

Unusual Presentations of Intracranial Solitary Fibrous Tumor with Malignant Transformation

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Abstract

Background: Intracranial solitary fibrous tumor was rare. However, most intracranial solitary fibrous tumors are benign, cerebral solitary fibrous tumor with malignant transformation was even more unusual.

Case present: Herein, we presented a 55 year-old-male who was diagnosed intracranial solitary fibrous tumor five years ago. Partial tumor excision was performed via the pterional craniotomy. Unfortunately, one year later, cerebral solitary fibrous tumor with recurrence on latest operation was diagnosed via follow-up Magnetic Resonance imaging. Reopen the craniotomy site, subtotal removal of tumor and stereotactic radiation surgery were performed. However, four years later, a recurrent tumor was seen via follow-up computed topographic (CT) scans of brain with contrast. A third craniotomy with subtotal removal of tumor was performed. Pathological examination revealed solitary fibrous tumor with malignant transformation. Postoperative CyberKnife stereotactic radiosurgery was performed. One year after resection and CyberKnife stereotactic radiosurgery, a follow-up brain MR images showed tumor regression.

Conclusion: Most of intracranial solitary fibrous tumors were benign lesions; however, physician should always keep in mind that malignant transformation may be found in intracranial solitary fibrous tumor.

Keywords: Intracranial solitary fibrous tumor; Malignant transformation

Introduction

Solitary fibrous tumors (SFTs) were initially described by Klemperer and Rabin [1] in 1931 as a focal pleural mass. Since then, SFTs have been detected in different extracranial sites, including the pericardium, nasopharyngeal sinuses, liver, prostate, thyroid, mesentery, urinary bladder, mediastinum, orbit, and nervous system. In the central nervous system, they usually present as dura-based masses; however, recently clinicians come to accept SFTs can present as primary central nervous system lesions [2]. Most of solitary fibrous tumors are benign, and a gross total resection is curative in most cases [3]. However, more aggressive variants have been associated with higher rates of recurrence and metastasis [4]. Herein, we presented a 55 year-old-male who was diagnosed intracranial solitary fibrous tumor five years ago. The patient received two operations of tumor resection and one stereotactic radiation surgery in past five years. Unfortunately, solitary fibrous tumor with malignant transformation was found on latest operation. However, Miettinen et al. show malignant transformation of the solitary fibrous tumor may be associated with trisomy 8 [5].

Case Present

Five years ago, a previously healthy 55 year-old-male sustained severely unstable gait and personal change for six months. The patient called at local clinic and took medicine, but the symptoms persisted. So, the patient called at our outpatient department. On neurological examination revealed no abnormal finding except to mild weakness of right limbs and personal change. Magnetic resonance (MR) imaging of brain with contrast showed an enhancing tumor at the left anterior medial parasellar region. Chest radiography revealed no significant abnormality, and laboratory data including blood cell count and blood chemistry, and some tumor markers were within normal ranges. Partial tumor excision was performed via the pterional craniotomy due to tumor invade into speech area. The pathological report revealed cerebral solitary fibrous tumor (Figure 1).

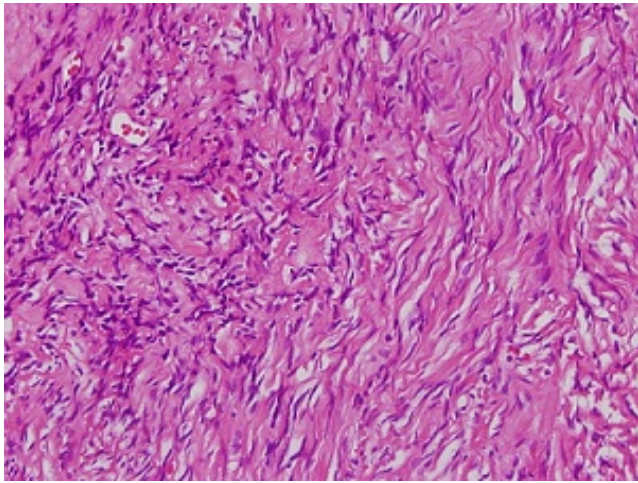


Figure 1: Photomicrograph illustrates fascicular arrangement of spindle-shaped tumor cells with attenuated intercellular collagen.

Unfortunately, one year later, follow-up MR imaging of brain with contrast presented with a recurrent enhancing tumor at the left anterior medial parasellar region (Figure 2).

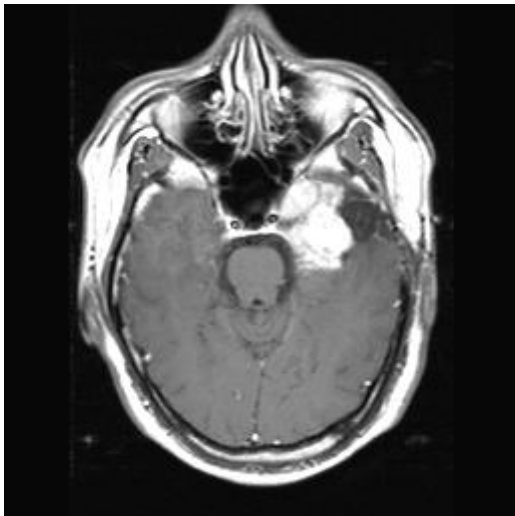


Figure 2: Magnetic resonance (MR) images of brain with contrast presented with an enhancing tumor at the left anterior medial parasellar region.

Then the patient received operation to reopen the craniotomy site and subtotal removal of tumor. Stereotactic radiation surgery (1000 cGy to 90% isodose volume) was performed one month later. However, four years later, computed topographic (CT) scans of brain with contrast revealed a recurrent tumor over left anterior temporal region (Figure 3).



Figure 3: Computed topographic (CT) scans of brain with contrast revealed an enhancing tumor over left anterior temporal fossa.

A third craniotomy with subtotal removal of tumor was carried out. Pathologic examination revealed solitary fibrous tissue with malignant transformation characterized by a spindle cell tumor with reticulin and areas of hypercellularity, large and pleomorphic tumor nuclei, and frequent mitotic figures (Figure 4a).

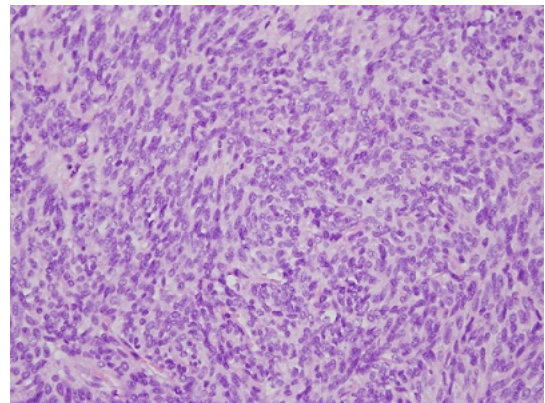


Figure 4(a): Photomicrograph illustrates fascicular arrangement of spindle-shaped tumor cells with attenuated intercellular collagen, areas of hypercellularity, large and pleomorphic tumor nuclei, and frequent mitotic figures (hematoxylin-eosin, 400X).

The specimen stained strongly and diffusely for CD34 (Figure 4b), but did not stain for epithelial membrane antigen, glial fibrillary acid protein, actin and S-100.

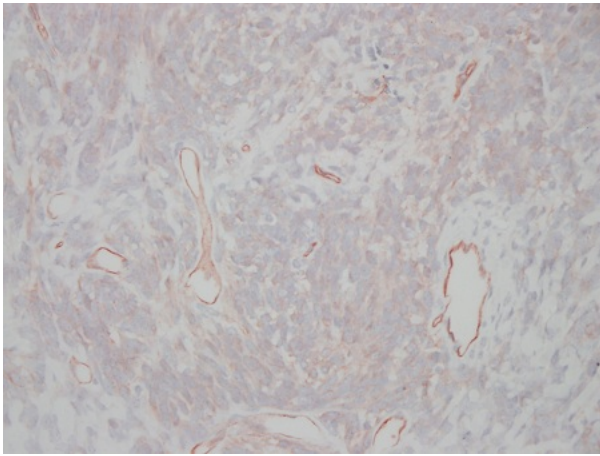


Figure 4(b): Photomicrograph illustrates CD34 immunopositivity in the tumor, which is compatible with a diagnosis of solitary fibrous tumor (400X).

Postoperative CyberKnife stereotactic radiosurgery was performed. One year after resection and CyberKnife stereotactic radiosurgery, a follow-up brain MR images showed tumor regression (Figure 5).

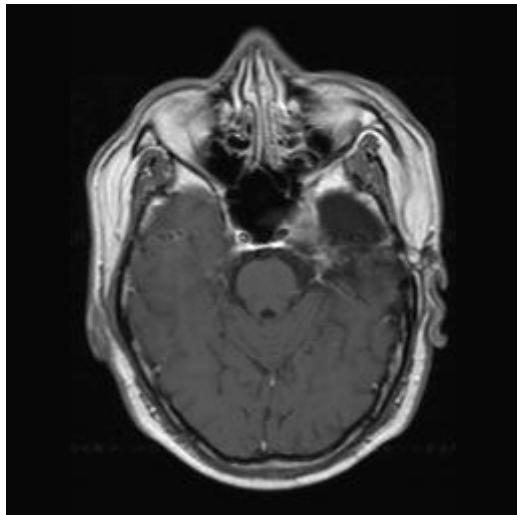


Figure 5: Magnetic resonance (MR) images of brain with contrast presented with a reduced tumor at the left anterior medial parasellar region.

Discussion

Intracranial SFTs are rare neoplasms but have well characterized in the pathological literature. Caroli E et al. [6] first reported intracranial SFT as a lesion distinct from fibrous meningioma in 1996. They usually present as dura-based masses and resemble meningiomas clinically and radiologically. These tumors can occur at any age and in most locations, regardless of proximity to the meninges, but the mesenchyme of the cerebral vasculature [7]. Most solitary fibrous tumors are benign, and a gross total resection is curative in most cases.

However, more aggressive variants (so-called ‘malignant solitary fibrous tumors’) have been associated with higher rates of recurrence and metastasis. We have presented here a case of meningeal SFT with malignant transformation, which locally relapsed two times over a period of five years.

SFT appears as a relatively well circumscribed, partially calcified heterogeneous mass on non-enhanced CT scan, but it usually demonstrates variable degrees of enhancement with intravenous contrast administration [7]. On MR imaging, the SFT is isointense with normal brain parenchyma on T1-weighted images, and hyperintense on T2-weighted images. However, it shows intense and homogeneous enhancement after intravenous administration of gadolinium [8]. MR spectroscopy shows high peaks of lipid and lactate, but normal choline and creatine peaks with unaltered ratio. [7] The imaging differential diagnoses of intracranial SFT include fibroblastic meningioma, meningeal hemangiopericytoma, neurofibroma, and schwannoma [9].

Histological examination usually disclosed cellular areas of spindle cells, thick collagen bundles with a hyaline appearance and staghorn-like vascular channels. Immunohistological analysis revealed tumor cells to be diffusely positive for vimentin and CD34 and completely negative for keratin and EMA. However, histological features associated with local or distant recurrence of extrathoracic SFTs are reported to include high cellularity, mitotic activity (>4/10 HPF), nuclear pleomorphism, and necrosis [4]. The results of a recent karyotyping study showing trisomy 8 to be associated with malignant behavior in the SFT of the pleura [10] might provide a new solution to this issue. However, intracranial SFT cases with malignant transformation are very low in number, and the follow-up duration has not been sufficiently long in most cases. Therefore, further clinicopathological analyses are required for clarification.

On histological examination, SFT can be similar to hemangiopericytoma or fibrous meningioma. On haematoxylin and eosin staining, meningeal solitary fibrous tumours usually have more prominent collagen deposition and are slightly less cellular than haemangiopericytomas. Hemangiopericytoma show weak positivity for CD34, rarely for EMA, and is negative for S-100 [10]. Moreover, fibrous meningioma characteristically express EMA and S-100 protein; CD34 reactivity is patchy and weak [11].

In conclusion, we have experienced a case of SFT derived from the meninges with malignant transformation. Regarding meningeal SFTs in the previous literature, there was no report of malignant transformation of a benign meningeal SFTs. De leval et al. reported malignant transformation of the solitary fibrous tumor of the pleural, they assumed the malignant transformation may be associated with trisomy 8 [11]. Although, SFTs with malignant transformation are very rare, neurosurgeons and neuropathologist should keep in mind.

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