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Late Progression of a Residual Solitary Fibrous Brain Tumor after Subtotal Resection and Adjuvant Radiotherapy: A Case Report

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Abstract Case Report

Background: Intracranial solitary fibrous tumors (SFTs) are rare mesenchymal neoplasms with an unpredictable clinical course. While gross total resection is associated with improved local control, residual tumor may persist and exhibit late progression, even after adjuvant radiotherapy. Case Presentation: We report a 35-year-old man who initially underwent subtotal resection of a tentorial SFT, followed by adjuvant fractionated radiotherapy. Regular MRI follow-up over four years showed stable residual disease. The patient later developed symptoms of intracranial hypertension, and imaging revealed significant tumor progression at the previous surgical site. A second subtotal resection was performed, confirming WHO grade II SFT. Postoperatively, the patient received three cycles of VAC chemotherapy, resulting in a reduction of residual tumor volume, with plans for additional cycles. Discussion: This case underscores the potential for late progression of residual intracranial SFTs, even in low-grade lesions and after adjuvant radiotherapy. Gross total resection remains the optimal management strategy, but anatomical constraints often limit resection. Systemic therapy may provide additional benefit in selected cases, while emerging targeted agents represent promising options for progressive or metastatic disease. Conclusion: Late progression of residual SFTs necessitates lifelong radiological surveillance. Early recognition of progressive disease allows timely surgical and medical interventions to optimize patient outcomes.

Keywords: Solitary fibrous tumor, intracranial, late progression, radiotherapy, chemotherapy.

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Introduction

Solitary fibrous tumors (SFTs) are uncommon mesenchymal neoplasms, initially described in the pleura by Klemperer and Rabin in 1931 [1]. Extrapleural SFTs, particularly those arising within the central nervous system (CNS), are exceedingly rare. Historically, they were considered part of the hemangiopericytoma (HPC) spectrum owing to overlapping histopathological and clinical characteristics [2]. The identification of the NAB2–STAT6 fusion gene has since clarified their nosological classification, establishing SFTs and HPCs as entities within a unified biological spectrum [3].

Intracranial SFTs usually present as dural-based lesions, radiologically indistinguishable from meningiomas. They predominantly occur in the cerebral convexities, falx cerebri, tentorium cerebelli, and spinal meninges [4]. Although many cases follow an indolent clinical course, SFTs are characterized by a significant

risk of local recurrence and, in some instances, extracranial metastasis—even decades after the initial diagnosis [5,6].

We report the case of a patient with a residual intracranial solitary fibrous tumor who developed late progression four years after subtotal resection and adjuvant radiotherapy. This case highlights the unpredictable natural history of CNS SFTs and underscores the importance of long-term surveillance. We also provide a review of the relevant literature focusing on clinical presentation, histopathological features, therapeutic approaches, and long-term outcomes.

CASE PRESENTATION

A 35-year-old man with no significant past medical history presented in 2020 with dizziness and symptoms of intracranial hypertension, including

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headaches and vomiting. Neurological examination was otherwise unremarkable.

Initial diagnosis:

MRI of the brain revealed a well-circumscribed lesion above and below the tentorium, with a large base of implantation on either side of the tent of the cerebellum on the right side, isointense on T1 and hyperintense on T2, intensely enhanced after injection measuring 73x38x40mm, with significant mass effect on the brainstem and V4 with upstream hydrocephalus and signs of transependymal resorption.

Treatment and histopathology:

The patient underwent ventriculocisternostomy followed by subtotal surgical resection. Histopathological examination revealed a solitary fibrous tumor (WHO grade II). The tumor showed dense sheets of proliferating cells without glandular or papillary structures. The cells had elongated to spindle-shaped nuclei with mild atypia and rare mitotic figures, in a background partially fibrous and rich in congested, thin-walled dilated vessels around which tumor cells were arranged. Some fragments contained only reactive glial tissue. No calcifications, vascular emboli, or true

rosettes were identified. Immunohistochemistry demonstrated diffuse CD34 positivity and nuclear STAT6 expression, confirming the diagnosis.

Adjuvant treatment and follow-up:

The patient received adjuvant fractionated radiotherapy (50.4 Gy in 28 fractions of 1.8 Gy) delivered with a Novalis TrueBeam linear accelerator, which was well tolerated. Regular MRI surveillance every six months showed stable residual disease.

Progression:

Four years later, the patient developed recurrent intracranial hypertension manifested by headaches and vomiting refractory to medical treatment. MRI demonstrated significant tumor progression at the previous surgical site, now measuring $5.3 \times 4.3 \times 9.1$ cm. The lesion infiltrated the occipital horn of the right lateral ventricle and exerted mass effect with early subfalcine herniation .

Second treatment and histopathology:

A second subtotal resection was performed. Histopathological analysis again confirmed a solitary fibrous tumor, WHO grade II (Figure 1).

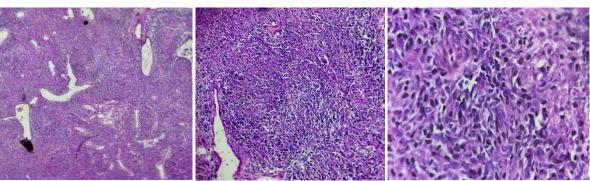


Figure 1: Microscopic examination reveals similar features. The tumor consists of a roughly rounded peripheral proliferation of moderately high cellular density. It is composed of sheets of rounded or slightly elongated cells. These cells exhibit nuclei of moderately increased size, slightly hyperchromatic, with a mitotic count estimated at fewer than 5 mitoses per 10 high-power fields. The cytoplasm is of moderate abundance and basophilic. The stroma is generally delicate and occasionally altered by foci of tumor necrosis. No intact glial tissue is identified

Adjuvant chemotherapy and outcome:

Postoperatively, the patient received three cycles of adjuvant chemotherapy with vincristine, actinomycin D, and cyclophosphamide (VAC regimen). Follow-up MRI demonstrated a reduction in the size of the residual tumor. The patient remains under close surveillance and is scheduled to complete an additional three cycles of chemotherapy.

DISCUSSION

Solitary fibrous tumors of the CNS are rare, accounting for less than 1% of intracranial tumors [4,6]. They were historically grouped with hemangiopericytomas until the discovery of the NAB2–STAT6 fusion, now considered pathognomonic, with STAT6 nuclear immunopositivity serving as a reliable diagnostic marker [3,7].

Radiologically, SFTs frequently mimic meningiomas [4,6]. Histologically, they display spindle to ovoid cells, a collagen-rich stroma, and branching "staghorn" vessels. The 2021 WHO classification stratifies them into three grades according to mitotic activity and necrosis [2]. Despite being grade II in our case, progression occurred four years later, illustrating the unpredictable course of these tumors.

Gross total resection remains the strongest prognostic factor for local control and overall survival [8,9]. However, as in our patient, location often limits complete resection. Adjuvant radiotherapy improves local control, especially after subtotal resection, but does not prevent recurrence [10].

The role of systemic therapy remains limited. Classical cytotoxic chemotherapy has modest efficacy

[11]. Recently, antiangiogenic agents and tyrosine kinase inhibitors (e.g., sunitinib, temozolomide plus bevacizumab) have shown promising results in advanced or metastatic disease [12–14].

Given the risk of late recurrence and extracranial metastasis—even decades after diagnosis—lifelong MRI surveillance is strongly recommended [5,6,15].

CONCLUSION

This case highlights the unpredictable natural history of intracranial solitary fibrous tumors, particularly in the context of residual disease. Even after subtotal resection and adjuvant radiotherapy, residual tumor may remain stable for several years before demonstrating late progression, emphasizing that histological grade alone cannot reliably predict long-term behavior. Management of progressive residual disease may require repeat surgery and, in selected cases, adjuvant chemotherapy, while emerging targeted therapies offer potential options for advanced disease. Lifelong radiological surveillance is essential to detect late progression and guide timely intervention.

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