Intracranial Solitary Fibrous Tumor with Concurrent Meningioma: Case Report and Review of the Literature

Tumor fibroso solitário intracraniano com meningioma concorrente: Relato de caso e revisão da literatura

Joaquin Vega Gonzales-Portillo1 Marco Gonzales-Portillo Showing2 Luis A. Huamán Tanta3 Sandro Casavilca Zambrano4

1 Universidad Peruana de Ciencias Aplicadas, Lima, Perú
2 Department of Neurosurgery, Instituto de Neurociencias de Lima, School of Medicine, Universidad Peruana Cayetano Heredia, Clínica San Felipe, Lima, Perú
3 Division of Neurosurgery, Hospital Nacional Dos de Mayo, Lima, Perú
4 Management Unit of the National Tumor Bank, Instituto Nacional de Enfermedades Neoplásicas, Lima, Perú

Address for correspondence Marco Gonzales-Portillo Showing, MD, IFAANS. Instituto de Neurociencias de Lima, Av. Del Pinar 198. Surco 15038. Lima, Perú (e-mail: marcogps@outlook.com).

Abstract

Introduction The present study describes a case of an intracranial solitary fibrous tumor (iSFT) concurrent with meningioma in different anatomical regions. Case Description A female patient, 64-years-old, presented with an 18-month history of progressive vision impairment in the right eye and no other neurological symptoms. The magnetic resonance imaging (MRI) revealed two solid and expansive lesions: one with right interhemispheric occipital location and dependent on the falx cerebri, and another located in the anterior skull base. We opted for a right frontotemporal craniotomy for the first tumor, and a right occipital craniotomy, 41-days later, for the second one, showing no postoperative complications. Histological and immunohistochemical findings confirmed the diagnosis of a grade-I fibrous meningioma and a grade-III SFT. After 9 months of follow-up, the patient showed vision improvement and no signs of neurological compromise or tumor recurrence in the last MRI. Conclusions The present study describes the first reported case of a patient with an intracranial SFT associated with a meningioma in different anatomical locations. The involved pathogenesis and evolution of both coexisting tumors are still unknown, which highlights the need for more case reports on them.

Keywords
► intracranial solitary fibrous tumor
► meningioma
► mesenchymal neoplasm
► case report
► surgical treatment

 recebeu February 17, 2024
aceptou May 16, 2024
ISSN 0103-5355.

© 2024. Sociedade Brasileira de Neurocirurgia. All rights reserved.
This is an open access article published by Thieme under the terms of the Creative Commons Attribution-NonDerivative-NonCommercial-License, permitting copying and reproduction so long as the original work is given appropriate credit. Contents may not be used for commercial purposes, or adapted, remixed, transformed or built upon. (https://creativecommons.org/licenses/by-nc-nd/4.0/)
Thieme Revinter Publicações Ltda., Rua do Matoaso 170, Rio de Janeiro, RJ, CEP 20270-135, Brazil
Introduction

Solitary fibrous tumors (SFTs) are fibroblastic neoplasms with a genomic inversion at the 12q13 locus, leading to the fusion of the genes NGFI-A-binding protein 2 (NAB2) and signal transducer and activator of transcription 6 (STAT6), as well as STAT6-gene nuclear expression.1

These tumors are commonly found in the mediastinum and visceral pleura, and in extra-pleural locations, such as the head and neck, pericardium, peritoneum, thyroid, liver, sinuses, and orbits, in a smaller proportion.1 In the central nervous system (CNS), SFTs are located intracranially, while a smaller proportion are intraspinal.2–4 The cerebellopontine angle, spinal dura, parasagittal area, meninges, and intraventricular region are common sites for this condition.5 Meningeal SFT is a rare form with greater aggressiveness and recurrence, being derived from smooth muscle pericytes surrounding the intraparenchymal microvasculature, also known as Zimmerman pericytes.6,7

Intracranial SFTs (iSFT) account for around 2.5% of all meningeal-based tumors and less than 1% of all intracranial ones, whereas meningiomas represent approximately 20% of primary intracranial tumors.8,9 Furthermore, SFTs are mostly diagnosed at around 50 to 60 years old, with equal gender prevalence.10–12

A recent cohort study of 31 patients diagnosed with iSFT, who underwent surgery from 2008 to 2021, exhibited a 1-year recurrence rate of 6.5% and a 5-year recurrence rate of 22.6%.13 Liu et al. reported that 38 iSFT patients, who underwent surgery from 2008 to 2020, exhibited a 3-, 5-, and 10-year progression-free survival of 82.2, 62.8, and 21.4%, respectively; and a 3-, 5-, and 10-year overall survival of 97.1, 86.9, and 64.2%, respectively.14

In 2016, the world health organization (WHO) granted the combination of both SFT and hemangiopericytoma (HPC) entities into SFT/HPC since these tumors, despite differing in terms of recurrence and aggressiveness, share the same genetic abnormality, a chromosomal inversion in the 12q13 locus, allowing a fusion of the NAB2 and STAT6 genes and, thus, the formation of the fusion gene NAB2-STAT6.15–18 Nonetheless, in the most recent classification published by the WHO in 2021, CNS5, the term SFT/HPC was discarded and SFT was established as the only terminology.19 Currently, the WHO classifies these tumors into three categories: benign, not otherwise specified (NOS) rarely metastasizing, and malignant.20

Meningiomas are the most common benign primary CNS tumor, whereas malignant meningiomas are an uncommon type of primary brain tumor.21 The WHO grading system classifies benign meningiomas with indolent behavior as grade I, whereas those with atypical–to–malignant histology are assigned grades II and III.21 Furthermore, a high Ki-67 proliferation index is associated with an increase in recurrence rate and a decrease in overall survival (OS), regardless of the tumor’s grade.22 The 10-year overall survival of grades I, II, and III are 83.7, 53, and 0%, respectively.17

The present study describes the first reported case of a patient with isSFT associated with a meningioma in different anatomical locations. We show histopathological findings that display the coexistence of a meningioma and a solitary fibrous tumor in different intracranial sites.

Case Report

A 64-year-old, Hispanic woman presented to the clinic with an 18-month history of progressive vision loss in the right eye and, in recent months, a decrease in the temporal field of the right eye. She had medical history of arterial hypertension. The neurological examination revealed a great compromise of visual acuity in the right eye, 0.3 on the Snellen test,
and right temporal hemianopsia. The left eye was normal. The rest of the neurological examination was normal.

A cerebral magnetic resonance imaging (MRI) revealed two expansive lesions: the first was a right interhemispheric occipital solid lesion, dependent on the falx cerebri, with well-defined lobulated borders with intermediate signal on T1 and T2 (►Fig. 1A and B), and intralcal serpiginous voids. The lesion demonstrated intense and heterogeneous enhancement following the intravenous administration of gadolinium (►Fig. 1C, D, and E), measuring 34 × 38 × 38 mm in the transverse, anteroposterior, and craniocaudal direction, suggestive of meningioma of the falx cerebri, causing compression of the cortical sulci of the right occipital lobe.

The second was a solid lesion with smooth, well-defined margins, and homogeneous enhancement following the administration of gadolinium, dependent on the dura mater and anterior floor of the skull base (►Fig. 1C, D, and E), measuring 28 × 34 × 20 mm, occupying the suprasellar cistern, compressing the infundibular stalk and prechiasmatic optic nerves and chiasm, suggestive of meningioma.

**Surgical Management**

The patient underwent a right frontotemporal craniotomy, and complete excision of the sellar tubercular meningioma was achieved.

The postoperative brain computed tomography (CT) scan showed signs of right frontal craniotomy and complete resection of anterior skull base lesion. There were also signs of intraparenchymal hemorrhage in the right parieto-occipital lobe, causing a mass effect on the midline and lateral ventricle (►Fig. 2).

No complications appeared following surgery. The patient was discharged 3 days later.

**Pathological Examination**

The pathologic examination of the first tumor showed a fibrous mass with tendency toward lobularity, compounded by fascicular cells with oval nuclei, with some vacuolated inclusions and occasional psammoma bodies. The mitoses are hard to find, and the immunostainings showed positivity to epithelial membrane antigen (EMA) antibodies and progesterone receptors, with a proliferative index evaluated with a KI 67 of 2%. The glial fibrillary acid protein (GFAP) immunostaining was negative when looking for brain parenchyma trapped, which confirmed the benign characteristics of the tumor, grade I on the WHO classification (►Fig. 3).

The pathologic examination of the second tumor showed a cellular mesenchymal solid tumor with a ramified hemangiopericytoma vascular pattern, compounded by cells with round and ovalated nuclei, with up to seven mitoses per mm², and a fibrous background. The immunostains are

![Fig. 1](https://example.com/fig1.png) The magnetic resonance imaging (MRI) of both brain tumors are isointense on T1 (A) and T2 (B) sequences; T1-weighted MRI exhibiting strong enhancement of the two brain tumors following gadolinium administration (C, D, and E).
positive for CD34 and STAT6 in cytoplasmic and nuclear patterns, respectively. A subpopulation of tumoral cells was positive for CD99, and the proliferative index evaluated with Ki67 was around 10%. Furthermore, the EMA, progesterone receptor, and glial fibrillary acidic protein (GFAP) immunostains were negative. This finding is consistent with the diagnosis of SFT WHO classification grade II (►Fig. 4). The second brain tumor was operated on 41 days after the first surgery. The lesion was approached through a right occipital craniotomy, and complete excision of the tumor was obtained. The postoperative course was uneventful, and the patient was discharged after a few days.

**Follow-up**

After a 9-month follow-up, the patient showed improvement in both visual acuity (Snellen test: 0.7 improvement) and visual fields on the right eye (full recovery) and did not develop any neurological deficits or impairments in life functioning. Additionally, the last brain MRI (►Fig. 5)
showed postsurgical changes in the right frontal lobe and occipital lobe with residual collection, compatible with sequelae encephalomalacia and thin capsule. The fluid-attenuated inversion recovery (FLAIR) and T2 showed no signs of recurrence, no restriction in the diffusion sequence, and no signs of peripheral gliosis in both operated areas. The MRI scan results showed no contrast enhancement (►Fig. 5-A, B).

**Discussion**

The concurrence of tumors could be considered purely coincidental. Besides our study, there are three other case reports of intracranial collision tumors of SFT and meningioma.\(^23\)\(^{-}\)\(^25\) Collision tumor is a lesion in which two histologically different neoplasms coexist in the same location.

Wang et al.\(^4\) report a meningioma component (WHO grade was not mentioned) and a WHO grade I SFT. The case report by Ashisawa et al.\(^23\) describes a WHO grade-I meningioma and a grade III SFT. Furthermore, Binello et al.\(^24\) reports a case with a WHO grade-I meningioma and a grade-II SFT.

Some hypotheses have been proposed to explain the association between different coexisting brain tumors in the same patient. Local tissue irritation from perilesional edema caused by the first tumor has been considered a factor...
that induces astrocyte or arachnoid cell transformation, causing future neoplastic proliferation.26,27 However, this hypothesis does not explain the presence of different tumors in distant places. Most reported cases have presented with common intracranial tumors that were not in juxtaposition.28 Other hypotheses have been proposed as a common genetic pathway, such as disruption of p53 and receptor tyrosine kinase signaling, molecules that have platelet-derived expression growth factor receptors (PDGFRs).29

Other theories have been proposed, stating that there may be unidentified carcinogens serving as stimuli that result in the development of tumors in different tissues, or that residual embryonic structures may instead form the basis of multiple lesions.30,31

The clinical manifestations of iSFTs are highly unspecific and associated with tumor location, with the most common being headaches, epilepsy, weakness of the extremities, paresthesia, visual impairment, anosmia, memory loss, dysphasia, hypotension, amenorrhea, and hypoglycemia.32 Location and histological subtype may influence the evolution and prognosis of patients presenting similar cases. Due to the meningioma’s location, size, and anatomical relationship with the surrounding structures, achieving a gross total resection (GTR) can be challenging. This is particularly true for skull-base meningiomas, in which a radical excision may represent a challenge and sometimes even be detrimental, especially when cranial nerve and vascular structures are involved.33

For an accurate iSFT diagnosis, the WHO has established as indispensable the confirmation of the NAB2/STAT6 fusion gene or the immunohistochemical confirmation of STAT6 protein.34 The STAT6 protein’s detection is regarded as a sufficient marker for routine diagnosis, since it is considered an effective diagnostic tool with a sensitivity of 98% and specificity greater than 85%.35–39 Furthermore, CD34 positivity and EMA negativity in SFT are useful in the differential diagnosis with meningioma, which is CD34 negative and EMA positive.40 However, 5 to 10% of SFTs were negative for CD34.12,41

Progesterone-receptor staining is more common in meningiomas but may be present in iSFT. Likewise, the K-b-67/MIB-1 index exhibits utility as both a risk of recurrence and a tumor activity greater than 85%.

Histologically, SFTs are comprised of atypical spindle cells with an unpatterned architecture, surrounded by dense stromal collagen with collagen bands and, often, a staghorn vascular pattern.25,42 However, although histological characteristics may be suggestive of SFT, they are not exclusive to it, since they can also be observed in other mesenchymal tumors.17,43 Meningiomas and schwannomas can also mimic the histological and radiological forms of SFTs, so it is important to consider differential diagnoses, as these similarities can make it difficult to identify these pathologies.40

The radiological differences between SFTs and meningiomas must be considered. The first generally present lobulated margins and frequent flow-related serpiginous voids, whereas meningiomas feature smooth margins and abundant calcification.44,45 In MRI imaging, iSFT’s unique features include a narrow base of attachment, irregular cross-leaf growth, intratumoral calcification absence, related osseous hyperostosis, bone erosion, and heterogeneous gadolinium contrast enhancement.46,47

Surgery is the gold standard for intracranial SFT treatment, combined with stereotactic and beam radiation therapy for tumor remnants or unresectable recurrences.3 Surgery following surgery, regardless of the resection degree.

Treatment for meningiomas can vary depending on clinical manifestations and tumor size, resulting in two main groups: asymptomatic tumors managed with routine surveillance imaging, and symptomatic or growing tumors managed with surgical resection.50 The goal for surgery in patients with grade-I meningiomas is GTR with routine follow-ups, or subtotal resection (STR) followed by rounds of stereotactic radiotherapy (SRT) or stereotactic radiosurgery (SRS). For grade-II meningiomas, the treatment is GTR with close follow-ups or STR with either SRT or SRS. In contrast, grade-III meningiomas require adjuvant radiotherapy following surgery, regardless of the resection degree.51

**Conclusion**

The SFT is an ultrarare mesenchymal ubiquitous tumor, with an incidence rate < 1 case/million people/year. The diagnosis of iSFTs relies on a comprehensive assessment encompassing clinical manifestations, imaging findings, pathological examination, immunohistochemical analysis, and molecular characteristics.

The present article illustrates a rare case of two different and simultaneous coexisting brain tumors: meningioma and SFT, emphasizing the rarity of primary iSFTs. This is the fourth reported case of meningioma and iSFT coexisting as primary intracranial tumors. Nonetheless, this case differs from the previously mentioned articles both in the meningioma’s histology subtype and tumor location. The current research exhibits a grade-I fibrous meningioma located at the base of the anterior skull, and a grade-II SFT with a right interhemispheric occipital location and dependent on the falx cerebri. The pathogenesis and evolution involved in the two coexisting tumors remain unclear, which highlights the necessity of more case reports about them.

**References**


WHO Classification of Tumours. Soft Tissue and Bone Tumours. 5th ed. Volume 3. IARC Press, Lyon, France 2020


Spallone A, Santoro A, Palatinsky E, Giunta F. Intracranial meningiomas associated with glial tumours: a review based on 54 selected literature cases from the literature and 3 additional personal cases. Acta Neurochir (Wien) 1991;110(3–4):133–139

Russell DS, Rubinstein LJ. Pathology of Tumors of the Nervous System. 5th edition Edward Arnold, London 1989

Myerson PG. Multiple tumors of the brain of diverse origin. J Neuropathol Exp Neurol 1942;1:406415


Intracranial Solitary Fibrous Tumor with Concurrent Meningioma

Gonzales-Portillo et al.