Case Report
Coexistence of neurofibroma and meningioma at exactly the same level of the cervical spine

Kai-Yuan Chen, Jau-Ching Wu, Shih-Cheih Lin, Wen-Cheng Huang, Henrich Cheng

*Department of Neurosurgery, Neurological Institute, Taipei Veterans General Hospital, Taipei, Taiwan, ROC
bDepartment of Pathology, Taipei Veterans General Hospital, Taipei, Taiwan, ROC
National Yang-Ming University School of Medicine, Taipei, Taiwan, ROC

Received May 22, 2013; accepted November 22, 2013

Abstract
We report a case of the coexistence of different spinal tumors at the same level of the cervical spine, without neurofibromatosis (NF), which was successfully treated with surgery. A 72-year-old female presented with right upper-limb clumsiness and weakness. Magnetic resonance imaging revealed an intradural, extramedullary tumor mass at the right C3–4 level with extradural extension into the intervertebral foramen. The extradural tumor was removed, and the pathology showed neurofibroma. After incision of the dura, the intradural tumor was removed, and was identified as meningioma in the pathological report. The patient did not meet the criteria of NF. Coexistence of neurofibroma and meningioma at exactly the same level of the spine without NF is extremely rare. Exploration of the intradural space may be necessary after resection of an extradural tumor if the surgical finding does not correlate well with the preoperative images.

Keywords: coexistence; meningioma; neurofibroma; neurofibromatosis; spinal tumor

1. Introduction
Multiple primary spinal tumors are not uncommon in patients with neurofibromatosis (NF), but are uncommon in the general population. These spinal tumors are rarely of different pathological natures. Moreover, the presentation of two histologically different primary tumors at exactly the same spinal segment has only been reported twice in patients without NF. We report, herein, a patient without NF and with both a neurofibroma and a meningioma (extradural and intradural, respectively), at exactly the same level of the cervical spine.

2. Case Report
2.1. Clinical history
A 72-year-old female presented with right-hand clumsiness with progressive weakness and numbness in the right upper and lower limbs for approximately 1 year. She had a past history of poliomyelitis at the age of 7 years, which had caused left leg weakness lasting until the present. She had been able to walk independently until recent deterioration of her right-side hemiparesis. Magnetic resonance imaging (MRI) demonstrated an enhancing intradural, extramedullary mass lesion measuring 24 mm × 12 mm × 34 mm in size at the right side of C3–4, with extradural extension into the intervertebral foramen (Fig. 1A and B). The computed tomography (CT) scan demonstrated little erosion or involvement of the surrounding bony structures (Fig. 2). Under the impression of a neurogenic tumor with deterioration of neurological function, surgery was recommended. During
Fig. 1. (A) Contrast-enhanced T1-weighted magnetic resonance imaging (MRI) demonstrates an oval, contrast-enhanced tumor (asterisk) at C3—4, sagittal view; (B) contrast-enhanced T2-weighted MRI demonstrates a suspicious intradural, extramedullary heterogeneous tumor (asterisk) compressing the spinal cord from the right aspect, with extension to the right interforamen space, axial view. It is difficult to discover the coexistence of intra- and extradural tumors in the preoperative MRI. (C, D) Follow-up MRI 12 months postoperatively demonstrates no local recurrence.

Fig. 2. (A) Sagittal computed tomography (CT) scan demonstrates a tumor with an enlarged intervertebral foramen between C3 and C4; (B) axial CT scan demonstrates a tumor between C3 and C4 with extension to the right intervertebral foramen. The pedicles of C3 and C4 were destroyed.
hospitalization, the physical examination demonstrated no café au lait spots, no vestibular schwannomas, or other signs of NF. Moreover, there was no family history of NF in her first- and second-degree relatives. A posterior cervical laminectomy was performed in a standard fashion in the prone position. A grayish extradural tumor was first exposed intraoperatively, and removed, with the sacrifice of the C3 nerve root. However, there was no continuation of this tumor into the intradural space according to the preoperative MRI. We decided to incise the dura sac at the same spinal level for exploration. Another small intradural tumor was also discovered and removed. Subsequent lateral mass screw fixation and posterior fusion with autograft were performed. The patient's neurological function improved significantly soon after the operation. There were no other complications during her hospitalization. Her neurological function continued to improve and was fully recovered to her postpolio status at 3 months postoperation. The pathology demonstrated a neurofibroma and a meningioma (extradural and intradural, respectively) (Fig. 3). At 12 months postoperation, the MRI demonstrated no recurrence (Fig. 1C and D).

2.2. Pathology

Two tumor parts, extradural and intradural, were resected and sent for pathology examination. Hematoxylin and eosin (H&E) stain of the extradural tumor demonstrated spindle cells with a loose myxoid background (Fig. 3A). The tumor cells had spindle-shaped nuclei, and some of them were curved. Deposition of collagen bundles in the stroma between the spindle tumor cells was seen. Parts of the tumor cells were immunoreactive to S-100 protein (Fig. 3B). The axons within the tumor were also demonstrated by Bodian's method (Fig. 3C). Therefore, the extradural tumor was a neurofibroma. For the intradural tumor, the H&E stain demonstrated solid sheets of meningothelial cells with a syncytial appearance. Psammoma bodies were occasionally encountered (Fig. 4A). The tumor cells were immunoreactive to epithelial membrane antigen (EMA) (Fig. 4B). The histopathological features of the intradural tumor were consistent with meningioma.

3. Discussion

This is the third reported case of two different primary spinal tumors occurring at the same level of the cervical spine. This 72-year-old female patient had an extradural neurofibroma and an intradural meningioma at the right C3–4 level. Successful resection of both tumors without recurrence was achieved. In our case, the preoperative MRI demonstrated an intradural tumor with extradural extension. However, an exactly extradural tumor without intradural continuation was found during the surgery, hence an intradural tumor was suspected. There would have been a high probability of inadequate surgical resection if the dural sac had not been opened. Therefore, the lesson learned here is that a high degree of suspicion of a coexisting intradural tumor after resection of an extradural tumor is warranted in these cases. Correlation of the surgical findings to the preoperative MRI is also warranted, and intradural exploration might be necessary.

There are seven cases of coexisting meningioma and nerve sheath tumor in the literature. Six of these reported cases were meningioma with schwannoma, and one was...
Menigioma coexisting with neurofibroma. The cervical spine was most commonly involved, followed by the thoracic and lumbar spine. Hokari et al. described the first case of coexistence of a primary meningioma and a neurofibroma at the C1–2 level in 2002. These cases are summarized in Table 1.

<table>
<thead>
<tr>
<th>Author &amp; year</th>
<th>Age/sex</th>
<th>Site</th>
<th>Spinal level</th>
<th>Histological type</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hokari et al (2002)</td>
<td>59/F</td>
<td>C1–2</td>
<td>Meningioma</td>
<td>Neurofibroma</td>
</tr>
<tr>
<td>Nakamizo et al (2012)</td>
<td>49/M</td>
<td>C2</td>
<td></td>
<td>Meningioma</td>
</tr>
<tr>
<td>Present case</td>
<td>72/F</td>
<td>C3–4</td>
<td>Meningioma</td>
<td>Neurofibroma</td>
</tr>
<tr>
<td>Araszkiewicz et al (1988)</td>
<td>22/F</td>
<td>T8–9</td>
<td>Meningioma</td>
<td>Neurofibroma</td>
</tr>
<tr>
<td>Nishihara et al (1989)</td>
<td>16/F</td>
<td>C6</td>
<td>Schwannoma</td>
<td></td>
</tr>
<tr>
<td>Dorizzi et al (1992)</td>
<td>70/F</td>
<td>C7</td>
<td>Schwannoma</td>
<td></td>
</tr>
<tr>
<td>Ogihara et al (2003)</td>
<td>54/F</td>
<td>T1</td>
<td>Meningioma</td>
<td></td>
</tr>
<tr>
<td>Different level</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

F = female; M = male.

Meningiomas are isointense to hypointense on T1WI and slightly hyperintense on T2WI. Enhancement of spinal meningiomas is strong and homogeneous. However, spinal meningiomas are typically small. Therefore, if an obvious tumor is already seen, as in our case, the meningioma can be easily missed and mimic a single-component tumor. Nakamizo et al. presented a case of concurrent schwannoma and meningioma with a slight difference in signal intensity on magnetic resonance cisternography. In the current case, there was little difference in the signal intensity on contrast-enhanced MRI. However, when retrospectively reviewed, the tumor was not perfectly oval in shape, which might give rise to suspicion of a lobulated tumor or even multiple tumors. Nevertheless, the diagnosis of the coexistence of neighboring intradural and extradural spinal tumors by preoperative MRI is not sensitive enough.

In conclusion, coexistence of neurofibroma and meningioma at exactly the same level of the spine without NF is extremely rare. Identification of such coexistence by preoperative images could be difficult. Although coexistence of intra- and extradural tumors at the same spinal level is rare, intradural exploration might be necessary if the surgical findings do not correlate well with the preoperative images after removal of the extradural tumor.

References