Unusual spinal cord enlargement related to intramedullary hemangioblastoma

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During the past 6 years, 60 intramedullary spinal cord tumors have been operated on at our institution, including a group of eight patients with spinal hemangioblastoma. Of special interest is the fact that seven of these eight patients displayed evidence of diffuse spinal cord enlargement over a considerable range above and below the level of the neoplastic growth. This observation seems to be unique to hemangioblastomas and appears to bear on their pathophysiology while having important implications in their surgical management.

Representative Cases

Case 1

This 24-year-old man was referred with a 14-month history of increasing lower-extremity weakness and progressive spasticity involving both legs. Examination showed hyperactive reflexes in all extremities and a spastic gait. A metrizamide myelogram showed widening of the entire spinal cord with the maximum diameter at the lower cervical and upper thoracic levels (Fig. 1 left). Spinal magnetic resonance (MR) imaging documented diffuse cord enlargement with several small cystic areas (Fig. 1 right). Spinal angiography demonstrated a solitary vascular lesion at T-2 (Fig. 2 left).

A laminectomy was performed from C-5 to T-5 and an intramedullary hemangioblastoma was completely removed from the T-2 level. Inspection of the spinal cord above the tumor revealed it to be gray and apparently edematous (Fig. 2 right). Postoperatively, the patient was ambulatory with a spastic gait and mild posterior column defect in the lower extremities.

Case 2

This 43-year-old man presented with a history of a progressive spinal cord syndrome. He initially noted paresthesias in the left arm and leg, followed by weakness of those extremities. Examination also revealed decreased pain and temperature sensation on the right side of the body below the shoulder. A myelogram showed marked widening of the entire cervical cord with dilated vessels (Fig. 3 left). An extensive laminectomy was performed from C-1 through T-1 with removal of an intramedullary hemangioblastoma confined to the C-2 level (Fig. 3 right). There was no cyst or other explanation for the extensive spinal cord widening.

Postoperatively, the patient had increased weakness and sensory changes in the left upper extremity. Follow-up myelography 3 months after surgery demonstrated that the cervical cord had returned to normal size. Six months after surgery, the patient had returned to work with only very mild left upper-extremity problems.
Spinal cord enlargement from intramedullary hemangioblastoma

Case 1

L: Metrizamide myelogram, anteroposterior view, showing the spinal cord from C-5 to T-4. Arrows mark the T-1 level. There is diffuse spinal cord enlargement over the entire area shown. R: Magnetic resonance image, sagittal view, of the spinal cord from the medulla to T-1. The upper arrows show the spinal cord margins at the C-2 level, and the lower arrows show the cord margins at C-5. There is diffuse enlargement over the cervical cord and medulla, with low-signal areas indicating edema.

Fig. 1. Case 1.

Case 2

L: Cervical myelogram, anteroposterior view. The black arrow indicates the C-2 level and the white arrow is at the T-1 level. There is widening over the entire cervical region of the cord. R: Intraoperative photograph of the C2-6 region showing a small tumor confined to the C-2 level (arrow). Diffuse widening of the spinal cord is demonstrated.

Fig. 3. Case 2.

Case 3

This 34-year-old man had a 3-year history of progressive weakness of the left side of the body and spasticity of both lower extremities. Spinal MR imaging demonstrated diffuse enlargement of the spinal cord from the medulla down to the midthoracic level, with a focal density change at C-4 (Fig. 4). A laminectomy was performed from C-3 to C-7 and an intramedullary hemangioblastoma, occupying two vertebral levels, was removed from the left dorsolateral portion of the cord. Postoperatively, the patient had decreased position sense in the left hand with improvement of his lower-extremity spasticity. Repeat MR imaging 4 months after surgery revealed diffuse atrophy of the cord with intramedullary cavitation.

Summary of Cases

There were eight patients in this series: seven males and one female. All patients presented with signs of progressive deterioration of spinal cord function, not unlike patients with other types of intramedullary spinal tumors. Seven patients experienced spastic paraparesis, and one patient came primarily for evaluation of sensory changes in the hands. Four patients had some degree of sensory abnormality or painful paresthesias involving the upper extremities, and three patients had early bowel or bladder dysfunction.

Table 1 outlines the features of spinal cord enlargement observed in these eight patients. The location of the tumors in the spinal axis was equally divided, with four in the cervical region and four in the thoracic spinal cord. Diagnosis was made in all cases with contrast myelography, and four patients also underwent preoperative MR imaging. Postoperative myelography.
TABLE 1

Characteristics of spinal cord enlargement in patients with hemangioblastomas

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Location of Tumor</th>
<th>Extent of Spinal Cord Widening</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>T-2</td>
<td>C1-T12</td>
</tr>
<tr>
<td>2</td>
<td>C-2</td>
<td>C1-T1</td>
</tr>
<tr>
<td>3</td>
<td>C4-5</td>
<td>medulla to T-6</td>
</tr>
<tr>
<td>4</td>
<td>T9-10</td>
<td>T9-10</td>
</tr>
<tr>
<td>5*</td>
<td>T-11</td>
<td>T1-T2</td>
</tr>
<tr>
<td>6</td>
<td>C2-3</td>
<td>C1-T2</td>
</tr>
<tr>
<td>7†</td>
<td>C-5</td>
<td>C1-T1</td>
</tr>
<tr>
<td>8</td>
<td>T-3</td>
<td>medulla to T-8</td>
</tr>
</tbody>
</table>

* Case 5 had multiple tumors, see text.
† An intramedullary cyst was found in Case 7.

R. A. Solomon and B. M. Stein

FIG. 4. Case 3. Magnetic resonance image, sagittal view, of the cervical region. The upper arrow indicates C-1 and the lower arrow points to a hemangioblastoma at C-4. There is extensive enlargement of the spinal cord, with edematous portions extending into the medulla.

was performed in two patients: the cord had returned to its normal size in one; the other patient demonstrated diffuse cord atrophy, whereas the preoperative study had shown enlargement. Three other patients underwent postoperative MR imaging, which showed in all cases that the cord had returned at least to its normal size or had become atrophic.

Only one patient in the series had definite evidence of multiple spinal hemangioblastomas. His symptomatic lesion was removed, and the other tumor nodules will be treated as they become symptomatic. Another patient had an associated cerebellar hemangioblastoma. Sagittal MR imaging revealed a large intramedullary cyst associated with the hemangioblastoma in one patient; the cyst extended rostrally from the tumor for three segments, yet the patient had cord widening for three to four segments caudal to the tumor.

All of the patients in this series underwent gross total removal of the spinal hemangioblastoma. In the immediate postoperative period, one patient had clear improvement of his preoperative neurological deficits, whereas four patients showed no change in their neurological condition, and three patients demonstrated an increase of their sensory or motor abnormalities. All of the patients have shown progressive neurological improvement following surgery over the follow-up period (ranging from 3 months to 6 years). All are ambulatory without aids, and six patients have returned to gainful employment. Overall, these results are better than the outcome statistics for the 60 patients with intramedullary spinal cord tumors.8,9

Discussion

Spinal cord enlargement beyond the focal intramedullary hemangioblastoma appears to be unique to this type of tumor. Several theories may be invoked to explain this phenomenon. The most likely explanation relates to the fact that these tumors are highly vascular, with arteriovenous shunts within the neoplasm resulting in prominent distended veins. These arterialized veins extend over the dorsal surface of the spinal cord for a considerable distance from the tumor, and more than likely induce marked venous congestion in the normal cord substance. Such congestion could lead to diffuse edema of the cord, and explain the grayish discoloration that is observed at operation.

This explanation appears credible, yet there is one very perplexing contradiction. We have not observed diffuse cord widening in a series of 10 intramedullary spinal arteriovenous malformations (AVM's) treated by us, even though these malformations produce a similar pattern of arterialized veins and venous congestion.8 In other series of spinal AVM's, diffuse cord enlargement has not been reported.9,13 This fact would indicate that either the flow characteristics of the arteriovenous shunting in hemangioblastomas are different from those of spinal AVM's or that the diffuse spinal cord enlargement is not related to the abnormal circulatory pattern.

The second plausible explanation for diffuse enlargement would involve the production by the neoplasm of an edema-promoting factor. Such a factor has been suspected to be associated with other types of tumors,7,9 and it is possible that hemangioblastomas secrete agents that induce increased capillary permeability. This possibility could be explored by careful neuropharmacological evaluation of cerebrospinal fluid and tumor cyst fluid from patients with hemangioblastomas.

It is significant that, in all five patients who underwent postoperative neuroradiological evaluation, either the spinal cord had returned to its normal size or the
Spinal cord enlargement from intramedullary hemangioblastoma
cord had become atrophic. Therefore, in no case could the diffuse enlargement be ascribed to the presence of unrecognized multiple tumors or large intramedullary cysts. In at least one patient (Case 2), the postoperative study demonstrated multiple cavitory lesions in the cord that had not been present preoperatively. This is of interest, because other authors have suggested that venous congestion associated with the Arnold-Chiari malformation and cervical spondylosis may be responsible for syringomyelia and neuronal dysfunction in distant parts of the spinal cord.\(^2\,\,12\) The incidence of hemangioblastomas in our series of intramedullary spinal cord tumors was 13%, much higher than the 1% to 3% noted in previous series.\(^4\,\,6\,\,14\) This high incidence may be fortuitous or a reflection of the nature of our university referral practice. However, it is possible that hemangioblastomas may have been overlooked in the past. Diffuse cord enlargement seen on conventional myelography may have been ascribed to infiltrating gliomas or syringomyelia. Spinal angiography is not routinely used for intramedullary processes and, even when it is performed, unless all the radicular vessels are injected, a hemangioblastoma could be missed. Furthermore, without proper preoperative guidelines, surgical exploration of the cord may not have included the region with the tumor. In our first case, we almost missed the tumor at the rostral end of the laminectomy which was centered over the cord enlargement (Case 2).

Modern imaging techniques such as computerized tomography and MR imaging are more precise than myelography alone in localizing a focal tumor nodule. The cases presented in this series were all diagnosed within the last 6 years, and six of the cases were seen in the last 2 years. Other recent series of spinal cord tumors will have to be analyzed to determine if the relative incidence of spinal hemangioblastoma compared to other types of tumors is higher than previously documented.

The surgical techniques for removal of spinal hemangioblastomas are similar to those described for treatment of other intramedullary spinal cord tumors.\(^10\,\,11\) The important difference is that hemangioblastomas must be removed as a single mass. Like AVM's, these tumors contain high-flow arteriovenous shunts, and torrential hemorrhage may result if the tumor is transected before it is disconnected from its blood supply. Although in several of the patients in this series the laminectomy was performed over multiple levels to encompass the areas of cord widening, we have learned that extensive laminectomy is unnecessary when the diagnosis of a solitary hemangioblastoma can be confirmed preoperatively. Laminctomy can be limited to the number of levels required to remove the solitary nodule of tumor. The diffuse cord widening will resolve after the responsible neoplasm has been eliminated.

References

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