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Case Report

Cervical intramedullary solitary fibrous tumor: Case report and review of the literature

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ABSTRACT

Background: Solitary fibrous tumors (SFTs) are benign tumors derived from mesenchymal tissues that predominantly occur in the pleura. Establishing the diagnosis of these very rare intramedullary spinal lesions, with no clear-cut pathognomonic radiographic characteristics, is particularly challenging.

Case Description: Two males, 30 and 41 years of age, presented with progressive cervical myelopathies attributed to a cervical intramedullary exophytic tumor with associated spinal cord edema. One patient showed that the lesion was highly vascularized. Both patients underwent surgical excision of firm, solid, focal, and, particularly in one of them, very vascular/hemorrhagic tumors; at surgery, there was some adherence between the tumors and the cord tissue, but gross-total resections were achieved in both cases, demonstrated on postoperative MR scans. Histological and immunohistochemical findings confirmed the diagnosis of SFT (WHO Grade I). After a 6-month postoperative period, both patients neurologically improved and had no MR evidence of tumor recurrence.

Conclusion: Intramedullary cervical exophytic SFTs are extremely rare. Although these solid tumors may present with hemorrhagic features and at surgery demonstrate significant adherence to the pial/cord surface, complete surgical resections are feasible resulting in good outcomes.

Keywords: Cervical, Intramedullary, Solitary fibrous tumor, Surgery, Vascularized

INTRODUCTION

Intramedullary cervical solitary fibrous tumors (SFTs) are extremely rare. Their low rate of presentation within the spinal cord itself makes the preoperative radiologic diagnosis and surgical planning, particularly challenging.[3-8] Gross-total surgical removal should be attempted to minimize the risk of tumor recurrence and improve patients' quality of life.[1,4] Here, we present two cases of cervical intramedullary SFTs that were completely removed with good clinical outcomes.

CASE REPORT # 1

A 30-year-old male presented a 3-month history of neck pain and a progressive cervical myelopathy. The MR showed an intramedullary exophytic well-circumscribed C2-C3 mass with

associated spinal cord edema inferior to the lesion. The tumor was isointense on T1-weighted image (T1WI), hyperintense on T2-weighted image (T2WI), and showed homogeneous gadolinium contrast enhancement. Digital subtraction angiogram (DSA) showed rich tumor vascularization with feeding branches from both of the vertebral arteries, including a right lateral spinal artery descending from the right posteroinferior cerebellar artery (PICA) [Figure 1a-e]. At surgery, the intramedullary tumor was solid/firm and hemorrhagic; it also demonstrated an exophytic component. Despite partial adherence to the cord, it was freed from the pia, allowing for piecemeal gross-total resection (GTR). The patient's postoperative course was uneventful. The immediate postoperative MR confirmed GTR [Figure 1f-g]. After 6 months, he fully recovered neurologically, and at 2 years postoperatively, the MR demonstrated no tumor recurrence.

Histology

The histological examination confirmed the diagnosis of a SFT. It revealed uniform spindle cells surrounded by moderate fibrous stroma with intermingled collagenous fibers and a few well-demarcated branched blood vessels ("staghorn" vasculature). The overall cell population barely had discrete cytological atypia and mitotic activity, and there was no evidence of necrosis. On immunohistochemistry, the neoplasm was diffusely and strongly positive for BcL-2, CD34, and CD99 and negative for S-100, glial fibrillary acidic protein (GFAP), and epithelial membrane antigen (EMA). The Ki-67 proliferation index was 1%, and the conclusive diagnosis of a SFT with no malignant features (WHO Grade I) was made.

CASE REPORT # 2

A 41-year-old male presented with a 2-month history of a slowly progressive cervical myelopathy. The MR demonstrated an intramedullary exophytic tumor at C5-C7. The lesion was isointense on T1WI, heterogeneous on T2WI (predominantly hypointense with alternating smaller hyperintense areas), and homogeneously enhanced with contrast [Figure 2a-c]. At surgery, this intramedullary lesion had exophytic components. Despite a thin attachment to the pia mater at the anterior left side of the mass and to the ipsilateral C6 exiting nerve root, a GTR was achieved. This was confirmed on the immediate postoperative MR [Figure 2d-e]. He was neurologically normal 7 months postoperatively, and no tumor regrowth was seen on the MR 3 years later.

Histology

The pathological analysis was consistent with an SFT. It showed uniform small, basophilic, ovoid to spindled cells with oval nuclei, and ill-defined cytoplasm, with numerous thin-walled ramifying blood vessels in certain areas. Necrosis and mitosis were not evident. Immunohistochemical staining indicated strong positivity for CD34 and Bcl-2 (C). The Ki-67 proliferation index was less than 1%. The final diagnosis of a SFT with no malignant features (WHO Grade I) was then established [Figure 3].

DISCUSSION

Inside the CNS, spinal cord occurrence of SFT is exceedingly uncommon; there have been 22 cases reported [Table 1].

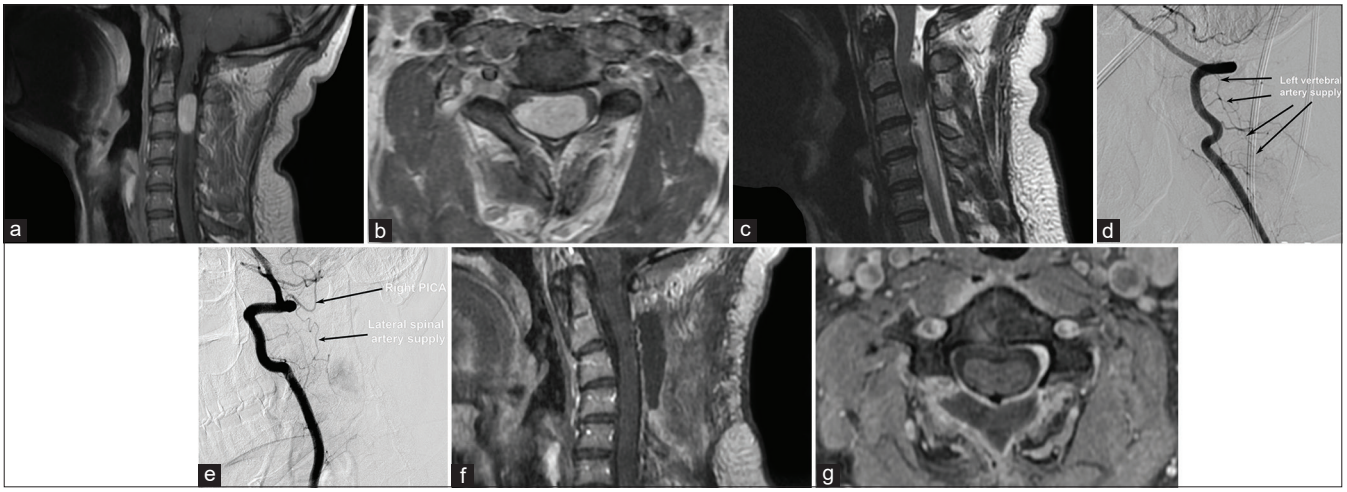

Figure 1: Sagittal (a) and axial (b) T1-weighted magnetic resonance image (T1WI) showing an intramedullary exophytic C2-C3 tumor with homogeneous gadolinium enhancement. (c) T2-weighted images showing a hyperintense mass with associated spinal cord edema inferior to the lesion with signal void images in the upper cervical spine representing prominent vessels within the cerebrospinal fluid. (d and e) Feeding branches from both vertebral arteries were visualized on digital subtraction angiogram (black arrows in d), including a right lateral spinal artery (black arrow on e) descending from the right posteroinferior cerebellar artery (black arrow on e). Sagittal (f) and axial (g) contrast T1WI confirming complete tumor resection.

Table 1: Summary of reported cases of intramedullary solitary fibrous tumors in the literature.

Author	Age/sex	Location	Neurologic presentation	Intraoperative findings				Follow-up	Long-term neurologic outcome	Recurrence
				Extramedullary exophytic component	Tumor/spinal cord clear interphase	Highly vasc.	Extent of resection			
Carneiro *et al.*, 1996	50/F	-	Bilateral lower limb pain, numbness, and weakness + incontinence	Yes	-	-	Subtotal	6 years	Improved	Yes
Alston *et al.*,1997	47/M	T4-T5	Brown-Sequard syndrome	No	Yes	No	Total	2 months	Improved	No
Kanahara *et al.*, 1999	62/M	C6-C7	Lower limb numbness	Yes	Yes		-	-	-	-
Mordani *et al.*, 2000	33/M	C5	Cervical myelopathy (sensory-motor deficit)	No	Yes	Yes	Total	18 months	-	No
Tihan *et al.*, 2003	-	-	-	No	Yes	-	-	-	-	-
Tihan *et al.*, 2003	-	-	-	No	Yes	-	-	-	-	-
Kawamura *et al.*, 2004	64/M	T2-T3	Brown-Sequard syndrome	Yes	No	-	Subtotal	6 months	Improved	No
Bohinski *et al.*, 2004	49/F	C4	Neck pain, arm dysesthesia	Yes	Yes	-	Total	10 months	Stable	No
Pizzolitto *et al.*, 2004	47/M	C4	Four-limb paresthesia; diminished light touch and pin-prick sensation	No	Yes	-	Total	12 months	Improved	No
Jallo *et al.*, 2005	41/M	C6-C7	Dysesthesias and upper limb weakness	Yes	No	No	Total	3.5 years	Stable	No
Jallo *et al.*, 2005	17/M	T5-T6	Back pain and spastic paraparesis	No	No	-	Total	1.6 years	Improved	No
Ishii *et al.*, 2009	63/F	C5	Sensory and motor deficit in right upper limb	No	Yes	Yes	Total	14 months	Improved	No
Ciappetta *et al.*, 2010	75/F	T6-T7	Bilateral lower limb weakness and paresthesia	Yes	-	Yes	Total	2 years	Improved	No
Fargen *et al.*, 2011	28/F	C2-C3	Face, neck, and shoulder pain + lower limb numbness	Yes	-	-	Total	2 years	No NL deficits	Nodular enhancement
Mariniello *et al.*, 2012	75/F	T6-T7	Bilateral lower limb weakness and paresthesia	No	-	Yes	Total	1 year	Improved	No
Robert *et al.*, 2014	49/F	T9-T10	Bilateral lower limb numbness and paresthesia	Yes	No	-	Partial	6 months	No NL deficits	No

(Contd...)

Table 1: *(Continued)*

Author	Age/sex	Location	Neurologic presentation	Intraoperative findings				Follow-up	Long-term neurologic outcome	Recurrence
				Extramedullary exophytic component	Tumor/spinal cord clear interphase	Highly vasc.	Extent of resection			
Hwang *et al.*, 2014	48/F	C7-T1	Sensory deficit right side below T5	Yes	No	Yes	Partial	6 months	Stable	No
Bruder *et al.*, 2015	83/F	T8-T9	Left lower limb sensory and motor deficits; incontinence	Yes	Yes	No	Total	8 months	Improved	No
Walker *et al.*, 2015	47/F	L1	Pain and weakness in the right lower limb	No	No	Yes	Total	1 year	Improved	No
Wang *et al.*, 2016	31/M	T1-T5	Bilateral lower limb weakness and paresthesia	-	-	-	Total	-	-	-
Mansilla *et al.*, 2019	48/F	T3-T4	Bilateral lower limb sensory deficit	Yes	Yes	-	Total	7 months	Stable	No
Yang *et al.*, 2019	35/F	C6-T1	Lower limb numbness and weakness; incontinence	No	No	Yes	Total	23 months	Improved	No
Present case	30/M	C3-C5	Bilateral upper limb motor and sensory deficits	Yes	Yes	Yes	Total	2 years	Improved	No
Present case	41/M	C5-C7	Weakness and numbness in lower limbs and hands; incontinence	Yes	Yes	No	Total	3 years	Improved	No

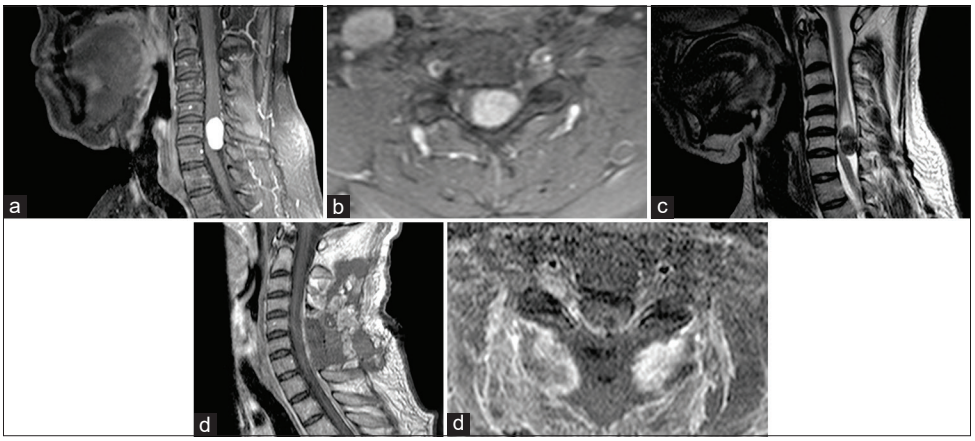

Figure 2: Sagittal (a) and axial (b) T1-weighted magnetic resonance image (T1WI) showing an intramedullary exophytic C5-C7 tumor with homogeneous gadolinium enhancement. (c) Slight heterogeneous aspect on T2-weighted images (predominantly hypointense with alternating smaller hyperintense areas). Sagittal (d) and axial (e) T1WI indicating complete removal of the tumor.

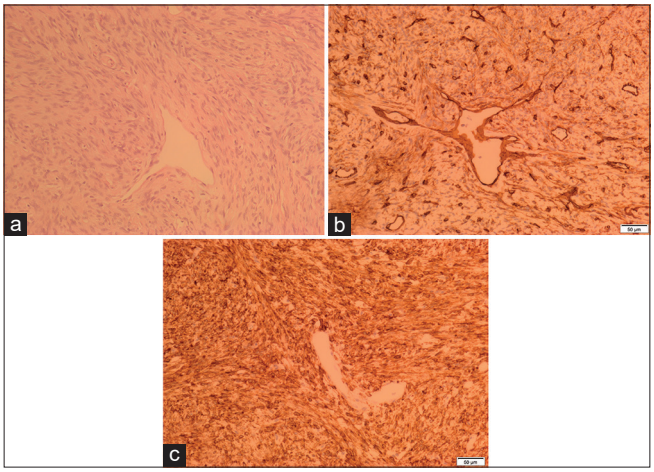

Figure 3: Representative pathologic images: (a) uniform small, basophilic, ovoid to spindle cells with oval nuclei and ill-defined cytoplasm, and numerous thin-walled ramifying blood vessels. (Hematoxylin and eosin stain). Strong staining is present for CD34 (b) and bcl-2 (c).

SFT occurs predominantly in middle-aged patients, with a modest male preponderance.[1] Clinical findings include localized pain and neurological deficits reflecting the level of intramedullary involvement. Since the majority of the SFT are considered benign, symptoms are attributed to their mass effect which can be best relieved with gross-total excision.

Radiographic findings

On MR, SFTs can be seen as a single, oval or irregular heterogeneous mass, sometimes having well-circumscribed margins, and tend to appear isointense on T1WI and hypointense on T2WI.[7,10] They markedly enhance with contrast (homogeneous or heterogeneous). Therefore, detailed examination and recognition of feeding branches (e.g., from vertebral arteries and PICA on the cervical region) on MRI, but specifically on DSA, are strongly recommended.

Histopathology

Histopathologically, SFT cells are encircled by dense collagen networks in fascicular, storiform, herringbone, or patternless arrangements on hematoxylin and eosin staining.[7] On immunohistochemistry, positivity for CD34, CD 99, vimentin, and BcL-2 and negativity for EMA, smooth-muscle actin, and S-100 are distinguishing features of SFT.[7,8]

Gross-total excision

Gross-total removal is the recommended treatment of spinal SFT (under intraoperative neurophysiological monitoring) [3-5,7] Here, we presented two well-circumscribed, firm, highly vascularized cervical SFT with strong adhesions to the spinal cord tissues, accompanied by exophytic components. These lesions may exhibit numerous feeding vessels from both vertebral arteries most readily diagnosed with DSA. In spite of their benign histology, and a 5-year survival rate of nearly 100%, recurrence (e.g., up to 2–14 years) has been noted with incomplete tumor resections.[1-3,8,9]

CONCLUSION

Here, we described two cases of intramedullary cervical exophytic SFTs that were completely excised and have not yet recurred, 2 and 3 years postoperatively.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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