9 Current Concepts of Surgical Therapy of Intradural and Intramedullary Tumours

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9.1 Introduction

Neoplasms of the spinal canal may be classified on the basis of their location into epidural, intradural-extramedullary and intramedullary tumours (Table 9.1). Whereas the majority of epidural neoplasms are metastatic in nature, the intradural tumours are often primary tumours. In neurosurgical practice, intraspinal neoplasms account for approximately 15% of all primary tumours of the central nervous system and its sheath elements (RUSSELL and RUBINSTEIN 1989). The relative frequency of epidural, intradural-extramedullary and intramedullary tumours is not exactly known. Based on available epidemiological data, metastatic spinal tumours are three to four times more common than primary spinal tumours (BYRNE and WAXMAN 1990). The average annual incidence of primary intraspinal neoplasms is reported to be 1.3 per 100,000 population, although figures ranging from 0.8 to 2.5 are quoted (FOGELHOLM et al. 1984). Metastatic disease to the spine is discussed in detail elsewhere in this book. This chapter will focus on the surgical treatment of intradural neoplasms.

Table 9.1. Classification of spinal neoplasms according to location

<table>
<thead>
<tr>
<th>Location</th>
<th>Tumours</th>
</tr>
</thead>
<tbody>
<tr>
<td>Epidural tumours</td>
<td>Metastasis, Multiple myeloma, Osteogenic sarcoma, Chordoma, Chondrosarcoma, Lipoma, teratoma</td>
</tr>
<tr>
<td>Intradural-extramedullary tumours</td>
<td>Meningioma, Nerve sheath tumour, Vascular malformation and tumour, Epidermoid and dermoid tumour, Lipoma, teratoma, Metastasis</td>
</tr>
<tr>
<td>Intramedullary tumours</td>
<td>Ependymoma, Astrocytoma, Ganglioglioma, Vascular malformation and tumour, Metastasis</td>
</tr>
</tbody>
</table>

9.2 Intradural-Extramedullary Tumours

The first successful surgical removal of an intraspinal neoplasm was that of an intradural-extramedullary tumour by Sir Victor Horsley in 1888. Large series have been reported since by ELSBERG (1925), LIST (1941), SLOOFF et al. (1964), GIUFFRE (1966) and LEVY et al. (1982). Intradural-extramedullary tumours are usually the histologically benign meningiomas and nerve sheath tumours, although (epi)dermoid cysts, teratomas, lipomas and metastatic neoplasms to the leptomeninges also occur.
9.2.1 Meningiomas

Meningiomas constitute one of the most common forms of spinal tumour. The literature remains divided as to whether meningiomas or nerve sheath tumours head the list. The large series of 1322 primary intraspinal tumours described by Slooff et al. (1964) recorded an incidence of 29% of schwannomas, 25.5% of meningiomas, 22% of gliomas and almost 12% of sarcomas. In other series, meningiomas made up 33–47% of primary spinal tumours (Byrne and Waxman 1990).

Meningiomas may arise from any of the cell elements that form the meninges but the majority stem from arachnoid cells. The tumour is usually firmly fixed to the lateral dura at the level of the dentate ligament and may extend anteriorly or posteriorly. In the cervical region an anterior location seems to be predominant. Meningiomas may occur at any level along the spinal axis, but it is generally accepted that the thoracic region is involved far more often than other levels. Occasionally the cervical segments are affected, whilst lumbar examples are rare. Spinal meningiomas are much more frequently encountered in women than in men. The most (approximately 80%) meningiomas in women occur in the thoracic region, whereas in men a nearly equal frequency of cervical (41%) and thoracic (47%) lesions has been reported (Levy et al. 1982). The reason for this predilection for the thoracic spine in women is unknown. Multiple spinal meningiomas may occur in von Recklinghausen’s disease.

The duration of symptoms arising from spinal meningiomas may be quite variable. Occasionally patients have an abrupt onset of symptoms, often precipitated by trauma. Usually, symptoms begin insidiously and progress over many months. As in most other spinal tumours, back pain is the most common presenting symptom and is usually progressive. Radicular pain is often a prominent complaint and may be present for months or years prior to diagnosis. In meningiomas the pain may be bilateral, contrary to neurofibromas where it is usually unilateral (Byrne and Waxman 1990). Subjective complaints of radicular paraesthesia appear to be common in cases of meningioma, with frequencies of 23–37% reported (Guidetti and Fortuna 1975). Sphincter disturbances are unusual early manifestations in intradural-extradural tumours unless the conus or cauda equina is involved. A meningioma en plaque, in which the tumour forms a diffuse collar-like mass around the cord, is rare. Occasionally, the spinal meningiomas are separate from the dura and deeply embedded in the cord.

Epidural meningiomas have been known to occur in up to 10% of cases (Byrne and Waxman 1990). It is noteworthy that epidural spinal meningiomas demonstrate a relatively high incidence in childhood and show a predominance in males (Kepes 1982). Epidural meningiomas are considered biologically more aggressive.

Abnormalities on plain spine radiographs occur in only 10% of patients with spinal meningiomas. The choice of further radiographic procedures may include myelography, CT scanning following intravenous contrast administration, CT myelography and MRI. The MRI scan is of course the most attractive because it is noninvasive, shows the entire spinal canal and surrounding structures in great detail, and can differentiate intramedullary neoplasms from syringomyelia, as well as demonstrate the relationship between the spinal cord and extramedullary lesions (Fig. 9.1). The characteristic radiographic features of meningiomas are discussed in detail elsewhere (Chap. 1).

9.2.2 Nerve Sheath Tumours

Russell and Rubinstein (1989) classified nerve sheath tumours into two main categories, which are usually readily distinguishable on both histological and electron microscopic grounds: the first, typified by a solitary encapsulated tumour of the nerve roots and peripheral nerves, is the schwannoma; the second is the neurofibroma, in which connective tissue fibres are a conspicuous element and nerve fibres within the substance of the tumour are often both more numerous and microscopically more obvious than in the schwannomas. This is not to say that neurofibromas are “unencapsulated” lesions, as previously thought. This characterisation was based upon histological discovery of nerve fibre elements within the tumour and not upon gross appearance or resectability. Most neurofibromas do have a capsule, and nerve fascicles are often peripherally enclosed in its layers (Donner et al. 1994).

Alternative designations for schwannoma still used today include “neurinoma” and “neurilemoma” (often spelt “neurilemmoma”). The name schwannoma is preferable as it explicitly identifies the cell type involved (Russell and Rubinstein 1989). While these peripheral nerve sheath tumours are two distinct entities, their clinical appearance and