Intradural teratomas, particularly cervicothoracic lesions, are extremely rare. Up to now only 6 cases of intramedullary cervical teratomas have been reported in adults, and all of these were histologically mature. The authors present the case of a 35-year-old man with progressive myelopathic symptoms who was admitted through an outpatient clinic and was surgically treated. The characteristics, diagnosis, epidemiology, and treatment of cervical intramedullary teratomas in adults are also reviewed. Postoperative MR imaging showed that the tumor had been near totally removed, and severely adherent tissue remained ventrocranially with tiny focal enhancement on follow-up MR imaging. Pathological examinations revealed immature teratoma without any malignant component. Adjuvant therapy was not performed. Although no change in neurological findings and symptoms was apparent postoperatively, lesion regrowth was demonstrated on MR imaging 4 months after surgery. At 8 months postoperatively, myelopathic symptoms had developed and a huge intramedullary tumor recurred according to MR imaging. This case is the seventh reported instance of intramedullary cervical teratoma in an adult, and the first case report of the immature type with malignant features. (DOI: 10.3171/2010.3.SPINE09461)

**Key Words**
- intramedullary immature teratoma
- cervical intramedullary tumor
- cervical intramedullary teratoma

**Intradural teratomas, particularly cervicothoracic lesions in adults, are extremely rare.** A meticulous 2006 review by Ak et al.¹ disclosed 3 cases from 1931, including 1 of their own. These 3 cases were later interpreted as adult intramedullary cervical teratomas³,⁸,¹² and all were histologically mature.

In this report, we present the first case of a spinal intramedullary immature teratoma located in the cervicothoracic region with malignant features, which rapidly recurred after near-total lesion removal. We also include a review of the 6 mature teratoma cases previously described and document the characteristics of cervical intramedullary teratoma in adults.

In 1954 Dereymaeker² reported the case of a 43-year-old woman with a teratoma between C-5 and T-2, which to our knowledge is the first instance of a cervical intramedullary mature teratoma described in an adult. In 1982 Padovani et al.¹¹ presented the case of a mature teratoma in a 21-year-old woman with a lesion located between the C-6 and T-1 levels. These 2 cases could not be evaluated with CT or MR imaging. Between 2004 and 2008, 4 additional cases of cervical intramedullary mature teratoma were reported¹,³,⁸,¹². Although 2 other cases of cervical spinal teratoma were reported by Sharma et al.,¹⁵ they involved intradural extramedullary lesions. Table 1 shows all 6 previously published cases and itemizes the characteristics of adult cervical intramedullary teratoma.

**Case Report**

**History and Examination.** This 35-year-old man was admitted through an outpatient clinic with symptoms of progressive quadripareisis over a period of 3 weeks, numbness below the lower abdomen, and a voiding difficulty of 2 weeks' duration. All clinical features showed a recent, progressive tendency. His medical and family histories were unremarkable, as was his physical examination; however, a neurological examination revealed quadriparesis. Bilateral upper and lower extremities showed Grade 4 (International Standards for the Classification of Spinal Cord Injury) with a Grade 3 right-hand grasp. Upper- and lower-extremity deep tendon reflexes were bilaterally hyperactive. In addition, Lhermitte sign, right ankle clonus, and paresthesia with numbness below the T-12 level were identified. Plain radiographs and CT scans were unremarkable; however, T2-weighted MR imaging revealed an intramedullary lesion at C6–7 with peritumoral heterogeneous high signal intensity at C3–T3 and scattered lesions with diverse high signal intensities. On T1-weighted images focal lesions with slightly
elevated signal intensity were observed. A Gd enhancement study showed a highly enhanced lesion with heterogeneous features (Fig. 1).

Operation. The patient underwent C6–7 total laminectomy with motor evoked potential monitoring. Intradural posterior myelotomy was also undertaken on the protruding portion of the lesion, which was rubbery, pink/gray in color, and distinctly separate from the surrounding cord. Although it provided a good dissection margin from the surrounding cord and was relatively easily excised by bipolar coagulation and sharp dissection, the lesion was severely adherent to the cord at some points, especially on the ventrocranial side of the tumor. After near-total tumor removal, a small ventrocranial hard and severely adherent region of the tumor with calcification was carefully coagulated by bipolar cautery so as not to damage the ventral cord. There was no definite cyst or hemorrhage in the lesion. During operation, no abnormal findings were detected on motor evoked potential monitoring.

Postoperative Course. Postoperatively the patient showed mild neurological improvement, and 8 days postoperatively he experienced no neurological symptoms other than mild neck pain and a patchy numbness in both legs, especially on the left side. Postoperative MR imaging showed that the peritumoral edema had markedly decreased. The tumor was near totally removed but extremely adherent tissue remained ventrocranially with tiny focal enhancement (Fig. 2A).

Microscopically the tumor was composed of a mixture of immature epithelial and stromal components. The epithelial components included some primitive glandular structures (Fig. 3A), which are immunohistochemically positive for α-fetoprotein (a marker of immature teratomas) and cytokeratin (Fig. 3C). Although neuroepithelial tissue is known to be the most frequent immature component in teratomas, no primitive neural tissue was observed in the featured case. The stromal tissue was composed mostly of primitive mesenchymal cells with small, spindle-shaped nuclei and scanty cytoplasm; however, some islands of immature cartilage were observed as well (Fig. 3B).

Immunohistochemically the stromal cells were positive for vimentin, and the cartilaginous component was also positive for S100 protein. The Ki 67 proliferation index was relatively high for both the epithelial and mesenchymal cells. The p53 expression was unremarkable. Furthermore, although no change in neurological findings or symptoms was observed, lesion regrowth was evident at 4 months postoperatively on MR imaging (Fig. 2B). At 8 months postoperatively, the patient showed myelopathic symptoms, a huge intramedullary tumor recurrence, and longitudinal cord edema, which had been suggested by postoperative MR imaging (Fig. 2C).

Discussion

Teratoma is one of the rarest intraspinal neoplasms and constitutes only 0.2–0.5% of all spinal cord tumors. In a literature review published in 2006, Ak et al. reported that 29 cases of adult intramedullary mature tera-

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**TABLE 1: Details of the 6 published cases of adult cervical intramedullary teratoma (including the present case)**

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs), Sex</th>
<th>Location</th>
<th>Accompanying Abnormalities</th>
<th>Neuropathy</th>
<th>Histology</th>
<th>Resection</th>
<th>Adjuvant Therapy</th>
<th>Follow-Up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dereymaeker, 1954†</td>
<td>43, M</td>
<td>C5–T2</td>
<td>bifid T2–3, nonunion of C-1</td>
<td>myelop</td>
<td>teratoma‡</td>
<td>subtotal</td>
<td>no</td>
<td>no comment</td>
</tr>
<tr>
<td>Padovani et al., 1982</td>
<td>21, F</td>
<td>C6–T1</td>
<td>not obvious on plain roentgenogram</td>
<td>progressive rad &amp; myelop</td>
<td>teratoma‡</td>
<td>subtotal</td>
<td>no</td>
<td>neurologically unmodified until 4 years</td>
</tr>
<tr>
<td>Paterakis et al., 2004</td>
<td>63, M</td>
<td>C2–5</td>
<td>vertebral body fusion C3–5, nonunion of C-1</td>
<td>progressive rad &amp; myelop</td>
<td>mature</td>
<td>subtotal</td>
<td>no</td>
<td>died 3 mos later due to respiratory failure unrelated to teratoma</td>
</tr>
<tr>
<td>Ak et al., 2006</td>
<td>43, F</td>
<td>C2–4</td>
<td>midline nodular soft skin mass connecting into dura at C-3 level</td>
<td>progressive rad &amp; myelop</td>
<td>mature</td>
<td>total</td>
<td>no</td>
<td>no comment</td>
</tr>
<tr>
<td>Makary et al., 2007</td>
<td>46, F</td>
<td>C1–2</td>
<td>bifid C-2, vertebral body fusion C2–3, scoliosis C3–4, dermal sinus connecting into dura</td>
<td>progressive rad &amp; myelop</td>
<td>mature</td>
<td>total</td>
<td>no</td>
<td>6 mos after op: no neurological evidence of recurrence; no follow-up MRI</td>
</tr>
<tr>
<td>Babak et al., 2009</td>
<td>34, M</td>
<td>C4–6</td>
<td>bifid C-6</td>
<td>progressive rad</td>
<td>mature</td>
<td>total</td>
<td>no</td>
<td>no evidence of recurrence on 2-year follow-up MRI</td>
</tr>
<tr>
<td>current case</td>
<td>35, M</td>
<td>C6–T1</td>
<td>absence</td>
<td>progressive myelop</td>
<td>immature</td>
<td>near total</td>
<td>no</td>
<td>rapid &amp; huge recurrence lesion on serial follow-up MRIs until 8 mos after surgery</td>
</tr>
</tbody>
</table>

* myelop = myelopathy; rad = radiculopathy.
† Article in French.
‡ No comment on subtype.
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tomas had been identified. The average patient age was 36.2 years, and lesions were predominantly located in the lower thoracic and thoracolumbar regions. To our knowledge, our case is the seventh instance of adult intramedullary cervical teratoma to be described in the literature. Teratomas are composed of cells of pluripotent origin that undergo disorganized organogenesis and display mixed tissue types derived from ectoderm, endoderm, and mesoderm. In our case, however, only 2 germinal layer elements were present—that is, endoderm and mesoderm—but this composition does not rule out a diagnosis of teratoma because derivatives of 1 or 2 germ layers can overgrow others. Teratomas are classified as:

1) mature teratomas if they are characterized by differentiated tissues; 2) immature teratomas if they contain some immature, nonmalignant tissues; and 3) malignant teratomas if they contain features of a yolk sac tumor, that is, if choriocarcinoma or embryonal carcinoma are encountered among the differentiated tissues. Of the previously reported 6 cases, the first 2 did not mention the mature or immature subtypes. In subsequently reported cases, however, pathological examinations revealed mature teratomas with fully differentiated components of diverse mature tissues but no evidence of immature or malignant components. Our case contained definite primitive mesenchymal tissue and immature cartilage (Fig. 3A–C). To our knowledge, this case is the first reported instance of cervical intramedullary immature teratoma.

Many theories have been proposed concerning the development of intraspinal teratomas, but the most accepted embryogenetic theory concerns the displacement of primordial germ cells into the dorsal midline during their normal migration from the primitive yolk sac to gonadal ridges. In this context, there is a well-known association between spinal cord teratomas and dysraphic congenital spinal malformations, such as spina bifida, dermal sinus, split cord, meningomyelocele, lipomeningomyelocele, syrinx, and a thickened terminal filum. As shown in Table 1, improved CT and MR imaging precision has allowed associated abnormalities of cervical intramedullary teratomas, including vertebral body fusion, bifid or nonunion of bony structure, and skin lesions (soft mass, dermal sinus communicating to the dura mater), to be discovered in later cases. In our case, however, no associated abnormality was found. Thus, although such cases are extremely rare, teratoma should be ruled out when a cervical intramedullary tumor is encountered with the above-mentioned associated malformations.

The role of plain radiography is limited to the detection of changes in vertebral bodies, such as erosion and widening of the interpedicular space with or without vertebral abnormalities at the lesion level. Furthermore, detailed CT evaluations of the bony structure can also show variable tumor density or calcification. Magnetic resonance imaging generally shows mixed high/low signal intensities, which correspond to the cystic and solid portions of teratomas. Although the lesions were not restricted to cervical intramedullary teratomas, Sharma et al. reported fat signals and areas of calcification in one-third of patients studied. Heterogeneous signal intensities on routine T1 and T2 signal MR images as a result of fat, calcification, or cysts with or without hemorrhage are one of the major radiological clues of teratoma. In our case, we were initially convinced that this cervical intramedullary lesion was an astrocytoma or ependymoma based on considerations of incidence alone, and we believed that the severe heterogeneous and high signal intensities observed on T1-weighted images were caused by multistage hemorrhagic features of previous cystic components, because the patient’s symptoms had a recent onset and...
were progressive. Therefore, these images may have been attributable to mixtures of fat, calcification, and microcystic components in the teratoma, although low signal intensities due to calcification were not prominent on T1- or T2-weighted images (Fig. 1). Although such cases are extremely rare, when severe heterogeneous signal intensities are present on T1- and T2-weighted images or when high signal intensity is present on T1-weighted images, fat suppression MR images might be helpful because they can be used to designate fat tissue and calcified materials more precisely.

Generally speaking, mature and immature teratomas usually demonstrate benign clinical behavior, but they can recur with potentially malignant features. Some authors have described immature teratomas as aggressive tumors that tend to recur frequently, with primitive undifferentiated components resembling “fetal” tissues. As for teratomas in other locations, total resection is the primary treatment modality. However, in patients with a spinal intramedullary lesion, this approach may be impossible as a result of tight adhesions with neural tissue. Subtotal resection can be performed by removing as much of the tumor as possible, while preserving neural function, because of the extremely slow growth of teratomas. In fact, in a study the difference between recurrence rates after total and subtotal resection was not significant (9% vs 11%, respectively). In pediatric cases, however, Lo Curto et al. found that incomplete resection and a female sex are important risk factors for relapse or death and that these risk factors were more important than an immature histology, although not all of their cases had a spinal location. Adjuvant radiotherapy is indicated when teratomas contain malignant elements, but the role of chemotherapy has not been proven. In the presence of a benign histology, it would seem appropriate to avoid radiotherapy after initial surgery. Furthermore, it has been suggested in cases of tumor recurrence or progression that the possibility of further surgery should be explored before considering radiotherapy, which even in this situation may have doubtful efficacy. No reports have been issued on spinal immature teratoma or on treatment modalities. In cases of intracranial immature teratoma, additional chemotherapy and radiotherapy can be administered based on evidence of residual disease. In our case, although a tiny tumor remnant was suspected based on postoperative MR imaging studies (Fig. 2A), the tumor was confirmed to be the immature pathological type without a malignant component, and thus adjuvant radiotherapy was not performed because second surgery was considered the primary option. However, on follow-up serial MR imaging, rapid and marked progression was shown within 8 months (Fig. 2B and C), which was contrary to our expectations. Furthermore, this malignant feature has not been reported for spinal teratoma. We are currently planning secondary surgery for removal and differential diagnosis and subsequent adjuvant radiochemotherapy. Although this case is the first reported instance of adult cervical intramedullary immature teratoma, it should remind surgeons that intramedullary immature teratoma can manifest aggressive, malignant features.

Disclosures

The authors have no conflict of interest to report concerning the materials or methods used in this study or the study findings.
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Author contributions to the study and manuscript preparation include the following. Conception and design: Park, Moon, Joo Han Kim. Acquisition of data: Moon. Analysis and interpretation of data: Moon, Shin. Drafting the article: Moon. Critically revising the article: Park, Moon, Joo Han Kim. Reviewed final version of the manuscript and approved it for submission: all authors. Statistical analysis: Moon. Administrative/technical/material support: Moon, Kwon, Chung. Study supervision: Park, Joo Han Kim, Jong-Hyun Kim.

References


Fig. 3. The tumor was composed of a mixture of immature epithelial and stromal components. A: Photomicrograph demonstrating epithelial components, including some primitive glandular structures (circle). B: Photomicrograph showing stromal tissue composed mostly of primitive mesenchymal cells with small, spindle-shaped nuclei and scanty cytoplasm, although some islands of immature cartilage (rectangle) also can be observed. C: Photomicrograph demonstrating epithelial components, such as some primitive glandular structures, which are immunohistochemically positive for α-fetoprotein, a marker of the immature type. Original magnification ×40 (A) and ×200 (B and C).