



Case report

Solitary fibrous tumor of the pineal region in the elderly: A case report

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ABSTRACT

Introduction: Solitary fibrous tumor (SFT) is a mesenchymal tumor with spindle cells that is often detected in the subcutaneous area and rarely in the central nervous system. Intracranial SFTs rarely occur in individuals older than 80 years, and there are only a few cases reports involving the pineal region.

Presentation of case: The present case was an 80-year-old man with lightheadedness, headache, and vomiting. Magnetic resonance imaging showed a tumor in the pineal region extending along the falx and tentorium cerebelli with homogenous enhancement. Subtotal resection was performed via the occipital transtentorial approach. A histopathological examination showed the proliferation of oval- to spindle-shaped tumor cells with bland nuclei arranged in a haphazard pattern and accompanied by staghorn-like branching vessels. Immunohistochemically, tumor cells were positive for CD34 (focal), CD99, and STAT6, but negative for epithelial membrane antigen (EMA) and S-100. Based on these findings, the tumor was diagnosed as SFT (WHO grade I).

Discussion: Although difficulties are associated with differentiating SFT from meningioma on imaging, recurrence and metastasis are more common with SFT than with meningioma; therefore, histological and immunohistochemical analyses are important. A correlation has been reported between postoperative adjuvant radiotherapy and longer progression-free survival; however, this needs to be confirmed in further studies.

Conclusion: SFT involving the pineal region is rare in the elderly, but needs to be considered as a preoperative diagnosis. Since high rates of postoperative recurrence and metastasis have been reported, long-term follow-ups are required after surgery.

1. Introduction

Solitary fibrous tumors (SFTs) are rare in the central nervous system (CNS), and were initially reported in 1996 [1]. Intracranial SFTs occur at a median age of 42.5 years, and rarely in individuals as old as 80 years [2]. CNS SFTs are most commonly located along the falx cerebri, occipital and spinal dura, tentorium cerebelli, and at the cerebellopontine angle, with only a few cases reports involving the pineal region [3,4]. We herein present a case of SFT in the pineal region of an 80-year-old man and review the literature. This work has been reported in line with the SCARE Criteria [5].

2. Presentation of case

The patient, an 80-year-old man, was admitted to our hospital with lightheadedness, headache, and vomiting for two weeks. His medical history was unremarkable, and no abnormal findings were detected in a neurological examination. Magnetic resonance imaging (MRI) revealed a 38 × 31 × 38 mm lesion in the pineal region with attachments along the falx and tentorium cerebelli, which showed hypointensity signals on T1-weighted images (WI) and mixed hypo-hyperintensity signals on T2WI, a clear boundary, and homogeneous gadolinium enhancement pattern (Fig. 1). Cerebral angiography revealed a tumor stain from the internal carotid artery (ICA) tentorial branch, bilateral middle meningeal arteries, and meningeal arteries from the posterior cerebral artery

Abbreviations: SFT, solitary fibrous tumor; CNS, central nervous system; ICA, internal carotid artery; EMA, epithelial membrane antigen; HPC, hemangiopericytoma; OS, overall survival; PD-L1, programmed cell death ligand-1.

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and superior cerebellar artery. Based on the results of the preoperative examination, falco-tentorial meningioma was suspected and surgical resection with craniotomy was performed.

Surgery was conducted in the left lateral position using the left occipital transtentorial approach. The tumor was pale reddish in color, highly vascularized, and mostly well demarcated; however, the border with the normal dura mater at the transition to the falx cerebri was unclear. The tumor was removed in a piecemeal manner by internal decompression with coagulation and hemostasis. Although some of the falx attachment, which had an indistinct border, remained, most of the tumor was removed; therefore, subtotal resection was achieved (Fig. 2). A histopathological examination showed oval- to spindle-shaped tumor cells with enlarged bland nuclei arranged in a haphazard pattern with staghorn-like branching vessels on HE slides. Mitotic figures were not detected. Immunohistochemically, tumor cells were positive for CD34 (focal), CD99, and STAT6, but negative for epithelial membrane antigen (EMA), glial fibrillary acidic protein, and S-100. Ki-67 immunostaining showed a 14% proliferative index (Figs. 3, 4).

Based on pathological findings, the patient was diagnosed with SFT (WHO grade I). The patient developed transient left homonymous hemianopsia after surgery; however, it gradually improved and the patient was discharged. Postoperative MRI showed that the tumor was partially located inside the falx cerebri, and, thus, the patient received local radiation therapy (54 Gy/27 Fr). Five months after the operation, imaging showed no increase in the residual tumor. The patient is being carefully followed up to detect any increases in the residual tumor or metastasis.

3. Discussion

CNS SFTs are rare and have only been sporadically reported since their initial description in 1996 [1]. SFTs and hemangiopericytomas (HPCs) were originally classified separately; however, in WHO 2016, the previous classification of low-grade SFTs, high-grade HPCs, and anaplastic HPCs was unified into SFT/HPC and categorized as grades I-III [6]. In WHO 2021, the term "HPC" has been removed and is now collectively termed "SFT" [7]. CNS SFTs grade I are typically considered benign, while their grade II and III have higher rates of recurrence and extracranial metastasis [8].

Intracranial SFTs were previously detected in patients aged between 13 and 69 years, with a median age of 42.5 years, and rarely in those as old as 80 years, the age of the present case [2]. CNS SFTs are most commonly located along the falx cerebri, occipital and spinal dura, tentorium cerebelli, and at the cerebellopontine angle, and often have an attachment to the meninges [4]. However, SFTs in the pineal region are rare, with only five cases, including the present case, being reported to date [3].

On MRI, intracranial SFTs show mixed hypo- to isointensity signals on T1WI. On T2WI, the solid region of tumors shows a mixed hypo-

hyperintensity signal, and sometimes a cord-like or patchy hypointensity signal called the "black and white sign" [9,10]. On gadolinium-enhanced MRI, tumors are heterogeneously enhanced, solitary lesions with an often irregular edge, clear boundary, and lobulated contour. SFTs with a large cystic area in the periphery of the lesion have also been detected [7].

Cerebral angiography shows that SFTs often have feeders from the ICA or a branch of the vertebral artery, and their tumor stain is long-lasting, which is useful for differentiating them from meningioma with a sunburst pattern [11,12].

In the present case, the tumor was located in the pineal region and had an attachment to the falx and tentorium cerebelli. In addition, MRI showed that the tumor was homogeneously enhanced with no cystic lesions, and difficulties were associated with differentiating the tumor from meningioma. However, cerebral angiography revealed that the tumor had a feeder from the ICA branch, the tumor stain was long-lasting, which is characteristic of the SFT cases reported to date, and there was no sunburst pattern. Therefore, cerebral angiography may be useful for differentiating meningioma.

Histopathologically, SFTs are characterized by oval- to spindle-shaped cells arranged in a pattern with staghorn-like vessels and an intervening collagenous stroma [13]. Immunohistochemically, tumor cells are positive for CD34, vimentin, CD99, and Bcl-2, and negative for S-100 and EMA [14-16]. SFTs and HPCs were originally considered to be histopathologically and immunohistochemically similar, and their differentiation was unclear [17]. However, in 2013, a gene fusion of the transcriptional repressor NAB2 with the transcriptional activator STAT6 was detected and found to be expressed in both SFTs and HPCs [18]. In WHO 2016, they were collectively referred to as SFT/HPC and categorized as grades I-III depending on the pathologic features [6]. In WHO 2021, the term "HPC" was removed and termed only "SFT", which is the same as the nomenclature for soft tissue [7].

Surgical resection is the first-line treatment for SFT [19]. Preoperative embolization may also be considered because SFTs are highly vascularized tumors [11]. A previous study reported a correlation between adjuvant radiotherapy and longer progression-free survival, but not overall survival (OS), in patients with SFT grade II². A relationship was also reported between radiotherapy and longer OS for HPCs [20].

The present case was diagnosed as SFT grade I, subtotal resection was performed, and the patient received postoperative radiotherapy (54 Gy/27 Fr). Although more case studies are needed to clarify the effects of radiotherapy, it may be indicated for patients based on the extent of resection performed and the WHO grade. The rates of recurrence and extracranial metastasis of SFT were previously reported to be 31.7 and 2.6-56%, respectively; therefore, long-term imaging evaluations, including the extracranial region, are necessary [2]. In addition, a previous report showed that intracranial SFTs frequently express the immune checkpoint protein, programmed cell death ligand-1 (PD-L1), and diffuse or intense expression of PD-L1 may be associated with early

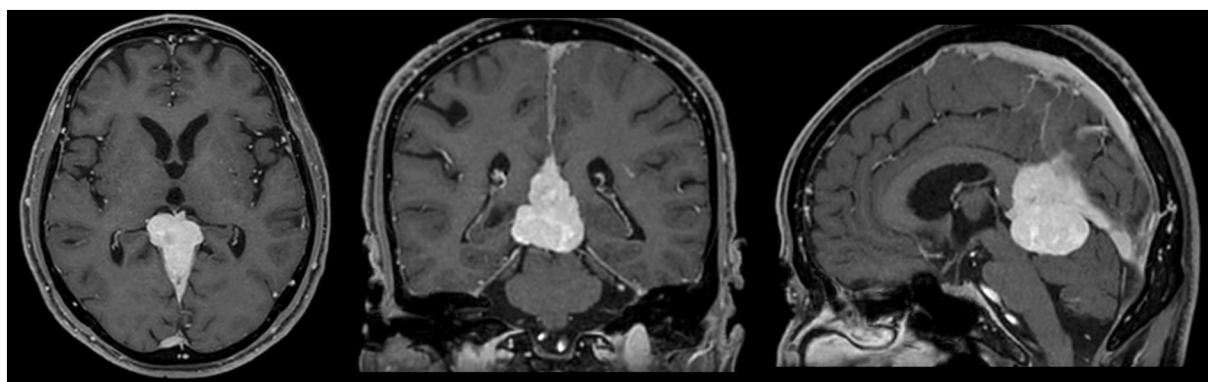


Fig. 1. T1-weighted magnetic resonance images with contrast showing strong enhancement in the pineal region.

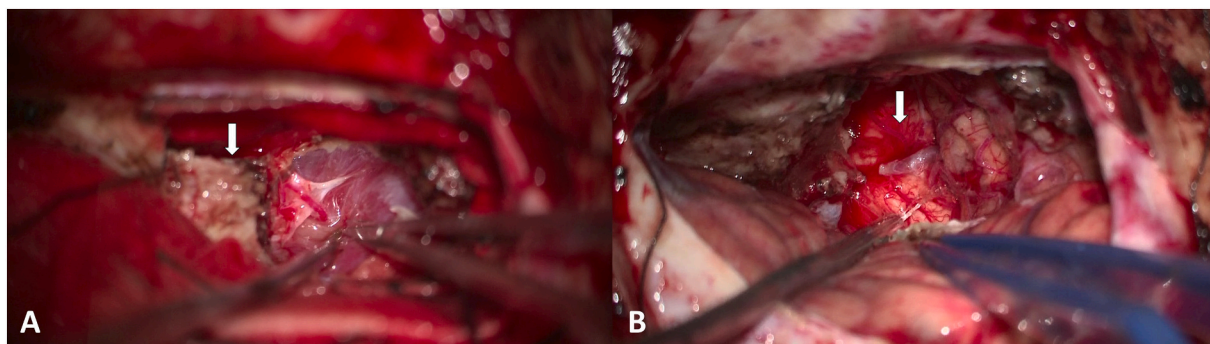


Fig. 2. Intraoperative photographs from the microscope: Left occipital transtentorial approach for tumor removal. (A) Tumor with pale reddish in color and highly vascularized has attachments to the falx and tentorium cerebelli (Arrow shows the falx cerebelli). (B) Following tumor removal (Arrow shows the tectum of midbrain).

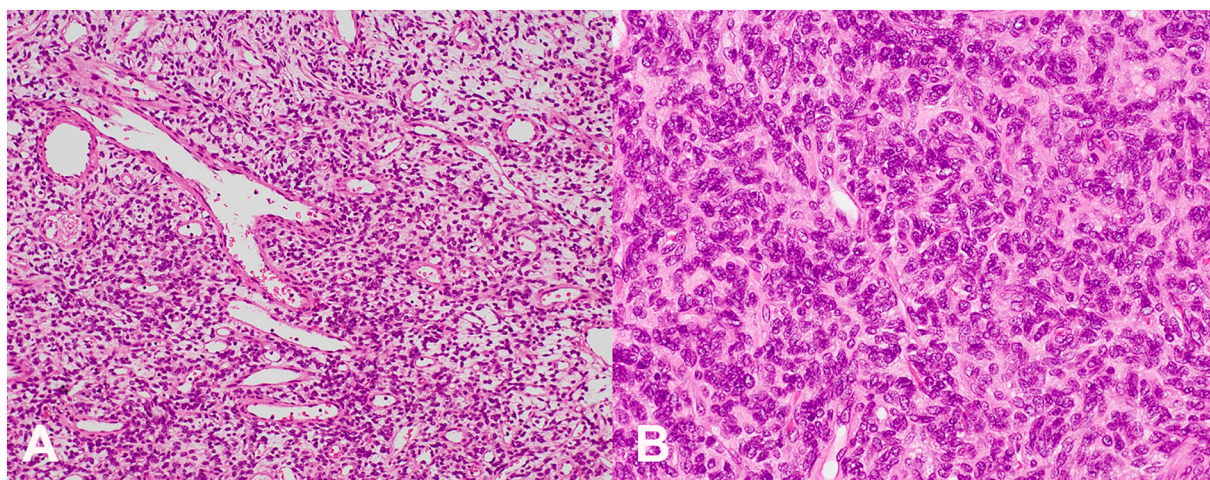


Fig. 3. A tumor composed of oval- to spindle-shaped cells with enlarged and bland nuclei arranged in a haphazard pattern and accompanied by staghorn-like branching vessels. Mitotic figures are not present (HE). A: 200 \times , B: 400 \times .

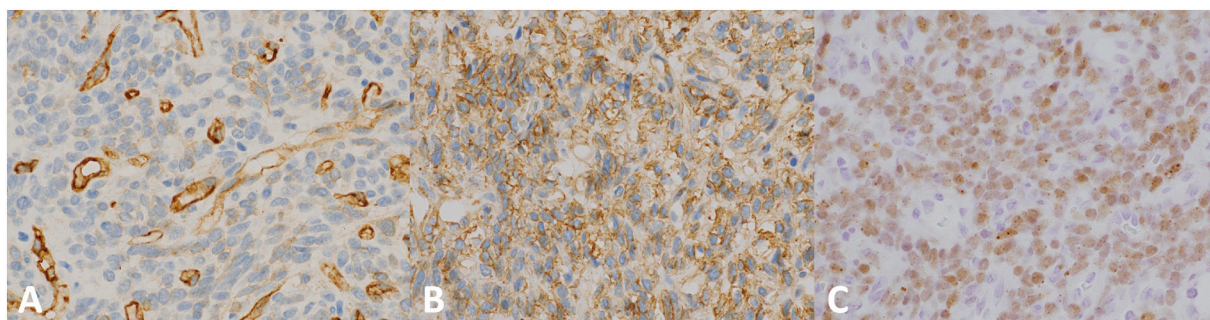


Fig. 4. Immunohistochemically, tumor cells are positive for A: STAT6, B: CD34 (focal), C: CD99.

development of extracranial metastasis [21]. Although further studies are needed, PD-L1 may play an important role in predicting extracranial metastasis of intracranial SFTs.

4. Conclusion

We herein presented a rare case of SFT in the pineal region of an 80-year-old man. SFT involving the pineal region in the elderly is rare, but needs to be considered as a preoperative diagnosis. However, difficulties are associated with differentiating it from meningioma in preoperative imaging examinations, and, thus, postoperative histopathological and

immunohistochemical analyses are important. Since high rates of post-operative recurrence and metastasis have been reported, adjuvant radiotherapy needs to be considered. However, the number of cases is small and, thus, further analyses are warranted. Moreover, long-term follow-ups with imaging, including the extracranial region, are required after surgery.

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Ethical approval

The study is exempt from ethical approval.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Hiroshi Oketani, Sadao Onaka and Takaharu Nakamura designed the study, analyzed preoperative data and performed the operation. Mizuki Handa and Yoshinao Oda analyzed pathological findings. All authors read and approved the final manuscript.

Research registration

None.

Guarantor

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Declaration of competing interest

The authors declare no conflicts of interest.

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