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Solitary fibrous tumor of the pineal region with delayed ectopic intracranial metastasis: A case report and review of the literature

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Abstract

Rationale: Solitary fibrous tumors of central nervous system are rare spindle-cell mesenchymal tumors. Although most are benign in nature, malignant transformation and extracranial metastasis have been reported. Up to now, only one case of CSF dissemination was described. Here we described an extremely rare case of intracranial Solitary fibrous tumors arising from the pineal region with a delayed ectopic metastasis.

Patient concerns: A 35-year-old female presented with double vision, memory disturbance and unsteady gait was referred to our center. MRI showed an irregular mass in the pineal region.

Diagnoses: The patient was diagnosed as pineal tumor, with unknown pathology.

Interventions: Gross total resection was achieved and the pathologic studies confirmed a solitary fibrous tumor. Thirty-nine months later local recurrence occurred and gamma-knife radiotherapy was offered. Seven months later, MRI found a metastasis in the left temporal lobe. Surgical resection was conducted and pathological analysis revealed changes in cell morphology, counts and Ki-67 level, confirmed the diagnosis of solitary fibrous tumor/hemangiopericytoma (WHO Grade III). The patient received post-operational radiotherapy.

Outcomes: The patient was followed up for 7 months with no signs of recurrence.

Lessons: Here, we report an extremely rare case of primary solitary fibrous tumor of pineal region with delayed intracranial ectopic metastasis, together with literature review of metastatic solitary fibrous tumors. Strict surveillance is strongly recommended, considering the malignant potential of this seemingly benign disease entity. Complete resection of the tumor is the treatment of first choice and radiotherapy might be an effective adjuvant therapy for high grade SFT/HPCs.

Abbreviations: Bcl-2 = B-cell lymphoma 2, CNS = central nervous system, EMA = epithelial membrane antigen, HPC = hemangiopericytoma, HPF = high power field, MRI = magnetic resonance imaging, SFT = solitary fibrous tumor.

Keywords: central nervous system, recurrence, solitary fibrous tumor/hemangiopericytoma

1. Introduction

Solitary fibrosis tumors (SFTs) are rare spindle-cell mesenchymal tumors, which were first reported by Klemperer in 1931.[1] It is most commonly found in the visceral pleura, but also has been reported in extrapleural sites such as thoracic wall, mediastinum, abdominal cavity and deep soft tissue of extremities. Rarely, SFTs occur in the central nervous system (CNS).[2,3] As reported, the majority of CNS SFTs are dura-based masses, originating from parasagittal region, intraventricular region and vertebral canal.[4] Although it has been known that more than 90% of the intracranial SFTs are benign and seldom metastasize, cases of delayed extracranial metastases to lungs, liver, pancreas and bone, have been reported.[5–7] Intracranial metastasis of primary CNS SFTs is extremely rare and only one case of CSF dissemination was reported.[8] Here, we described an interesting case of intracranial SFT arising from the pineal region with a delayed ectopic metastasis at the left temporal lobe after gross total resection, together with a literature review of metastatic intracranial SFTs.

2. Case report

A 35-year-old woman suffering from progressive headache with double vision, memory impairment and unsteady gait for 1

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YW and JZ contributed equally to this work and should be considered co-authors.

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The authors declare that they have no conflict of interest.

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month was admitted to our hospital on July 29, 2011. Magnetic resonance imaging (MRI) revealed a solid lesion with cystic component in the pineal region with tentorium attachment, measuring 52mm×35mm×32mm in size, exhibiting hypo- and iso-intensity on both T1 and T2-weighted images, and greatly enhanced with contrast (Fig. 1A–C). These radiological findings were highly indicative of meningioma. Her past medical history was unremarkable. Vital signs were stable. Upon neurological examination, the patient walked slowly despite intact muscle strength, and had difficult with up-gazing.

Surgical resection was performed in prone position via right occipital transtentorial approach. The tumor appeared as a grayish white, firm, well-defined and highly vascularized mass. Gross total resection was achieved and confirmed by postoperational MRI (Fig. 1D-E). Under microscope the tumor was a mixture of hypercellular and hypocellular areas containing short spindle-shaped cells separated by thin bands of collagen and staghorn-like vessels (Fig. 1F). Immunohistochemically, the tumor cells displayed strong immunoreactivity with vimentin, CD99 and B-cell lymphoma 2 (Bcl-2), and a 15% positive rate of Ki-67, while negative with epithelial membrane antigen (EMA) and CD34 (Data not shown). Diagnosis of a pineal SFT was confirmed based on pathological analysis.

Thirty-nine months after the first surgery, a routine follow-up MRI indicated local tumor recurrence (Fig. 2A-B). The patient was symptom-free and was treated with gamma-knife radiotherapy. Seven months later, the patient returned to the clinics complaining of an intense and persistent headache for 10 days. MRI revealed a new lesion in the left temporal lobe, displaying mixed intensity on both T1 and T2-weighted images with strong inhomogeneous enhancement (Fig. 2C-D). Craniotomy and total resection were conducted (Fig. 2E-F). Postoperational histological analyses revealed similar morphological characteristics with the previous pineal mass, except that the tumor cells were more of an ovale to round shape and the mitosis counts were more than 50/10 high power field (HPF) (Fig. 2G). Tumor cells expressed weak CD34 and S-100, diffusive vimentin, variable levels of EMA, whereas no immunoreactivity with CD99 (Fig. 2H-I). The Ki-67 index, however, increased to 20% to 40% (Fig. 2J). According to 2016 WHO classification system of CNS tumors, a final diagnosis of solitary fibrous tumor/hemangiopericytoma (WHO Grade III) was made. The patient recovered uneventfully. Considering the increased malignancy of the recurrent tumor, traditional radiotherapy was offered targeting both the primary and metastasis sites. The patient was followed up routinely for 7 months with no signs of recurrence.

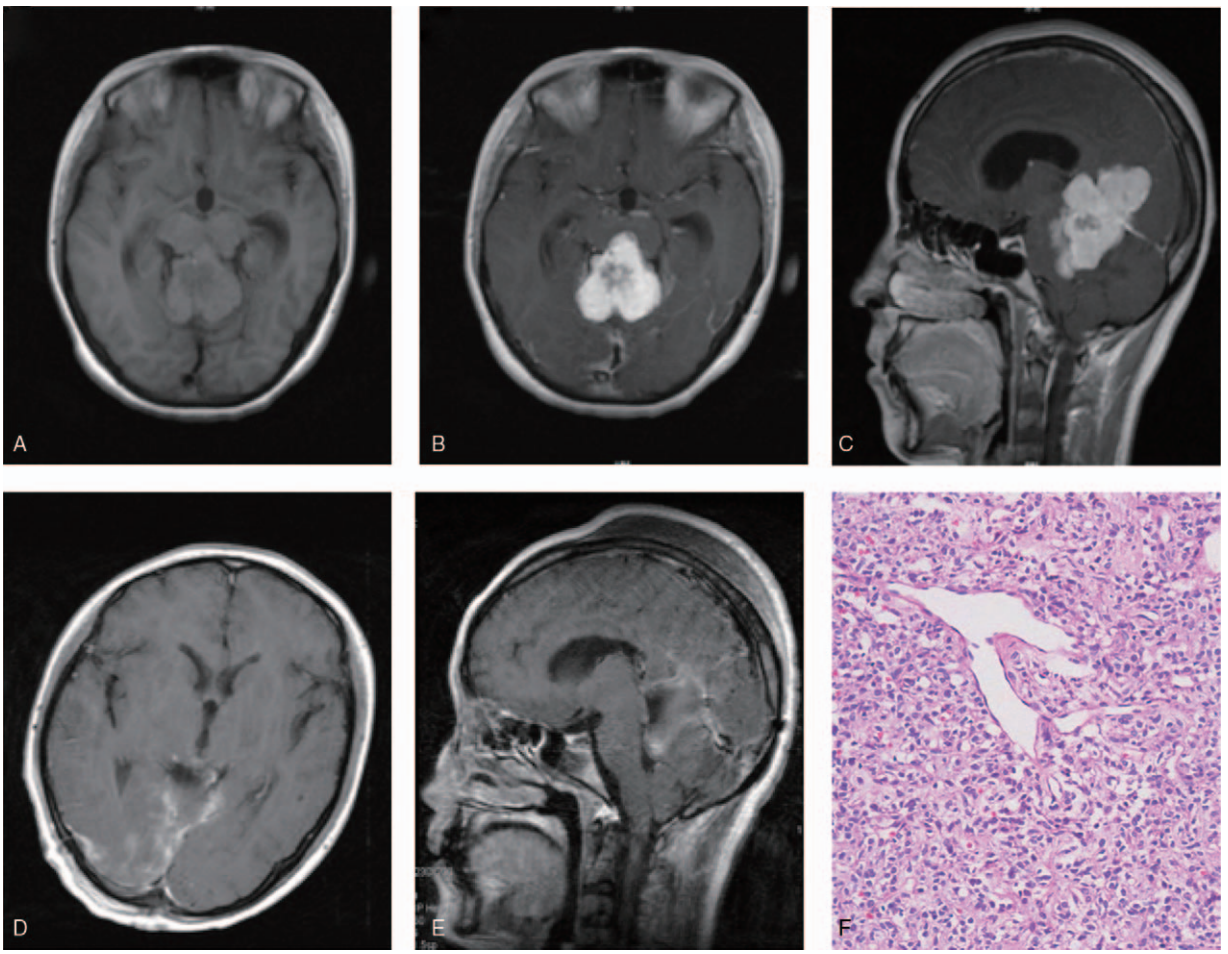

Figure 1. MRI scanning and pathological analysis of the solitary fibrous tumor in pineal region. (A–C) preoperative MRI. The tumor was a well-circumscribed mass, exhibiting hypo- to iso-intensity on T1 images (A). Axial (B) and sagittal (C) T1-weighted images with contrast showed a strong enhancement of the lesion with tentorium attachment. Postoperative axial (D) and sagittal (E) MRI images indicated gross total resection. (F) the tumor was a mixture of hypercellular and hypocellular areas containing short spindle-shaped cells separated by thin bands of collagen and staghorn-like vessels. (H&E, 100×).

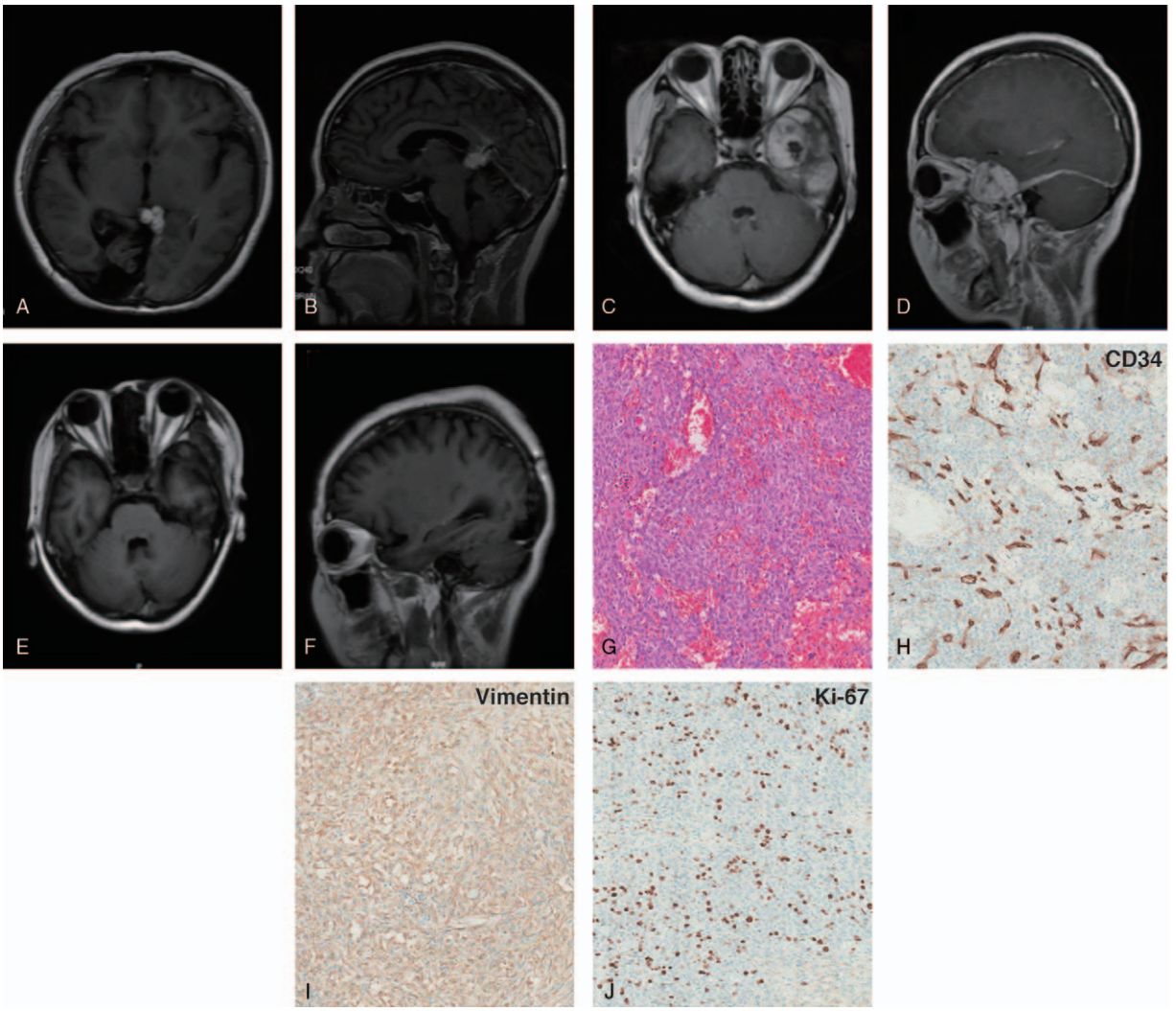

Figure 2. MRI scanning and pathological analysis of the in situ recurrent and ectopic metastatic masses. (A–D) preoperative MRI of the recurrent and metastatic masses. Axial (A) and sagittal (B) T1-weighted image with contrast showed a new lesion with strong and homogeneous enhancement in the pineal region. Axial (C) and sagittal (D) T1-weighted image with contrast revealed a new lesion in the left temporal lobe, exhibiting strong and heterogeneous enhancement. Postoperative axial (E) and sagittal (F) MRI images confirmed gross total resection of the temporal lobe mass. (G–J) Immunohistochemical features of the ectopic metastatic tumor. (G) The tumor was composed of short-to-ovale shaped cells with micro-hemorrhage and hypercellularity. (H&E, 100×) Immunohistochemically, tumor cells show weak reactivity for CD34 (H) (100×), strong diffuse reactivity for vimentin (I) (100×), and increased Ki-67 index (J, 40%) (100×) compared to the primary SFT in pineal region (not shown).

3. Discussion

The CNS SFTs are rare spindle-cell tumors and were first described as a distinct entity by Carneiro et al in 1996.[9] Till now, only about 200 cases have been reported.[10] They often arise from extra-axial space, with a dura-based attachment, and mostly involved the tentorium, posterior fossa, ventricular system and spine.[11,12] The SFT involving the pineal region, as described in our case, is extremely rare and only 4 cases have been reported. Zhang et al proposed that the SFT in this area might originate from the velum interpositum of the third ventricle.[13] SFTs in the pineal region have to be differentiated from pineal parenchymal tumor, germ cell tumor, astrocytoma and meningioma. The case we encountered, however, is confused with meningioma due to the obvious dural attachment.

Radiologically, CNS SFTs exhibit unspecifically as hypo- or hyper-intensity on T1 and T2WI images, with diffuse or heterogeneous enhancement after gadolinium injection, and when displaying dural-tail sign, they can be easily misdiagnosed as meningioma.[12,14] Therefore, definite diagnosis mainly depends on the pathological analysis. Classically, SFTs are distinguished by the presence of fascicles of elongated cells with staghorn vessels that alternate with paucicellular collagen-rich tissue.[15] (Solitary fibrous tumors of the central nervous system: clinicopathological and therapeutic considerations of 18 cases) On immunohistochemistry, SFTs are characterized by diffusive staining of CD34, vimentin, CD99 and Bcl-2, while always negative for S-100 and EMA expression, which can be easily distinguished from meningioma.[2,16,17] Negative CD34 as in our case, however, has been reported and considered as a potential indicator of poor prognosis.[5,12,18,19]

Historically, CNS SFTs have to be differentiated from hemangiopericytomas (HPCs), both are thought to originate from mesenchymal cells with pericytic differentiation.[20] HPCs

Table 1

Literature review of metastatic CNS SFTs and WHO I SFT/HPC.

Author	Year	Age/Gender	Location	GTR	Recurrence	Radiotherapy	Metastasis	Site
Kamamoto[26]	2018	62/female	transverse sinus	NK	3.57y	NK	7.23y	NK
Han[6]	2016	43/female	right frontal and temporal	yes	no	yes	10.3y	liver, breast, lung
Osuga[27]	2014	46/female	NK	NK	9y	yes	16y	pancreas
Wu[5]	2015	23/male	right occipital lobe	NK	1y	yes	2y	bilateral lung, multiple bone
Jiang[10]	2016	27/male	L3-S1	yes	no	no	3m	NK
NG[28]	2000	55/female	Posterior fossa	yes	1y	yes	9y	soft tissues and lungs
Ogawa[29]	2004	44/female	left tentorial	yes	12y	no	26y	lung
Metellus[15]	2007	34/male	tentorium cerebelli	no	9.08y	yes	10.42y	lung and liver
Muñoz[30]	2008	35/male	left sacrum	yes	6.5y	no	4.58y	lung and liver
Miyashita[8]	2004	48/female	falcotentorial	NK	7y	yes	10y	CSF dissemination
Hu[31]	2009	54/male	left occipital	no	1.33y	no	10m	lung
Gessi[32]	2013	53/female	right occipital	NK	6y	no	12y	lung
Degnan[33]	2016	29/male	left temporal	NK	no	no	10y	renal, pancreas, liver
present case	2018	35/female	pineal region	yes	3.25y	yes	3.83y	temporal lobe

are composed of ovale-to-round cells with enlarged nuclei in a jumbled pattern. Similar with SFTs, tumor cells are diffusely reactive with vimentin and Bcl-2, but only weak with CD34.[21,22] Specifically, HPCs display evidence of increased mitotic activity, nuclear atypia, necrosis, hemorrhage and hypercellularity, and have a much higher tendency to recur and metastasize compared with SFTs.[18,23] Due to their similar pathogenesis and common pathological features, differential diagnosis between SFTs and HPCs remained controversial.[4,18,19,21,22] In 2013, genetic overlaps were identified due to the discovery of NAB2-STAT6 fusion gene in both tumors.[24] Therefore, in 2016 WHO classification of CNS the low-grade SFT, high-grade HPC and anaplastic HPC were combined as a single category as SFT/HPC, and recognized as a spectrum of tumors with similar origin but with different malignancy and prognosis.[25]

Although SFTs as a whole are indolent, CNS SFTs are reported to be more malignant and have a tendency of extracranial metastasis.[3,6] A thorough literature review focusing on metastatic SFT in the pre-WHO 2016 era and WHO I SFT/HPC in the post-WHO 2016 era revealed a total of 14 cases, referred in particular as malignant SFTs (Table 1).[5,6,8,10,15,26–33] Data analysis showed no gender predominance (male: female, 3:4). The median age at initial tumor onset was 42 years, and there was a mean lag time of 8.74 years before metastasis took place. The primary intracranial sites were common location for CNS SFTs except pineal region in our case, and the most common metastatic target was lung, followed by liver. It needs to be pointed out that metastasis occurred without sign of local recurrence in 3 cases, and prior to recurrence in 2 cases. Intracranial metastasis was rare, with one case of CSF dissemination after multiple surgeries and sessions of radiotherapy, and our case of ectopic metastasis to a distant lobe. The mechanism behind the preference towards extracranial metastasis remains unknown, and might be related to the mesenchymal origin of SFTs and disruption of blood-brain barrier.

The pathological analysis of the ectopic metastatic lesion in our case demonstrated changes in cell morphology, necrosis, hypercellularity, higher Ki-67 index (40% vs 15%) and mitoses compared with the primary lesion, implying dedifferentiation and progression in malignancy after relapse. All these features supported the diagnosis of a SFT/HPC (WHO Grade III), which corresponded to what was previously called anaplastic hemangiopericytoma.[25] This phenomenon of sequential deterioration in malignant and/or proliferative features has been reported.[29,32] Therefore, strict and long-term surveillance was strongly required, even with totally resected low-grade SFT.[5] Considering the tendency of extracranial metastasis, it is recommended to screen lung, abdomen and bone periodically.

Surgical resection is the treatment of first choice for CNS SFTs.[11,34] Complete resection is preferred and considered as an effective approach to reduce local recurrence and metastasis.[15,34] Up to now there lacks evidence guiding the application of radiotherapy or gamma knife in improving the long-term prognosis of CNS SFTs. However, for CNS Grade II or III SFT/HPCs, recent studies indicated that radiotherapy were associated with longer recurrence-free interval, but no significant improvement in overall survival was observed.[35–37] Cumulative radiation, however, might result in dedifferentiation of low-grade SFT into malignant fibrosarcoma.[38] Further studies are needed to investigate the role of radiotherapy in the treatment of CNS SFT/HPCs. For SFT with multiple metastasis, systematic chemotherapy has been tried and might be effective.[15,30] In our case, the patient underwent total resection the first time but unfortunately suffered from local recurrence and ectopic metastasis consecutively 3 years later. Gamma knife radiosurgery was conducted for the in situ relapse and traditional radiotherapy for the ectopic metastasis as adjuvant treatment after second surgery. Fortunately for this patient, the size of the local recurrent tumor decreased and no new symptoms were reported during routine follow-up.

4. Conclusion

In summary, we report here an extremely rare case of a primary intracranial SFT in the pineal region with delayed ectopic metastasis in the temporal lobe. Despite low WHO grade, a small subgroup of CNS SFTs has malignant potential. Careful and thorough histopathological analysis is of vital importance to discriminate SFTs from other intracranial disease entities, and strict surveillance was strongly required. Complete resection of the tumor is the first choice for treatment and radiotherapy might be an effective adjuvant therapy for high grade SFT/HPCs.

Author contributions

Wang and Zhang drafted the first manuscript and made a contribution to acquisition and interpretation of data. Qichang

Liu and Fuyi Liu performed the clinical work-up and literature search. Zhang revised the language and grammar of the manuscript. Zhu revised the manuscript that led to the final approval of the current submission. All authors read and approved the final manuscript.
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