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.¹⁻³ 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.

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 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].
| Author               | Age/sex | Location | Neurologic presentation                                      | Extramedullary exophytic component | Intraoperative findings | Follow-up | Long-term neurologic outcome | Recurrence |
|---------------------|---------|----------|-------------------------------------------------------------|-----------------------------------|-------------------------|-----------|------------------------------|------------|
| Carneiro et al., 1996 | 50/F    | -        | Bilateral lower limb pain, numbness, and weakness + incontinence | Yes                               | No                      | Subtotal  | 6 years                      | Improved   | Yes                     |
| Alston et al., 1997  | 47/M    | T4-T5    | Brown-Sequard syndrome                                      | No                                | Yes                     | Total     | 2 months                     | Improved   | No                      |
| Kanahara et al., 1999| 62/M    | C6-C7    | Lower limb numbness                                         | No                                | Yes                     | -         | -                            | -          |
| Mordani et al., 2000 | 33/M    | C5       | Cervical myelopathy (sensory-motor deficit)                 | No                                | Yes                     | Total     | 18 months                    | -          |
| Tihan et al., 2000   | -       | -        | -                                                           | No                                | Yes                     | -         | -                            | -          |
| Kawamura et al., 2004| 64/M    | T2-T3    | Brown-Sequard syndrome                                      | No                                | Yes                     | 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    | Dysesthasias and upper limb weakness                         | Yes                               | 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                     | Total     | 14 months                    | Improved   | No                      |
| Ciappetta et al., 2010| 75/F   | T6-T7    | Bilateral lower limb weakness and paresthesia                | 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                      |
| Author          | Age/sex | Location | Neurologic presentation                                                                 | Intraoperative findings                        | Follow-up | Long-term neurologic outcome | Recurrence |
|-----------------|---------|----------|----------------------------------------------------------------------------------------|-----------------------------------------------|-----------|-------------------------------|------------|
| 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          |           |                               |            |
SFT occurs predominantly in middle-aged patients, with a modest male preponderance. 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 with well-circumscribed margins, and tend to appear isointense on T1WI and hypointense on T2WI. 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. On immunohistochemistry, positivity for CD34, CD99, vimentin, and Bcl-2 and negativity for EMA, smooth-muscle actin, and S-100 are distinguishing features of SFT.

**Gross-total excision**

Gross-total removal is the recommended treatment of spinal SFT (under intraoperative neurophysiological monitoring). 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.

**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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Conflicts of interest
There are no conflicts of interest.

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