Subcutaneous sacrococcygeal ependymoma with inguinal lymph node metastasis

Case report

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A patient with a subcutaneous sacrococcygeal ependymoma and metastasis to the inguinal lymph nodes is presented and his treatment is described. Previous reports on sacrococcygeal ependymoma are reviewed.

KEY WORDS • ependymoma • spinal neoplasm • sacrococcygeal neoplasm • lymph node • metastasis

THE ependyma is a single layer of columnar cells lining the ventricular system of the central nervous system (CNS). Ependymomas are tumors originating from the ependymal cells and are mostly confined to the CNS. In rare cases a tumor occurs with extraneural primary origin. Ependymomas outside the CNS originate from a persistence of the neurenteric canal (especially in a presacral location), from heterotopic ependymal cells (especially nasal) or from the coccygeal medullary vestige and ependyma-lined cavity in the caudal part of the neural tube beneath the skin of the postnatal pit. The myxopapillary ependymoma is the most common type of ependymoma in the sacrococcygeal region.

Tumors from neural origins represent 2% to 10% of all retrorectal or presacral tumors. One in every 40,000 tumors arising in this area is an ependymoma. Subcutaneous myxopapillary ependymomas are also rare and, until now, only 49 cases have been reported (Table 1). The female: male ratio in affected patients is nearly equal. The age at which patients present with symptoms varies from 10 months to 47 years. Ependymomas arising in the CNS, cauda equina, and presacral region are associated with neural dysfunction, whereas these symptoms are absent in purely subcutaneous myxopapillary ependymomas. A slowly progressing, sometimes painful mass in the intergluteal fold is often misdiagnosed preoperatively as a pilonidal cyst or teratoma.

Case Report

This 35-year-old man originally presented at another clinic in December, 1965, at the age of 15 years because of a tumor measuring 3.5 × 1.5 × 1.5 cm located above the sacrococcygeal region which had been slowly growing during the previous year. Surgical excision was performed with the clinical diagnosis of pilonidal cyst. At histological examination, however, it appeared to be a myxopapillary ependymoma. The resection margins were free of tumor cells. In December, 1974, a recurrent tumor measuring 6.5 × 3 × 3.5 cm was completely excised, and in February, 1984, a second recurrent tumor, 7 × 6 × 4.5 cm in size, was excised. This time the margins were not free of tumor, but no additional treatment was given. The pathologist described for the first time a few mitoses and infiltration into the muscle and connective tissues. In July, 1985, the patient came to the St. Radboud Hospital for evaluation of a tumor, again in the intergluteal fold, which had been enlarging slowly since January, 1985. He did not complain of neurological dysfunction.

Examination. A 10 × 5 × 4.5-cm tumor was found above the sacrococcygeal region; it was movable from the surrounding area and the sacrum. A mobile solid lymph node with a diameter of 2 cm was palpable in the right groin. Neurological examination showed no abnormalities with the exception of a congenital myotonia (Thomsen’s disease). No signs of primary or sec-
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TABLE 1
Summary of reported cases of sacrococcygeal located ependymomas*

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs)† &amp; Sex</th>
<th>Preop Duration of Symptoms</th>
<th>Preop Diagnosis</th>
<th>Therapy‡</th>
<th>Metastasis</th>
<th>Local Recurrence</th>
<th>Follow-Up Period</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mallory, 1902</td>
<td>44, F</td>
<td>none</td>
<td>surgery</td>
<td>unknown</td>
<td>unknown</td>
<td>unknown</td>
<td>5 yrs</td>
</tr>
<tr>
<td>Cooper, et al., 1951</td>
<td>30, M</td>
<td>6 yrs</td>
<td>surgery</td>
<td>unknown</td>
<td>unknown</td>
<td>unknown</td>
<td>2 yrs</td>
</tr>
<tr>
<td>Anderson, 1966</td>
<td>8, F</td>
<td>unknown</td>
<td>surgery</td>
<td>no</td>
<td>no</td>
<td>persistent</td>
<td>14 yrs</td>
</tr>
<tr>
<td>Anderson, 1966</td>
<td>16, F</td>
<td>6 yrs</td>
<td>pilonidal cyst</td>
<td>surgery</td>
<td>no</td>
<td>persistent</td>
<td>14 yrs</td>
</tr>
<tr>
<td>Anderson, 1966</td>
<td>32, F</td>
<td>&lt; 1 yr</td>
<td>pilonidal cyst</td>
<td>surgery</td>
<td>no</td>
<td>persistent</td>
<td>14 yrs</td>
</tr>
<tr>
<td>Anderson, 1966</td>
<td>3, M</td>
<td>pilonidal cyst</td>
<td>surgery &amp; RT</td>
<td>no</td>
<td>persistent</td>
<td>persistent</td>
<td>14 yrs</td>
</tr>
<tr>
<td>Anderson, 1966</td>
<td>34, M</td>
<td>pilonidal cyst</td>
<td>surgery</td>
<td>no</td>
<td>persistent</td>
<td>persistent</td>
<td>14 yrs</td>
</tr>
<tr>
<td>Hendren &amp; Hardin, 1963; &amp; Morantz, et al., 1979</td>
<td>16, M</td>
<td>9 yrs</td>
<td>none</td>
<td>surgery (6), RT, &amp; chemotherapy for mets</td>
<td>yes</td>
<td>yes</td>
<td>6 yrs</td>
</tr>
<tr>
<td>Tennelkoon, 1968</td>
<td>34, F</td>
<td>5 yrs</td>
<td>chordoma</td>
<td>surgery</td>
<td>unknown</td>
<td>unknown</td>
<td>2 yrs</td>
</tr>
<tr>
<td>Prabhakar, et al., 1969</td>
<td>15, M</td>
<td>none</td>
<td>surgery (2)</td>
<td>no</td>
<td>yes</td>
<td>2 yrs</td>
<td>14 yrs</td>
</tr>
<tr>
<td>Scharrer &amp; Heiming, 1974</td>
<td>7, F</td>
<td>cyst, teratoma</td>
<td>surgery</td>
<td>unknown</td>
<td>unknown</td>
<td>unknown</td>
<td>2 yrs</td>
</tr>
<tr>
<td>Woyke &amp; Czerniak, 1978</td>
<td>20, M</td>
<td>pilonidal cyst</td>
<td>surgery (3)</td>
<td>yes</td>
<td>yes</td>
<td>2 yrs</td>
<td>14 yrs</td>
</tr>
<tr>
<td>Vagaiwala, et al., 1979</td>
<td>36, M</td>
<td>20 yrs</td>
<td>surgery (6), chemotherapy for mets</td>
<td>yes</td>
<td>yes</td>
<td>4 yrs</td>
<td></td>
</tr>
<tr>
<td>Bale, 1980</td>
<td>4, F</td>
<td>1 mo</td>
<td>pilonidal cyst</td>
<td>surgery</td>
<td>no</td>
<td>no</td>
<td>14 yrs</td>
</tr>
<tr>
<td>Timmerman &amp; Bubrick, 1982</td>
<td>29, F</td>
<td>16 yrs</td>
<td>teratoma</td>
<td>surgery</td>
<td>no</td>
<td>no</td>
<td>14 yrs</td>
</tr>
<tr>
<td>Dormal &amp; Baye, 1984</td>
<td>22, F</td>
<td>pilonidal cyst</td>
<td>surgery (21/pt)</td>
<td>no</td>
<td>no</td>
<td>no</td>
<td>1 yr</td>
</tr>
<tr>
<td>Helwig &amp; Stern, 1984</td>
<td>17 F &amp; 15 M</td>
<td>pilonidal cyst</td>
<td>chemotherapy for mets in 4 pts</td>
<td>yes (4)</td>
<td>yes (4)</td>
<td>&gt; 5 yrs, 5 yrs, &gt; 14 yrs, 15 yrs</td>
<td></td>
</tr>
<tr>
<td>Kramer, et al., 1988</td>
<td>10, M</td>
<td>1 yr</td>
<td>pilonidal cyst</td>
<td>surgery (4), RT for mets</td>
<td>yes</td>
<td>yes</td>
<td>20 yrs</td>
</tr>
</tbody>
</table>

* RT = radiotherapy; mets = metastasis.
† Age at first operation.
‡ Numbers in parentheses refer to number of operations.

Secondary tumor sites could be detected in the CNS on computerized tomography (CT) of the brain, myelography, or cerebrospinal fluid (CSF) analysis. Blood chemistry findings and chest x-ray films were normal. A CT scan of the abdomen and pelvis showed only the enlarged node in the right groin. Cytological examination of aspirate obtained from the inguinal lymph node strongly suggested metastasis from an ependymoma.

Operation. The sacrococcygeal mass was found to be surrounded by fibrotic tissue and was connected to the sacrum by a stalk. It was closely related to the anal sphincter. Excision was possible together with amputation of the os coccygis and the lower part of the sacrum. The inguinal node was also excised.

Postoperative Course. Because of the previous tumor recurrences, the high histological grade of the tumor, the infiltration of the skin, and the inguinal metastasis, the patient was treated with irradiation over the sacral region and the inguinal and iliac lymph nodes. He received 18 MV photons via two opposing fields, with a total dose of 50.4 Gy in fractions of 1.80 Gy over 6½ weeks.

Pathological Examination. Gross examination of the excised specimen showed a grayish-white tumor measuring $7 \times 6 \times 4.5$ cm under the overlying skin. It was well encapsulated and solid. On microscopic examination (Fig. 1 left), solid clusters of medium-sized cuboid cells were seen partially lining open spaces which were filled with mucinous material. Infrequent papillary growth was observed. The cytoplasm stained acidophilic. Nuclei were generally uniformly round or ovoid, with occasional anisokaryosis and hyperchromatism. In these regions few mitoses were seen. Rosettes were sparse. Microscopically, there was infiltration of the skin, but no invasion of the bone. The margins were free of tumor cells. The lymph node showed the same histological features (Fig. 1 right), but with signs of dedifferentiation and higher numbers of mitotic figures.

Discussion

Physical examination for a suspected subcutaneous myxopapillary ependymoma is usually confined to the lesion itself. When the disease involves the anal region a rectal as well as a pelvic examination is obligatory.
Special attention should be paid to the inguinal lymph nodes. A normal CSF protein value and a normal myelogram exclude conus medullaris and cauda equina locations. Magnetic resonance (MR) imaging may eventually supersede myelography as a diagnostic tool but has not yet been adequately evaluated. Compared with contrast-enhanced radiography, MR imaging is more convenient for the patient and gives information about the entire CNS. When metastatic disease is suspected, analysis should include full blood chemistry studies, chest x-ray films, CT scans of the entire abdomen and pelvis, and if possible intravenous urograms and total skeletal scintigraphy. To rule out metastasis in normal-sized lymph nodes on CT, bipedal lymphangiography is advised.

In nine patients with metastatic subcutaneous myxopapillary ependymomas described in the literature (Fig. 2), most metastases were found in the lungs (six cases), followed by the lymph nodes (four cases), the pleura (three cases), bone (one case), and subcutaneous areas (one case). No metastases have been reported in the liver. There is probably a correlation between local recurrence and distant metastases.

The preferred treatment for subcutaneous myxopapillary ependymoma is surgery. Most authors advise excision of the primary tumor, although others recommend a combination of coccygectomy and lymphadenectomy. Hendren and Hardin identified metastases in the inguinal lymph nodes of their patient and removed the positive nodes only; however, the patient developed a recurrence in the inguinal region on the same side. Bilateral radical groin dissection was carried out, but no pathological nodes were found on the contralateral side. Wolff, et al., also performed bilateral radical lymph node dissection in a patient with metastasis to one inguinal lymph node. No other site was found.

The role of radiotherapy in the treatment of subcutaneous myxopapillary ependymoma is controversial. Ependymomas are radiosensitive and radiotherapy prolongs survival in patients with intracranial involvement. Prolongation of life by radiotherapy has not been observed in cases of subcutaneous myxopapillary ependymoma. Several factors may be responsible for this failure, including a large tumor load and a low dosage level of radiotherapy. We believe the best treatment for these tumors is wide local excision. Coccygectomy is indicated when the tumor is attached to the bone. Postoperative radiotherapy at curative levels is necessary in cases where the tumor is incompletely resected or after removal of a local recurrence. The
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minimum tumor dose needed for a good result is 45.0 to 60.0 Gy given over 5 to 8 weeks. We would suggest a simple ipsilateral superficial inguinal lymphadenectomy when inguinal lymph nodes are involved and no pathological pelvic nodes are diagnosed. The advantage of prophylactic dissection of the pelvic lymph nodes is not clear. We would prefer to administer irradiation rather than radical bilateral lymphadenectomy, which is likely to have a higher morbidity rate. Radiotherapy is palliative in cases where the tumor is not resectable. Until now, chemotherapy has not proved to be effective in the treatment of subcutaneous myxopapillary ependymoma.9,25

References

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