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Review Article

Solitary fibrous tumor/hemangiopericytoma of the cervical spine: A systematic review of the literature with an illustrative case

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ABSTRACT

Background: In the WHO 2016 classification of central nervous system tumors, solitary fibrous tumors (SFT) and hemangiopericytomas (HPC) were considered part of the same category given a shared mutation. Nevertheless, since the new 2021 WHO classification, the term “hemangiopericytoma” has been retired, and SFT is considered an independent pathological entity.

Methods: We reviewed the literature following preferred reporting items for systematic reviews and meta-analyses guidelines focusing on the treatment options and prognosis of patients with cervical SFT. We also present a 68-year-old female with spinal intradural extramedullary SFT complicated by diffuse extension into paravertebral tissues and muscles.

Results: We found 38 cervical SFT in the literature. Patients averaged 47.3 years of age and 47.4% were female. Typically, these lesions spanned two spinal levels resulting in cord compression and most frequently exhibited benign features (i.e., diagnosed as Grade I SFTs). Interestingly, two patients exhibited distant metastases and had initial pathology consistent with grade II SFT.

Conclusion: SFT of the cervical spine is rare and its management varies according to the histological grade and the clinical behavior, generally warranting surgical excision and adjuvant radiation therapy and/or systemic chemotherapy.

Keywords: Adjuvant chemotherapy, Cervical spine, Hemangiopericytoma, Solitary fibrous tumor, Spinal tumor

INTRODUCTION

Solitary fibrous tumor/hemangiopericytoma (SFT/HPC) are since the new 2021 WHO classification of central nervous system tumors the same entity and account for 1.9–4%

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of intracranial neoplasms, with few being found in the spine.^[6] Despite gross-total surgical excision of spinal lesions (i.e., preferred treatment), higher grade lesions exhibit an increased survival rate but higher recurrence rate (i.e., especially for the WHO-2021 Grade III tumors) despite the utilization of adjuvant radiation or chemotherapy.^[8] Given the recent finding of STAT6 expression in more than 55% of the cases, it is now agreed on most authors that the tumor has a mesenchymal origin.^[3] Here, we present a patient with an intra-extradural cervical WHO-2021 Grade I SFT with diffuse paraspinal extension. As gross-total surgical resection was not feasible due to the high risk to adjacent neurological structures, sub-total tumor resection plus radiation therapy and chemotherapy increased the patient's quality of life and survival (i.e., durable disease control).

MATERIALS AND METHODS

A qualitative review of the literature was performed in compliance with the updated preferred reporting items for systematic reviews and meta-analyses guidelines using the electronic database MEDLINE/PubMed.^[7] The search used "solitary fibrous tumor," "hemangiopericytoma," and "cervical spine." Full-text screening for inclusion was operated by two independent reviewers (F.C. and N.P.F.). Finally, 31 papers were included in the qualitative analysis [Figure 1].

CASE DESCRIPTION

Clinical and radiological presentation

A 68-year-old female presented with a 6-month history of poorly localized cervicalgia, bilateral brachialgia, and

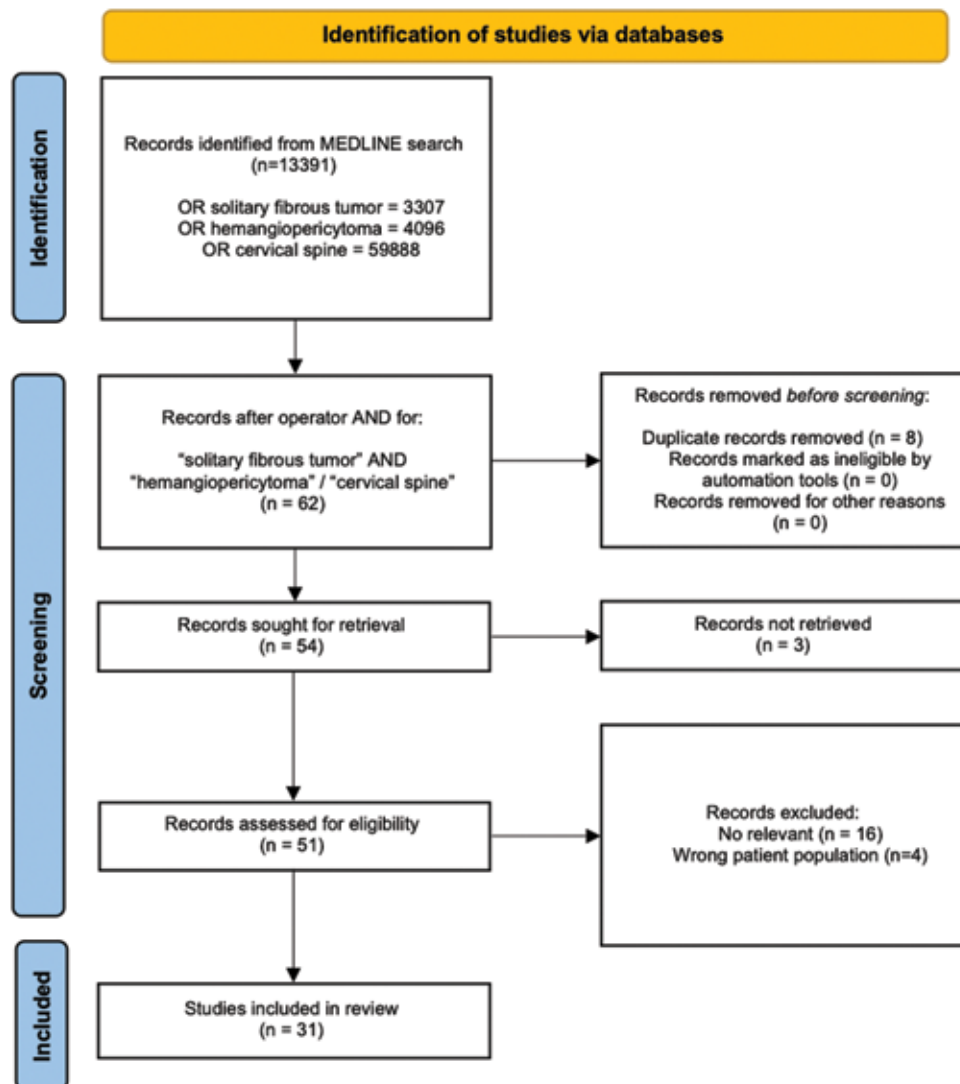


Figure 1: The preferred reporting items for systematic reviews and meta-analyses protocol used for the qualitative review.

hyposthenia of the upper extremities accompanied by progressive 2/5 paresis of the upper extremities with decreased sensation to touch and burning paresthesia in both arms (i.e., increased in the left hand). Proprioception and vibration were intact.

Radiological assessment

The entire spine was evaluated with magnetic resonance imaging (MRI). It revealed a cervical 8 cm, poorly circumscribed, intra-extradural tumor located between C0 and C5 level with epidural extension, and cord compression [Figure 2]. Posterior extension through the left neural foramina of C2–C3 and C3–C4 was also noted (i.e., 4 × 2.5 cm) with involvement of the paraspinal muscles. Anteriorly, it involved C2 and C3 vertebrae and the MRA showed a compromise of the left vertebral artery.

Surgical procedure

The patient agreed to a C2–C5 laminectomy with bilateral lateral mass resection and instrumented fusion to achieve a subtotal

tumor resection (STR) through a posterior-only approach. The paramedian dural incision revealed a white-gray-colored mass compressing the left anterolateral surface of the spinal cord shifting tumor to the right. Tumor was debulked/decompressed, as confirmed by the postoperative MRI [Figure 2e].

Postoperative course and pathology

Postoperatively, the patient improved demonstrating reduced muscular weakness and brachialgia; she was discharged on the 4th postoperative day and referred to a rehabilitation center. She refused a potential secondary anterior tumor resection due to the risk of increased morbidity.

The permanent histological sections documented a pattern consistent with the diagnosis of SFT [Figure 3 and Table 1].

Adjuvant therapy and follow-up

A month after surgical debulking, the patient received radiation therapy (24 Gray) but no further surgery. She also agreed to undergo adjuvant chemotherapy with four

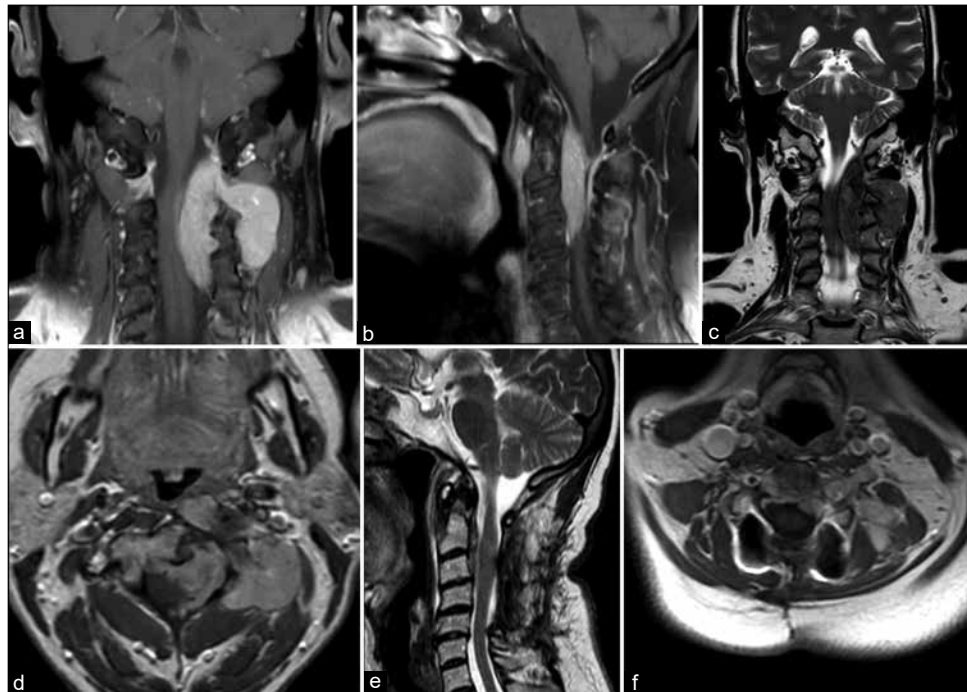


Figure 2: (a) Coronal and (b) sagittal T1-weighted images demonstrating the diffusely infiltrating hyperintense tumor causing marked compression on the left and anterior spinal cord surfaces. Of notice the extraspinal extension through the left third and fourth foramina and the anterior component of the lesion located anteriorly to the third and fourth vertebral bodies. (c) Sagittal T2-weighted scan showing the inhomogeneous hypointensity of the lesion. (d) The paraspinal and the intra-extradural components of the tumor exhibited homogeneous gadolinium enhancement. (e) Postoperative T2-weighted image documented successful decompression of the spinal cord and debulking of the intraspinal component of the tumor. (f) Axial T1-weighted image after the administration of contrast showing considerable reduction of the paraspinal component of the solitary fibrous tumors/hemangiopericytomas following the four cycles of chemotherapy.

cycles of Epirubicin. An MRI following the completion of chemotherapy revealed a sensible reduction of the residual extraspinal component of tumor-infiltrating the paraspinal muscles [Figure 2f]. At 2-year clinical follow-up, she had no symptoms/signs of SFT recurrence.

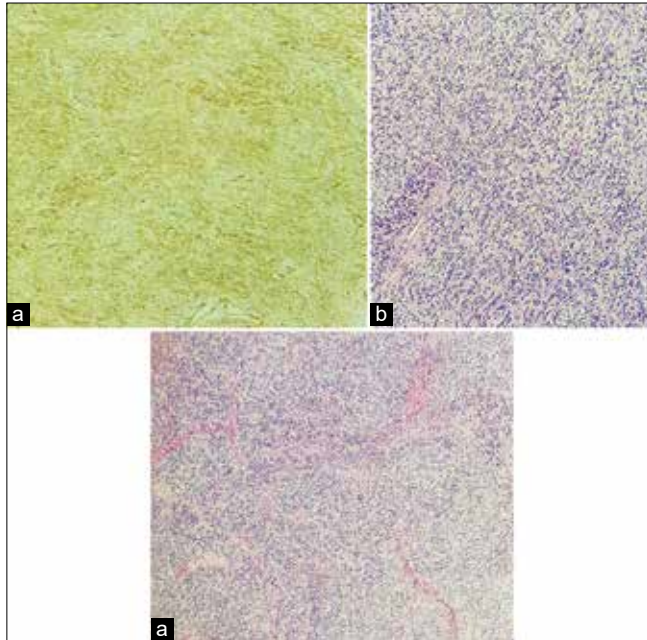


Figure 3: Histological images showing (a) a moderately cellular tumor exhibiting ovoidal and spindle cells arranged in small fascicles and embedded in a collagenous stroma (Hematoxylin eosin). (b) Malignant cells did not show neither signs of cytologic atypia nor necrosis and present a low mitotic count (Hematoxylin eosin). (c) Immunohistochemistry shows strong and diffuse positivity for CD34, a pattern consistent with the diagnosis of solitary fibrous tumors.

Table 1: Clinical data and histology with immunohistochemical summary.

Hematoxylin eosin	Immunohistochemical staining
Spindle cell proliferation without signs of cytologic atypia	Positivity for CD34, Bcl2, and CD99
Mitotic index <1/10 HPF	Negativity for actin, desmin, S100 protein, HMB45, CD31, and EMA

REVIEW OF THE LITERATURE

Patient characteristics

We evaluated 31 articles concerning 38 cases (including this one) of cervical SFT/HPC: 29 case reports and two case series (i.e., largest cohort of five patients). Patients averaged 48.2 years of age for Grade I, 45.7 years old with Grade II SFT, and 40.5 for Grade III HPC lesions. Seven tumors (18.4%) involved one level, while 21 (55.3%) occupied two levels of the spinal cord, three tumors (7.9%) extended to three levels, 5 (10.5%) involved four levels, and two tumors (5.3%) extended for >4 levels [Table 2].

Tumor characteristics and management

In 25 cases (65%), gross-total resection (GTR) of the tumor was performed as the sole treatment, whereas in three cases (7.9%), surgical treatment was followed by radiation therapy and in four patients (10.5%), adjuvant chemotherapy was also deployed.

DISCUSSION

Since 2021, the WHO classified SFT into three grades: a Grade I that corresponds to the highly collagenous, relatively low cellularity with typical spindle cells previously diagnosed as SFT; a Grade II, frequently more cellular and exhibiting less collagenous component with “staghorn” vasculature that was previously designated as HPC; and a Grade III that most often corresponds to what was termed anaplastic HPC.^[6]

In their systematic review, Giordan *et al.*^[2] analyzed a sample of 368 patients, histologically diagnosed with SFT/HPC of the CNS, and found that 70.4% of tumors were primarily intracranial with only 109 cases of spinal involvement. The majority of spinal SFT/HPCs were identified at the thoracic level (45.3%) followed by cervical and lumbar segments with the primary intradural localization reported in 27.5% of patients.

Microscopic examination of this neoplasm demonstrates a patternless architecture composed of spindle-shaped cells with different grades of atypia disposed between thick bands of collagen. Confirmation of the diagnosis is made with immunohistochemical staining, with abnormal cells exhibiting positivity for CD34, CD99, Bcl2, vimentine,

Table 2: Demographic and pathological characteristics of previously reported cases with associated treatment strategies.

SFT Grade	Median age (years)	Sex	Levels involved (number of patients)	Treatment
I (17)	48.2		1 level (7)	25 GTR
II (12)	45.7	18F	2 levels (21)	3 STR+RT
III (6)	40.5	20M	3 levels (3)	4 STR+RT+CHT
NA (2)	63		4 levels (5)	6 NA
			>4 levels (2)	

SFT: Solitary fibrous tumor; GTR: Gross-total resection STR: Sub-total resection RT: Radiation therapy CHT: Chemotherapy

and no signs of reaction for S100 protein and EMA.^[1] Morphological features range from more ill-circumscribed lesions associated with bony erosions, preferably seen in Grade III SFT, to well-defined ovoid or dumbbell-shaped tumors exhibiting less aggressive behavior [Table 1].^[4]

RADIOLOGICAL DIAGNOSIS

Frequently encountered radiological features of SFT include hypo/isointensity on T1-weighted images with a heterogeneous enhancement after the administration of contrast.^[5]

Prognosis

A recent retrospective study Sung *et al.*^[8] comparing treatment results based on the 2016 WHO classification of SFT/HPC revealed relatively benign and indolent courses for grade I lesions, with a 25% of recurrence rate, which was strongly dependent on the volume of the residual tumor after resection. However, more aggressive lesions exhibited a significantly shorter overall survival regardless of the extent of surgical resection or the adjuvant therapy performed.

CONCLUSION

SFT of the cervical spine is rare and relatively benign tumors, whose preferable treatment lays within a combination of surgical tumor resection, radiation therapy, and adjuvant chemotherapy.

Declaration of patient consent

Patients' consent not required as patients' identities were not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest

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