Intradural chordoma without bone involvement

Case report and review of the literature

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Chordomas are most commonly of extradural origin and associated with bone destruction. The authors describe a rare case of an intradural chordoma. To the best of their knowledge, this is the first case in which multiple lesions were found intradurally without bone involvement; the lesions were disseminated through the thoracolumbosacral spinal cord and extended into the paraspinal muscles, and metastatic lesions in the cervical cord were also present.

KEY WORDS • chordoma • intradural lesion • lumbosacral spine • paraplegia

C HORDOMAS are rare neoplasms of the axial skeleton that arise from the remnants of the primitive notochord (chorda dorsalis).6,17 They are mostly low-grade malignant tumors accounting for 1 to 4% of all malignant bone neoplasms.4 Approximately 50% arise in the sacroccygeal region, and 35% arise in the skull base, where they typically involve the clivus.19 The remaining 15% occur in the midline along the path of the notochord, primarily involving the cervical vertebrae.2,10,14 This malignant mass is thought to be essentially extradural in nature and is generally associated with extradural extension and bone destruction. If left to grow for long periods, however, it may invade the dura mater, extending intra- and extradurally.13 The incidence of metastasis, which has been reported to be as high as 25%, is greater than previously believed and may be related to its long clinical history. In order of most to least frequent, the common sites of distant metastases are the lungs, liver, and bone.2

We describe the rare case of a patient with a primary intradural chordoma, located in the dorsolumbar and lumbosacral spinal cord.

Case Report

History, Multiple Presentations, and Clinical Course

This 50-year-old woman had a 28-year history of chronic low-back pain. Eight years before presentation she complained of an increase in pain after long periods of sitting. This pain was associated with severe headaches, deterioration of vision, and numbness and loss of strength in the right leg.

October 1997. The clinical examination showed a positive Lasègue sign on the right and hypesthesia in the right foot. An axial CT scan of the lumbar spine revealed a wide spinal canal but no other lesions. Magnetic resonance imaging demonstrated multiple soft-tissue lesions, with intradural and extramedullary extension. The largest masses were observed at the thoracolumbar level. There was no neuroimaging evidence of bone involvement (Fig. 1).

November 1997. The patient underwent a T10–L2 decompressive laminectomy and total excision of a partly intramedullary, mostly extramedullary tumor that infiltrated the paraspinal muscles and the pleura. Intraoperatively, the intraspinal part was vaporized and removed as two voluminous masses. To the right, a voluminous extraspinal mass, which had not been visualized preoperatively on CT or MR imaging, and which infiltrated the muscles, was detected. The dissection was extended into the muscles. A large (12 × 7–cm) intramuscular mass was removed entirely. Because massive invasion of the pleura was detected, a thoracotomy was performed to remove the intrapleural portion. This intrapleural part could only be removed partially because of the extreme-lateral extension. Anatomicopathological examination indicated a diagnosis of a chordoma in all resected specimens.

February 1998. The patient’s symptoms recurred. On MR imaging a large cyst was visualized at L3–4. On puncturing of the cyst, 10 ml of dark yellow fluid was aspirated. This procedure did not relieve the patient’s pain. Examination of the aspirated fluid showed some groups of cells, histologically and immunohistochemically characteristic of chordoma.

Abbreviations used in this paper: CT = computerized tomography; MR = magnetic resonance.
March 1998. In March 1998 a second operation was performed. An L4–S2 laminectomy was performed as was subtotal resection of a cauda equina chordoma. Intraoperatively, an enormous arachnoiditis was detected. The dura was very fragile and a purple mass shone through it. Between two fibers of the cauda equina an intratumoral cyst was seen as a green mass. Only a subtotal resection could be performed because the fibers of the cauda equina were adherent to the tumor capsule.

June 1998. In June 1998 MR imaging was again performed. The study demonstrated multiple intradural cystic structures, a total loss of delineation between the fibers of the cauda equina, and the lumbar mass that extended around the sacral nerve roots. No bone involvement was demonstrated. The tumor was inoperable. One month later electromyography demonstrated an L-5 radiculopathy bilaterally and an S-1 radiculopathy on the left side with significant effects on contractions. There were no motor potentials found in the left anterior tibial muscle and the long extensor muscle of the toes.

August 1998. The patient was admitted to the rehabilitation center, suffering from incomplete T-12 paraplegia (American Spinal Injury Association Grade D). She could walk with assistance for 10 m. She had a drop foot on the left side. Sensory function was seriously impaired in both legs.

October 1998. Magnetic resonance imaging of the full spine was performed to determine the cause of functional deterioration. The L5–S1 chordoma remained unchanged, but the multiple contrast-enhancing lesions in the spinal canal at the thoracolumbar level had grown. A small C-3 nodular lesion and a small lesion at the craniocervical junction were newly detected. There were signs of fibrosis with formation of cysts on the lumbar and thoracic levels. All lesions were located intradurally.

Because metastatic cervical lesions were present, head MR imaging was performed but revealed no parenchymal lesions. The clivus appeared normal on these images.

Ultrasonography of the liver demonstrated no metastatic lesions, and an axial CT scan of the thorax revealed no lesions in the lungs. We observed only a small nodular lesion of the paraspinal pleura on the right. Bone scintigraphy demonstrated no metastases.

April 1999. Because the tumor progression was slow and because of its wide spread, we decided to perform repeated MR imaging evaluation 6 months later. This study demonstrated numerous (at least seven) new small intradural spots in the cervical and high-thoracic spinal cord. Those in known locations remained unchanged.

May 1999. Clinical evaluation revealed no new neurological deficits. The patient could still walk with assistance for a distance of approximately 10 m.

Because of the absence of clinical deterioration, the slow tumor progression, and the wide tumor spread, we decided not to undertake radiotherapy, because it is not effective for such an extended chordoma. When and if clinical deterioration occurs, a new MR imaging study

FIG. 1. Sagittal MR image revealing an intradural tumor without bone involvement at the thoracolumbar level.

FIG. 2. Photomicrographs of the intradural tumor showing a lobulated structure composed of physaliphorous cells on a chondroid background. H & E, original magnification × 100 (upper) and × 200 (lower).
Histological Findings

In November 1997 a tumor sample was obtained, and histological examination showed a lobular growth pattern. The lobules consisted of a proliferation of large cells with multiple vacuoles (physaliphorous cells) on a chondroid background. The cells contained a small round nucleus, occasionally with a nucleolus included. More cells were positive for epithelial membrane antigen. More cells were positive for KL1 and all cells are positive for S-100.

Discussion

Including the present case, 15 cases of intradural chordomas have been reported (Table 1). Their locations included the preoptine region in seven cases, the suprasellar region in two cases, the spinal region in three cases, and the intrasellar region, foramen magnum, and tentorium cerebelli in one case each. The intracranial region is overwhelmingly the most common site for intradural extraosseous chordomas, especially the preoptine region.13

To our knowledge, this report represents the first published case in which multiple lesions were found intradurally without bone involvement, and were disseminated through the thoracolumbosacral spinal cord, invading into the paraspinal muscles and the pleural cavity, and in which metastatic lesions were present in the cervical spinal cord.

In all other 14 cases the chordoma was found in an entirely intradural location; an intradural chordoma is significantly less aggressive than one of an osseous origin. In all other cases the tumors remained intradural without extension to the extradural space. There was no bone involvement and no metastases were found. Therefore, a new classification was proposed by Jallo, et al.,7 to serve as a better predictor of the prognosis. In their system, Type I chordomas are osseous extradural (the majority); Type II are extrasosseous extradural; Type III are osseous intradural; and Type IV chordomas are extrasosseous intradural.8 In their system chordomas are separated into four categories based on the space they occupy as well as the presence/absence of an osseous connection. Type II and IV lesions are thought to have a better prognosis because complete resection is more likely (due to their more limited local extension) and because of their less aggressive behavior. Metastases have never been associated with intradural extraosseous chordomas.

Our case is unusual in several respects. First, because an intradural location without bone involvement is rare in cases of chordoma (only 14 previous cases reported). Second, this intradural chordoma was located in the spinal cord, whereas the most common location is intracranial (11 of the 14 previously reported chordomas). Third, it is an unusual case because of the presence of the multiple lesions in the spinal cord and, furthermore, because of the tumor’s extension through the dura into the paraspinal

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs), Sex</th>
<th>Location</th>
<th>Size (cm)</th>
<th>Symptoms &amp; Signs</th>
<th>Resection</th>
<th>Radiotherapy</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dublin &amp; MacCarty, 1952</td>
<td>ND, M</td>
<td>preoptine</td>
<td>2 * 1 * 0, 2</td>
<td>asymptomatic</td>
<td>no</td>
<td>no</td>
<td>died of unrelated disease</td>
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<tr>
<td>Bartolini, 1974</td>
<td>57, M</td>
<td>preoptine</td>
<td>1/2 size of nut</td>
<td>headache, dysartria, cranial nerve palsies, hyperreflexia, mydriasis, hemianopsia</td>
<td>no</td>
<td>no</td>
<td>died of SAH</td>
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<tr>
<td>Mathews &amp; Wilson, 1974</td>
<td>41, M</td>
<td>intrasellar</td>
<td>ND</td>
<td>acromegaly, myxedema, bitemporal hemianopsia</td>
<td>subtotal</td>
<td>yes</td>
<td>normal visual field</td>
</tr>
<tr>
<td>Mapstone, et al., 1983</td>
<td>26, M</td>
<td>preoptine</td>
<td>large</td>
<td>cranial nerve palsies, pt hemiparesis, cerebellar sign</td>
<td>subtotal</td>
<td>ND</td>
<td>ND</td>
</tr>
<tr>
<td>Vaquero, et al., 1983</td>
<td>39, F</td>
<td>suprasellar</td>
<td>ND</td>
<td>headache, apathy, confusion, changes in emotional state</td>
<td>total</td>
<td>ND</td>
<td>died 3 wks postop</td>
</tr>
<tr>
<td>Ramiro, et al., 1986</td>
<td>33, F</td>
<td>thoracic spine</td>
<td>2 * 2</td>
<td>paraparesis, Babinski sign, hyperreflexia, T-6 sensory level</td>
<td>total</td>
<td>ND</td>
<td>no neurological deficit</td>
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<td>Yuhi, et al., 1986</td>
<td>28, M</td>
<td>preoptine</td>
<td>ND</td>
<td>headache, papilledema, cranial nerve palsies, gait disturbance</td>
<td>total</td>
<td>ND</td>
<td>no neurological deficit</td>
</tr>
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<td>Katayama, et al., 1991</td>
<td>30, F</td>
<td>foramen magnum</td>
<td>ND</td>
<td>coma, paraparesis, hypesthesia</td>
<td>total</td>
<td>no</td>
<td>diminished paraparesis, hypesthesia</td>
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<tr>
<td>Warnick, et al., 1991</td>
<td>58, M</td>
<td>tentorium cerebelli</td>
<td>ND</td>
<td>lethargy, dysarthria, limited upgaze, nystagmus</td>
<td>subtotal</td>
<td>ND</td>
<td>Left hemianopsia</td>
</tr>
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<td>Hardie, 1992</td>
<td>28, F</td>
<td>preoptine</td>
<td>ND</td>
<td>headache, hearing loss, diplopia</td>
<td>ND</td>
<td>ND</td>
<td>rt hearing loss</td>
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<td>Tashiro, et al., 1994</td>
<td>56, F</td>
<td>suprasellar</td>
<td>3, 5 * 3</td>
<td>paraparesis, cranial nerve palsies</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
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<tr>
<td>Wainbuchi, et al., 1994</td>
<td>54, M</td>
<td>suprasellar</td>
<td>4 * 4 * 3</td>
<td>left homonymous hemianopsia</td>
<td>total</td>
<td>no</td>
<td>hemianopsia resolved</td>
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<td>Vaz, et al., 1995</td>
<td>37, F</td>
<td>C5–T2</td>
<td>ND</td>
<td>paresthesias of arms, abnormal tendon reflex</td>
<td>subtotal</td>
<td>no</td>
<td>paresis of rt hand</td>
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<tr>
<td>Nishigaya, et al., 1998</td>
<td>28, M</td>
<td>preoptine</td>
<td>4, 5 * 4 * 2</td>
<td>headache, cerebellar sign</td>
<td>subtotal</td>
<td>no</td>
<td>rt moderate hearing loss</td>
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<td>present study</td>
<td>50, F</td>
<td>lumbar, thoracolumbar, cervical metastasis</td>
<td>multiple lesions, largest 12 * 7</td>
<td>back pain, headache, loss of strength &amp; sensibility in rt leg</td>
<td>subtotal</td>
<td>no</td>
<td>incomplete T-12 paraplegia, L5–S1 radiculopathy</td>
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</tbody>
</table>

* ND = not described; SAH = subarachnoid hemorrhage.

will be performed, and focal radiotherapy will be considered when surgery is impossible.
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muscles. Finally, the case is unique because metastases developed in the cervical spinal cord, whereas intradural extraosseous tumors are thought to have a less aggressive behavior.

This case does not fit into the classification proposed by Jallo, et al.7 because intradural and extradural components, multiple lesions, and cervical metastases were present. Their classification is based on very few cases (three cases of Type II and 13 cases of Type IV). Therefore, their system must be regarded as speculative. Additionally, significant long-term follow-up data are not available. We believe that the benefit of their classification is more theoretical than practical.

The findings in this case indicate that an intradural chordoma, when left untreated for a long period of time (several years), can finally penetrate the dura and extend into other tissues; additionally we found that metastases may develop.

References


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