Intradural extramedullary spinal metastasis

A report of 10 cases

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The management of 10 patients with symptomatic localized intradural extramedullary spinal metastasis is reviewed. The single most common primary source was carcinoma of the breast (four cases). The initial symptom in nine patients was pain, with five patients reporting a characteristically severe cramping discomfort with radicular distribution. All patients underwent laminectomy decompression. At the time of surgery, six of the patients were weak but ambulatory and four were bedridden. Following surgery, four patients enjoyed some measure of pain relief, seven patients became ambulatory, and three remained bedridden. Two patients achieved a "satisfactory" result, and were walking and continent 6 months after surgery. Secondary brain tumors were demonstrated or implicated in nine patients, supporting the concept that the spinal metastases represented tertiary deposits following dissemination via the cerebrospinal fluid.

Symptomatic intradural extramedullary spinal metastasis causes a virulent clinical syndrome with poor prognosis and disappointing outcome after treatment. Given the high incidence of associated cerebral metastatic involvement, total neuraxis radiation and/or chemotherapy should be considered when symptomatic spinal metastasis is discovered to be intradural and extramedullary.

KEY WORDS • spinal cord compression • intradural extramedullary metastasis • metastatic tumor • spinal metastases • brain tumor • pain

SYMPTOMATIC spinal metastasis represents a serious complication of systemic cancer. The devastating morbidity associated with relentlessly progressing cord compression due to spinal metastases is well recognized. The vast majority of symptomatic spinal metastases occur in the extradural space. Intradural extramedullary metastases are unusual by comparison. Intradural extramedullary spinal metastasis causing spinal cord and/or root compromise is seldom encountered and has rarely been reported. We describe here the management of 10 such cases.

Summary of Cases

Clinical Material

Among 200 patients treated with symptomatic spinal metastases by the Division of Neurosurgery at the Wellesley Hospital, spinal cord or cauda equina compression was due to extradural spinal metastasis in 189 cases. An intramedullary spinal metastasis was confirmed at surgery in one patient. Localized intramedullary tumor was discovered in 10 cases, and these are the subject of this report.

Our series includes four men and six women, ranging in age from 39 to 68 years (with a mean age of 55 years). The culpable primary sites were the breast in four cases, the lung in three, the skin (melanoma) in two, and the uterus in one.

Signs and Symptoms

The clinical features in this group were similar to the manifestations of symptomatic extradural spinal metastases. Pain was the initial symptom in nine of the 10 patients. The pain was often worse at night. Five patients reported a radicular component, which was characteristically described as a "severe cramping" and episodic "shock-like" discomfort. The pain was present an average of 2 months before surgery. Local tenderness was present over the involved spinal segments in eight of the 10 cases. Motor function was impaired in all patients. Six were weak but ambulatory, and four were bedridden at the time of surgery. A clear-cut sensory level was present in all cases. Bowel and bladder dysfunction was recorded in eight patients preoperatively (Table 1).
TABLE 1
Relative frequency of clinical features in 10 cases

<table>
<thead>
<tr>
<th>Feature</th>
<th>Percent of Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>pain</td>
<td>90</td>
</tr>
<tr>
<td>motor function</td>
<td>60</td>
</tr>
<tr>
<td>weak (4/5)</td>
<td>40</td>
</tr>
<tr>
<td>bedridden</td>
<td>100</td>
</tr>
<tr>
<td>sensory level</td>
<td>80</td>
</tr>
</tbody>
</table>

TABLE 2
Results of spinal decompression in 10 cases

<table>
<thead>
<tr>
<th>Result</th>
<th>Percent of Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>satisfactory*</td>
<td>20</td>
</tr>
<tr>
<td>pain improved</td>
<td>40</td>
</tr>
<tr>
<td>motor function</td>
<td>70</td>
</tr>
<tr>
<td>no improvement</td>
<td>30</td>
</tr>
</tbody>
</table>

* Continent and able to walk 6 months after surgery.

The symptoms of spinal involvement were preceded by manifestations of intracranial metastases in three patients. A solitary melanoma deposit, presenting at the cortex, was removed 4 months earlier in one patient, and fluctuating focal neurological deficits were recorded in two others. Following surgery, intracranial metastatic disease was either suspected or confirmed in six patients.

Investigations and Surgery

Plain x-ray films were of little help in localizing the spinal lesion; myelography, on the other hand, was positive in all cases. A complete block was demonstrated in seven patients, and a partial block was shown in the remaining three. The distribution of intradural extramedullary spinal metastasis is shown in Fig. 1. In one case the metastasis occurred in the midthoracic region (T-5), and in nine it was found entangled in the nerve roots about the thoracolumbar junction (T10–L3). Cerebrospinal fluid (CSF) cytology was positive in two patients.

Surgical treatment involved extensive laminectomy, decompression, and intradural exploration at the involved segments. An attempt was made to remove as much of the extramedullary tumor as possible, with meticulous dissection from the cord and roots. Nerve roots were generally matted down in tumor, and little could be accomplished beyond local decompression. Primary dural closure was achieved in two cases, a dural graft was used in three, and the dura was left open in five patients.

Operative Results

No patient was worse as a result of spinal decompression. There was no operative mortality (that is, within 30 days of surgery).

Four patients reported improvement in preoperative pain, but only one enjoyed pain relief. The remaining six patients required high doses of analgesics following surgery, and one underwent chordotomy because of continuing, characteristically cramping, dysesthetic pain. Seven patients were ambulatory and three bedridden following surgery (compared with six patients who were ambulatory and four bedridden before surgery), representing a net categorical gain in one case (Table 2).

A "satisfactory" result has been defined as patients who are continent and able to walk 6 months following surgery. Only two patients qualified for this distinction in the present series.

Discussion

Intradural extramedullary spinal metastasis originating from a primary tumor outside the central nervous system (CNS) is rare. Few isolated case reports have appeared in the literature. Our series includes 10 patients in whom spinal cord and/or nerve root compression was due to localized intradural extramedullary spinal metastasis.

The spread of primary brain tumors by CSF dissemination is well recognized. A similar mechanism...
Intradural extramedullary spinal metastasis can account for the majority of intradural extramedullary spinal metastases in our series. Secondary brain tumors were demonstrated, or implicated, in nine of our 10 cases, supporting the concept that the spinal metastases represented tertiary deposits following dissemination via the CSF. Furthermore, CSF cytology was positive in both patients whose CSF was examined for malignant cells. It has been suggested that free tumor cells arising from CNS secondary sites settle out in the CSF by gravity, giving rise to the tertiary deposits. Consistent with such a mechanism is the observation that the intradural extramedullary metastases were found entangled among the cauda equina roots in the majority (90%) of our cases.

Other theoretical possibilities for the pathogenesis of intradural spinal metastases include spread by transdural invasion, hemogenous dissemination, or extension along perineural lymphatic ducts of spinal nerve roots (very rarely). Spread by CSF would seem to be the most likely mechanism in our experience.

Symptomatic spinal metastases generally present with a characteristic clinical syndrome. Local back or neck pain is the earliest complaint in about 80% of patients and is followed by weakness, sensory loss, and sphincter dysfunction.

When we compare the clinical features of extradural with intradural extramedullary spinal metastases, a number of distinctions are apparent. Intradural extramedullary metastases are relatively rare (5% in our experience of 200 consecutive cases with neurosurgically treated symptomatic spinal metastases). While pain is the earliest and most prominent complaint in the vast majority of patients with intradural extramedullary metastasis, radicular distribution is a prominent feature and the pain is characteristically severe and crampy in nature. Furthermore, clear-cut sensory loss and sphincter dysfunction is present approximately twice as often with intradural extramedullary spinal metastases. A “winking owl” sign (the most common finding on plain films of the spine in patients with symptomatic extradural metastasis) was not seen when the metastasis was intradural and extramedullary. Clinical deterioration occurs more rapidly and surgical decompression is much less effective in cases with intradural extramedullary spinal metastasis. Less than half of such patients obtained any measure of pain relief, and a satisfactory result was obtained in only 20% of our series.

The association of intradural extramedullary spinal metastasis with overt or latent CNS secondary disease suggests that total neuraxis radiation and/ or chemotherapy should be a major consideration in the treatment of these patients.

Conclusions

Spinal cord and cauda equina involvement by intradural extramedullary metastasis is a rare complication of systemic cancer and results in devastating morbidity and mortality. The signs and symptoms to some extent resemble those associated with symptomatic extradural spinal secondary tumors; however, the clinical course is more virulent, the prognosis is significantly worse, and the outcome following treatment is less satisfactory in the case of intradural extramedullary metastasis. Given the high incidence of associated cerebral metastatic involvement, total neuraxis radiation and/or chemotherapy should be considered when symptomatic spinal metastasis is discovered to be intradural and extramedullary.

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References


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