

# Peritorcular meningeal solitary fibrous tumor: A case report

## Tumor fibroso solitario meníngeo peritorcular: reporte de un caso

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### Resumen

**Objetivo:** Con el fin de contribuir a la epidemiología latinoamericana de estos tumores, presentamos un caso colombiano de un tumor fibroso solitario meníngeo (TFSm) peritorcular, demostrando los hallazgos histológicos, los reparos quirúrgicos y la anatomía relevante para la resección quirúrgica. **Reporte de caso:** Una mujer hispana de 47 años se presentó al servicio de urgencias con un historial de 1 mes de cefalea severa que no respondía a la medicación. La resonancia magnética demostró una lesión hipointensa en T2 de 4 x 4 x 4,7 cm en la región occipital izquierda con edema peritumoral, con realce homogéneo en el T1 tras la administración de gadolinio. La resección total macroscópica se realizó mediante una craneotomía parasagital occipital derecha con un abordaje sencillo. El examen patológico informó la proliferación de células fusiformes con tinción nuclear STAT6 positiva intensa y difusa, lo que confirma el diagnóstico histológico de un mSTF. **Conclusiones:** Presentamos la resección de un TFSm, contribuyendo a la epidemiología y presentación de este raro tumor en América Latina.

**Palabras clave:** Tumor cerebral, tumor fibroso solitario, torcula, STAT6.

### Abstract

**Objective:** To contribute to the Latin American epidemiology of these tumors, we present a Colombian case of a peritorcular meningeal solitary fibrous tumor (mSFT), demonstrating the histological findings, surgical nuances, and relevant anatomy for surgical resection. **Case report:** A 47-year-old Hispanic woman presented to the emergency department with a 1-month history of severe headache that was unresponsive to medication. Magnetic resonance imaging demonstrated a 4 x 4 x 4.7 cm hypointense T2 lesion in the left occipital region with peritumoral edema, with homogeneous enhancement in T1 after gadolinium administration. Gross total resection was performed through a right occipital parasagittal craniotomy with a straightforward approach. Pathology examination reported the proliferation of spindle cells with diffuse and intense positive nuclear STAT6 staining, confirming the histological diagnosis of an mSTF. **Conclusions:** We demonstrate a feasible resection of a peritorcular mSTF, contributing to the epidemiology and presentation of this rare tumor in Latin America.

**Key words:** Brain tumor, solitary fibrous tumor, torcula, STAT6.

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## Introduction

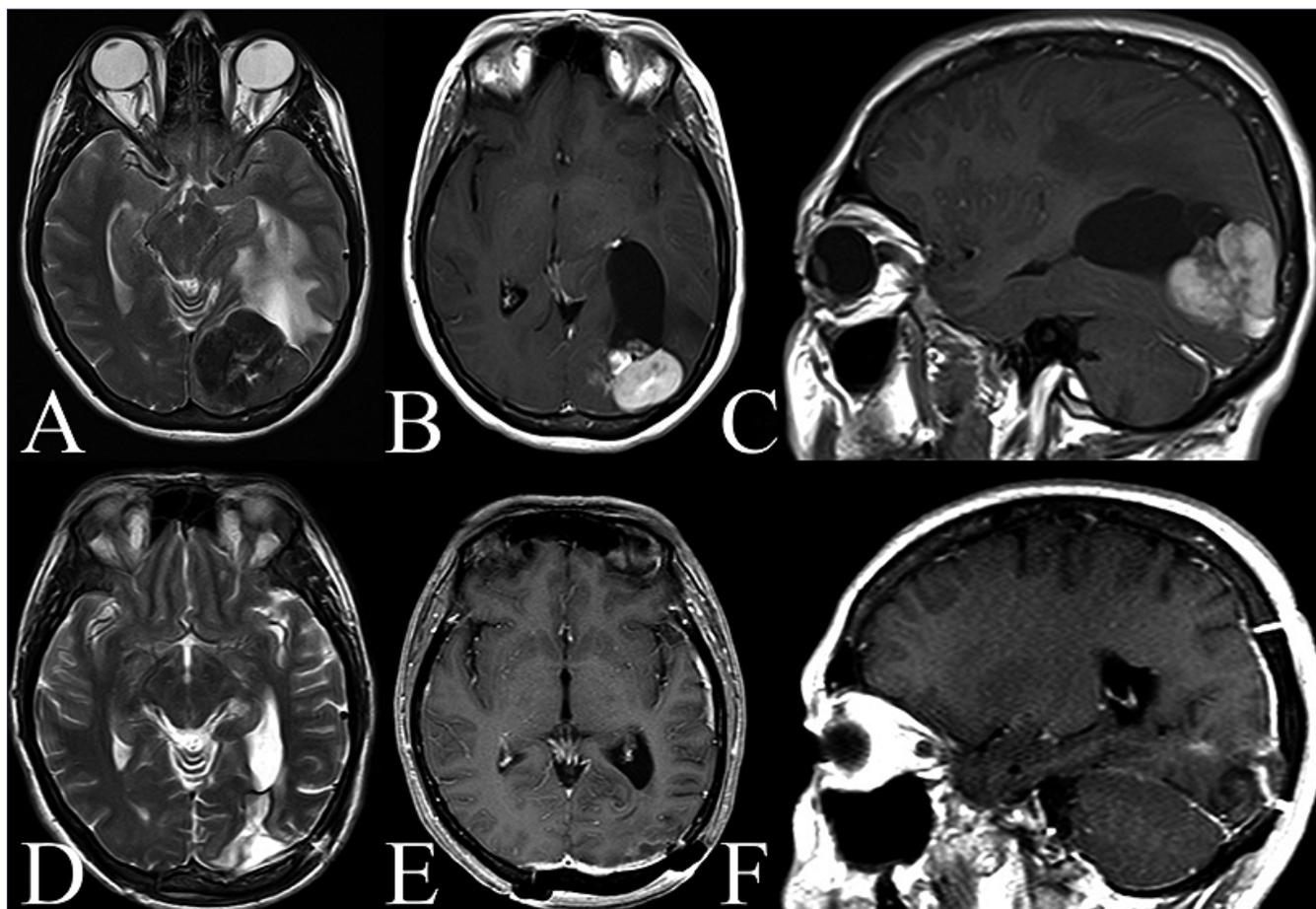
A solitary fibrous tumor (SFT) is a rare sarcoma-type neoplasm characterized by *NAB2-STAT6* gene fusion. Nearly 78-88% of SFT cases are benign, while 12-22% are malignant. They are both oval and spindle-shaped lesions within highly variant collagen stroma. Despite SFT is usually benign and managed by surgical treatment alone, about 34% of cases may develop distant metastasis<sup>1,2</sup>. Pleura is the most common site of origin, but it may occur in any other location, such as the central nervous system (CNS) and extremities<sup>3</sup>. Additionally, extrapleural SFTs are usually more aggressive than pleural. Among extrapleural, meningeal SFT (mSTF) has been reported as a typical primary tumor in this localization<sup>4</sup>.

mSFT may present at any age, but the onset was reported to peak between the fourth and fifth decades. Headache is the most common reason for consultation. Although neuroimaging typically demonstrates well-defined masses, it is still challenging to discriminate mSFT from meningioma pre- and intraoperatively<sup>5,6</sup>. Even though the clinical course is rarely aggressive, with an average recurrence of 42.9%, mSFT may be unpredictable, and its behavior relies on phenotype<sup>7</sup>.

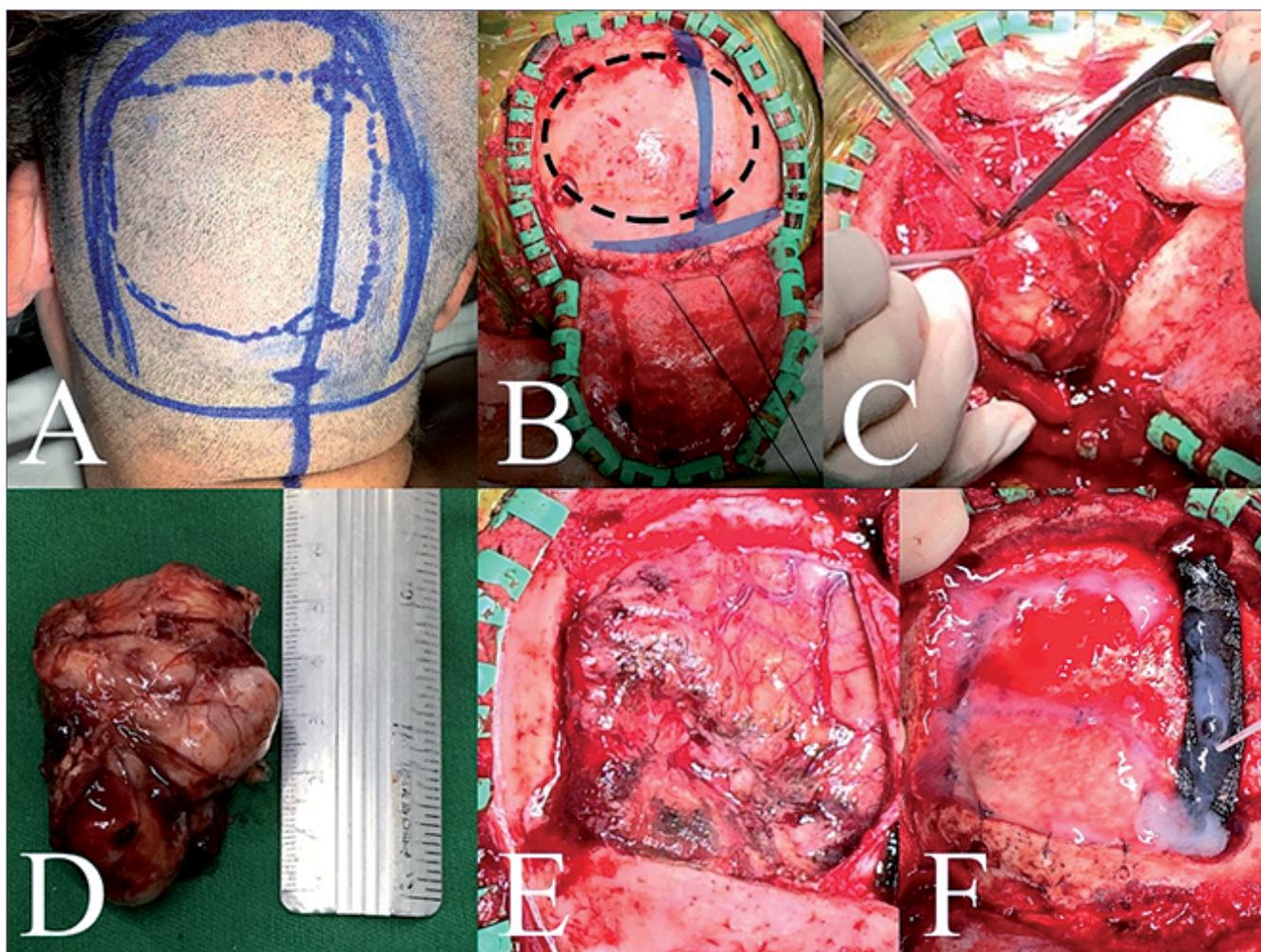
STAT6 nuclear staining is detected in most mSTF (60-95% of tumor cells), making it an excellent specific biomarker for diagnosis<sup>8,9</sup>. Also, CD34 is significant for diagnosis and classification and other biomarkers such as vimentin and CD99. Furthermore, Ki67 is positively expressed in about 10% of cases<sup>2</sup>. To contribute to the Latin American epidemiology of these tumors, we present a Colombian case of a peritorcular mSTF, demonstrating the histological findings, surgical nuances, and relevant anatomy for surgical resection.

## Case report

A 47-year-old Hispanic woman presented to the emergency department with a 1-month history of severe headache that was unresponsive to medication. Besides headache, the patient had no relevant medical antecedents. Neurological examination revealed right homonymous hemianopsia. T2-weighted magnetic resonance imaging (MRI) demonstrated a 4 x 4 x 4.7 cm hypointense lesion in the left occipital region with peritumoral edema, with homogeneous enhancement after gadolinium administration. Distant metastasis was not



**Figure 1.** Pre- and postoperative MRI demonstrating gross total resection of a large occipital fibrous solitary tumor. (A) Axial T2, axial post-contrast T1; (B) and sagittal; (C) post-contrast T1 MR images demonstrate an extra-axial occipital mass, with mixed solid and cystic components and remarkable perilesional brain edema with a consequent left-to-right midline shift; (D) Axial T2 and post contrast T1 axial; (E) and sagittal; (F) MR images demonstrate gross total resection, with improvement of brain edema and ventricular compression.



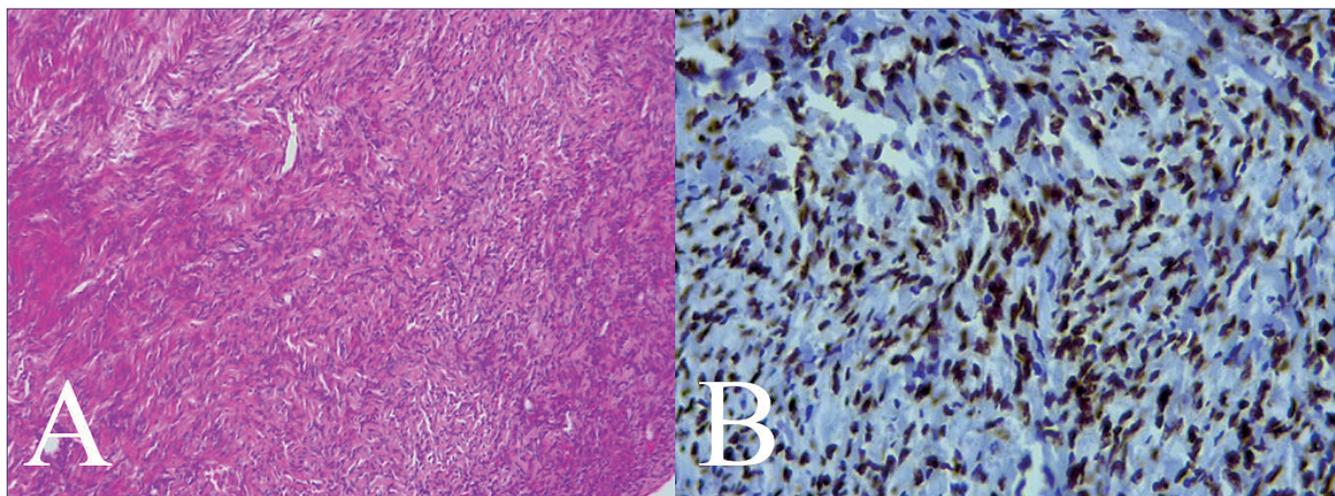
**Figure 2.** Intraoperative images of a peritorcular meningeal solitary fibrous tumor resection. (A) Patient is positioned in prone, with the head rotated 30° degree to the right. The incision, as well as the craniotomy and the superior sagittal sinus are marked; (B) An occipital horseshoe incision is performed, preserving irrigation from both occipital arteries. Both the sagittal and the transverse sinuses are illustrated with blue lines; (C) The dura is opened protecting the sagittal sinus. Therefore, tumor is exposed, coagulated and resected in a piecemeal fashion; (D) An *en-bloc* resection was performed, including the solid and the cystic portion of the tumor; (E) Hemostasis is performed over the residual brain parenchyma and (F) a hermetic dural closure is performed, covered with Tisseel to achieve epidural hemostasis and a hermetic watertight closure.

identified. The lesion was oval-shaped, well-circumscribed, and entrapment of the left lateral ventricle's posterior horn was also detected (Figure 1); therefore, a typical meningioma was initially suspected. An *en bloc* gross total resection was performed through a right occipital parasagittal craniotomy with a straightforward approach (Figure 2). Pathology examination reported the proliferation of spindle cells with diffuse and intense positive nuclear STAT6 staining (Figure 3), confirming the histological diagnosis of an mSTF while ruling out a classic meningioma. The patient was discharged on the fourth day after resection with an adequate postoperative neurological examination. She was readmitted a day after discharge with signs of an adverse drug reaction to phenytoin with eosinophilia and was newly discharged after a 3-day hospitalization after corticoid treatment for the adverse reaction to the medication. After one year, the follow-up MRI revealed no evidence of tumor recurrence, and the clinical follow-up showed no neurological sequelae.

## Discussion

### mSTF Clinical and Radiological Features

mSFTs are rare mesenchymal tumors with a challenging diagnosis but with a mostly mild course<sup>4</sup>. Although its etiology remains unknown, histopathological characterization advances have allowed discriminating this neoplasm from other brain tumors with similar imaging patterns like the meningioma<sup>10</sup>. *NAB2-STAT6* fusion has been reported in about 89% of mSFTs enabling diagnosis by detecting nuclear expression of *STAT6*<sup>11</sup>. The current edition of the World Health Organization Classification of Tumors of the Central Nervous System (WHO CNS) recognizes SFT and hemangiopericytoma (HPC) in a single designation since both share the 12q13 inversion, which is responsible for *NAB2-STAT6* fusion, leading to nuclear *STAT6* expression<sup>12</sup>. There are an estimated less than six cases of meningeal SFT per million people making



**Figure 3.** Histological findings consistent with a solitary fibrous tumor. (A) Characteristic histological pattern of the solitary fibrous tumor conformed by proliferation of spindle cells alternated with abundant collagen fibers and vascular structures; (B) Immunohistochemistry sample shows STAT 6 intense positivity, which are consistent findings of a solitary fibrous tumor.

it challenging to study this malignancy<sup>13</sup>. This report presents the first case of meningeal SFT in the Colombian population with successful management through a complete radiological resection.

Headache was the reason for consultation in this case, as previously described in prior reports<sup>6</sup>. Although the MRI scan revealed a typical contoured lesion, the diagnosis was made only after positively confirming nuclear *STAT6*. Meningioma was the primary differential diagnosis, only excluded after the pathology report. Despite the value of neuroimaging in deciding surgical management, the best approach for differential diagnosis of SFTs against meningiomas when a case is suspected is through the determination of immunohistochemical markers<sup>14</sup>. Challenging management occurs in patients over 55 years old, with tumors sized over 15 cm or with a significant mitotic rate (higher than 4/10 high-power fields)<sup>3</sup>.

In most cases, only surgery is effective in managing mSFT. Radiation therapy, for instance, is not associated with significant clinical improvement<sup>15</sup>. Gross total resection (GTR) is achieved in approximately 75.0% of patients, while subtotal resection is performed by about 25.0%. Nevertheless, STR increases 16-fold the risk of recurrence compared with GTR<sup>16,17</sup>. After surgery, about 68.2% of patients have no residual tumor in follow-up imaging investigation, and the overall 5-year survival reaches 95.0%<sup>18</sup>. Prognosis depends on tumor location, age, tumor size, and extent of resection<sup>19</sup>. In this patient, GTR was performed without complications and no recurrence despite its size and intimate relation to the venous sinuses. Even though the patient did not complain about vision impairment at the consultation, right homonymous hemianopia was found at the neurological examination due to the mass effect on the visual pathways. Both headache and vision loss resolved after surgery, and a follow-up after nine months revealed no neurological sequelae neither recurrence on MRI. Results were expected since this case corresponds to the benign course of disease with no significant risk factors for not considering GTR<sup>18</sup>. Besides some mSFTs reported in the literature, this report contributes as to our best knowledge as

the first of its nature to report a satisfactory surgical treatment of a confirmed mSFT in the Colombian population. Low-to-middle-income countries like Colombia remain in developing adequate surgical techniques and training of specialized pathologists to adequately diagnose this type of rare tumor. Additionally, the former characterization of demographics and epidemiology on these rare tumors in Latin-American population remains scarce due to the ability to translate information from Spanish literature.

#### Anatomical Key-points for resection

Surgical planning for peritortular tumors includes a delicate analysis of the imaging, including the analysis of the venous relations to the tumor, as the main risk is to have an intraoperative rupture of the venous sinuses. A straightforward approach may seem to be the more reasonable approach, and a great precaution must be taken into account when performing the occipital craniotomy. As a venous sinus rupture is imminent during the calvarium opening, the neurosurgeon has to decide whether to cross over the midline or limit opening to the tumor's ipsilateral side. Craniometric points, including the vertex and theinion, are the most remarkable critical points for an adequate approach. The distance from the Torcula, the transverse sinus, and the superior sagittal sinus are critical. As the surface of mSTF may be challenging to differentiate from the normal brain cortex, it is of paramount importance to start arachnoid dissection from the falx cerebri or the superior aspect of the tentorium whenever possible. If possible, an *en bloc* resection must be made, as it is for all extra-axial tumors. A cleavage plane must be conserved during all the procedures, and an early devascularization must be made coagulating the tumor capsule. Finally, for closure, special attention must be paid to prevent cerebrospinal fluid leaking. If necessary, the dura should be excised. However, whereas for meningioma, it is necessary to resect the compromised meninges, for mSFT, the compromise seems to be less aggressive, and a complete resection can be achieved while preserving the adjacent dura.

## Conclusion

mSFT is a rare neoplasm with a mostly benign behavior that may be treated by surgery alone. The outcome is also favorable in most cases, but neurosurgeons must consider metastasis when a case is suspected. Although meningioma is usually the primary differential diagnosis on imaging investigation, histopathological characterization allows discriminating both entities by the nuclear staining of *STAT6*. Physicians are encouraged to report long-term to follow up this pathology's behavior to improve knowledge of its clinical outcome. We present the first Colombian mSTF in the English literature to our knowledge, contributing to the epidemiology and presentation of this rare tumor in Latin America.

**Informed consent:** Written informed consent was obtained from the patient for the submission of this case report.

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**Conflicts of interest:** There are no conflicts of interest.

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