Caso Clínico

Peritorcular meningeal solitary fibrous tumor: A case report

Tumor fibroso solitario meníngeo peritorcular: reporte de un caso

Edgar G. Ordóñez-Rubiano¹, Paula V. Robayo-Manrique², Enrique Acosta-Medina¹, Gabriel E. Acelas-González², Jorge Racedo³

¹Department of Neurosurgery, Hospital de San José, Sociedad de Cirugía de Bogotá, Fundación Universitaria de Ciencias de la Salud (FUCS). Bogotá, Colombia.
²School of Medicine, Fundacion Universitaria de Ciencias de la Salud (FUCS). Bogotá, Colombia.
³Department of Research and Clinical Trials, IMAT Oncomédica. Montería, Colombia.

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 craniotomía 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 mSFT. Conclusions: We demonstrate a feasible resection of a peritorcular mSFT, contributing to the epidemiology and presentation of this rare tumor in Latin America.

Key words: Brain tumor, solitary fibrous tumor, torcula, STAT6.
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\(^1,2\). 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\(^3\). Additionally, extrapleural STFs are usually more aggressive than pleural. Among extrapleural, meningeal STF (mSTF) has been reported as a typical primary tumor in this localization\(^4\).

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 masse, it is still challenging to discriminate mSFT from meningioma pre- and intraoperatively\(^5,6,8\). 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\(^7\).

STAT6 nuclear staining is detected in most mSTF (60-95% of tumor cells), making it an excellent specific biomarker for diagnosis\(^8,9\). 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\(^2\). To contribute to the Latin American epidemiology of these tumors, we present a Colombian case of a peritumoral 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](image)

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.
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 curse. Although its etiology remains unknown, histopathological characterization advances have allowed discriminating this neoplasm from other brain tumors with similar imaging patterns like the meningioma. NAB2–STAT6 fusion has been reported in about 89% of mSFTs enabling diagnosis by detecting nuclear expression of STAT6. 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. There are an estimated less than six cases of meningeal SFT per million people making...
it challenging to study this malignancy. 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. 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. 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).

In most cases, only surgery is effective in managing mSFT. Radiation therapy, for instance, is not associated with significant clinical improvement. 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. 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%. Prognosis depends on tumor location, age, tumor size, and extent of resection. 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. 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 peritocular 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. Cranioometric points, including the vertex and the inion, are the most remarkable critical points for an adequate approach. The distance from the Torcular, 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 mSFT 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.

Financial support and sponsorship: None.

Conflicts of interest: There are no conflicts of interest.

References

1. Demicco EG, Wagner MJ, Maki RG, et al. Risk assessment in solitary fibrous tumors: Validation and refinement of a risk stratification model I. Mod Pathol. 2017;30(10):1433-1442. doi:10.1038/modpathol.2017.54
2. Zhanlong M, Haibin S, Xiangshan F, Jiacheng S, Yicheng N. Variable solitary fibrous tumor locations: CT and MR imaging features. Med (United States). 2016;95(13):e3031. doi:10.1097/MDD.0000000000003031
3. Demicco EG, Park MS, Araujo DM, et al. Solitary fibrous tumor: A clinicopathological study of 110 cases and proposed risk assessment model. Mod Pathol. 2012;25(9):1298-1306. doi:10.1038/modpathol.2012.83
4. Ronchi A, Cozzolino I, Zito Marino F, et al. Extrapleural solitary fibrous tumor: A distinct entity from pleural solitary fibrous tumor. An update on clinical, molecular and diagnostic features. Ann Diagn Pathol. 2018;34:142-150. doi:10.1016/j.anndiagpath.2018.01.004
5. Zhij R, Pang J, Yang C, Huo Z. Solitary Fibrous Tumors/ Hemangiopericytomas of the Maters (Meninx): A Clinicopathologic Analysis. Acta Acad Med Sin. 2019;41(4):512-516. doi:10.3881/j.issn.1000-503X.10845
6. Ohba S, Murayama K, Nishiyama Y, et al. Clinical and Radiographic Features for Differentiating Solitary Fibrous Tumor/ Hemangiopericytoma From Meningioma. World Neurosurg. 2019;130:e383-e392. doi:10.1016/j.wneu.2019.06.094
7. Gubian A, Ganau M, Cebula H, et al. Intracranial Solitary Fibrous Tumors: A Heterogeneous Entity with an Uncertain Clinical Behavior. World Neurosurg. 2019;126:e48-e56. doi:10.1016/j.wneu.2019.01.142
8. Zhang X, Cheng H, Bao Y, Tang F, Wang Y. Diagnostic value of STAT6 immunohistochemistry in solitary fibrous tumor/meningeal hemangiopericytoma. Zhonghua Bing Li Xue Za Zhi. 2016;45(2):97-101. doi:10.3760/cma.j.isn.0529-5807.2016.02.006
9. Fritchie KJ, Jin L, Rubin BP, et al. NAB2-STAT6 gene fusion in meningeal hemangiopericytoma and solitary fibrous tumor. J Neuropathol Exp Neurol. 2016;75(3):263-271. doi:10.1093/jnen/niw026
10. Schweizer L, Koelsche C, Sahm F, et al. Meningeal hemangiopericytoma and solitary fibrous tumors carry the NAB2-STAT6 fusion and can be diagnosed by nuclear expression of STAT6 protein. Acta Neuropathol. 2013;125(5):651-658. doi:10.1007/s00401-013-1117-6
11. Fritchie K, Jensch K, Moskalev EA, et al. The impact of histopathology and NAB2–STAT6 fusion subtype in classification and grading of meningeal solitary fibrous tumor/hemangiopericytoma. Acta Neuropathol. 2019;137(2):307-319. doi:10.1007/s00401-018-1952-6
12. Louis DN, Perry A, Reifenberger G, et al. The 2016 World Health Organization Classification of Tumors of the Central Nervous System: a summary. Acta Neuropathol. 2016;131(6):803-820. doi:10.1007/s00401-016-1545-1
13. Darlix A, Zouaoui S, Rigau V, et al. Epidemiology for primary brain tumors: A nationwide population-based study. J Neurooncol. 2017;131(3):525-546. doi:10.1007/s11060-016-2318-3
14. Macagno N, Figarella-Branger D, Mokthari K, et al. Differential Diagnosis of Meningeal SFT-HPC and Meningioma. Am J Surg Pathol. 2015;40(2):1. doi:10.1097/PAS.00000000000000526
15. Champeaux C, Khan AA, Wilson E, Thorne L, Dunn L. Meningeal haemangiopericytoma and solitary fibrous tumour: a retrospective bi centre study for outcome and prognostic factor assessment. J Neurooncol. 2017;134(2):387-395. doi:10.1007/s11060-017-2538-1
16. Sung KS, Moon JH, Kim EH, et al. Solitary fibrous tumor/hemangiopericytoma: Treatment results based on the 2016 WHO classification. J Neurosurg. 2019;130(2):418-425. doi:10.1016/j.jns.2017.10.004
17. Fargen KM, Opalach KJ, Wakefield D, Jacob RP, Yachnis AT, Lister JR. The central nervous system solitary fibrous tumor: A review of clinical, imaging and pathologic findings among all reported cases from 1996 to 2010. Clin Neurol Neurosurg. 2011;113(9):703-710. doi:10.1016/j.clineuro.2011.07.024
18. Champeaux C, Rousseau P, Devaux B, Nafat F, Tauziede-Espariat A. Solitary fibrous tumours and haemangiopericytoma of the meninges. A retrospective study for outcome and prognostic factor assessment. Neurochirurgie. 2018;64(1):37-43. doi:10.1016/j.neuchi.2017.10.004
19. Boyett D, Kinslow CJ, Bruce SS, et al. Spinal location is prognostic of survival for solitary-fibrous tumor/hemangiopericytoma of the central nervous system. J Neurooncol. 2019;143(3):457-464. doi:10.1007/s11060-019-03177-0