

A solitary fibrous tumor with concurrent meningioma at the same site: A case report and review of the literature

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Abstract. The present study describes a case of a solitary fibrous tumor (SFT) concurrent with meningioma in the same anatomical region. The patient was admitted to Tianjin Huanhu Hospital (Tianjin, China) presenting with progressive eyesight impairment, dizziness and right hemiparesis. Cranial magnetic resonance imaging revealed two primary tumors co-occurring at the same site. One lesion was a solid lesion located in the left frontal convex with homogeneous enhancement, and was closely associated with the dura mater; thus, it was suspected that the lesion was a meningioma. The second lesion was cystic and solid with an irregular shape, and was located next to the first tumor; this lesion was believed to be a hemangiopericytoma or astrocytoma. The patient underwent a left temporoparietal craniectomy and a complete excision of the two tumors was achieved. Subsequent pathological examination of the resected tissues confirmed that the two tumors were a secretory meningioma and a SFT, respectively. Immunohistochemistry is important in differentiating SFTs from other tumors. Currently, a total tumor resection is the optimal treatment strategy when managing these rare lesions, often with no requirement for adjuvant post-operative therapy; however, long-term follow-up is essential to detect any signs of recurrence. The possibility of multiple tumors should be taken into consideration when performing clinical examination. To further understand the mechanisms underlying the occurrence of multiple intracranial tumors, further research is required, alongside an increased number of case reports.

Introduction

Meningioma is a common brain tumor accounting for ~20% of all primary intracranial neoplasms (1), while schwannoma is a

type of nerve sheath tumor. Both meningiomas and schwannomas may imitate intracranial solitary fibrous tumors (SFTs) histologically and radiologically. SFTs are spindle-cell mesenchymal neoplasms. Concurrence of intracranial SFTs and other tumor types is particularly rare, and SFTs are easily misdiagnosed due to a lack of typical symptoms and imaging features. By contrast, meningiomas, which arise from cells covering the arachnoid layer of the dura mater or from the intraventricular choroid plexus, present with a typical dural tail sign upon magnetic resonance imaging (MRI) (2,3). Histologically, both meningiomas and SFTs are composed of interlacing fascicles of spindle or ovoid tumor cells with intervening collagen bands. Surgery is the first choice of therapy for SFTs, with a good prognosis. In particular, stereotactic and external beam radiation therapy may be recommended for postsurgical tumor remnants and for unresectable recurrences (4). Analysis of the literature identified ~220 cases of SFTs, of which the majority were intracranial. In decreasing frequency, intracranial tumors involved the supratentorial and infratentorial compartments, the pontocerebellar angle, the sellar and parasellar regions, and the cranial nerves (4). The current study describes the case of a patient who presented with two primary intracranial tumors that originated from different cell types. The case report is followed by a discussion of the pathogenesis of multiple intracranial tumors and a brief literature review. Written informed consent was obtained from the patient.

Case report

A 71-year-old woman was admitted to the Tianjin Huanhu Hospital (Tianjin, China) on September 7, 2012. The patient presented with progressive eyesight impairment, dizziness and right hemiparesis. Routine biochemical and hematological tests were within normal limits.

MRI (MAGNETOM Trio, A Tim System 3 Tesla; Siemens AG, Munich, Germany) revealed two primary tumors that were in close proximity (Fig. 1). The first was a solid lesion, measuring 20x16x14 mm in size, with a clear boundary and visible peritumoral edema. The tumor had originated from the left frontal convex and was adhered to the dura mater, connecting to the adjacent skull with a wide base, with associated bone hyperplasia. The lesion was isointense to the brain parenchyma on T1- and T2-weighted images (Fig. 1A and B). The tumor demonstrated intense and homogeneous enhancement following the intravenous administration of gadolinium

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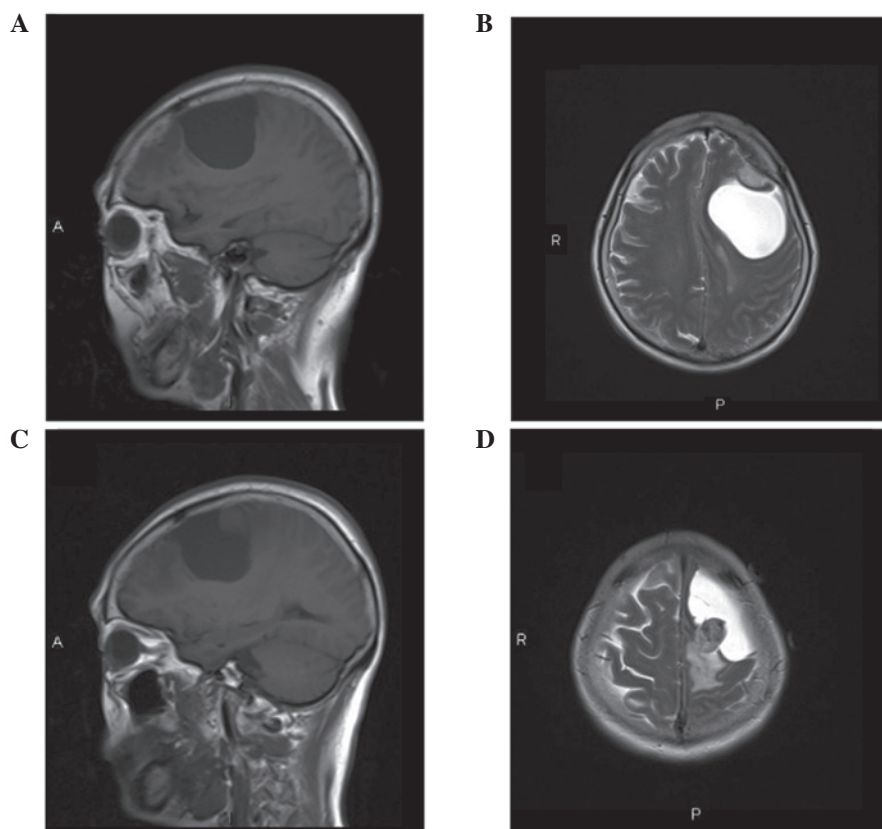


Figure 1. Magnetic resonance images. The first tumor exhibited isointensity to brain parenchyma on (A) T1-weighted sagittal and (B) T2-weighted axial scans. The second tumor was located next to the first lesion and consisted of solid and cystic regions; the cystic region of the mass revealed (C) hypointensity on T1-weighted images and (D) hyperintensity on T2-weighted images. The solid region of the mass exhibited (C) isointensity to the adjacent brain tissue on T1-weighted images and (D) iso- or hyperintensity on T2-weighted images.

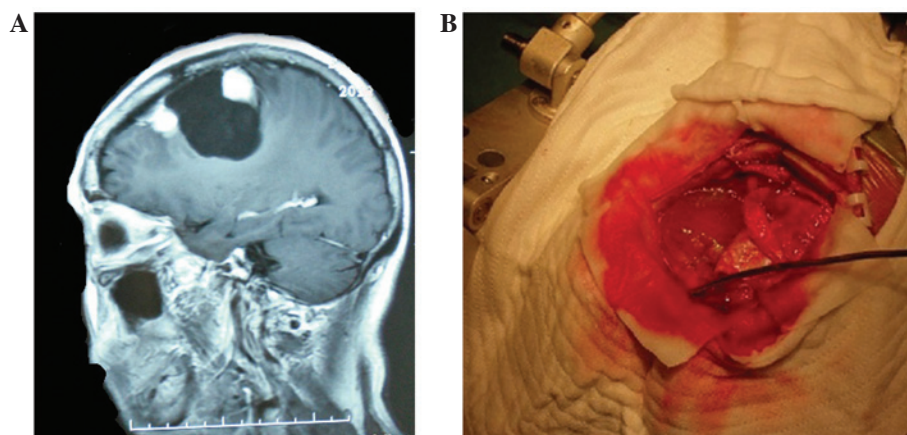


Figure 2. An MRI scan of each lesion and intraoperative image of the meningioma. (A) T1-weighted sagittal MRI exhibiting strongly-enhanced, solid regions of the two tumors following gadolinium administration. (B) Intradural exploration showing the cystic region of the second tumor containing yellow liquid. MRI, magnetic resonance imaging.

(Fig. 2A). The radiological and clinical features were highly indicative of a meningioma. The second lesion was located in close proximity to the first lesion, and was cystic and solid with an irregular shape, measuring 45x46x66 mm in size. The cystic region of the mass exhibited hypointensity on T1-weighted images and hyperintensity on T2-weighted images. The solid region of the mass exhibited isointensity to adjacent brain tissue on T1-weighted images and iso- or hyperintensity on T2-weighted images (Fig. 1C and D). In

addition, the mass demonstrated intense and homogeneous enhancement following the intravenous administration of gadolinium (Fig. 2A). The clinical features were suggestive of a hemangiopericytoma or astrocytoma.

The patient underwent a left temporoparietal craniectomy, and complete excision of each tumor was achieved. A well-defined, 20x16x14 mm, solid tumor, which was located in the left frontal convex, was extirpated along with the attached dura mater. Following excision, the tumors were placed in

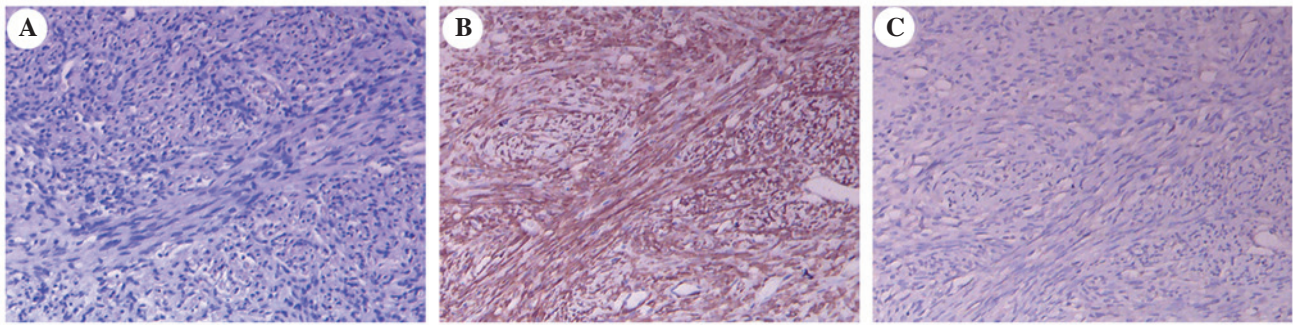


Figure 3. (A) Histological examination determined that the solitary fibrous tumor was composed of proliferating spindle cells (staining, hematoxylin and eosin). Immunohistochemistry demonstrated that the tumor tissue was (B) positive for B-cell lymphoma 2 (magnification, x100) and (C) negative for epithelial membrane antigen (magnification, x100).

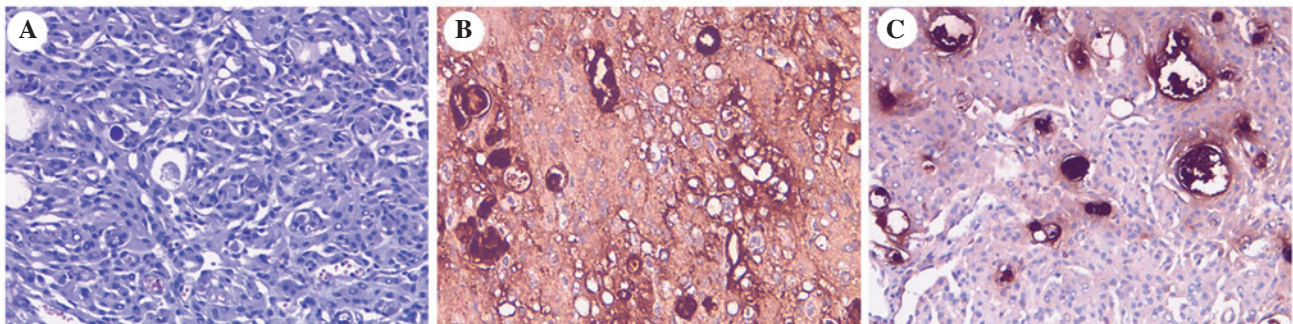


Figure 4. (A) Histological examination revealed evidence of multifocal epithelial cell differentiation and an intraepithelial microcavity containing eosinophil pseudopsammoma bodies. Immunohistochemistry demonstrated that the tumor tissue was positive for (B) epithelial membrane antigen (magnification, x200) and (C) carcinoembryonic antigen (magnification, x100).

normal saline and sent to the Department of Pathology in Tianjin Huanhu Hospital for pathological, histological and immunohistochemical analysis. Pathological examination confirmed that this mass was a secretory meningioma. The second solid mass was encapsulated, contained yellow cystic liquid and was located in close proximity to the meningioma (Fig. 2B). This lesion was located in a capsule wall, measured 45x46x66 mm in size and was separated from the dura mater. Pathological examination confirmed a diagnosis of an SFT.

Following histological analysis of the specimens, it was noted that the SFT was composed of proliferating spindle cells (Fig. 3A). Immunohistochemistry determined that the SFT cells were positive for cluster of differentiation (CD)34, vimentin, B-cell lymphoma 2 (Bcl-2) (Fig. 3B) and CD117, and negative for epithelial membrane antigen (EMA) (Fig. 3C) and S-100, with a Ki-67 proliferation labeling index of ~2.5%. Histological examination of the secretory meningioma demonstrated evidence of multifocal epithelial cell differentiation and an intraepithelial microcavity containing eosinophil pseudopsammoma bodies (Fig. 4A). Immunohistochemistry determined that the secretory meningioma cells were positive for EMA (Fig. 4B), vimentin and carcinoembryonic antigen (Fig. 4C), with a Ki-67 proliferation labeling index of ~2.3%. Periodic acid-Schiff staining was positive. No complications appeared following surgery. The patient was followed-up at 4 and 8 months and every 12 months subsequent to surgery. At the 8-month follow-up, there were no signs of recurrence.

Discussion

Multiple primary intracranial neoplasms were first described in 1938 (5), and since then, an increasing number of cases have been reported. However, the majority of cases report the incidence of common intracranial tumors, including glioma and meningioma (6). The current study introduces a case that presented with the co-occurrence of mixed intracranial tumors. The tumors consisted of a secretory meningioma, a relatively uncommon subtype of meningioma, and an intracranial SFT, which is extremely rare. To the best of our knowledge, this is the first case of its type to be reported in the literature.

Although various theories have been proposed to explain the occurrence of multiple primary intracranial neoplasms of diverse germinal origins in the same individual, none of these have yet been proven. The concurrence of the tumors could be considered as purely coincidental. The majority of reported cases have presented with common intracranial tumors that were not in a close juxtaposition (7). If one tumor is close to or intermixed with another, there may be an association between them. The present study proposes that an initial tumor may form and function as an irritating agent, subsequently inducing and stimulating the excessive growth of a second lesion (8). It is generally considered that the relatively slow growth of benign stimulation induced the malignant tumor. With regard to the current case, it was hypothesized that the meningioma functioned as a stimulus source, which subsequently induced the SFT.

Other theories have been proposed stating that there may be certain unidentified carcinogens serving as stimuli, which result in the development of tumors in different tissues (9), or that residual embryonic structures may instead form the basis of multiple lesions (10).

It has also been hypothesized that common genes may be implicated in the development and progression of concurrent tumors. According to Black *et al* (11), deletion of chromosome 22 in patients with type 2 neurofibromatosis, and in up to 50% of solitary meningiomas, is associated with the appearance of multiple meningiomas (11). Previously, a meningioma-associated tumor suppressor gene was identified on the long arm of chromosome 14, determined as N-myc downstream-regulated gene 2, which was commonly inactivated in clinically aggressive meningiomas (12). However, only 1 case of an SFT of the central nervous system (CNS) has been detected by DNA analysis and flow cytometry, and 2 cases have been detected by molecular analyses (4). Therefore, further research is required to draw reliable conclusions.

Currently, no etiological association has been identified between meningiomas and SFTs. A review of the literature demonstrated that there have been no cases reported that are similar to the present case. The theory of stimulation may account for this pattern of tumoral linkage, but an increased number of similar cases in the future may enable identification of a potential association between such tumors.

With regard to the present case, a pre-operative diagnosis was challenging. According to the clinical and imaging features alone, the lesions were diagnosed as meningioma and hemangiopericytoma or astrocytoma. As the diagnosis of SFT proved to be difficult, it is necessary to include a brief literature review for intracranial SFT in the present study.

An SFT is a rare, mesenchymal neoplasm, which was first described as a pleural lesion by Klemperer and Rabin in 1931 (13). SFTs of the meninges were originally described by Carneiro *et al* in 1996 (14). The origin of SFTs has been a subject of controversy; they are typically dura-based, but may also present as intraventricular masses arising from cranial nerves or ubiquitous CD34-positive, dendritic, fibroblastic cells, which do not have an apparent association with the meninges (15,16). The World Health Organization classification of tumors of the CNS states that mesenchymal, non-meningothelial tumors originate from submesothelial, mesenchymal, fibroblast-like cells as opposed to developing from the mesothelium itself (17). The spine and posterior fossa are the most frequent locations for SFTs to develop (18). These tumors primarily occur following the third decade of life, with patient ages ranging from 33-75 years (19), and demonstrate a slight female preference, with a male to female ratio of 1:1.5 (19,20).

There are no reliable neuroradiological signs of an SFT, therefore, the pre-operative diagnosis is challenging. SFTs are generally isointense on T1-weighted MRI and hyperintense on T2-weighted MRI. Cystic lesions commonly exhibit peripheral enhancement (21). In the present case, the SFT appeared isointense to adjacent brain tissue on T1-weighted MRI and iso- or hyperintense on T2-weighted images. Following intravenous contrast administration, the tumor exhibited homogeneous enhancement.

In the current case, radiological evaluation could not provide an accurate diagnosis, and detailed histopathological

and immunohistochemical examinations were required. Histologically, SFTs are composed of interlacing fascicles of spindle to ovoid tumor cells, with intervening bands of collagen (21). Immunohistochemically, the tumor cells demonstrate strong positivity for CD34, vimentin and the antiapoptotic marker Bcl-2, and are typically negative for EMA and S-100 protein. By contrast, meningiomas are usually positive for EMA and negative for CD34 (22). In the present case, the immunohistochemical findings were consistent with the features of SFTs.

Regarding the treatment of SFTs, surgery is the preferred choice of management. The tumors are typically well-circumscribed and therefore amenable to gross total resection. Radiotherapy, including external beam radiation therapy or gamma-knife radiosurgery, is administered in cases that experience incomplete (partial or subtotal) resection, or in certain cases with malignant histology or recurrence (23). If the proliferation rate is high, the chemotherapeutic agent, toremifene, may also be administered (23). In the present case, the tumor was totally resected and no further treatment was required.

Due to the limited available data, the clinical behavior of these tumors is unpredictable. Although the majority of SFTs behave in a benign manner, recurrence, cerebrospinal fluid dissemination and malignant variants with distal metastasis have been reported (24). With regard to recurrence, the Ki-67/MIB-1 labeling index (>5%) is a useful marker of the risk of recurrence and tumor grade in the prognostication of SFTs of the CNS. Although the Ki-67 proliferation labeling index was particularly low (~2.5%) in the present case, long-term follow-up is essential to detect any signs of recurrence.

In conclusion, to the best of our knowledge, the current case is the first of its type to report of an SFT with concurrent meningioma. Despite SFT being rare, it should be considered in the neuroimaging differential diagnosis. Immunohistochemical examination is particularly important in aiding the differentiation between SFT and the more prevalent meningioma and schwannoma, which may imitate SFT histologically and radiologically. Surgical removal is considered as the optimal therapeutic strategy in managing this rare entity. As such lesions typically exhibit benign histological behavior, generally no adjuvant post-operative therapy is required; however, long-term follow-up is essential to detect any signs of possible recurrence. The possibility of the coexistence of multiple tumors at two sites should be taken into consideration. In order to understand the mechanisms underlying the development of multiple intracranial tumors, further research and a greater number of case studies are required.

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