Lipomas of the spinal cord and cauda equina

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This report presents 18 cases of lipoma of the spinal cord or cauda equina, four of which were unassociated with congenital anomalies of the spine and 14 with such anomalies. Review of the literature shows that unassociated cases are rare, with only 100 cases reported, while those associated with congenital anomalies are more common. The patients ranged in age from 2 months to 5 years. Fourteen patients had external evidence of an abnormality in the form of a mass or dimple over the vertebral column. Plain x-ray films showed evidence of an intraspinal lesion in all cases except one. The treatment was surgical in all cases, with nine good results, eight satisfactory, and one unsatisfactory. The exact pathogenesis of spinal cord lipomas is unclear, although the high incidence of associated congenital defects, age of onset, and location of the tumors suggest a developmental origin. The tumors occupy a juxta medullary-subpial location and are composed of firm, lobulated adult fat. Early surgery consisting of generous laminectomy, incision of the pia, and subtotal removal of the tumor is felt to be the treatment of choice.

Key Words: spinal cord tumor - lipoma - spina bifida - cauda equina tumor - conus tumor - congenital defect

True lipomas, unassociated with spina bifida, comprise about 1% of all spinal cord tumors. Hamby found the incidence to be somewhat higher in children; of 214 intraspinal tumors in patients under the age of 14 years, 10 were lipomas, an incidence of 4.7%. The majority of reports have concentrated on purely intradural forms. Stookey reviewed the literature up to 1927 and added one case located in the cervicothoracic area. He excluded those having an associated spina bifida. Ehni and Love in 1945 reviewed the literature and found only 26 cases of intradural lipomas and added four of their own, excluding those associated with spina bifida. In addition, they reviewed extradural lipomas, 17 from the literature and three of their own. Caram in 1957 reviewed 49 cases of intradural lipoma and added two. The total number in the literature was brought to 100 by Giuffré in 1966 who reviewed 99 cases and added one. These authors all excluded any case in which there was a dural, bony, or muscular defect connecting the subdural space with the subcutaneous adipose tissue. On the other hand, Lassman and James presented 26 patients with subcutaneous lipomas over the lumbosacral spine. All had spina bifida and widening of lumbosacral spinal canal on plain x-ray. In 18 of these the lipoma was continuous with an intradural lipoma. Swanson and Barnett presented nine similar cases in children, all having midline superfi-
intraspinal lipomas which were continuous with the intraspinal contents through a congenital anomaly of the spine.

Case Material

To facilitate systematic consideration of the problem, this paper presents 18 cases of intraspinal lipoma that include examples of both types, with and without assorted congenital anomalies of the spine. The groups are divided as follows:

Group A. These four patients had a spinal lipoma but no associated congenital anomaly. In three, the lipoma was exclusively intradural and in the other it was entirely extradural but totally intraspinal.

Group B. These 14 patients had a spinal lipoma plus an associated congenital anomaly of the spine or its contents. This group is subdivided according to the type of anomaly:

Type B1: Lipoma. These 10 cases had a subcutaneous lipoma which extended intradurally through a spina bifida occulta.

Type B2: Lipomeningocele. This single case had a subcutaneous lipoma that was continuous with the cauda equina. The meningeal sac was dilated and extended through a spina bifida, but the neural elements were not out of the spinal canal.

Type B3: Lipomyelomeningocele. These three cases were similar to Type B2 except that the neural elements herniated into the meningocele sac. Although many myelomeningoceles have fatty tissue in the sac, in these three cases, the lipoma was clearly the major pathological problem.

Clinical Features

The patients ranged in age from 2 mos to 55 years. There were 10 males and 8 females. Clinical evidence of a lesion was a mass in six patients, paralysis in eight, bowel or bladder dysfunction in two, and pain in one. Four patients were asymptomatic neurologically, and sought attention because of a lumbosacral mass. The signs and symptoms of the 14 symptomatic patients were suggestive of a tumor of the spinal cord or cauda equina. These symptoms included gait abnormalities, weakness, abnormalities in leg tone, and abnormal bowel and bladder function. Only one patient complained of pain as a major symptom. This was a 22-year-old woman with an intradural extradural lipoma involving the cauda equina and conus, who had bilateral radicular leg pain.

Fourteen patients had external evidence of a lesion in the form of a mass or dimple. Those who did not were the three patients with completely intradural tumors and the one with a completely extradural tumor.

Examination of these patients revealed neurological signs typical of any mass lesion involving the spinal axis. Motor symptoms ranged from mild weakness in the legs to marked paraparesis or quadriplegia. The reflex changes and sensory disturbances predominantly involved the lower dermatomes; common findings were absent ankle jerks and/or saddle anesthesia. The high percentage of patients with bowel and bladder disturbance reflects the common site of these lesions in the conus and cauda equina (Table 1).

Radiological evidence of a lesion was present on plain x-ray films in all cases except the one patient with a completely extradural lipoma in the thoracic region. The three patients who had completely intradural tumors demonstrated widening of the bone canal and pedicular erosion in the area of the tumor. The patients with extradural extensions of intradural lesions had an associated spina bifida in addition to the widening and erosion (Figs. 1 and 2). In eight patients myelography was performed. All demonstrated some abnormality, usually widening of the sac, a mass lesion, or low termination of the spinal cord (Fig. 3).

Therapy

The treatment was surgical in all cases, for there really is no other form of therapy. The only points of contention are the timing of surgery in asymptomatic patients and the

TABLE 1

<table>
<thead>
<tr>
<th>Symptom</th>
<th>No. of Cases</th>
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<tr>
<td>weakness; gait abnormal</td>
<td>11</td>
</tr>
<tr>
<td>reflex changes</td>
<td>13</td>
</tr>
<tr>
<td>sensory deficit</td>
<td>10</td>
</tr>
<tr>
<td>bowel and bladder deficit</td>
<td>9</td>
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</tbody>
</table>

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aggressiveness governing removal. At the time of surgery the tumor involved the conus and/or cauda equina in 14 patients. In eight of these it was impossible to tell whether the major attachment was primarily to one or the other. In three patients it was apparent that the attachment was primarily to the conus and in three it was primarily to the cauda equina. In the four remaining cases, the attachment was to the filum terminale in one, in the thoracic cord in two (one intra- and one extradural), and in one the lipoma was intradural and involved the entire length of the spinal cord from C-2 to the conus.

Pathology

The pathology of these tumors in all cases was adult fat. In three patients there was a persistent dermal sinus extending from a skin dimple to the lipoma. There was no evidence of malignancy or primitive cell types in any of the tumors.

Results of Treatment

There were 21 operations on 18 patients. The follow-up period ranged from 3 mos to 20 years. The results of treatment were classified as “good,” “satisfactory,” or “unsatisfactory.” “Good” included those patients who were neurologically asymptomatic preoperatively and remained so postoperatively, and those who had a discernible neurological deficit preoperatively and had a significant improvement postoperatively. “Satisfactory” included those who had no further progression of neurological deficit. “Unsatisfactory” included those whose condition deteriorated after surgery. Under this classification, there were nine “good” results, eight “satisfactory,” and one “unsatisfactory.” Table 2 shows the results in each group.

The site of the lesion could not be correlated with ultimate prognosis. The one patient with an unsatisfactory result had tumor attached to both the cauda equina and the conus (Table 3).

There was no correlation between clinical symptoms or significant findings and the site of tumor attachment (conus, cauda, or both). This agrees with the finding of Lassman and James,19 who also found no correlation between the symptoms and signs, and findings at surgery, with the possible exception of a high association of bladder involvement with lesions of the filum terminale in...
Discussion

The exact pathogenesis of spinal cord lipomas is unclear although the high incidence of associated congenital defects, the age of onset, and location of the tumors suggest a developmental origin. This is especially evident in those with a midline congenital anomaly of the bone canal, which was the situation in 14 of the 17 patients in this report. These tumors are not infrequently associated with other congenital anomalies in the nervous system, either in the same area (dermal sinus, cholesteatoma, abnormal filum, diplomyelia, diastematomyelia) or more distant (hydrocephalus).9,11 Anomalies involving other organ systems have been reported by various authors to occur in 30% to 50% of the cases.8,12 These anomalies include pilonidal sinuses, cataracts, club feet, pseudohermaphroditism, absent kidney, and Klippel-Feil syndrome. One of the patients in this series sought medical attention because of an imperforate anus and rectovaginal fistula; one kidney was also absent. A teenage boy in our series with a completely intradural lipoma of the thoracic cord died 5 years after surgery for cerebral hemorrhage. At autopsy he was found to have multiple intracranial lipomas, an aneurysm of the basilar artery, and a hemispheral arteriovenous malformation.

In 1863 Virchow9 described fat cells in the leptomeninges. However, it is known that adult fat cells do not reproduce themselves, so they are an unlikely source of these tumors. Ehni and Love9 in their excellent article on this subject in 1945 discussed the theories of histogenesis that had been proposed to that time and offered another explanation. They believed that these lipomas are of mesenchymal rather than neural origin and arise because of local failure of normal control over the formation of fat from normally present multipotential peri-capillary mesenchymal cells. Another theory that is attractive is that of Bostroew9 who stated that the lesion was due to an inclusion of misplaced elements within the closing lips of the neural groove, and he grouped dermoids, epidermoids, and lipomas in a category of "inclusion tumors" stemming from embryonic error before the fifth somite stage.

Whatever the exact histogenesis may be, the ultimate outcome is a tumor in a juxta-medullary/subpial location.6,8,12,13 The tumor is composed of adult fat that is firm, lobulated, and surrounded by a layer of connec-
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TABLE 2

<table>
<thead>
<tr>
<th>Location</th>
<th>Group</th>
<th>Good</th>
<th>Satis.</th>
<th>Unsatis.</th>
<th>Total</th>
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<tr>
<td>intradural</td>
<td>A</td>
<td>1</td>
<td>2</td>
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<tr>
<td>extradural</td>
<td>A</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>intra and extradural</td>
<td>B1</td>
<td>6</td>
<td>3</td>
<td>1</td>
<td>10</td>
</tr>
<tr>
<td>lipomeningocele</td>
<td>B2</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>lipomyelomeningocele</td>
<td>B3</td>
<td>0</td>
<td>3</td>
<td>0</td>
<td>3</td>
</tr>
<tr>
<td>total</td>
<td></td>
<td>9</td>
<td>8</td>
<td>1</td>
<td>18</td>
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</tbody>
</table>

tive tissue derived from the pia with septae of fibrous tissue and fat cells extending into the cord. At the margins this connective tissue becomes continuous with the pia above and below. The situation is the same when the tumor involves nerve roots, the fibrous capsule in this instance being derived from the endoneurium, which is the continuation of the pia. The lipomas that are both intra- and extradural have the same intimate connection with neural tissue as do the completely intradural tumors. Thus, although the bulk of these tumors with extradural connections is easily removed, the surgical principles involved in removing the part that is involved with neural tissue are the same as with the completely intradural tumors. Although no lipoma has been found that was completely surrounded by neural tissue, these tumors are firmly adherent and intimately connected to neural tissue by fibrous septae. It is important to be aware of this anatomical fact when removing these tumors.14

As briefly mentioned earlier, the argument regarding treatment of these tumors is not whether surgery should be done, but when and how much. Although various authors mention a cleavage plane that sometimes exists between the tumor and the spinal cord or nerve roots, in the majority of cases this does not exist.3,4,15 There are only 10 cases of total removal of intradural lipomas reported in the literature. One is a case by Elsberg,7 who totally removed a tumor of the cervicothoracic cord. The patient was left with a transverse myelopathy at that level. Elsberg thereafter emphasized a conservative approach to these lesions. Crosby, et al.,4 presented a case of thoracic lipoma that was totally removed in two stages. Caram, et al.,3 reported one case in which they were able to accomplish total removal. Even these authors who were able to obtain total removal in a single case emphasize that the best mode of treatment is a generous laminectomy, incision of the pia, and subtotal or partial removal, if there is no readily discernible cleavage plane. The long-term results are independent of the extent of the tumor removal.3 In Caram's review of the literature of intradural lipomas, 10 total removals resulted in nine patients improved, and 14 partial removals in 13 patients improved. Of 12 cases treated by laminectomy and biopsy, only six improved. Dubowitz, et al.,5 advocated partial resection and freeing up of fibrous bands that restrict the movement of the cord, and obtained satisfactory results in 10 of 12 patients by this treatment. In Swanson and Barnett's15 series of children, all with bladder dyfunction except one showed improvement following incomplete removal.

Our experience supports these findings. In only one intradural tumor was a total removal accomplished. This was in a 4-year-old boy whose tumor was attached only to the filum. In the rest of the intradural tumors the tumor was firmly attached to neural structures. These attachments were always firm, the tumor blending imperceptibly into

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TABLE 3

<table>
<thead>
<tr>
<th>Site</th>
<th>Total</th>
<th>Good</th>
<th>Satis.</th>
<th>Unsatis.</th>
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<tbody>
<tr>
<td>cauda</td>
<td>3</td>
<td>2</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>conus</td>
<td>3</td>
<td>2</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>both</td>
<td>8</td>
<td>2</td>
<td>5</td>
<td>1</td>
</tr>
<tr>
<td>filum</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>thoracic cord</td>
<td>2</td>
<td>1</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>entire cord</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
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<td>18</td>
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<td>8</td>
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neural tissue, and no cleavage plane could be delineated. Generous laminectomy and removal of all tumor possible without undue manipulation of the cord was accomplished. Where the tumor was in intimate contact with spinal cord or nerve roots, it was removed by sharp dissection, leaving a small portion of the tumor.

The timing of surgery is important, especially in the asymptomatic patients. Arguing for early treatment, Dubowitz, et al., pointed out that most of these tumors will cause neurological symptoms sooner or later. They cite several instances in the literature that tend to support this argument. For example, Bassett reported the cases of two infants each 2 months old and asymptomatic. One had surgery immediately and remained free of symptoms. The other had surgery deferred, and at age 16 months showed gross deficits (paraparesis and loss of sphincter control). Surgery then produced no improvement. Two of the nine children in Swanson and Bassett’s series were seen before 6 months of age without symptoms. They developed symptoms at 4 and 10 years. Lassman and James had four patients with lumbosacral lipomas who did not develop symptoms until their teens. Brickner reported two adults who first developed bladder symptoms at age 18. We have three patients in our group who developed symptoms after age 15. Although some improvement can be obtained in these patients, it is generally concluded that results are not as good once neurological involvement has occurred. Because there is no way to tell with certainty which asymptomatic patients will become symptomatic and which will not, we believe that the best treatment is to assume that all will sooner or later develop symptoms and thus advocate early surgery. For technical reasons we would wait, if possible, until an asymptomatic child is 1 year old.

Summary

Eighteen cases of lipoma of the spinal axis have been presented; 14 were combined intradural and extradural tumors, three completely intradural, and one was extradural. The histogenesis, pathology, and anatomy have been discussed, and early removal advocated.

References

2. Brickner WM: Spina bifida occulta: (1) with external signs, with symptoms. (2) with external signs, without symptoms. (3) without external signs, with symptoms. (4) without external signs, without symptoms. Amer J Med Sci 155:473–502, 1918

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