Primary Intradural Conus and Cauda Equina Tumors: Long-time Outcome With 14 Years Follow-up

ABSTRACT
OBJECTIVE: This clinical study evaluated mixed tumor pathologies in the lumbar dural sac. Currently, there are few published clinical studies with long-term follow-up in the literature.

METHODS: This is a retrospective review of the clinical presentation, diagnosis and management of 24 patients with primary intradural conus and cauda equina tumors (ICCET). We operated on 24 ICCET patients during the last 14 years.

RESULTS: The largest pathology group of 9 (37.4%) cases was ependymomas. The rest were 5 (20.8%) neurinomas, 4 (16.6%) meningiomas, 1 (4.16%) epidermoid (with dermal sinus tract), 4 (16.6%) dermoids (one case with adult tethering cord and dermal sinus tract) and 1 (4.16%) hemangioblastoma. Two of the cases (ependymoma and neurinoma) had giant tumors, which extended from L1 to L5. Low back pain and urinary symptoms were the most common initial symptoms of the cases. The diagnosis was made with standart myelography (2 cases), computed tomographic myelography (4 cases) and magnetic resonance imaging (18 cases). All patients underwent tumor resection through a single posterior approach. The neurological conditions of 22 (91%) patients improved. Two patients did not improve and the postoperative Nurick Grade was grade 5. Postoperative radiotherapy was used in one case with malignant neurinoma. There was a rest tumor in one patient who had a dermoid tumor (4.6%). The mean follow-up period was 75.5 months.

CONCLUSION: We report the effect of surgical treatment for ICCET as excellent in nineteen out of 24 cases. ICCET are generally slow-growing, benign lesions. They are mostly surgically curable and radiotherapy is not usually necessary. The prognosis after surgical treatment depends not only on histology but also on the degree of neurological deficit before the surgery, and the importance of early diagnosis and subsequent treatment is well recognized.

KEY WORDS: conus, cauda equina, intradural, primary, tumor.

INTRODUCTION

Primary tumors of the intradural conus and cauda equina (ICCET) region are uncommon and consist mainly of neurofibromas, ependymomas and neurinomas, and less commonly meningiomas, (7,10,14,16) dermoids, epidermoids, paragangliomas, high grade gliomas and hemangiofibromas (4,5,6). A high proportion of tumors in this region are benign (4). Intramedullary conus gliomas are rare and will not be discussed here. They have been considered to cause a progressive neurological deficit by compression of the surrounding intramedullary conus rather than infiltration into the surrounding neural tissues such as cauda equina nerves. In addition, they have been recognized as neoplasms that are amenable to complete surgical resection with
acceptable morbidity and mortality and a low incidence of recurrence. However, some cases associated with surgical morbidity have been reported.9 The functional results after microsurgical resection of ICCET are directly related to the preoperative neurological status.

The emphasis of this paper is on the clinical presentation, indications, radiological and surgery-related findings and outcome of the tumors of this region.

**PATIENTS AND METHODS**

A total of 230 intradural spinal tumors were histologically diagnosed between 1990 and 2004 and 24 (11%) were ICCET. In the last fourteen years, 24 patients with ICCET were treated with the microsurgical posterior approach at our neurosurgery clinic. There were 16 males and 8 females with a mean age of 36.2 years (range 13 to 68 years). No correlation was found between tumor type and patient age. There was no significant relationship between neurological abnormalities and tumor size.

The localization of the tumor, patient’s age at presentation, and duration of symptoms prior to admission are listed in (Table I).

The main preoperative complaints included minor motor weakness in 12 out of the 24 patient (50%), paresthesia in 7 patients (29.4%) and pain in 20 patients (84.8%). The back pain became more severe at night or when the patient assumed a recumbent position for any length of time in most patients. The pain was only mitigated when the patient stood or sat up. The patients frequently reported sleeping in the sitting position. Bladder dysfunction was observed in 18 (75%) patients who had 100 cc of postvoiding residual urine. The duration of symptoms before the initial diagnosis ranged from 2 to 105 months (mean duration, 29 months).

Outcome after operation was assessed from the clinical records, symptomatically in terms of pain, paresthesia, subjective lower limb weakness and sphincter disturbance, and by objective neurological examination.

**Preoperative evaluations**

All patients underwent both myelography and computed tomography between 1990 and 1995. We have only one case investigated by myelography (Figure 1) and this tumor was a myxopapillary ependymoma. Magnetic resonance imaging (MRI) was performed for the patients admitted after 1994. MRI showed well-demarcated ICCET in the 18 patients studied. Both solid (Figure 2) and cystic (Figure 3) components of the tumor were well represented in myelography pictures. The youngest patient was a 12-year-old girl with dermoid tumor (Figure 4). Most tumors were found to be extend for 1 or 2 vertebral segments, while the tumor extended over 5 vertebral segments in 2 cases. The long segmental spinal enrollment was demonstrated in (Figure 5) (ependymoma) and (Figure 6) (neurinoma). There was no dumbbell neurinoma in this location and all cases were single or solitary (Figure 7). One case had malignant neurinoma. One case who had hemangioblastoma was admitted with paraplegia, and was the only patient operated as an emergency. The location of the conus level was L1 except one patient who had a dermoid tumor with adult tethered cord syndrome (Figure 8). Meningiomas were the most recognized tumor in the preoperative period (Figure 9). Multisegmental spinal tumors were found in one case (4.6%). There were no cases with high-grade gliomas. All patients were evaluated preoperatively according to the grading system presented by Nurick (22):

**Table I. Data at presentation in 24 cases of conus and cauda equina tumors**

<table>
<thead>
<tr>
<th>Tumor type</th>
<th>No of case (%)</th>
<th>Age (yr)</th>
<th>Duration of Symptoms (Year)</th>
<th>Nurick grade postoperative</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Mean</td>
<td>Range</td>
<td>M:F</td>
</tr>
<tr>
<td>Ependymoma</td>
<td>9 (37.4)</td>
<td>34</td>
<td>23-59</td>
<td>1.55</td>
</tr>
<tr>
<td>Neurinoma</td>
<td>5 (20.8)</td>
<td>39</td>
<td>22-68</td>
<td>4.5</td>
</tr>
<tr>
<td>Meningioma</td>
<td>4 (16.6)</td>
<td>42</td>
<td>38-46</td>
<td>3.4</td>
</tr>
<tr>
<td>Epidermoid</td>
<td>1 (4.16)</td>
<td>43</td>
<td>32</td>
<td>2.5</td>
</tr>
<tr>
<td>Dermoid</td>
<td>4 (16.6)</td>
<td>43</td>
<td>13-54</td>
<td>7.1</td>
</tr>
<tr>
<td>Hemangioblastoma</td>
<td>1 (4.16)</td>
<td>35</td>
<td>-</td>
<td>0.2</td>
</tr>
</tbody>
</table>
Figure 1: Male, 33 years old, conus tumor is demonstrated by myelography. This lesion was caused a partial obstruction at the between L3 level. The histopathological diagnosis was myxopapillary ependymoma.

Figure 2: Male, 38 years old, T1- and T2-weighted sagittal image showing an intradural conus tumor (ependymoma).

Figure 3: T1- and T2-weighted sagittal 3A and axial 3B images showing an intradural cystic ependymoma.

Figure 4: Female, 13 years old, intradural conus dermoid tumor. Typical appearance of a dermoid tumor on a sagittal T1-weighted MR image.

Figure 5 A: Female, 56 years old, sagittal T2-weighted image showing an intradural mass (ependymoma). B: T2-weighted enhanced sagittal imaging not showing postoperatively in the conus levels.
Figure 6: Female, 66 years old, T1 and T2 weighted enhanced sagittal imaging showing an intradural giant tumor (neurinoma). The tumor was iso-hyperintense, the associated necrosis and cyst formation had low signal intensity on the T1-weighted image.

Figure 7: Male, 23 years old, T1-weighted sagittal (7A) and axial (7B) MR images after contrast: the tumor was remarkably enhanced, with tumor margin sharpened. The tumor was completely removed surgically and the histological diagnosis was neurinoma.

Figure 8: Female, 52 years old, sagittal T2-weighted MR image demonstrated tethered cord (arrow), dermal sinus tract (striped arrow) and dermoid tumor complex (small arrow).

Figure 9: Male, 45 years old, sagittal T1-weighted MR image post-contrast demonstrated an intradural conus meningioma (arrow).
Grade 1: Neurologically normal or normal walk
Grade 2: Slight difficulty walking
Grade 3: Disability limiting normal work or more severe neurological deficit
Grade 4: Requires assistance with walk/gait or severe deficit
Grade 5: Bedridden or wheelchair bound

Surgical techniques and results
A midline surgical approach was used for all our patients. After laminectomy and opening the dura, the tumors were resected under the microscope. In one case (13 years old) the tumor was resected by laminoplasty. Osteoblastic laminoplasty was preferred because of the age and the pathology (dermoid). In this case, we decided that laminectomy may cause delayed instability and myelopathy. This matter is very important at the thoracolumbar junction region and in this age.

Myelotomy was performed after the midline dural incision if the tumor was intramedullary by gentle spreading of the posterior spinal cord column and cauda equina nerve roots using a microdissector and forceps. The tumors were always different in colour and softer than spinal cord tissue, and they were well-demarcated and encapsulated (such as neurinomas).

The number of resected nerve roots was one root in 6 cases, two roots in 2, and three roots in one. Two dermal sinuses with dermoid tumor tracts were excised.

Meningioma was the most easily resectable tumors in ICCET because of its extramedullary location. We did not observe prominent feeding vessels on the ventral surface of the tumors. A microsurgical approach was used in all cases and no blood or blood product transfusion was required.

Complete removal of the tumor was achieved in 23 (95.4%) patients and subtotal removal was achieved in one case with a dermoid tumor. One case with dermoid tumor pathology was operated in two separate stages as residual tumor was identified after the first operation. The histological diagnoses were as follows:

- Ependymoma in 9 (37.4%) cases,
- Neurinoma in 5 (20.8%) cases (one case was malignant neurinoma)
- Meningiomas in 4 (16.64%) cases,
- Epidermoid in 1 (4.16 %) case,
- Dermoid in 4 (16.64 %) cases,
- Hemangioblastoma in 1 (4.16 %) case (Table II).

No deaths occurred after a mean follow-up period of 54 months. One patient who had malignant neurinoma underwent radiation therapy after surgery. A syrinx was not detected in any of our cases.

The myelotomy was not closed in any of the patients.

After the surgical procedures, 2 (8.32 %) out of the 24 patients experienced a decrease of neurological function, and this persisted during the follow-up period (2 patients; grade 2). Except two cases with paraplegia, these neurological deficits included minor motor deficit, urinary dysfunction, and posterior column dysfunction. One patient had acute paraplegia due to intratumoral bleeding (hemangioblastoma) and emergency decompression was performed with subtotal tumor excision. The postoperative neurological function of this case (Nurick grade 5) was the same as the preoperative status. One patient experienced repeated growth or residue of a dermoid tumor in the conus causing partial cauda equina syndrome and did not show

Table II: Clinical presentation (preoperative period) related to tumor histology

<table>
<thead>
<tr>
<th>Tumor Type</th>
<th>Back Pain</th>
<th>Radiculopathy</th>
<th>Motor Weakness</th>
<th>Upper Motor Neuron Sign</th>
<th>Spincter Involvement</th>
<th>Sensory Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ependymomas</td>
<td>9 (100)</td>
<td>5 (55.5)</td>
<td>8 (88.8)</td>
<td>7 (77)</td>
<td>2 (22.2)</td>
<td>9 (100)</td>
</tr>
<tr>
<td>Neurinomas</td>
<td>3 (60)</td>
<td>3 (60)</td>
<td>4 (80)</td>
<td>4 (80)</td>
<td>1 (20)</td>
<td>4 (80)</td>
</tr>
<tr>
<td>Meningioma</td>
<td>4 (100)</td>
<td>2 (50)</td>
<td>2 (50)</td>
<td>-</td>
<td>-</td>
<td>3 (75)</td>
</tr>
<tr>
<td>Epidermoid</td>
<td>1 (100)</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Dermoid</td>
<td>4 (100)</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>1 (100)</td>
<td>-</td>
</tr>
<tr>
<td>Hemangioblastoma</td>
<td>-</td>
<td>1 (100)</td>
<td>1 (100)</td>
<td>-</td>
<td>1(100)</td>
<td>-</td>
</tr>
</tbody>
</table>
improvement months after the surgery (Nurick grade 5). However, neurological status improved thereafter. Twenty-two patients were neurologically intact at the time of the most recent follow-up examination (mean duration: 3 year).

All the patients were given methylprednisolone and antibiotics in the preoperative period.

DISCUSSION

The ICCET are rare lesions, and a large clinical series with long follow-up has not been reported before in the neurosurgical literature. We have operated on 230 spinal intradural tumors in our clinic between 1990 and 2004. Twenty-four (10.5%) cases were localized to the conus. These tumors were further subdivided as extramedullary (5 cases; 20.8%) and intramedullary (19 cases; 79.4%).

The few population-based studies that have been published give annual incidence rates of 1 new tumor per 100,000 population. Attempts to distinguish conus from cauda equina lesions have been made clinically, but many authors believe the distinction is of little value. The predominant symptoms and signs of these tumors are well documented. These tumors usually have benign histology. The prognosis after surgical treatment depends not only on histology but also on the degree of neurological deficit before the surgery, and the importance of early diagnosis and subsequent treatment is well recognized.

The conus medullaris, which is the terminal segment of the adult spinal cord, lies at the inferior aspect of the L1 vertebra. The segment above the conus medullaris is termed the epiconus. Nerve roots travel caudally from the conus medullaris as the cauda equina.

Clinical findings at the time of presentation to a specialist are documented in Table 2. Back pain was present in all patients and bilateral radiculopathy was also present in over half. Sensory findings were present in 18 (74.8%) patients. Bladder dysfunction was observed in 18 (74.8%) patients who had 100 cc of postmixture residual urine. Two patients had sphincter involvement (8.32 %) and did not improve postoperatively. Uchiyama et al. have reported that more than 83 % of the patients have urinary symptoms.

Ependymomas of the conus and cauda equina almost exclusively have a myxopapillary histology (8,16). Conus medullaris ependymomas originate from the ependymal cell cluster of the ventriculus terminalis in the filum (4,14,16,23). There are four histological types of ependymomas papillary, cellular, epithelial, or mixed. Myxopapillary ependymoma is most common ependymoma at the conus level. Cystic degeneration is observed in 40 %. Rostral and caudal cysts are seen in 90% of all patients; all cysts were are on T1-weighted images and hyperintense on T2-weighted images. This study included only one case of cystic ependymoma (Figure 3). Tumor-related cysts can be classified into three types (4,7,8). The first type is cystic tumors, most likely resulting from tumor necrosis or hemorrhage. The second type is syrinx formation, and the third type is the tumor-associated cyst, which is the most commonly seen. Ependymomas have a higher incidence of bleeding than other conus tumors. All ependymomas were slightly hyperintense on T2-weighted MR images and contrast enhanced. Precontrast, they were isointense on T1-weighted MR images. On histologic examination, these tumors contain rings of ependymal cells that surround cores of blood vessels and mucin (16). Histological examination revealed a benign ependymoma in all our patients. There is usually a plane of cleavage between tumor and spinal cord and complete excision can generally be achieved.

Neurinomas or schwannomas can present with macroscopic cyst formation (7). We have 5 cases of neurinoma, one of which was a malignant neurinoma (20 %). Giant neurinomas of this region are very rare (4). This study included only one such a case (Figure 6). These patients usually have multiple cysts. Total excision could not be achieved. Despite incomplete removal of the tumor, the risk of recurrence is low (13). They must be distinguished from lumbar disc herniations. Neurinomas usually appear hypointense in comparison to the spinal cord on unenhanced T1-weighted images and hyperintense on T2-weighted images. Typically, meningiomas appear isointense on both T1- and T2-weighted images. Neurinomas often have smoother margins but more irregular enhancement in comparison to meningiomas, and neurinomas have cystic degeneration and internal necrosis.17 Two of our patients underwent myelography, three patients CT myelography, while 20 patients were investigated by MRI.

Meningiomas arising in the lumbosacral region are uncommon, and represent only 2% of all spinal
cord meningiomas. Overall, spinal cord meningiomas occur more frequently in females, but the sex incidence is equal in the lumbosacral region (13). Meningiomas can be completely removed. All our meningiomas were the meningothelial subtype. For these tumors, it is important to remember that even patients who have a severe deficit preoperatively can have a dramatic improvement after removal of the intradural meningioma. These tumors have a classic appearance on MRI (Figure 9).

Epidermoid and dermoid tumors are probably developmental in origin, and are lined by keratinizing squamous epithelium. Dermoid tumors also contain cutaneous adnexae, which accounts for their greasy contents, often admixed with hair. They can be microscopically totally excised in up to 40% of cases (13). This is reasonable, provided it is not associated with substantial morbidity, as partial removal can effectively relieve symptoms, often for many years and permanently in some cases. All of these tumors were intramedullary. They comprise approximately 10% of all intradural spinal cord tumors. They arise most frequently at the lumbar site. Two of the 5 patients with dermoid and epidermoid tumors had dermal sinus (Figure 8); one of these patients had an improvement in symptoms, while the other case had incomplete removal and no improvement. These tumors (midline congenital tumors) had a poor prognosis in this study.

Spinal intradural hemangioblastomas make up between 2 and 6% all spinal cord tumors in most series (17). The origin of hemangioblastomas has not been clearly delineated. Hemangioblastomas may arise sporadically as single lesions or in the setting of Von Hippel-Lindau disease (VHL). Our case had VHL and was extramedullary. An association with VHL is found in only 30% of all spinal hemangioblastomas (17). We recommend complete excision because incomplete removal usually results in recurrence as in our case where regrowth was seen 8 years later. We used irradiation for treatment in this case. On unenhanced T1-weighted and T2-weighted MR images, the tumors are usually isointense. They can also be hyperintense on T2-weighted MR images. In post-contrast study, the tumor nodules of hemangioblastoma often have stronger enhancement (17).

Lumbar X-rays of our two patients whom has tumors of conus and cauda equina, occasionally demonstrates widening of the interpedicular distance, erosion of the posterior vertebral body, or enlargement of a neural foramen (scalloping). Because of their subarachnoidal location, these lesions are readily demonstrated by myelography or MRI (1,4,6-11,19,23,26). All clinically significant intradural-extramedullary tumors will be demonstrated by myelography, while small intramedullary conus tumors can be missed. When an intramedullary tumor produces complete obstruction, differential diagnosis from extramedullary tumors may be difficult. More frequently, the tumor produces a partial block, displacing the contrast column laterally; this appearance merely represents enlargement of the spinal cord and distinction from myelitis, intramedullary hematoma or syringomyelia may be impossible (2,21,25). In one of our cases, ICCET produced partial obstruction, displacing the contrast column laterally (fig1), not detected by other investigation methods as CT and MRI. More recent studies found MRI to be equal to or better than a CT myelogram. CT myelogram carries the risk of contrast dye which may induce nephrotoxicity, worsening neurologic function due to positioning for the procedure, and bleeding complications from the lumbar puncture. The advantages for MRI are multifold in the radiological evaluation for spinal cord tumors. In addition, MRI better identifies lesions for surgical interventions, and better differentiates extradural and intradural lesions. Contraindications to MRI include patients with pacemakers, non-compatible metallic implants, and severe claustrophobia (12,20). Contrast-enhanced MRI improves tumor characterisation and delineation. Almost all tumors enhance, and gadaolinium helps to separate the solid portion of tumors from adjacent edema and associated cysts (3).

All of the tumors were totally excised except one case (96%). Sphincter dysfunction was present in four patients on admission and improved by following surgery in all except one. Back pain was the most important complain, present in 21(84%). There was bowel, bladder and sexual organ dysfunction in 6 cases, and only urodynamic dysfunction in 19 (79 %) patients on admission and these were improved. In addition, spinal stability was rarely compromised.

The surgical management of ICCET is a complex decision making process. The main determinants for surgical candidacy are tumor histobiology,
neurologic condition, and general medical condition. Surgery for ICCET is performed from the dorsal approach via Th11-12-L1 laminectomies. We generally resect ICCET tumors with the aid of the operating microscope and microneurosurgical techniques. Many surgeons do not utilize evoked potential monitoring for ICCET. We could not use peroperative spinal cord monitoring because it is unavailable. Peroperative frozen section must be used in conjunction with the clinical appearance and we used it for all of our patients. If a CSF leak is determined postoperatively, which usually occurs within the first week of surgery, a lumbar subarachnoidal drain may be placed at the time. If CSF diversion via a lumbar drain is not adequate, then surgical reexploration and closure of the dura may be necessary (15). CSF leak was determined in 2 of our cases postoperatively in Day 2 and a lumbar drain inserted was adequate for 3 days. There were no another surgical complications in this study.

Spinal cord tumors account for 15% of all nervous system neoplasms, with 10% of these found in the conus and cauda equina (1). They do not metastasize except myxopapillary ependymomas that can rarely do so (9). The frequency of tumor type was different in our series. The predominant tumor was ependymoma, but neurofibromas are the most common tumor in the conus region in other series (4,18). We did not detect a neurofibroma in our cases. The roles for radiotherapy in the management of these benign tumors are limited. The correct diagnosis of ICCET and the definition of their subtype are always made postoperatively from the histological study of these tumors. Best functional outcome depends on duration of the symptoms and total removal of the tumor. The remaining 19 patients at long-term follow-up study (mean duration 75.5 months) were neurologically stabl, while remaining 4 cases minimal or moderate disability.

CONCLUSION

Long-term outcomes becomes more favourable by the development of new technologies for the diagnosis and treatment of ICCET. These developments have allowed more accurate preoperative diagnosis and safer, more effective operative interventions. Although follow-up surveillance may be needed, a majority of patients with solitary conus and cauda equina lesions can be cured, with gross total removal of the lesion. Probably the introduction of MRI will allow an early diagnosis of these lesions and the use of new surgical instruments. Early diagnosis gives excellent results and the prognosis is apparently directly related to the length of the preoperative clinical history.

REFERENCES