Intramedullary spinal neurinomas

Report of two cases

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Two cases of intramedullary neurinoma are presented in patients with no signs of von Recklinghausen’s disease. Only 16 similar cases have been published so far. The relevant literature is reviewed.

KEY WORDS • neurinoma • intramedullary tumor • pathogenesis • von Recklinghausen’s disease • spinal cord tumor

EXTRAMEDULLARY spinal neurinomas are a frequent occurrence, but the intramedullary variety is very rare in patients not presenting signs of von Recklinghausen’s disease. Only 16 such cases have been reported to date. This study describes two cases of intramedullary neurinoma found in a series of 145 patients with spinal neurinomas and 173 intramedullary spinal gliomas operated on at the Institute of Neurosurgery, University of Rome, from 1955 to May, 1981. The relevant literature is reviewed.

Case Reports

Case 1

The first case has already been described elsewhere. This 57-year-old man gave a history of thoracolumbar vertebral pain radiating to the right lower limb. The pain occurred mostly when the patient was in the recumbent position. Neurological examination on admission elicited a right-sided Laségue’s sign with ankle dorsiflexion at 10°, and no right ankle reflex. Lumbar puncture with manometry revealed a total block on Queckenstedt’s test, and examination of the cerebrospinal fluid (CSF) yielded 4 gm% albumin. Plain films of the thoracolumbar spine were normal, but Pantopaque myelography by a suboccipital route revealed a total block at T-12.

A T12-L1 laminectomy was performed, with complete removal of a well encapsulated tumor that lay partly within the conus and partly outside in contiguity with the roots of the cauda equina. Histological examination showed that the tumor was a neurinoma (Fig. 1). The postoperative course presented no problems apart from slight urinary incontinence, which cleared within a few days. The patient was discharged after 20 days, with mild superficial hypesthesia in a saddle distribution.

FIG. 1. Case 1. Photomicrograph of the tumor showing bipolar cells with arrangement of the nuclei in palisades. H & E, × 250.
FIG. 2. Case 2. Myelogram, anteroposterior view, showing widening of the cervical cord at the level of C-7.

FIG. 3. Case 2. Intraoperative photographs. Left: A well circumscribed tumor can be seen in the left dorsolateral portion of the cord. Center: The tumor is totally removed. Right: The tumor bed can be seen inside the cord.

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Case 2

This 54-year-old woman had a 2-year history of cervical pain and paresthesias of the left arm, spreading some months later to the left lower extremity. Eight months before admission, she felt a loss of strength in the left limbs. Neurological examination on admission showed weakness of the left extremities, increased tendon reflexes with ankle clonus on the same side, superficial hypesthesia from C-6 to T-4, and hypoalgesia as far as T-2, mainly on the left side. Lumbar puncture with manometry revealed total block on Queckenstedt’s test. On cytochemical examination of the CSF, the albumin value was found to be 2 gm%. Plain films showed a cervical canal that was wider than normal, without alterations of the peduncles or vertebral bodies. Myelography with metrizamide administered by a lumbar route showed widening of the spinal cord at C6-7, with block of the contrast medium at C-6 (Fig. 2).

Laminectomy was performed at C-3 to C-7, and a well encapsulated tumor was found occupying the left posterolateral portion of the cord at the C3-5 level and extending to the surface (Fig. 3). The posterior roots of C-3 and C-4, which were involved in the tumor, were cut, and the tumor was totally removed. Histological examination showed that the tumor was a neurinoma (Fig. 4). The postoperative course was uneventful, and the patient was discharged without change in her neurological status, apart from improvement in the strength of the left leg.

Discussion

Neurinomas are fairly common spinal tumors, accounting for 30% of all such tumors. The classic site is extramedullary, either intradural or extradural, or a combination of the two. The rarity of the intramedullary variety is proved by the fact that only 16 cases have been described in the world literature to date (Table 1).

Intramedullary neurinomas are found mainly in men. Only two of the previously reported patients were women; our Case 2 is the third. The portion of cord most affected is the cervical segment, followed by the thoracic and lumbar segments (Table 1).

<table>
<thead>
<tr>
<th>Authors, Year</th>
<th>Sex, Age</th>
<th>Vertebral Level</th>
<th>Site</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kernohan, 1932</td>
<td>M, 12</td>
<td>C4-7</td>
<td>intramedullary center of cord</td>
<td>subtotal removal</td>
<td>died from meningitis; postmortem diagnosis</td>
</tr>
<tr>
<td>Roka, 1951</td>
<td>M, 30</td>
<td>high cervical</td>
<td>center of cord</td>
<td>biopsy negative</td>
<td>died from “cardiac disease”; postmortem diagnosis</td>
</tr>
<tr>
<td>Riggs &amp; Clary, 1957</td>
<td>M, 60</td>
<td>C4-5</td>
<td>behind ependymal canal</td>
<td>exploratory surgery negative</td>
<td>recurrence, reoperation, improvement</td>
</tr>
<tr>
<td>Ramamurthi, et al., 1958</td>
<td>M, 35</td>
<td>T-2</td>
<td>cord incised</td>
<td>removal</td>
<td>deterioration</td>
</tr>
<tr>
<td>Lang &amp; Bridge, 1959</td>
<td>M, 2nd decade cervical</td>
<td>intramedullary, surfacing</td>
<td>total removal</td>
<td>improvement</td>
<td></td>
</tr>
<tr>
<td>Scott &amp; Bentz, 1962</td>
<td>F, 35</td>
<td>T3-4</td>
<td>4 mm deep cord incised</td>
<td>subtotal removal</td>
<td>?</td>
</tr>
<tr>
<td>Lu, et al., 1963</td>
<td>M, 32</td>
<td>C1-6</td>
<td>cord incised</td>
<td>total removal</td>
<td>improvement</td>
</tr>
<tr>
<td>M, 43</td>
<td>C2-5</td>
<td>cord incised</td>
<td></td>
<td>subtotal removal</td>
<td>?</td>
</tr>
<tr>
<td>McCormick, 1964</td>
<td>M, 62</td>
<td>L-2 (medullary)</td>
<td>rt posterior funiculus, surfacing</td>
<td>total removal</td>
<td>deterioration</td>
</tr>
<tr>
<td>Mason &amp; Keigher, 1968</td>
<td>M, 37</td>
<td>T8-9</td>
<td>2 mm deep lt posterior funiculus, pia mater incised</td>
<td>total removal</td>
<td>deterioration</td>
</tr>
<tr>
<td>Cambier, et al., 1974</td>
<td>M, 60</td>
<td>C2-3</td>
<td>lt posterior funiculus, surfacing</td>
<td>total removal</td>
<td>deterioration</td>
</tr>
<tr>
<td>Schmitt, 1975</td>
<td>M, 68</td>
<td>conus</td>
<td>multiple small (max 0.5 mm) nodules in parenchyma</td>
<td>symptoms of intraspinal tumor; myelography negative; no treatment</td>
<td>death from “heart attack”; postmortem diagnosis</td>
</tr>
<tr>
<td>Wood, et al., 1975</td>
<td>M, 48</td>
<td>C1-3</td>
<td>rt posterior funiculus, surfacing</td>
<td>radiotherapy</td>
<td>thrombophlebitis in lower limb; pulmonary embolism; postmortem diagnosis</td>
</tr>
<tr>
<td>Pardatscher, et al., 1979</td>
<td>M, 41</td>
<td>C4-T8</td>
<td>multiple</td>
<td>removal of one &amp; decompression</td>
<td>death 8 months later after progressive deterioration</td>
</tr>
<tr>
<td>Vailati, et al., 1979</td>
<td>F, 40</td>
<td>T-9</td>
<td>2.5 mm deep posterior surface</td>
<td>total removal</td>
<td>deterioration</td>
</tr>
<tr>
<td>Cantore, et al., 1982</td>
<td>M, 57</td>
<td>conus-cauda</td>
<td>posterior surface</td>
<td>total removal</td>
<td>recovery</td>
</tr>
<tr>
<td>F, 54</td>
<td>C4-6</td>
<td>lt posterior funiculus, surfacing</td>
<td>total removal</td>
<td>recovery</td>
<td></td>
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</table>

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cross section, the structures most affected are the posterior funiculi in the cervical cord and the dorsal columns in the thoracic cord.

Histologically, all the published cases and our own have been typical Antoni type A neurinomas. The pathogenesis of this type of tumor is still under discussion, because Schwann cells are not normally present inside the spinal cord. Kernohan, who was the first to describe an intramedullary neurinoma, argued that it originated from the Schwann cells forming part of the small nerve fiber tracts that accompany vessels perforating the cord. This hypothesis has been supported by Riggs and Clary and by Scott and Bentz, and it explains the anterior location of some neurinomas. Other authors have demonstrated the presence of aberrant nerve fibers within the spinal cord from which the tumor might originate. Ectopia of Schwann cells that derive from the neural crest (namely, a shift toward the midline during closure of the neural tube) has been suggested by Ramamurthi, et al., as a possible origin of the tumor. Differentiation of mesenchymal cells into Schwann cells must also be mentioned as a pathogenetic factor. These hypotheses would account for the location of neurinomas in the central part of the spinal cord. On the other hand, the tumor may arise from the Schwann cells of the spinal roots, which enter the cord for a short distance, and from pial cells of neuroectodermal derivation at the point at which the posterior roots lose their sheath on entering the pia mater. Mason and Keigher have called this a "critical area." In these cases, the site of the neurinoma is posteriorly posterosilateral or posterior. The pathogenesis of the tumor in our Case 1 might be explained on the basis of a central intramedullary site, since the tumor was not continuous with the spinal roots. However, in Case 2 the presence of two roots in the tumor mass suggests that the tumor originated from the Schwann cells of the roots as they cross the pia.

It is important to distinguish intramedullary neurinomas from another distinct entity, namely, the proliferation of Schwann cells from reticulin and myelin fibers inside the spinal cord, which Russell and Rubinstein called "schwannosis." The latter condition does not present clinically as a tumor growth but is found chiefly in patients who have sustained severe traumatic lesions of the cord, in diseases of long duration, or in aged patients. For this reason, this is not regarded as a primary disease but as a local reaction of the spinal cord secondary to other diseases. There have been occasional examples of the condition in healthy cords.

From a histological viewpoint, the differential diagnosis rests on the abundance of axons and especially of myelin (not present in neurinomas), in which are found Verocay bodies and palisading nuclei (not encountered in schwannosis). Schwannosis of the "zona terminalis" of Lissauer, described by Hori in patients not bearing signs of von Recklinghausen's disease and considered by the author to be a forme fruste of this disease, is likewise to be considered as a pathological entity sharply distinct from intramedullary neurinoma.

References

3. Cowen D: Cited in Reference 1 (see p 730)
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