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Case Report

Meningeal malignant solitary fibrous tumor with multiple recurrence, extracranial extension, cervical lymph node metastases: case report and review of the literature

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Abstract

Introduction Solitary fibrous tumor (SFT) of the central nervous system (CNS) is a spindle cell neoplasm originating from mesenchymal tissue. SFT is prone to local recurrence or distant metastasis. The main sites of metastasis include the liver, lung, and bone. However, the tumor direct extracranial extension via natural skull base foramina to cervical region is rare.

Case presentation A 43-year-old male presented with an incidentally discovered left temporo-occipital region mass spanning the tentorium cerebelli identified during head and neck CT angiography, initially suspected as meningioma. The patient underwent gross total resection, with histopathological confirmation of a grade III malignant solitary fibrous tumor (WHO CNS 2007) originating from the left tentorium cerebelli. Immunohistochemical analysis demonstrated tumor cell positivity for CD34, vimentin, and Ki-67 (approximately 10%), while negative for EMA, PR, and P53. Subsequent disease progression manifested as multiple local recurrences with sacral and left pubic metastases. Multimodal management included adjuvant radiotherapy—Intensity-modulated radiation therapy (IMRT) and Gamma Knife radiosurgery(GKRS)—and concurrent bone-modifying agents (Ibandronate Sodium) for skeletal metastases. In June 2020 re-evaluation prompted by a palpable left neck mass revealed magnetic resonance imaging (MRI)-documented multifocal recurrence involving the left mastoid process and tentorium cerebelli, with extended through the jugular foramen into the left parapharyngeal space and involved the left cervical lymph nodes IMRT was performed with a prescribed dose of 54 Gy in 30 fractions. After completion of the treatment course, significant regression of most lesions was observed. However, the patient discontinued clinical follow-up after July 2021. Subsequent telephone contact confirmed expiration in September 2022 secondary to disease progression.

Conclusions We report the first case of extracranial extension and cervical lymph node metastases from meningeal malignant SFT. This finding provides novel insights into the dissemination patterns of intracranial SFT. Surgical resection is the gold standard for the treatment. Postoperative radiotherapy (PORT) whether gross total resection (GTR) or subtotal resection (STR) may be the optimal treatment strategy, but PORT dose < 60 Gy with IMRT or marginal dose < 15 Gy with GKRS may be insufficient. Close and long-term follow-up, especially in the first five years after diagnosis, is essential to manage such patients because of high risk of recurrence and metastasis.

Mingying Geng and Jungang Ma have equally supervised this study.

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Keywords Solitary fibrous tumor · Extracranial extension · Lymph node metastase · Radiotherapy

1 Introduction

Solitary fibrous tumor (SFT) is a spindle cell neoplasm originating from mesenchymal tissue. SFT was first reported in the visceral pleura, but subsequent studies have revealed that SFT can arise in any part of the body. SFT can occur at any age, particularly between 50 and 60 years of age, with a similar incidence in both genders [1].

Intracranial SFT was first described in 1996 by Carneiro SS [2]. This disease is extremely rare accounting for 0.4% of primary central nervous system (CNS) tumors [3]. Intracranial SFT radiologically mimics meningioma and therefore is frequently misdiagnosed on initial presentation. The 2021 WHO Classification of CNS Tumors simplifies the histological grading of SFT into three tiers based on two criteria: mitotic count (per 10 high-power fields, HPFs) and necrosis: Grade 1: Mitotic count < 5/10 HPFs; Grade 2: Mitotic count ≥ 5/10 HPFs without necrosis; Grade 3: Mitotic count ≥ 5/10 HPFs with necrosis [4]. This tumor was historically classified as benign lesion but has been demonstrated to exhibit aggressive behavior [5]. Surgical resection is the first-line treatment for localized SFT, with a 5-year overall survival (OS) rate of 84.5% following complete excision of skull base SFT [6]. However, some studies have reported high rates of local recurrence and distant metastasis. The metastatic sites predominantly involves the bone, lung, and liver (accounting for approximately 70% of all distant metastases), kidney, breast, adrenal gland, pancreas, retroperitoneum, peritoneum, soft tissues and skin site had also been reported with less frequency [7–9]. To date, aside from an isolated case report of intracranial SFT involving the jugular foramen, no additional cases have been documented demonstrating direct extracranial extension via natural skull base foramina. Here, we present a case of meningeal SFT with direct extracranial extension through the left jugular foramen to the left parapharyngeal space and involved the left cervical lymph nodes.

2 Case presentation

A 43-year-old male presented to the traumatology department following a motor vehicle accident, sustaining a right tibial fracture and closed head injury. Incidentally, head and neck computed tomography (CT) angiography revealed a left temporo-occipital region mass spanning the tentorium cerebelli measuring 4.2 × 2.5 × 3.8 cm (Fig. 1a, b). The lesion demonstrated mild hyperdensity on non-contrast imaging with marked heterogeneous enhancement post-contrast. Notably, the patient denied any tumor-related symptoms (e.g., headaches, seizures, or focal neurological deficits) prior to the trauma and had no significant medical history, including prior neoplasms or genetic disorders.

Subsequent cranial magnetic resonance imaging (MRI) (Fig. 1c–h) further characterized the lesion. On non-enhanced sequences, the mass exhibited predominantly isointense signal intensity with mild heterogeneity on T1-weighted imaging (T1WI), T2-weighted imaging (T2WI), and fluid-attenuated inversion recovery (FLAIR), while diffusion-weighted imaging (DWI) showed no restricted diffusion. The tumor displayed a well-defined boundary but irregular margin, broad-based attachment to the left tentorium cerebelli, and scattered intralesional vascular flow voids. Mild peritumoral edema was observed without parenchymal invasion. Post-gadolinium sequences revealed intense homogeneous enhancement across axial, sagittal, and coronal T1WI sequences. The tumor measured 4.5 × 2.9 × 4.2 cm in maximal dimensions. Based on these radiological features, the provisional diagnosis favored meningioma.

The craniotomy via a left temporo-occipital approach for intracranial tumor resection was delayed by 1 month due to prioritized management of traumatic injuries, with subsequent achievement of gross total resection (GTR). Intraoperatively, it was observed that the lesion originated from the left tentorium cerebelli, demonstrating a firm consistency, dark-red coloration, and rich vascularity. The tumor exhibited infiltrative margins with microscopic adherence to adjacent cerebellar parenchyma, necessitating meticulous microsurgical dissection under neuromonitoring. Post-resection, the excised gross tumor mass measured 6 × 5 × 4 cm. Histopathological evaluation (Fig. 1i, j) revealed a hypercellular neoplasm composed of monomorphic spindle-to-oval cells arranged in a patternless architecture interspersed with staghorn-shaped, hyalinized vessels. Mitotic activity reached 5/10 HPFs. Immunohistochemistry demonstrated diffuse strong positive for CD34 (Fig. 1k), vimentin and Ki-67 (approximately 10%) (Fig. 1l), but negative for EMA (epithelial membrane antigen), PR (progesterone receptor) and P53. These findings, combined with elevated mitotic activity and infiltrative growth pattern, confirmed the diagnosis of a grade III malignant solitary fibrous tumor (WHO CNS 2007) arising from the left tentorium cerebelli.

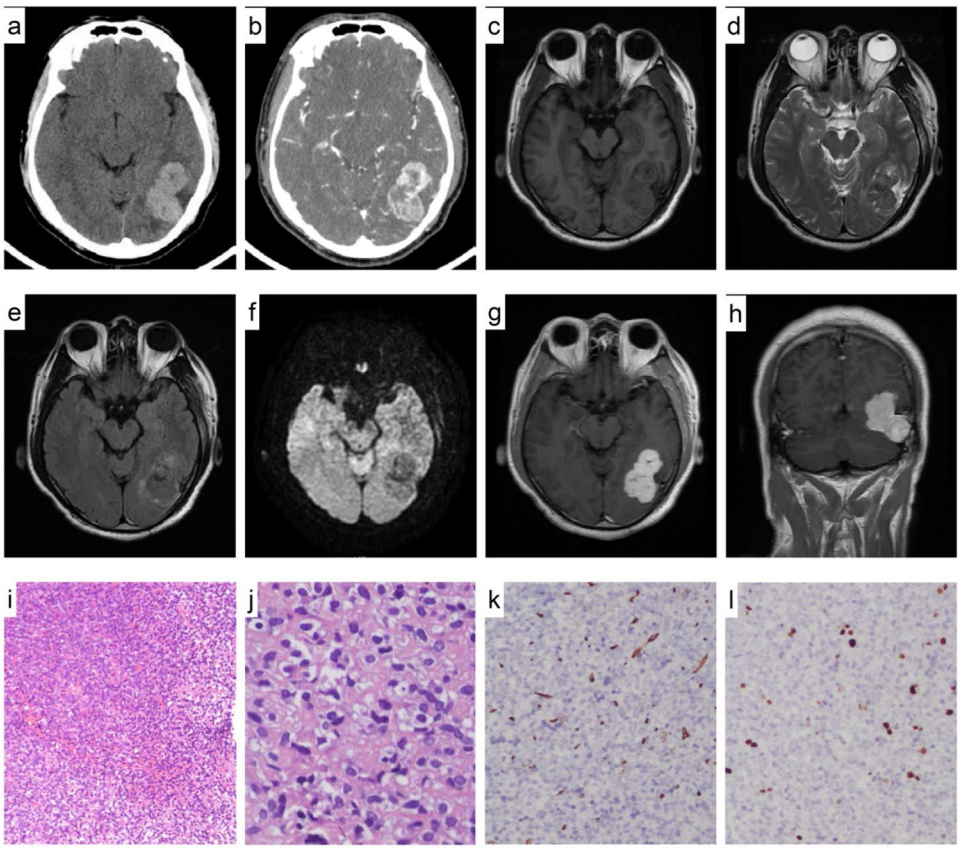

Fig.1 Imaging and pathological findings of the tumor in the left temporo-occipital region. **a** Axial non-contrast CT, **b** Axial post-contrast CT; **c** Axial non-contrast T1WI, **d** Axial non-contrast T2WI, **e** FLAIR, **f** DWI, **g** Axial post-contrast T1WI, **h** Coronal post-contrast T1WI; **i** H&E × 100, **j** HE × 400). **k** CD34 (× 200); **l** Ki-67 (I: × 200)

The patient did not make regular follow-up due to poor compliance and remained asymptomatic until 34 months postoperatively, when he presented to our outpatient department with new-onset intermittent left occipital traction-related pain and ipsilateral hearing loss. Cranial MRI was performed promptly and revealed local tumor recurrence at the primary resection site (Fig. 2a), demonstrating a new lesion with 3.8 × 2.5 × 4.6 cm in diameters appearing heterogeneously enhancing mass with dural tail sign. The tumor resection was performed via the prior left temporo-occipital surgical approach, with intraoperative findings demonstrating dense adherence of the tumor to the tentorium cerebelli and partial involvement of the transverse sinus and sigmoid sinus. Given the risk of venous infarction and cranial nerve injury, subtotal resection (STR) was performed. Intraoperative frozen section and subsequent histopathological examination confirmed a grade III recurrent SFT (WHO CNS 2007) characterized by focal hypercellularity and a mitotic count of 4/10 HPFs. The lesion exhibited a characteristic immunoprofile with diffuse CD34 positivity and a 10% Ki-67 proliferation index. The patient did not received any postoperative adjuvant therapy due to poor compliance.

Gamma Knife radiosurgery (GKRS) was performed at 8 months and 22 months following the second surgery due to local recurrence of the lesion (Fig. 2b). The administered radiotherapy dose was 12 Gy delivered to the tumor margin and maximum doses of 30 Gy and 24 Gy to the tumor core respectively in both treatments. The patient remained asymptomatic until 36 months post-reoperation, when he presented to the neurosurgery outpatient department with a 1-month history of headache, dizziness with nausea and vomiting. Cranial MRI showed a 5.3 × 3.6 cm heterogeneously enhancing mass, with irregular margin, obscure boundary in the left cerebellopontine angle, moderate peritumoral edema and prominent vascular flow voids (Fig. 2c). The third surgical operation was performed and the tumor was basically removed except for a residual dural nodule (Fig. 2d yellow arrow). Subsequent histopathological examination confirmed recurrent SFT characterized by STAT6 positivity staining. The residual dural nodule and tumor bed received adjuvant radiotherapy (54 Gy in 27 fractions). Serial contrast-enhanced MRI demonstrated marked regression of the lesion in volume at the 4th month (Fig. 2e) and the 17th month (Fig. 2g). Moreover, the residual dural nodule was assessed as complete response (CR) at the time of the last follow-up (RECIST v1.1 criteria). However, multiple newly arisen nodules recurred in the left occipital region at the 17th month after the third surgical operation (Fig. 2f red arrow). No surgical intervention was recommended due to extensive local meningeal invasion after multidisciplinary team (MDT) discussion. Although

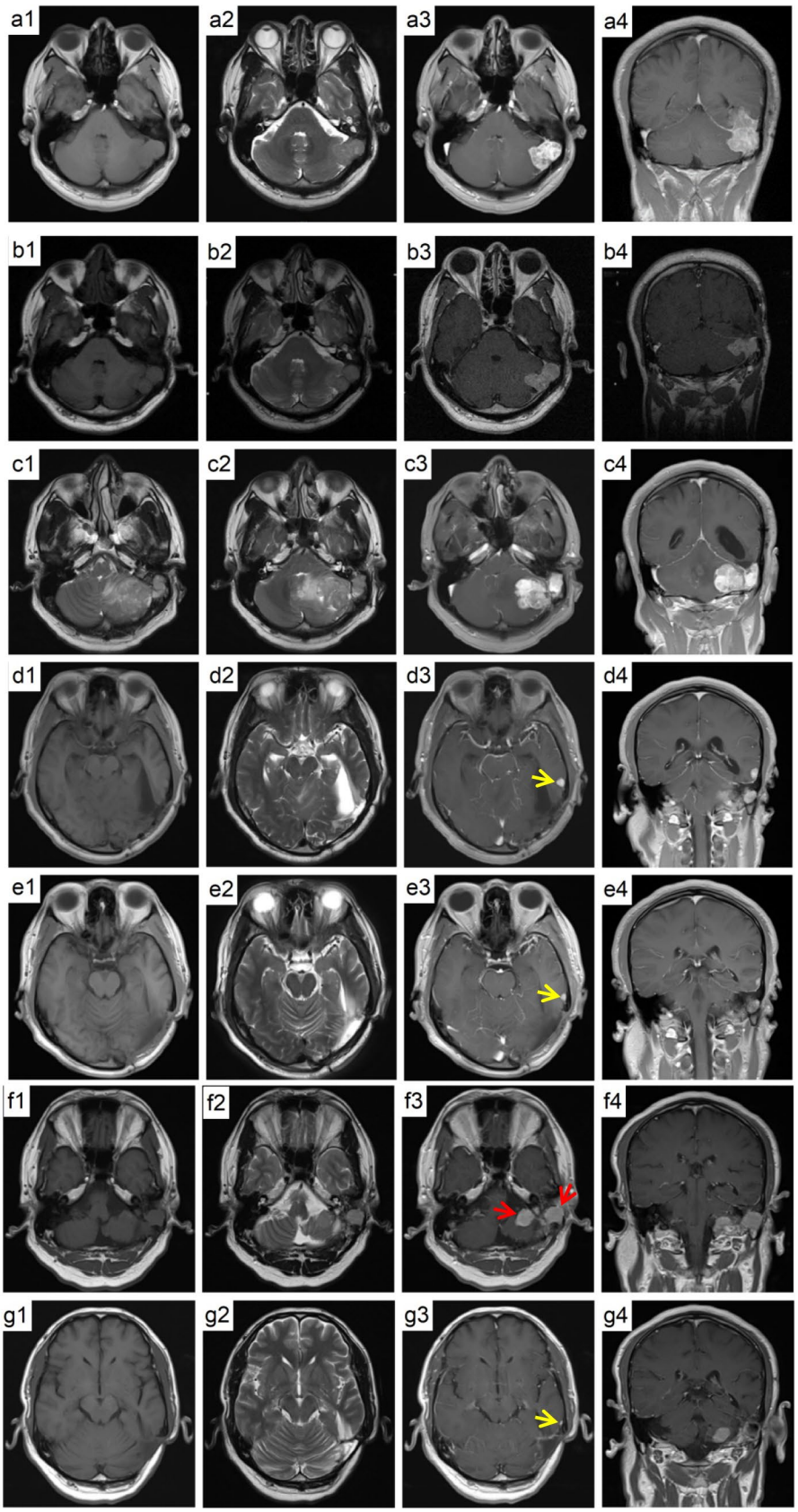

Fig.2 Cranial MRI of recurrent lesion in the left temporo-occipital region (**a**, **b**, **c**, **f**) and residual dural nodule in the left temporal region (**d**, **e**, **g**). **a** pre-reoperation; **b** prior to the initial GKRS; **c** prior to the third operation; **d** 1 month after the third operation; **e** 4 months after the third operation; **f**, **g** 17 months after the third operation. (1.Axial non-contrast T1WI, 2. Axial non-contrast T2WI, 3.Axial post-contrast T1WI, 4.Coronal post-contrast T1WI)

next-generation sequencing (NGS) guided targeted therapy was proposed to identify actionable mutations, the patient declined any examinations and interventions due to financial reasons.

In April 2017, the patient incidentally touched a painless, slowly growing left inguinal region mass but deferred medical evaluation. Over the ensuing 16 months, the lesion exhibited progressive enlargement, prompting pelvic MRI in August 2018. Imaging revealed two osteolytic lesions in the sacrum and left pubis (one lesion had 5.4 × 6.7 × 5.8 cm in diameters and another had 5.8 × 5.2 × 7.0 cm in diameters, respectively). Each of the two lesions was associated with oval-shaped soft tissue masses demonstrating invasive features into adjacent musculature. Heterogeneous hypointensity on T1WI, mild hyperintensity with internal septations on T2WI, obvious heterogeneous enhancement was observed on enhanced scans (Fig. 3a, b). Contrast-enhanced CT further delineated a delayed venous-phase predominance of enhancement (vs. arterial phase), suggesting hypervascularity even though delayed-phase imaging was omitted (Fig. 3c, d). Notably, a 5-mm lytic focus with sclerotic margins was identified in the right ilium (Fig. 3c: red arrow). Given the patient's history of multiple SFT recurrences, these multifocal osteolytic lesions were highly suspicious for osseous metastases.

Ultrasound-guided core needle biopsies of the sacral vertebrae and left pubic lesions confirmed pathologically a grade III metastatic SFT (WHO CNS 2016) exhibiting STAT6 positivity staining (Fig. 4a, b). The patient received palliative radiotherapy (50 Gy in 25 fractions) to the pelvic lesions along side bone-modifying agents (Ibandronate Sodium, 4 mg/month). At the last follow-up on July 22, 2020, pelvic CT manifested stable disease (SD) (Fig. 5a, c). However, the last time spinal MRI on 2021-07-30 demonstrated enlarged sacral lesion and was assessed as progressive disease (PD) (Fig. 5b). At that time, left pubic lesion assessment was precluded due to incomplete anatomical coverage and right iliac lesion remained stable (no radiotherapy delivered) (Fig. 5d).

In June 2020, the patient re-presented with a progressively enlarging left cervical mass. MRI of the head and neck demonstrated increased enlargement of the left temporo-occipital lesion with invasion of the left mastoid process. The tumor extended through the jugular foramen into the left parapharyngeal space and involved the left cervical lymph nodes (Figs. 6, 7a). Additionally, a newly developed nodular lesion was observed along the medial aspect of the tentorium cerebelli. Following re-evaluation by the MDT, surgical intervention was deemed inadvisable due to tumor encasement

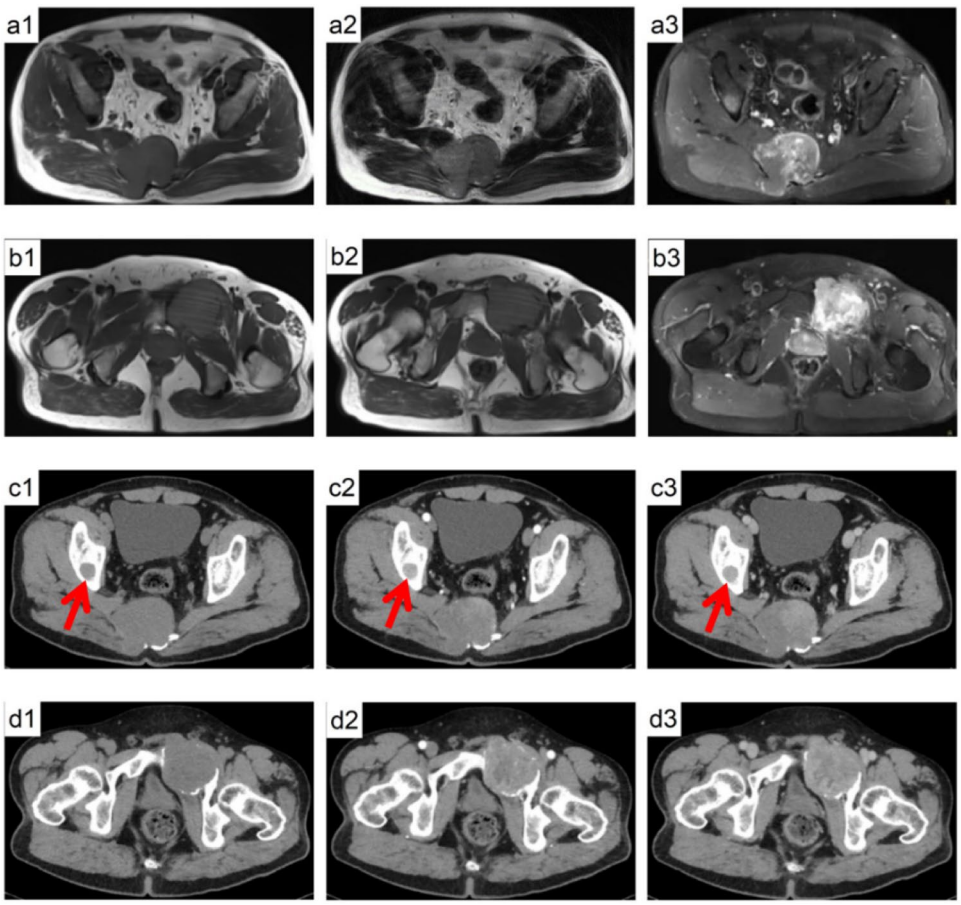

Fig.3 Pre-treatment osseous lesions involving the sacral vertebrae (**a**, **c**) and left pubis (**b**, **d**). **a**, **b** 1.Axial non-contrast T1WI, 2. Axial non-contrast T2WI, 3.Axial post-contrast T1WI. **c**, **d** 1.Axial non-contrast CT, 2. Axial contrast CT(arterial phase); 3. Axial contrast CT(venous phase). c1-3 red arrow: lytic leision in the right ilium

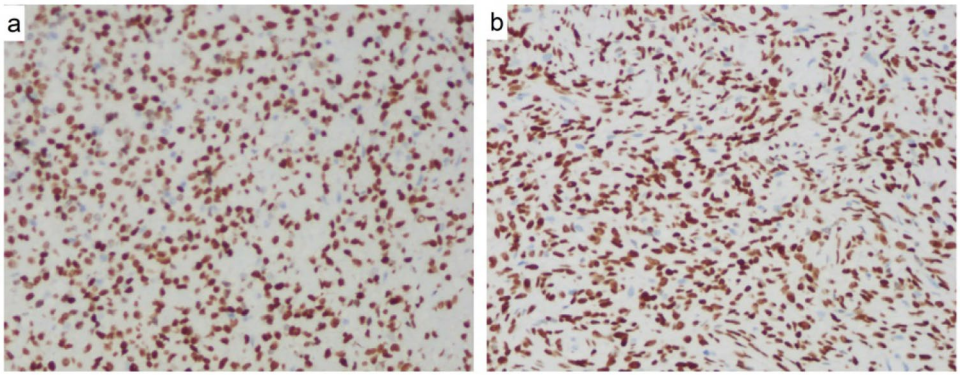

Fig.4 STAT6 were positive by immunohistochemistry in sacral vertebrae (**a**) and left pubic lesions (**b**)

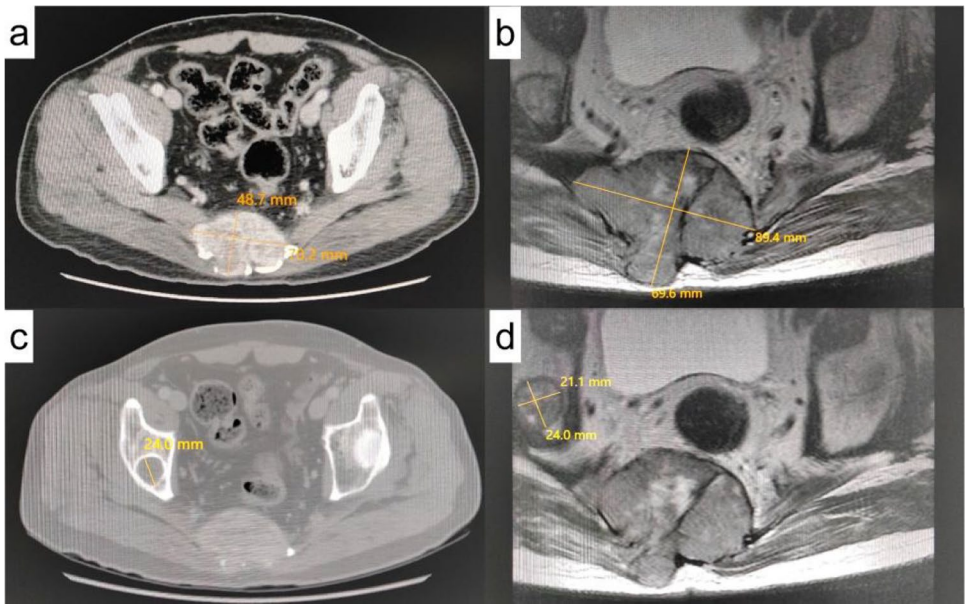

Fig.5 Comparative Post-Treatment Imaging of the sacral lesion and the right iliac lesion. **a** 21-month post-treatment of the sacral lesion on the pelvic CT. **b** 33-month post-treatment of the sacral lesion on the spinal MRI. **c** 21-month post-treatment of the right iliac lesion on the pelvic CT. **d** 33-month post-treatment of the right iliac lesion on the spinal MRI

of the internal carotid artery and complex regional anatomy. Therefore, IMRT (54 Gy in 30 fractions) was performed. One month post-radiotherapy, the left parapharyngeal and cervical lymph node lesions had shrunk evaluated as partial response (PR) based on both clinical palpation and imaging evaluation (Fig. 7b). Notably, the last time cranial MRI follow-up on 2021-07-29 revealed ongoing regression of these lesions (Fig. 7c yellow arrow). The left parapharyngeal space and cervical lesions changes could only be roughly assessed based on coronal views of the cranial MRI because cervical MRI was not performed. However, the nodular opacity along the medial aspect of the tentorium cerebelli had increased in size and the treatment response was assessed as PD (Fig. 7c red arrow). The patient discontinued clinical follow-up after July 2021. Subsequent telephone contact confirmed expiration in September 2022 secondary to disease progression. A comprehensive summary of the patient's major therapeutic interventions, including chronological time points, anatomical treatment sites, treatment modalities (with radiation doses specified in Gy), and RECIST-based response evaluations with an 152 months OS is systematically presented in Fig. 8.

3 Discussion

In the 2016 WHO Classification of Tumors of the Central Nervous System (CNS), SFT and hemangiopericytoma (HPC) were classified as a unified diagnostic entity (SFT/HPC) based on their shared origin as rare mesenchymal spindle cell neoplasms. However, the 2021 WHO Classification of CNS Tumors has eliminated the HPC designation, now recognizing SFT as a distinct diagnostic entity supported by molecular genetic evidence [4]. Originally described as a primary pleural neoplasm by Klemperer and Rabin in 1931 [10], SFT is now recognized as a ubiquitous mesenchymal tumor. Subsequent

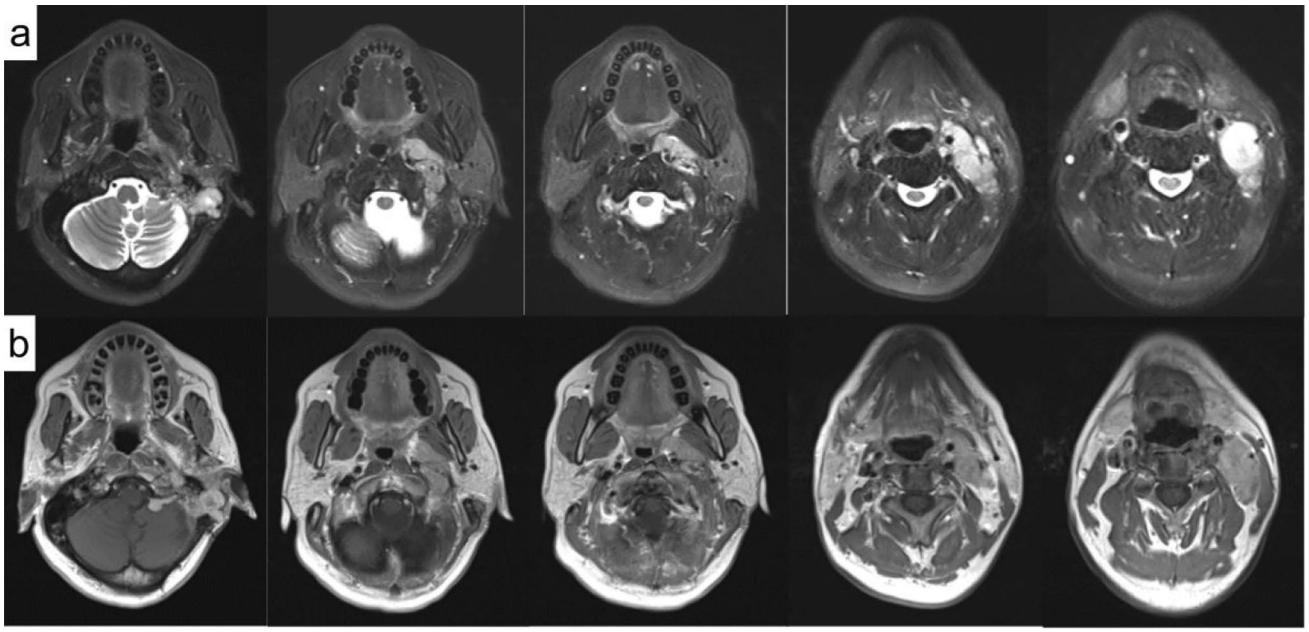

Fig.6 MRI of the head and neck revealed multiple soft tissue masses in the left mastoid process and tentorium cerebelli, with transjugular foramen extension into the left parapharyngeal space and cervical lymph nodes on T2WI sequence (**a**) and obvious heterogeneous enhancement on T1WI enhanced sequence (**b**)

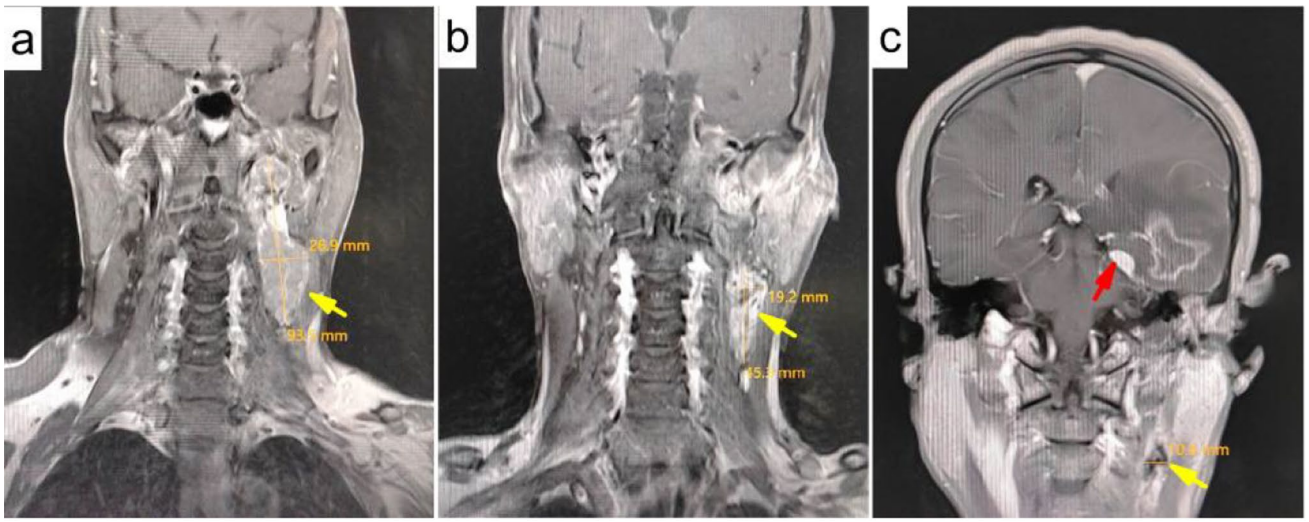

Fig.7 Comparative Pre-/Post-Treatment Imaging of Cervical Lesions (Coronal T1WI MRI). **a** Baseline cervical MRI at initial diagnosis of cervical lesions(yellow arrow). **b** 1-month post-radiotherapy cervical MRI follow-up. **c** 1-year post-radiotherapy cranial MRI surveillance

studies have documented its occurrence in virtually every anatomical sites encompassing thoracic structures (pleura, mediastinum, lung), abdominal/pelvic regions (peritoneum, omentum, liver, bladder, kidney), endocrine organs (thyroid, breast) and head/neck territories (nasal cavity, sinus and intracranial cavity) [11, 12]. SFT can occur at any age, particularly between 50 and 60 years of age, with a similar incidence in both genders or female predominance [1, 13–16].

Patients may present tumor related compression symptoms such as headache, dizziness, hearing loss and so on [17]. However, some patients have no any symptoms, the lesion is accidentally found in physical examination by cranial MRI or CT, this phenomenon is particularly exemplified by the present case who had too small lesions to exert any perceptible pressure on the surrounding tissues.

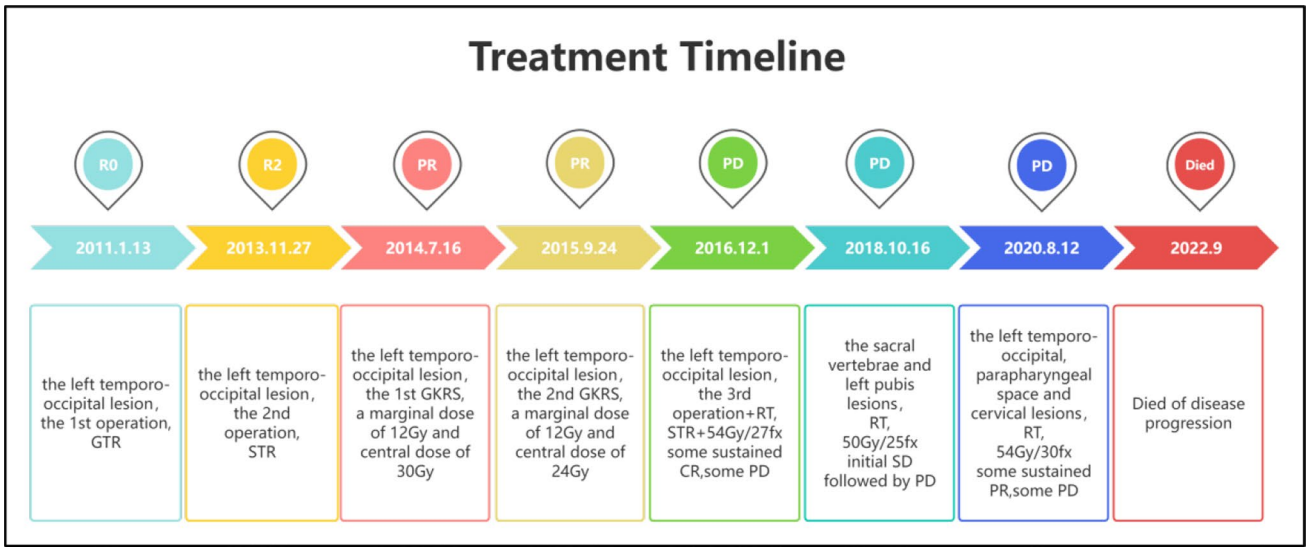

Fig.8 The comprehensive summary of the patient's major therapeutic interventions

CNS SFT often grows in or close to the meninges and has similar radiographic appearance with meningioma. So CNS SFT is often misdiagnosed as meningioma especially fibrous/angiomatous meningioma which accounts for 37% of the primary intracranial tumors [18–21]. This patient we reported was also misdiagnosed as meningioma at the time of initial MRI diagnosis while it was pathologically confirmed as meningeal SFT. Although CNS SFT is rare, many researchers reported its distinguishable radiographic characteristics [19, 22–24]. Some studies found that it usually appears to be large and well-defined but marginal lobulated soft tissue mass [19, 22, 24]. The tumor often grew spanning the falx or the tentorium cerebelli [19]. An obviously heterogeneous signal was often found on MRI particularly on T2WI sequence which represent iso-intensity or slightly high signal and mixed banded or slender or patchy low signal, so-called 'black and white sign' or 'yin-yang sign' [23]. The degree of enhancement was different in various reports with mild, moderate and markedly heterogeneous enhanced which may be related to cell density, content of collagen fiber and tumor blood supply. However, markedly enhancement was noted in most patients [25]. Moreover, Clarençon et al. reported that the tumor showed gradual enhancement especially in the low signal area on T2WI, which may be related to content of collagen fiber [22]. The bone lesions of this case also exhibited progressive enhancement. It is reported that 'dural tail sign' presented in a few patients make this disease easily to be misdiagnosed as meningioma [26]. This may be one of the reasons why our case was misdiagnosed as meningioma at the time of the first diagnosis. In addition, other symptom such as peritumoral edema and bone destruction was also reported but without specificity. Li et al. [19] found that cystic degeneration or necrosis could be a potential feature. The present case was generally consistent with the aforementioned features except lack of bone destruction and cystic degeneration or necrosis in primary site.

Pathologically, SFT is most composed of spindle-cells with intercellular collagen. Immunohistochemically, meningeal SFT is strongly positive for CD34, vimentin, Bcl-2 and CD99. Especially, CD34 IHC evaluation can yield 81% sensitivity and 96% specificity in the diagnosis for meningeal SFT [27]. In contrast, meningeal SFT is negative for SMA, S-100 and EMA protein [28]. The landmark discovery in 2013 identified the NAB2-STAT6 gene fusion can operates as a transcriptional coactivator for a specific set of enhancers and promoters that are normally targeted by the EGR1 transcription factor resulting in the carcinogenesis of SFT. This genetic alteration drives constitutive nuclear translocation of STAT6 protein, which is now established as the diagnostic gold standard for SFT through immunohistochemical nuclear positivity [29]. Moura DS et al. identified a novel NFIX-STAT6 fusion gene in a patient with pelvic SFT that provides new insights into the pathogenesis of this tumor [30]. In addition, aldehyde dehydrogenase 1 (ALDH1) and glutamate receptor 2 (GRIA2) have been reported to be diagnostic markers of meningeal SFT. However, GRIA2 was rarely used because it was also positive in meningiomas [27, 31]. Most of patient with meningeal SFT showed high Ki-67 expression which is considered a poor prognostic factor [32]. In the line with these previously published studies, the present case were positive for CD34 and STAT6 confirming the diagnosis of SFT.

Surgical resection remains the therapeutic cornerstone for meningeal SFT, with GTR historically considered definitive treatment. While conventional paradigms recommend adjuvant RT primarily for STR cases [33], emerging

evidence challenges this dogma. Contemporary clinical series demonstrate that PORT significantly improves 5-year local control (LC) rates from 61.5% to 92.8% and progression-free survival from 46.7% to 88.7% despite achieving macroscopically complete resection [34]. This patient experienced in situ recurrence shortly after the first surgery despite with GTR, which underscores the necessity of the use of PORT independent of conventional margin assessment criteria in all SFT patients. Furthermore, the optimal dose and method of radiotherapy are still unclear. It is reported that radiotherapy dose ≥ 60 Gy can improve LC compared with < 60 Gy [35]. In this patient, despite implementation of IMRT targeting the tumor bed and bone lesions after third surgical operation, poor LC was still observed possibly to be attributable to a subtherapeutic total radiation dose (< 60 Gy). Notably, a landmark study revealed that intracranial SFT patients treated with definitive radiotherapy (RT) at 60 Gy can achieve an objective response rate (ORR) of 75% with 5-year LC rate exceeding 80% and OS approaching 90%. This finding challenges the historical perception of considering SFT as a radioresistant entity [35]. This paradigm-shifting evidence may catalyze further exploration of RT as a potential definitive treatment modality for SFT, particularly in cases where complete surgical resection is anatomically constrained. The significant and durable regression of residual lesions in the left temporal region, parapharyngeal space, and cervical lymph nodes of this patient suggests that radiotherapy doses ≥ 60 Gy may achieve curative outcomes for SFT, which is consistent with the report previously published by Haas RL [1]. Gou et al. showed that PORT IMRT can improve the DFS than PORT SRS because the former can provide dose gradient irradiation, not just tumor bed irradiation [36]. Despite twice GKRS sessions (both achieved a marginal dose of 12 Gy) carried out for the intracranial lesion, the patient experienced recurrent local progression. This therapeutic resistance may be attributable to suboptimal radiation dosing, as cumulative evidence indicates that marginal dose > 15 Gy is associated with superior LC rates (74.5% vs 65.7% at 5 years) in intracranial SFT [37]. Systemic therapy is frequently utilized in patients with metastatic or inoperable disease, though its therapeutic efficacy remains suboptimal. Receptor tyrosine kinases (RTKs) are other therapeutic alternation that have been employed in the treatment of SFT patients in recent years. For example, pazopanib has demonstrated actionable activity and a significant improvement in median PFS and OS compared to conventional chemotherapy with values of 12.1 months and 49.8 months versus 4.6 months and 11.5 months, respectively [38].

CNS SFT demonstrates a high propensity for locoregional recurrence and distant metastasis, documented metastatic incidence of 28% in clinical cohorts [8]. While extracranial metastasis represents the predominant metastatic pattern, intracranial dissemination remains exceptionally rare [39]. Notably, direct extracranial invasion through anatomical foramina has not been previously reported. This case is the first report of intracranial SFT demonstrating direct extracranial invasion, with the lesion extending extracranially through the left jugular foramen and infiltrating the ipsilateral parapharyngeal space and cervical lymph nodes. Although Hayenga et al. reported a SFT case with jugular foramen involvement, that patient exhibited intraspinal dissemination via cerebrospinal fluid pathways rather than direct extracranial invasion [40]. Therefore, this present case report provides novel insights into the dissemination patterns of intracranial SFT, particularly highlighting its capacity for direct extracranial extension and a previously undocumented biological behavior that expands our understanding of tumor progression mechanisms in neurogenic mesenchymal neoplasms.

Giordan et al. [41] reported that the recurrence and metastasis of meningeal SFT often occur within the first 5 years following initial diagnosis, with a recurrence rate estimated at 60%. In present case, the recurrence occurred 3 years after initial diagnosis and metastasis occurred more than 6 years after initial diagnosis. The apparent delay in metastasis detection may be attributed to three primary factors: First, the absence of standardized follow-up protocols potentially leads to missed opportunities for early identification of recurrent lesions. Second, clinical diagnosis of recurrence often occurs only after substantial tumor growth triggers symptomatic presentation of larger tumor masses. Third, despite the typically indolent growth pattern observed in these lesions, metastasis may already occur during the initial five-year period following primary diagnosis. For these reasons, an early, close and long-term follow-up including clinical and imaging follow-up is recommended during the disease course especially in the first five years after diagnosis, but optimal surveillance intervals and modality combinations require validation through prospective multicenter trials, particularly given the tumor's unpredictable biological behavior.

This case report has several limitations. The patient's financial constraints and poor compliance prevented regular follow-up examinations, resulting in the inability to accurately determine the time of recurrence or evaluate treatment efficacy. The lack of comprehensive molecular or genetic analyses beyond STAT6 hindered the implementation of effective targeted therapy. Further research is required to identify driver mutations and develop corresponding targeted therapeutic strategies.

4 Conclusion

In conclusion, we present a case of extracranial extension and cervical lymph node metastases from meningeal malignant SFT particularly highlighting its capacity for direct extracranial extension-a previously undocumented biological behavior that expands our understanding of tumor progression mechanisms in neurogenic mesenchymal neoplasms. Surgical resection is the gold standard for the treatment, but radiotherapy dose ≥ 60 Gy may achieve curative outcomes for SFT. PORT whether GTR or STR may be the optimal treatment strategy,but radiotherepy dose < 60 Gy with IMRT or marginal dose < 15 Gy with GKRS may be insufficient. Close and long-term follow-up, especially in the first five years after diagnosis, is essential to manage such patients because of high risk of recurrence and metastasis.

Author contributions Rong He, Peng Zhong, Juntao Hu, Mingying Geng, Jungang Ma contributed to the literature reviewing, writing and manuscript editing. He Xiao, Guangkuo Guo, Lin Lei, Yun Liu provided radiographic data and interpretation. All authors read and approved the final manuscript.

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Data availability All data generated or analyzed during this study are included in this published article.

Code availability Not applicable.

Declarations

Ethics approval and consent to participate This study was approved by the Medical Ethics Committee at Institute of Surgery Research, Third Affiliated Hospital, Army Medical University (Third Military Medical University). A written consent to participate this study was obtained from the patient.

Consent for publication A written consent to permit publication of clinical data was obtained from the patient.

Competing interests The authors declare no competing interests.

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