Case Report

Spinal intradural solitary fibrous tumor/hemangiopericytoma with intramedullary invasion mimicking a hemangioblastoma

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ABSTRACT

Background: Solitary fibrous tumor/hemangiopericytomas (SFT/HPCs) are rare mesenchymal tumors of nonmeningothelial origin that comprises <1% of all central nervous system tumors.

Case Description: A 45-year-old male presented with sleep apnea (apnea-hypopnea index was 17.1 events/hour) and dysesthesias of the right upper and lower extremities. The magnetic resonance demonstrated a heterogeneous intradural extra-axial C1 mass with syringobulbia and syringomyelia. The right vertebral angiography revealed a hypervascular mass (i.e., intense tumor staining). With the preoperative diagnosis of a spinal hemangioblastoma, the patient underwent tumor removal. However, intraoperative findings demonstrated that the ventral component of the tumor was intramedullary without a dural attachment. Further, the histological diagnosis was consistent with SFT/HPC (HPC phenotype). The postoperative course was uneventful, and the patient's symptoms and the syrinxes spontaneously regressed.

Conclusion: A 45-year-old male presented a rare spinal intradural lesion at C1 appeared to be a spinal hemangioblastoma, but proved to be SFT/HPC (HPC phenotype) with intramedullary invasion.

Keywords: Hemangioblastoma, Hemangiopericytoma, Intramedullary invasion, Solitary fibrous tumor

INTRODUCTION

Hemangiopericytoma (HPC) and solitary fibrous tumor (SFT) are rare intradural mesenchymal tumors that are difficult to differentiate from meningiomas, schwannomas, ependymomas, astrocytomas, and hemangioblastomas, and constitute <1% of all central nervous system (CNS) tumors. Although SFTs are generally considered benign and HPCs are more malignant, in 2016, the World Health Organization (WHO) combined these two as they share common "inversions" at the genetic site 12q13 (i.e., leading to STAT6 nuclear expression). Here, a 45-year-old male presented with a C1 intradural lesion documented on magnetic resonance (MR) with accompanying syringobulbia/syringomyelia that proved to be intramedullary in location and histologically consistent with a SFT/HPC (HPC phenotype).
CASE REPORT

Clinical presentation

A 45-year-old male presented with numbness in his right forearm, hand, and leg of 2 months’ duration. He was found to have sleep apnea (i.e., polysomnography [PSG] showed apnea-hypopnea index [AHI] of 17.1 events/hour [normal range: <5/h]) and a history of hypertension, hypercholesterolemia, asthma, and a lumbar disc herniation. On examination, he had dysesthesias in the right occipital region, the right C5–C7 distributions, the medial right lower leg, and the medial sole of the foot.

MR evaluation

The cervical MR imaging (MRI) revealed a heterogeneously enhancing right-sided dorsal intradural/extradural mass with pial/subpial invasion most consistent with a spinal hemangioblastoma at the C1 level. It was isointense on T1 MR and moderately hyperintense on T2 MR studies; flow voids surrounded the tumor [Figure 1]. There was accompanying syringomyelia and syringobulbia, and there was no enhancement of the cyst/syrinx walls [Figure 1]. The right vertebral angiography documented a hypervascular C1 lesion fed by the C1/C2 radicular arteries [Figure 2].

Surgery

A suboccipital craniotomy, with laminectomy of the atlas, and cephalad laminotomy of C2 (C0-C2) were performed utilizing neurophysiological monitoring. Once the dura was opened, a bright red vascular lesion was identified on the dorsal aspect of the cord. However, the tumor's ventral component was intramedullary in location. Once the pial feeding vessels were coagulated, the tumor was circumferentially separated and resected en bloc [Figure 3].

Histopathology

Histopathology confirmed that the tumor was a WHO Grade II SFT/HPC (HPC phenotype). It demonstrated round/spindle cell proliferation and compact chromatin-stained nuclei that were relatively uniform without mitotic figures (i.e., consistent with a benign lesion). Immunohistochemistry was positive for CD34 and CD31 and negative for Bcl-2; the Ki-67 index was 2%. In addition, many small and large blood vessels with characteristic Staghorn vasculature were evident throughout [Figure 4].

Postoperative course

Postoperatively, the numbness of the right upper and lower extremities improved. The postoperative PSG showed...
improvement as his AHI decreased to 0.4 events/h, and his dysphagia disappeared. No further radiation was scheduled due to the complete tumor excision. Eight years later, he has exhibited no tumor recurrence [Figure 5].

**DISCUSSION**

**Case summary**

SFT/HPC, nonmeningothelial mesenchymal tumors that are rarely intradural/extramedullary account for <1% of all CNS tumors [Table 1].[^1] Here, the MRI revealed a well-demarcated and intensely enhancing mass on the dorsal surface of the

**Table 1: Summary of spinal intradural SFT/HPC with subpial invasion in the literature.**

| Characteristics                  | n  |
|----------------------------------|----|
| Total patients[^1-8]             | 34 |
| Age (years)                      |    |
| Mean                             | 48.1|
| Range                            | 15–83|
| Sex                              |    |
| Male                             | 21 |
| Female                           | 13 |
| Original diagnosis[^1-8]         |    |
| SFT                              | 23 |
| HPC                              | 11 |
| Location[^1-8]                   |    |
| Cervical                         | 12 |
| Cervicothoracic                  | 2 |
| Thoracic                         | 18 |
| Lumbar                           | 2 |
| Preoperative diagnosis[^1-8]     |    |
| Meningioma                       | 7 |
| Schwannoma                       | 6 |
| Hemangioblastoma                 | 5 |
| Ependymoma                       | 3 |
| Neurofibroma                     | 3 |
| Astrocytoma                      | 2 |
| Others                           | 7 |

HPC: Hemangiopericytoma, N: Total number of patients, SFT: Solitary fibrous tumor

[^1]: [3]
cord surrounded by vascular flow voids (i.e., hypervascular on vertebral angiogram) with accompanying syringobulbia/ syringomyelia prompting the preoperative diagnosis of a hemangioblastoma.[4] However, at surgery, this 45-year-old male's C1 lesion was both intradural and intramedullary (i.e., subpial invasion), and pathologically proved to be a SFT/HPC (HPC phenotype).

**Literature summary**

We found 34 cases of SFT/HPC cases (i.e., including this case) in our review of case studies from the literature [Supplemental Table 1].[1-8] Patients averaged 48.1 years of age, and there was a 1.6:1 male-to-female ratio [Table 1].[1-8] Tumors were typically located in the lower cervical region followed by the upper and midthoracic regions, but were only rarely encountered in the lumbar spine [Figure 6]. T1 HPC: Hemangiopericytoma, MRI: Magnetic resonance imaging, SFT: Solitary fibrous tumor, WI: Weighted image

MR findings for SFT/HPC were nonspecific. However, the hypointensity on T2 MR best correlated with the collagen-rich content, hypocellularity, and calcification of the SFT [Table 2].[2] The additional findings of syrinx/edema in the spinal cord were also common with intradural SFT/HPC and subpial invasion [Table 2].

**Treatment and long-term prognosis**

Following WHO 2016 guidelines, gross total resection (GTR) with radiotherapy is the mainstay for the treatment of the CNS SFT/HPC that typically result in good outcomes [Table 3].[3] In contrast subtotal resections usually have high recurrence and metastasis rate and are fatal.[7,8] Due to paucity of strong evidence, the role of postoperative radiotherapy in recurrence-

**Table 2:** Radiological characteristics of the spinal intradural SFT/HPC with subpial invasion reported in the literature.

| MRI | SFT | HPC | SFT/HPC |
|-----|-----|-----|---------|
|     | T1WI | T2WI | T1WI | T2WI | T1WI | T2WI |
| Isointense (%) | 69.6 | 0 | 100 | 18.2 | 79.4 | 5.9 |
| Hypointense (%) | 26.1 | 82.6 | 0 | 9.1 | 17.7 | 58.8 |
| Hyperintense (%) | 4.3 | 13 | 0 | 72.7 | 2.9 | 32.4 |
| Syrinx/edema (%) | 8.7/69.6 | 27.3/36.4 | 14.7/58.8 |
| Homogeneous enhancement (%) | 87 | 63 | 79.4 |

0 2 4 6 8

Figure 5: The postoperative T2-weighted magnetic resonance imaging 6 years after the operation shows complete tumor resection with a resolution of syringobulbia and syringomyelia.

Figure 6: Anatomic location of the tumor. The tumor was most commonly located in the thoracic region followed by the cervical and lumbar levels.

Figure 7: Anatomic location of the tumor. The tumor was most commonly located in the thoracic region followed by the cervical and lumbar levels.
free survival is yet to be established.[2,5,6] Our patient, GTR was made possible by a clear plane between the tumor and cord, allowing for the 8-year follow-up without tumor recurrence.

CONCLUSION

Here, we reported a 45-year-old male with a C1 spinal intradural SFT/HPC (HPC phenotype) with an intramedullary invasion that resulted in MR-documented syringomyelia and syringobulbia. Following GTR, the patient remained asymptomatic 8 years later.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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| No | Author | Year | Age | Sex | Phenotype | Location | Treatment | Recurrence | FU month | Preoperative diagnosis | Angio-gram | MRI T1WI | MRI T2WI | MRI Gd-DTPA | MRI remark |
|----|--------|------|-----|-----|-----------|----------|-----------|------------|----------|------------------------|-------------|----------|----------|------------|------------|
| 1  | Kanahara | 1999 | 62  | M   | SFT       | C6-C7    | NA        | NA         | NA       | NA                     | NA          | Hypo     | Hypo     | Marginally enhanced Edema |
| 2  | Mordani | 2000 | 33  | M   | SFT       | C5       | GTR       | No         | 18       | Schwannoma, neurofibroma, meningioma, astrocytoma, ependymoma | NA          | Hypo     | Iso      | Heterogeneous Edema       |
| 3  | Kawamura | 2004 | 64  | M   | SFT       | T2-T3    | STR       | No         | 6        | Schwannoma, neurofibroma, meningioma, astrocytoma, ependymoma | NA          | Hypo     | Hypo     | Homogeneous Edema          |
| 4  | Bohinski | 2004 | 49  | F   | SFT       | C4       | GTR       | No         | 10       | Schwannoma, neurofibroma, meningioma, astrocytoma, ependymoma | NA          | Iso      | Hypo     | Homogeneous Edema          |
| 5  | Jallo   | 2005 | 59  | M   | SFT       | T5       | GTR       | No         | 56       | Intradural tumor       | NA          | Iso      | Hypo     | Homogeneous NA             |
| 6  | Jallo   | 2005 | 37  | F   | SFT       | T2-3     | GTR       | No         | 60       | Intradural tumor       | NA          | Iso      | Hypo     | Homogeneous NA             |
| 7  | Jallo   | 2005 | 41  | M   | SFT       | C6-C7    | GTR       | No         | 41       | Intradural tumor       | NA          | Iso      | Hypo     | Homogeneous Edema          |
| 8  | Jallo   | 2005 | 17  | M   | SFT       | T5-T6    | GTR       | No         | 18       | Intradural tumor       | NA          | Iso      | Hypo     | Homogeneous Edema          |
| 9  | Ogawa   | 2005 | 63  | F   | SFT       | T11      | GTR       | No         | 18       | NA                     | NA          | Iso      | Hypo     | Homogeneous Edema          |
| 10 | Ogunbbo | 2005 | 53  | M   | SFT       | C3-C4    | GTR       | NA         | NA       | Hemangioblastoma        | NA          | Iso      | Hypo     | Homogeneous Syrinx         |
| 11 | Kashiwazaki | 2007 | 31  | M   | HPC      | T4-T6    | GTR       | No         | 36       | Hemangioblastoma, subpial schwannoma | NA          | Iso      | Hyper   | Heterogeneous Edema flow void |
| 12 | Endo    | 2008 | 62  | M   | HPC      | L1       | GTR       | NA         | 48 days  | Primary neurogenic tumor, meningioma Schwannoma, neurofibroma, meningioma, astrocytoma, ependymoma | NA          | Iso      | Hyper   | Homogeneous Edema          |
| 13 | Chou    | 2009 | 80  | M   | HPC      | T10      | GTR       | No         | 36       | Primary neurogenic tumor, meningioma Schwannoma, neurofibroma, meningioma, astrocytoma, ependymoma | NA          | Iso      | Hyper   | Homogeneous Edema          |
| 14 | Ishii   | 2009 | 63  | F   | SFT       | C5       | GTR       | No         | 14       | Schwannoma, neurofibroma, meningioma, astrocytoma, ependymoma | NA          | Iso      | Hyper   | Homogeneous Edema          |
### Supplemental Table 1 (Continued)

| No.  | Author           | Year/Year   | Age | Sex | Phenotype  | Location | Treatment | Recurrence | FU | Preoperative diagnosis | Angio-gram | MRI T1WI | MRI T2WI | GAD-TPA | MRI remark | Remark |
|------|------------------|-------------|-----|-----|------------|----------|-----------|------------|----|------------------------|------------|----------|----------|----------|------------|--------|
| 15   | Ciappetta        | 2010        | 75  | F   | SFT        | T6-T7    | GTR       | No          | 24 | NA                     | 15         | Hypo     | Iso      | Hypo     | Homogenous | Edema  |
| 16   | Ackerman         | 2011        | 58  | M   | HPC        | T10      | GTR       | No          | 24 | NA                     | 16         | Hypo     | Iso      | Hypo     | Homogenous | Edema  |
| 17   | Moscovici        | 2011        | 20  | F   | SFT        | C4-C7    | GTR       | No          | 12 | NA                     | 17         | Hypo     | Iso      | Hypo     | Homogenous | Edema  |
| 18   | Mariniello       | 2012        | 67  | M   | SFT        | C7-T1    | GTR       | No          | 12 | NA                     | 18         | Hypo     | Iso      | Hypo     | Homogenous | Edema  |
| 19   | Ackerman         | 2011        | 67  | M   | HPC        | T5-T6    | GTR       | No          | 24 | NA                     | 19         | Hypo     | Iso      | Hypo     | Homogenous | Edema  |
| 20   | Liu/2013         | 2013        | 58  | M   | Meningioma, schwannoma, neurofibroma, ependymoma | No | 20 | Iso | Hypo | Homogeneous, Edema | |
| 21   | Liu/2013         | 2013        | 32  | F   | HPC        | T5-T6    | GTR+RT    | No          | 6  | NA                     | 21         | Hypo     | Iso      | Hypo     | Homogenous | Edema  |
| 22   | Moscovici        | 2011        | 67  | M   | SFT        | C4-C7    | GTR       | No          | 12 | NA                     | 22         | Hypo     | Iso      | Hypo     | Homogenous | Edema  |
| 23   | Robert           | 2014        | 49  | F   | HPC        | T9-T10   | GTR       | No          | 6  | NA                     | 23         | Hypo     | Iso      | Hypo     | Homogenous | Edema  |
| 24   | Tur/2015         | 2015        | 19  | F   | HPC        | C2       | GTR       | No          | 6  | NA                     | 24         | Hypo     | Iso      | Hypo     | Homogenous | Edema  |
| 25   | Tur/2015         | 2015        | 15  | F   | HPC        | C7-T1    | GTR       | No          | 6  | NA                     | 25         | Hypo     | Iso      | Hypo     | Homogenous | Edema  |
| 26   | Bruder           | 2015        | 83  | M   | SFT        | T8-T9    | GTR       | No          | 8  | Meningioma             | 26         | Hypo     | Iso      | Hypo     | Homogenous | Edema  |
| 27   | Walker           | 2015        | 47  | F   | SFT        | L1       | GTR       | No          | 12 | Ependyma, cavernoma    | 27         | Hypo     | Iso      | Hyper   | Heterogeneous | Edema |
| 28   | Wang/2016        | 2015        | 31  | M   | HPC        | T9-T10   | GTR       | No          | 6  | Meningioma             | 28         | Hypo     | Iso      | Hyper   | Heterogeneous | Edema |
| 29   | Y 2019           | 2019        | 35  | F   | HPC        | C4-C7    | GTR       | No          | 6  | Meningioma             | 29         | Hypo     | Iso      | Hyper   | Heterogeneous | Edema |
| 30   | Chungyang        | 2016        | 80  | F   | HPC        | T4-T5    | GTR       | No          | 10 | NA                     | 30         | Hypo     | Iso      | Hyper   | Homogeneous | Edema  |
| 31   | Rodriguez-Mena   | 2016        | 30  | M   | SFT        | C2-C3    | GTR       | No          | 24 | NA                     | 31         | Hypo     | Iso      | Hyper   | Homogeneous | Edema  |
| 32   | Rodriguez-Mena   | 2016        | 41  | M   | SFT        | C5-C7    | GTR       | No          | 36 | NA                     | 32         | Hypo     | Iso      | Hyper   | Homogeneous | Edema  |
| 33   | Yokoyama         | 2020        | 47  | F   | SFT        | T1-T2    | GTR       | No          | 6  | Meningioma             | 33         | Hypo     | Iso      | Hyper   | Homogeneous | Edema  |
| 34   | Present case     | 2021        | 31  | F   | SFT        | C6-C7    | GTR       | No          | 6  | Meningioma             | 34         | Hypo     | Iso      | Hyper   | Homogeneous | Edema  |

HPC, hemangiopericytoma; GTR, gross total resection; MRI, magnetic resonance imaging; NA, not available; OP, operative; SFT, solitary fibrous tumor; WI, weighted imaging; CT, computed tomography; Angio-gram, angiogram.