, and abdominal cavity[7]. Notably, extracranial metastasis may occur years after treatment of the primary lesion, underscoring the importance of long-term surveillance[8].

Due to its rarity and overlap with meningiomas, recognizing HPC is crucial for neurosurgeons and neuropathologists. The purpose of this article is to present a rare case of intracranial solitary fibrous tumour/hemangiopericytoma and highlight its diagnostic challenges, histopathological features, and the need for long-term clinical follow-up.

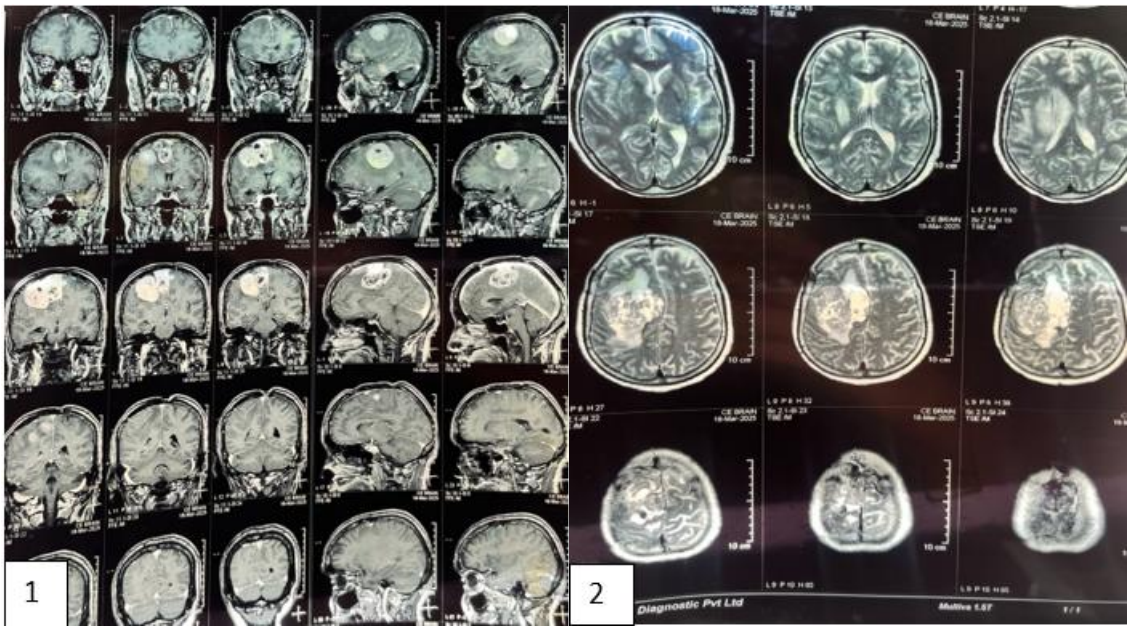
2.Case Presentation

A 65-year-old female presented to the neurosurgery outpatient department with complaints of left-sided weakness for one year and a confused state for the past month. She had a history of craniotomy with decompression of a tumor mass in 2013, although the medical records of that surgery were unavailable. Routine laboratory investigations were within normal limits, and viral markers were negative.

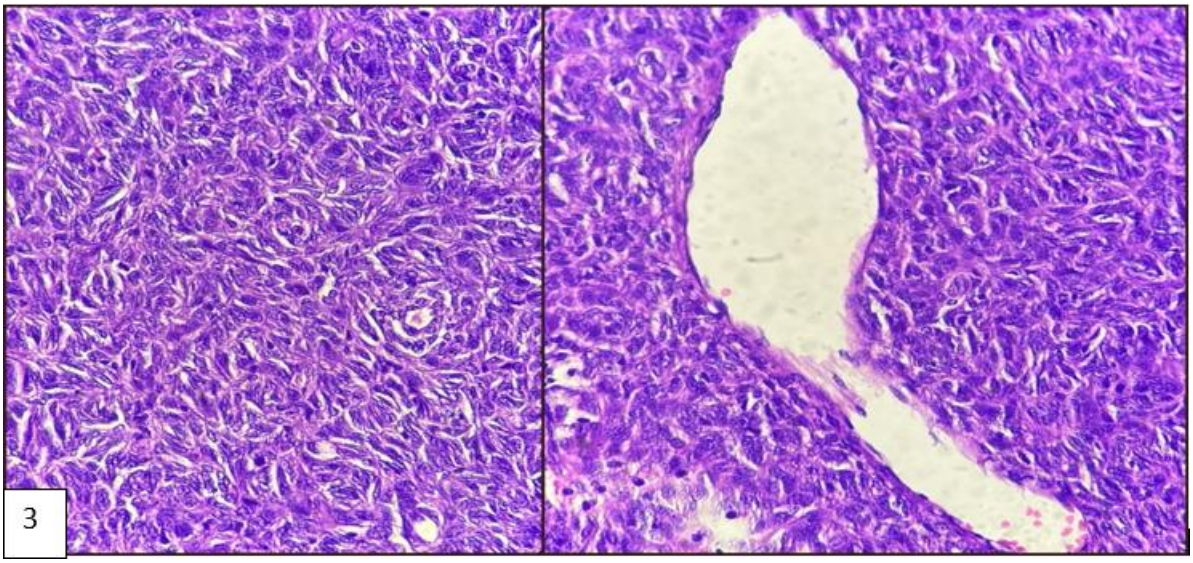
Non-contrast CT (NCCT) of the brain revealed an ill-defined, lobulated, iso- to hypodense extra-axial lesion with cystic and necrotic areas in the right high frontal region, abutting the falx and minimally extending into the left frontal lobe. A provisional diagnosis of meningioma was made. The patient underwent right frontoparietal craniotomy with gross total excision of the tumor. Intraoperatively, the lesion appeared as a soft, suckable mass involving the anterior part of the superior sagittal sinus and falx cerebri.

In the pathology department we received multiple grey yellow to grey brown soft to firm tissue pieces measuring together 3.5x3x0.7 cm which was processed whole. Microsections examined show a tumour composed of oval to spindle shaped cells with cart wheel pattern nuclei, foci of nuclear hyperchromasia and mild to moderate pleomorphism including few mitoses admixed with interspersed staghorn vessels with infiltration into the adjacent meninges. Areas of fibrosis, collagenization and calcification also seen. Differential diagnosis of anaplastic meningioma and hemangiopericytoma were made. Immunohistochemistry profiling was employed; these cells were vimentin, CD34, BCL2 and CD99 positive and EMA was focal positive while CD56, TLE, SMA and S100 were negative. Ki67 proliferating index was around 5-10%.

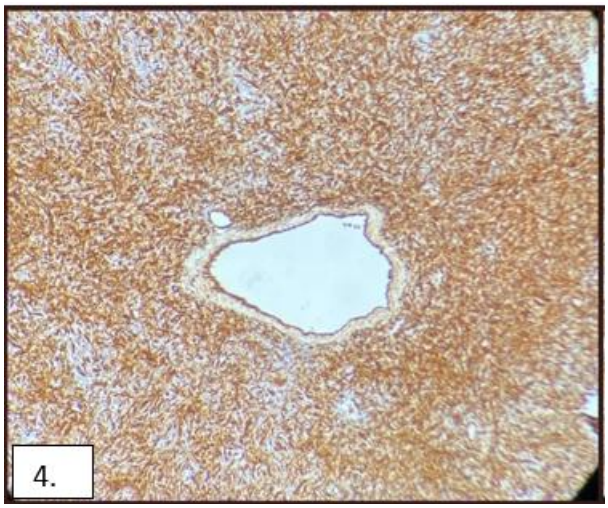
Histomorphological features and immunohistochemical study favour the diagnosis of hemangiopericytoma (WHO II).



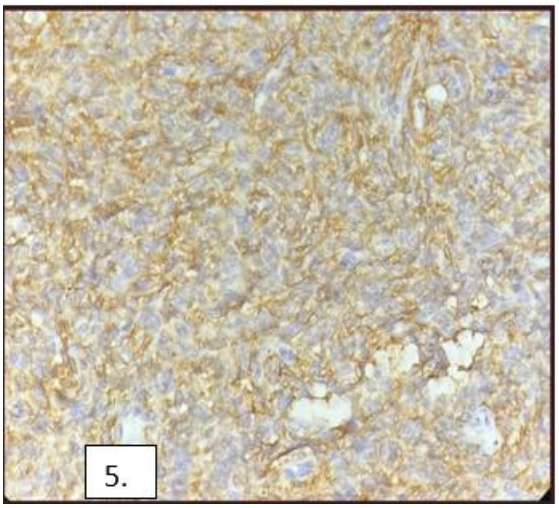
Photograph 1 & 2: Non contrast CT Brain- Sagittal and Axial; showing an extra-axial lesion in Right frontal region, abutting the falx



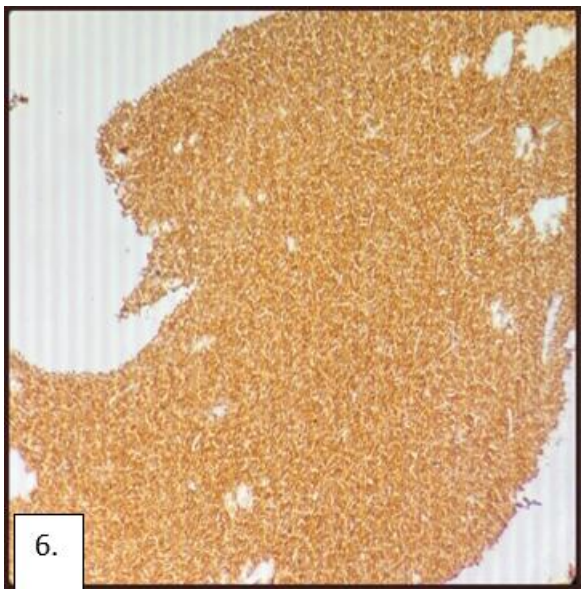
Photograph 3: H&E-stained slide (400x) showing a tumor composed of oval to spindle shaped cells with cart wheel pattern nuclei along with a staghorn vessel



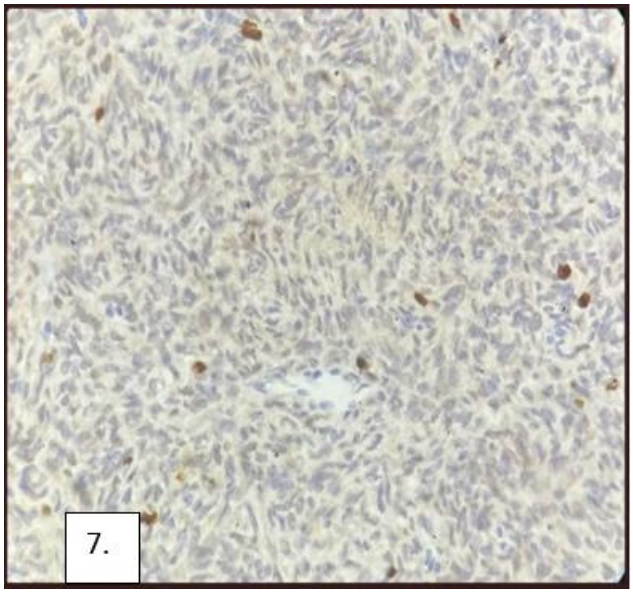
Photograph 4: Immunhistochemical stain of Vimentin showing diffuse cytoplasmic positivity



Photograph 5: Immunohistochemical stain of CD99 showing membranous positivity



Photograph 6: Immunohistochemical stain of BCL2 showing diffuse cytoplasmic positivity



Photograph 7: Immunohistochemical stain of Ki67 showing proliferative index of <5%

3.Discussion

This case emphasizes the importance of distinguishing HPC from meningioma due to differing management approaches and long-term prognoses.

HPC of the central nervous system is an uncommon spindle-cell neoplasm with a wide morphological spectrum, ranging from bland, fibrous lesions to highly cellular, mitotically active tumors. Historically, these tumors were regarded as an angioblastic subtype of meningioma due to overlapping morphologic features, but their distinct biological behaviour prompted reclassification. Stout and Murray in 1942 described hemangiopericytomas as vascular tumors derived from perivascular Zimmerman pericytes[3]. Subsequent molecular studies demonstrated recurrent NAB2-STAT6 gene fusions and STAT6 nuclear expression, leading the WHO in 2016 to unify SFT and HPC as a single entity. The 2021 WHO classification now applies a three-tier grading system based on mitotic activity and cellular features, rendering the term "hemangiopericytoma" obsolete[3].

Clinically, intracranial HPC presents with nonspecific neurological symptoms such as headache, seizures, and motor deficits, largely dependent on tumor size and location[9].These tumors most frequently occur in supratentorial region along the falx and dural sinuses, but posterior fossa and spinal involvement are also documented [10]. Compared with meningiomas, they tend to arise in younger patients, show slight male predominance, and carry a much poorer prognosis [11].

From a pathologist's standpoint, the main diagnostic challenge is distinction from meningioma. On histology, HPCs demonstrate a hypercellular proliferation of uniform to moderately pleomorphic cells arranged around branching "staghorn" vessels, with prominent pericellular reticulin. Mitotic count remains a key prognostic parameter: tumors with >5 mitoses per 10 high-power fields are classified as WHO grade III, previously termed anaplastic HPC [4]. Immunohistochemistry is indispensable: strong nuclear STAT6 positivity is highly sensitive and specific, while CD99 and bcl-2 are often co-expressed [12].

A hallmark feature of HPC is its high propensity for local recurrence and extracranial metastasis, reported in 14–50% of cases[9]. The most frequent metastatic sites are bone, liver, and lung. Spinal metastases are distinctly rare, with only nine reported cases to date, occurring after a mean latency of 11.5 years from the initial intracranial tumour. The vertebral body is the most commonly affected site, attributed to its abundant vasculature and marrow content [9]. The hematogenous route, facilitated by the valveless paravertebral venous plexus and azygos system, is considered the principal metastatic pathway[13].

Interestingly, metastatic deposits may display histological progression compared to the primary lesion. In reported cases, metastatic spinal tumours often reveal increased cellularity, higher mitotic activity, and elevated Ki-67 labelling indices (>10%) compared to their intracranial counterparts, thereby meeting WHO grade III criteria[3].

For long-term monitoring, gadolinium-enhanced MRI is the most sensitive modality for detecting spinal metastasis, while bone scans, though useful, may yield false-negative results and should be repeated when clinical suspicion remains[14]. Recognition of NAB2-STAT6 fusions and STAT6 nuclear staining has strengthened diagnostic precision and could contribute to future targeted therapies.[15]

In conclusion, intracranial solitary fibrous tumour/hemangiopericytoma remains a diagnostically challenging yet clinically critical entity due to its aggressive behaviour and potential for delayed metastasis. Accurate histopathological diagnosis, supported by immunohistochemical markers, is essential in differentiating it from meningioma. The case discussed illustrates the importance of long-term follow-up and reinforces the role of pathology in guiding appropriate treatment strategies.

HPCs are rare but clinically significant CNS tumors with distinctive histological and molecular features. Their diagnostic overlap with meningiomas underscores the critical role of pathology in accurate classification. The potential for late recurrence and metastasis, including to the spine, highlights the need for prolonged surveillance in all patients with a history of HPC, even after seemingly complete surgical excision.

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