Case report

Solitary fibrous tumour with intramedullary component: Case report and review of the literature

Thomas Robert a, *, Christophe Duc b, Diego San Millán Ruíz c, Marc Morard a

a Department of Neurosurgery, Hôpital de Sion, Réseau Santé Valais, Sion, Switzerland
b Department of Pathology, Institut Central des Hôpitaux Valaisans, Sion, Switzerland
c Department of Radiology, Hôpital de Sion, Réseau Santé Valais, Sion, Switzerland

1. Introduction

Solitary fibrous tumour (SFT) is a rare neoplasm of mesenchymal origin that arises most commonly in the visceral pleura [1]. Solitary fibrous tumour can occur in a variety of other sites [2–5], including the central nervous system (CNS). Solitary fibrous tumours in CNS are usually dural based, meningioma-like masses [6–8]. Only few isolated cases of intramedullary SFT are described in the literature [9–17]. We report a case of intramedullary SFT with extramedullar component and review the literature.

2. Case report

A 49-year-old Caucasian woman presented for over one-year history of bilateral lower extremities numbness and paresthesia predominately on the right side associated with progressive inability to walk. She also described nocturnal back pain which forced her to stand up. General physical examination was normal. Neurological examination revealed decreased pain, light touch and joint position sensations below the T12 level. The legs were severely spastic, rendering the walk difficult. Increased deep tendon reflexes were demonstrated...
bilaterally in knees and ankles. There were no subjective or objective sphincter disturbances.

Magnetic resonance imaging (MRI) revealed an intradural mass measuring 13 mm longitudinally at the level T9–T10 (Fig. 1). The cranial component of the tumour seemed to be intramedullary and involved the posterior horns bilaterally. The caudal part was extramedullary and bulged posteriorly. There was no apparent infiltration of the dura mater or the posterior nerve roots. The mass was strongly hypointense to the spinal cord on spin-echo T2-weighted images and echo-gradient T2-weighted images with no blooming effect suggesting high cellularity. The mass appeared mildly hypointense to the spinal cord on T1-weighted images with homogenous enhancement after gadolinium administration. The lesion was surrounded by intramedullary vasogenic oedema predominantly around the upper aspect of the tumour, extending over one vertebral level. No dural attachment and no “duro–tail” were demonstrated on the gadolinium-enhanced studies.

Surgery was performed by a T9–T10 laminectomy approach. The tumour was definitely intramedullary in origin but was eccentric at the caudal aspect. The tumour appeared immediately, covered by an arachnoid layer. The tumour arose clearly

---

Fig. 1 – (A) Sagittal T2-weighted magnetic resonance (MR) image demonstrates a low-intensity intramedullary tumour surrounded by oedema. (B) Sagittal T1-weighted MR image confirms the presence of isointense tumour. Post-gadolinium sagittal (C) and axial (D) MR images reveal homogeneous enhancement.
from inside the medulla which seemed to split in the midline. The cranial part of the tumour was imbedded in the medulla while the caudal part extruded and could appear as extra-medullary. The tumour was very hard and ultrasonic aspirator could not aspirate it. We performed a piecemeal intracavitary excision. There was no cleavage plane between the tumour and the surrounded medulla. According to this, a complete excision of the tumour would have harboured a tremendous risk of neurological worsening and we decided to do a partial excision (Fig. 2).

The histological examination of the tumour demonstrated a solid proliferation of spindle-shaped cells with a patternless or fascicular growth pattern associated to a collagenous matrix background (Fig. 3). The immunohistochemical staining showed a strong and diffuse positivity for CD34 and Bcl-2. There was no mitosis and no necrosis. The proliferation index of the tumour, analysed with Mib-1 (Ki-67), was smaller than 1%. The tumour cells were negative for keratin, EMA, CD117, DOG1 and S-100 protein. The negativity for S-100 argued against nervous tumour such as schwannoma or neurofibroma; the negativity for EMA – against perineurinoma or meningioma. The overall morphology of the tumour was consistent with a SFT.

There was worsening of the joint position sense and the walk on the first postoperative week, but this had totally recovered in one month. Postoperative MRI demonstrated partial removal of the lesion with increased intramedullary oedema. It increased even if the patient has been operated.

Six months postoperatively, the neurological examination was normal and the patient was back to normal life and to work.

### 3. Discussion

Solitary fibrous tumour is a mesenchymal tumour that arises most commonly in the visceral pleura [1]. Histologically, the
<table>
<thead>
<tr>
<th>Described by</th>
<th>Year</th>
<th>Age</th>
<th>Sex</th>
<th>Location</th>
<th>Signs</th>
<th>Aspect</th>
<th>Treatment</th>
<th>Clinical outcome</th>
<th>Recurrence</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Carneiro et al. [9]</td>
<td>1996</td>
<td>50</td>
<td>M</td>
<td>Conus</td>
<td>Cono-caudal syndrome</td>
<td>Intra- and extramedullary</td>
<td>Total removal</td>
<td>Totally improved</td>
<td>Yes</td>
<td>5 years</td>
</tr>
<tr>
<td>Alston et al. [10]</td>
<td>1997</td>
<td>47</td>
<td>M</td>
<td>T4–T5</td>
<td>Brown-Sequard syndrome</td>
<td>Purely intramedullary</td>
<td>Total removal</td>
<td>Partially improved</td>
<td>No</td>
<td>2 months</td>
</tr>
<tr>
<td>Kanahara et al. [11]</td>
<td>1998</td>
<td>62</td>
<td>M</td>
<td>C6–C7</td>
<td>Cervical myelopathy</td>
<td>Dorsal portion, partially</td>
<td>Total removal</td>
<td>ND</td>
<td>Not described</td>
<td>Not described</td>
</tr>
<tr>
<td>Mordani et al. [12]</td>
<td>2000</td>
<td>33</td>
<td>M</td>
<td>C5</td>
<td>Cervical myelopathy</td>
<td>Dorsal portion, predominantly</td>
<td>Total removal</td>
<td>ND</td>
<td>No</td>
<td>18 months</td>
</tr>
<tr>
<td>Jallo et al. [14]</td>
<td>2005</td>
<td>59</td>
<td>M</td>
<td>T5</td>
<td>Dorsal myelopathy</td>
<td>ND</td>
<td>Total removal</td>
<td>ND</td>
<td>No</td>
<td>4.8 years</td>
</tr>
<tr>
<td>Jallo et al. [14]</td>
<td>2005</td>
<td>37</td>
<td>F</td>
<td>T2–T3</td>
<td>Paresthesias</td>
<td>ND</td>
<td>Total removal</td>
<td>ND</td>
<td>No</td>
<td>5 years</td>
</tr>
<tr>
<td>Jallo et al. [14]</td>
<td>2005</td>
<td>41</td>
<td>M</td>
<td>C6–C7</td>
<td>Cervical myelopathy</td>
<td>Eccentric, partially</td>
<td>Total removal</td>
<td>ND</td>
<td>No</td>
<td>3.5 years</td>
</tr>
<tr>
<td>Jallo et al. [14]</td>
<td>2005</td>
<td>17</td>
<td>M</td>
<td>T5–T6</td>
<td>Scoliosis</td>
<td>Right-sided, partially</td>
<td>Total removal</td>
<td>No neurological deficit</td>
<td>No</td>
<td>1.6 years</td>
</tr>
<tr>
<td>Ogungbo et al. [15]</td>
<td>2005</td>
<td>53</td>
<td>M</td>
<td>C3–C4</td>
<td>Cervical myelopathy</td>
<td>Two-thirds intramedullary</td>
<td>ND</td>
<td>Improvement in numbness and gait</td>
<td>ND</td>
<td>ND</td>
</tr>
<tr>
<td>Ishii et al. [16]</td>
<td>2009</td>
<td>63</td>
<td>F</td>
<td>C5</td>
<td>Paresthesias</td>
<td>Purely intramedullary</td>
<td>Total removal</td>
<td>Totally improved</td>
<td>No</td>
<td>14 months</td>
</tr>
<tr>
<td>Ciappetta et al. [17]</td>
<td>2010</td>
<td>75</td>
<td>F</td>
<td>T6–T7</td>
<td>Dorsal myelopathy</td>
<td>Right-sided, partially</td>
<td>Total removal</td>
<td>Totally improved</td>
<td>No</td>
<td>24 months</td>
</tr>
<tr>
<td>Present case</td>
<td>2011</td>
<td>49</td>
<td>F</td>
<td>T9–T10</td>
<td>Dorsal myelopathy</td>
<td>Dorsal portion, partially</td>
<td>Partial removal</td>
<td>Partially improved</td>
<td>No</td>
<td>6 months</td>
</tr>
</tbody>
</table>

ND – Not Described.
The majority of SFT seems to be benign; however, SFT with atypical features has been described [5–8,18–21]. The completeness of resection and the histological appearance are the most important recognized prognostic factors [5–8]. Since the first case of pleural SFT described by Klemperer and Rabin [1] in 1931, a wide variety of extra-pleural locations of SFT such as liver [22], thyroid [23] and orbit [24] have been reported [25].

Solitary fibrous tumour of the CNS is included in the 2000 World Health Organization classification in the category of mesenchymal neoplasm [6]. SFT of the CNS is a distinct clinicopathologic entity with similarities to fibrous meningioma and hemangiopericytoma [5,7]. The pathological diagnosis of SFT is a strong and diffuse positivity for CD34 and Bcl2 without any mitoses or necrosis. Differential diagnosis such as meningioma, schwannoma or neurofibroma is based on the negativity of the other immunohistochemical staining [26]. SFT of the CNS generally affects adults and tend to arise predominantly in the posterior fossa and the spinal column [6]. Five cases [5–7,18,21] of intraspinal and intracranial SFT with multiple locations have been described and Tachonouchi [18] reported a case of drop metastases after complete resection of a right fronto-parietal SFT.

Only few isolated cases [9–17] of intramedullary SFT are described in the literature (Table 1). First reported by Carneiro et al. in 1996, we reviewed 12 cases by a systematic analysis of Pubmed support. The median age at presentation is 50 years (range: 17–75 years) and sex ratio (M/F) is 2.25. The cervical (38%) and upper thoracic (46%) spine are the most frequent locations. Ninety-two percent (12/13) of the cases disclosed progressive neurologic deficits and 69% (9/13) presented an intradural extramedullary extension with dural-based attachment seen at surgery. In 76% (10/13), total resection could be accomplished with only one case of tumoral recurrence.

In studies about SFT of the CNS, these tumours seemed to be successfully managed by surgery alone [5–7,27–29]. Published data suggest that hypercellularity, marked nuclear atypia, high mitotic activity and a high proliferation index are risk factors for recurrence [1–4,30,31]. These findings were based on isolated cases of intramedullary SFT [9–17] as no case series are reported in the literature. Total removal of intramedullary SFT without worsening of neurological signs is, however, less easily achieved than in others locations. In the majority of reports of intramedullary SFT, the margins between the tumour and the spinal cord were not clear at surgery [10,12,13,16] as in our case where no cleavage plane existed between the cranial portion of the lesion and the spinal cord, precluding complete resection of the lesion. So, we would like to highlight that complete removal of intramedullary SFT could be technically difficult and could complicate the following management.

There are no clear data about the efficacy of adjuvant therapy (chemotherapy and radiotherapy) on recurrence, regrowth and progression of the lesion in case of intramedullary SFT [5–8]. The place of postoperative radiotherapy in partially resected or histologically atypical CNS SFTs is still unknown but is often chosen as analogous to the treatment of current meningial tumours. Very few cases of SFT of the CNS treated by radiotherapy have been described [5–8] in the literature. According to the literature, long follow-up is recommended after MRI due to the unpredictable behaviour of some SFT [1–8,25]. Few studies [5–9,18,21,32] reported recurrence of SFT of the CNS several years after the first clinical manifestation. Magnetic resonance imaging of the neuraxis is recommended every year for the follow-up, although complete resection has been performed. The duration of follow-up is difficult to establish but we could propose a minimum of ten years regarding data about SFT in the CNS.

No data are available in the literature about management of recurrence or regrowth of the lesion. We think that the more appropriate is a second-look surgery without complement radiotherapy.

4. Conclusion

Although SFTs are generally benign lesions, malignant cases of SFTs have been reported. In such cases, recurrence and metastasis may take place even after complete resection. Because of the rarity of reports and the lack of long-term follow-up, the course of intramedullary SFT is unknown and careful long-term follow-up is recommended. Magnetic resonance imaging of the neuraxis is the imaging of choice for the follow-up.

Conflict of interest

None declared.

Acknowledgement and financial support

None declared.

Ethics

The work described in this article has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans; Uniform Requirements for manuscripts submitted to Biomedical journals.

REFERENCES
