Surgical management of giant sacral schwannomas

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Thirteen cases of giant sacral schwannomas with erosion of the anterior aspect of the sacrum and associated intrapelvic extension are reviewed. These tumors showed no sex predilection; the patients' mean age was 38.6 years at the time of diagnosis, and their symptoms predated the diagnosis by an average of 5.2 years. The most common symptoms were low-back pain and lower-extremity dysesthesiae. Plain roentgenograms, myelography, and computerized tomography constituted essential and complementary studies in the preoperative assessment. Choice of surgical approach (anterior transabdominal vs. posterior transsacral) was dependent upon the amount of sacral destruction, intrapelvic extension, and sacroiliac joint involvement. Microscopic examination revealed classic features of benign schwannoma in all but three cases, which were classified as cellular schwannomas. Patients who presented with pain and dysesthesiae reported immediate and complete relief of symptoms following surgery. In addition, all 13 patients were ambulatory and able to resume their routine daily activities postoperatively. At the last reported follow-up examination, which ranged from 5 months to 33 years and 3 months (mean 9 years) after surgery, two patients had died of unrelated causes, two reported return of preoperative symptoms, and the remainder were asymptomatic. This experience suggests that these histologically benign but neurologically devastating tumors should be aggressively resected with the intent of complete extirpation, and that this goal may be accomplished with minimal risk and an excellent prognosis.

Summary of Cases

Thirteen patients with giant sacral schwannoma that had eroded through the anterior sacrum and extended into the retrorectal space were surgically treated at the Mayo Clinic between May, 1952, and January, 1985. Each case was analyzed for clinical presentation, diagnostic evaluation, surgical intervention, and quality of survival.

Clinical Presentation

The clinical data are summarized in Table 1. The group consisted of seven male and six female patients. Age at diagnosis ranged from 16 to 57 years, with an average of 38.6 years. Pain was a presenting symptom for 12 patients. Nine individuals reported pain localized to the lumbosacral region, and five of these patients complained that the discomfort was also referable to the distribution of one or more lumbar or sacral nerve roots. Three patients presented strictly with lower-extremity radicular pain. Dysesthesiae of the lower ex-
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TABLE 1  
Clinical summary of 13 cases of giant sacral schwannomas*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Presenting Symptoms &amp; Signs</th>
<th>Operative Approach</th>
<th>2nd Approach &amp; Elapsed Time</th>
<th>Follow-up Duration &amp; Condition</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>34, M</td>
<td>low-back pain, sciatica, diminished reflexes</td>
<td>posterior</td>
<td>—</td>
<td>5 mos: asymptomatic; residual tumor on CT</td>
</tr>
<tr>
<td>2</td>
<td>31 M</td>
<td>low-back pain, sciatica, diminished reflexes</td>
<td>posterior</td>
<td>—</td>
<td>1 yr 9 mos: return of low-back pain &amp; sciatica; residual tumor on CT</td>
</tr>
<tr>
<td>3</td>
<td>20, F</td>
<td>low-back pain, sciatica, diminished reflexes</td>
<td>anterior combined, 10 mos</td>
<td>2 yrs 7 mos: pain-free, voluntary bladder &amp; bowel control, flat-footed gait; no recurrence on CT</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>57, M</td>
<td>low-back pain, sciatica, S-1 hypesthesia, diminished reflexes, urinary hesitancy</td>
<td>anterior</td>
<td>—</td>
<td>10 mos: asymptomatic; residual tumor on CT</td>
</tr>
<tr>
<td>5</td>
<td>27, F</td>
<td>low-back pain, sciatica, diminished reflexes</td>
<td>posterior anterior, 7 mos</td>
<td>1 yr 7 mos: pain-free, wears rt foot-drop brace; no recurrence on CT</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>31, F</td>
<td>low-back pain, sciatica</td>
<td>anterior† anterior, 22 mos</td>
<td>7 yrs 1 mo: asymptomatic; no recurrence on CT</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>50, M</td>
<td>sciatica, diminished reflexes, S-1 hypesthesia, L-5 &amp; S-1 weakness (unilateral)</td>
<td>posterior anterior, 84 mos</td>
<td>1 yr 10 mos: asymptomatic; no recurrence on MRI</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>16, M</td>
<td>low-back pain, sciatica, L-5 weakness, diminished reflexes</td>
<td>posterior</td>
<td>—</td>
<td>16 yrs 1 mo: return of low-back pain &amp; sciatica</td>
</tr>
<tr>
<td>9</td>
<td>47, F</td>
<td>sciatica</td>
<td>posterior</td>
<td>—</td>
<td>17 yrs: asymptomatic</td>
</tr>
<tr>
<td>10</td>
<td>46, M</td>
<td>sciatica, constipation</td>
<td>posterior</td>
<td>—</td>
<td>10 yrs 10 mos: asymptomatic; died from unrelated cause</td>
</tr>
<tr>
<td>11</td>
<td>47, F</td>
<td>low-back pain, sciatica, S2-5 hypesthesia, diminished reflexes</td>
<td>posterior</td>
<td>—</td>
<td>3 yrs 7 mos: return of low-back pain; died from unrelated cause</td>
</tr>
<tr>
<td>12</td>
<td>47, M</td>
<td>asymptomatic</td>
<td>posterior</td>
<td>—</td>
<td>20 yrs 3 mos: asymptomatic</td>
</tr>
<tr>
<td>13</td>
<td>49, F</td>
<td>low-back pain</td>
<td>posterior</td>
<td>—</td>
<td>33 yrs 3 mos: asymptomatic</td>
</tr>
</tbody>
</table>

* Cases 7, 9, and 11 had hypercellular schwannomas (see text). CT = computerized tomography; MRI = magnetic resonance imaging.
† Initial operation was performed at another institution.

Tremors accompanied or developed shortly after the onset of pain in five cases. One patient had urinary hesitancy and retention, while another had difficulty with constipation. One patient did not present with neurological symptoms but was found to have a presacral mass during a routine examination. None of the patients complained of motor deficit as a component of their initial symptomatology. Duration of symptoms ranged from 2 months to 23 years, with an average of 5.2 years.

Upon initial neurological evaluation, three patients displayed diminished perianal or lower-extremity sensation, two had lower-extremity weakness, and eight patients had decreased deep-tendon reflexes as verifiable signs. Percussion tenderness was elicited from five patients over the lumbosacral spine, and three had positive straight-leg raising tests. A distinct retrorectal mass was palpated on digital examination in 11 of the 13 patients.

**Diagnostic Investigations**

Plain roentgenograms revealed irregular defects of the anterior aspect of the sacrum with sclerotic margins, indicative of sacral erosion, in 11 patients (Fig. 1). Plain films also demarcated the tumor mass as a homogeneous soft-tissue density in 11 of the 13 patients. Computed tomography (CT) scanning was carried out in seven patients, and demonstrated each tumor to be a destructive expansile lesion with a homogeneous presacral extension variably displacing the normal pelvic structures (Fig. 2). Myelography was performed on seven individuals, and showed intradural extension of the tumor in four (Fig. 3), epidural involvement in two, and a normal study in one. Three patients underwent barium enemas, resulting in two studies that were consistent with an extrinsic retrorectal mass; the remaining examination was unremarkable. Proctoscopy was performed in four patients and in each case confirmed the presence of a posteriorly situated extrarectal mass. One of the three patients who were assessed by intravenous pyelography showed displacement of the ureters and the bladder. Electromyographic examinations of five patients were consistent with irritation of multiple nerve roots in three and were considered normal in the remaining two. Cerebrospinal fluid protein levels were elevated in all six patients sampled. Values ranged from 49 to 795 mg%, with a mean of 321 mg%. Two patients underwent needle biopsy prior to surgery. The first patient’s biopsy was performed transrectally; the second patient had percutaneous needle biopsy at another institution prior to referral. Both tissue specimens obtained were diagnostic for schwannoma.
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FIG. 1. Preoperative plain roentgenograms, anteroposterior (left) and lateral (right) views, of a giant sacral schwannoma showing sacral destruction and a homogeneous presacral soft-tissue density.

Surgical Techniques

All schwannomas in this series were excised surgically. One of two distinct procedures was employed: an anterior midline abdominal approach or a posterior retrorectal transsacral method. A combination of the two techniques under one anesthetic was used in one patient. As originally described by MacCarty, et al., at our institution the combined services of a neurosurgeon, an orthopedist, and an abdominal surgeon are considered essential for successful resection of a mass involving the sacrum and the presacral space.

Anterior Approach. With the patient in a supine position, the abdomen is prepared and draped widely. A midline incision from the pubis to the epigastrium is made and the abdominal cavity entered. Following thorough exploration of the abdomen, the retrosigmoid mesentery is opened ventrally and the retrosigmoid colon is mobilized with blunt dissection, taking care to identify and protect the ureters and iliac vessels (Fig. 4a). The bowel is then retracted to the left. The tumor mass can often be identified posteriorly as it displaces the rectum, uterus, bladder, and ureters. Dissection is carried around the tumor in a circumferential manner to isolate it from the surrounding structures (Fig. 4b). This is accomplished by developing a plane between the highly vascularized loose areolar tissue, which surrounds the lesion, and the capsule of the tumor. The vascular supply originates from the medial and lateral sacral arteries and the hypogastric plexus (Fig. 4c). A very diffuse venous complex encompasses the mass and drains into the iliac veins. The extreme vascularity and the extensiveness of the venous channels pose a formidable obstacle to total resection since profuse bleeding may occur with minimal manipulation.

FIG. 2. Preoperative computerized tomography scans of four giant sacral schwannomas. a: Scan showing sacral destruction and a large multiloculated presacral tumor mass. The arrow points to the compressed contrast-filled bladder. b: An intrasacral tumor mass is revealed, with a presacral component and bilateral sacroiliac joint involvement. c: Scan showing sacral destruction with a modest presacral component and unilateral sacroiliac joint involvement. d: An intrasacral tumor mass is shown, with a presacral component and early unilateral sacroiliac joint involvement.
After isolation and control of the vascular supply has been achieved, the tumor capsule is incised. The schwannoma is gutted by piecemeal dissection as the capsule wall is mobilized from the sacrum. Multiloculated cysts exuding xanthochromic fluid will be encountered if tumor degeneration is present. If possible, the dissection is carried to the nerve root of origin and the mass is amputated. When the nerve of origin cannot be identified, then an attempt to excise all visible tumor intrasacrally should be made (Fig. 4d). Upon completion of the resection, the eroded sacral margins are curetted and hemostasis is achieved. Routine use of large drainage catheters placed in the presacral space is recommended. Finally, the retroperitoneum and abdomen are closed in anatomical layers.

**Posterior Approach.** The patient is placed in the flexed prone position (Fig. 5a) and the skin of the low back is prepared and draped from the upper lumbar region to the anus, excluding the anus from the sterile operative field. One lateral thigh should also be prepared in the event that a fascia lata graft is required for the operative field. One lateral thigh should also be prepared in the event that a fascia lata graft is required for the operative field. A midline incision from L-4 to the sacrococcygeal junction is made through the skin and subcutaneous tissues. Once the spinous processes have been exposed and the lumbodorsal fascia incised, the tumor capsule is incised. The dural closure is achieved, the tumor capsule is incised, and an intracapsular enucleation begun (Fig. 5d). Piecemeal removal is mandatory and sacrifice of many or all sacral roots may be necessary if recurrence is to be minimized (Fig. 5e).

Bilateral preservation of the first three sacral roots and the pudendal nerves is ideal. Unilateral third sacral root sectioning can be performed without significant neurological deficit; however, further nerve root injury will inevitably result in bladder and bowel dysfunction.

Cephalad extension above the sacrum usually occurs intradurally, requiring a dural opening and later reconstruction of the cul-de-sac when loss of dura or the quality of remaining dura precludes primary closure. Reconstruction with a fascia lata graft to accommodate viable lumbosacral nerve roots may be an artistic endeavor (Fig. 5f).

The caudal and anterior dimensions of the schwannoma will determine the need for a more aggressive sacral resection. The distal presacral space is approached by releasing the anococcygeal muscles from the coccyx, which is then removed. The retrorectal space is now accessible, allowing detachment of the piriformis and coccygeus muscles and separation of the rectum from the schwannoma. As outlined by MacCarty, et al., the sacral resection can be carried cephalad by dividing the sacrum at the selected foraminal level and sectioning the filum terminale (Fig. 6). Caution must be exercised in maintaining continuity of the nerve roots as they pass within the canal. After withdrawal of the sectioned sacrum, all remnants of tumor are excised. En bloc removal above the level of the sacroiliac joints is usually not possible, and precise fragmentary resection is required instead.

External drainage is always employed. In those instances necessitating an open dural procedure, meticulous attempts at a watertight closure are mandatory to minimize the risks of external drainage. Low-pressure bulb suction systems are recommended, and the surgeon is cautioned to refrain from using high negative-pressure systems when the cerebrospinal fluid space has been violated.

Closure is accomplished by firmly suturing the muscle and fascial layers in place and reattaching the anococcygeal ligament to the distal fascia. The skin and subcutaneous tissues are then closed in standard fashion.

**Fusion.** Occasionally, the continuity of the sacroiliac joints is disrupted, either as a result of extensive sacral resection or by direct tumor involvement. The stability of the joints must be preserved if useful function is to be maintained. If instability exists following tumor removal, then a fusion procedure is indicated. When the majority of the sacroiliac joint is intact, a simple posterior packing with an autologous or homologous bone graft is sufficient. More extensive violation of the joints may require internal fixation between the sacrum and ilium in addition to bone grafting (Fig. 7). Fusion should be performed as a separate procedure after the initial operation has had time to heal and permanent instability has been verified. Generally, 9 to 12 weeks between operations is a sufficient waiting period.
Operative Results

Ten patients underwent a posterior approach and three had an anterior resection as their initial surgical procedure (Table 1). In six cases, the nerve root from which the tumor arose could not be positively identified. A ventral S-1 nerve root sheath was considered the source of the schwannoma in three cases and a ventral S-2 nerve root was thought to be responsible in four patients. Four of the schwannomas extended intradurally, requiring open dural procedures, and nine were entirely extradural. Gross total removal was accomplished in four patients, all of whom were operated on with the posterior approach. Of the nine patients with subtotal resections, five remained free of symptoms during the follow-up period, whereas four required further surgery for recurrent disease. The elapsed time between the first and second operations for these four patients was 7, 10, 22, and 84 months. The second operation was an anterior resection in three patients.
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and a combined anterior-posterior procedure in the fourth. Postoperatively, two of these individuals were considered to have sacroiliac joint instability and underwent subsequent fusion procedures with autologous and homologous bone grafting. One of these two patients also required internal spinal fixation. Postoperative complications were found in three patients. One patient experienced prolonged wound drainage, another developed a seroma which required open drainage, and a third patient developed a superficial wound infection following a fusion procedure. There were no perioperative deaths.

Postoperative Results

All 13 patients reported resolution of preoperative pain following surgery. In addition, all of them were ambulatory and able to resume their routine daily activities. Postoperative examination revealed neurological deficits in six patients. Unilateral perianal sensory loss was present in four and bilateral perianal sensory deficit in one. One patient, who underwent extensive sacral resection and required a fusion procedure with internal spinal fixation, displayed complete motor and sensory loss in a unilateral L-5 and S-1 distribution. This patient and one other experienced decreased rectal sphincter tone and urinary retention as permanent deficits requiring self-catheterization and voluntary bladder and bowel control techniques.

Follow-up periods ranged from 5 months to 33 years and 3 months, with a mean of 9 years. Eleven of the 13 patients were asymptomatic and showed no evidence of tumor progression. One patient reported the gradual onset of mild bilateral lower-extremity dysesthesiae beginning more than 14 years after surgery. Another patient noted return of low-back and lower-extremity pain only 21 months postoperatively. Computerized tomography documented residual tumor in this latter case and in two additional asymptomatic patients. Two patients died of unrelated illness.

Pathological Findings

The microscopic sections from each case were examined to reconfirm the diagnosis. All except three tumors showed the characteristic features of schwannoma (neurilemoma), including monomorphous composition of elongated Schwann cells with variable "degenerative atypia" and nearly total absence of mitosis. Hyalinized vessels accompanied by perivascular hemosiderin deposits were noted in all cases (Fig. 8 upper left and right). In addition to the usual cytological and vascular features of schwannoma, three tumors displayed significant hypercellularity and focal mitotic activity, with up to three mitoses per 10 high-power fields in one instance (Fig. 8 lower left and right). These lesions correspond to the so-called "cellular schwannoma" of Woodruff, et al.27

FIG. 5. Artist's drawings illustrating the posterior transsacral approach. a: Patient positioning and location of the incision. b: Right lateral view showing the schwannoma's presacral portion, intradural extension, and erosive nature. c: The sacrum has been unroofed and an L-5 laminectomy performed to expose the full extent of tumor involvement. d: Intradural tumor resection. e: Extradural tumor resection. f: Completed tumor resection with preservation of a unilateral S-3 nerve root and "hand-in-glove" dural repair with a fascia lata graft.
FIG. 6. Artist's drawings illustrating the technique for extensive sacral resection. Depicted are the normal sacral anatomy (left), the method of radical sacral resection (center), and dural repair (right).

FIG. 7. Postoperative plain roentgenograms, anteroposterior (left) and lateral (right) views, showing extensive sacral resection and sacroiliac reconstruction with bone grafts and internal fixation.
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Discussion

The growth of cervical, thoracic, and lumbar schwannomas is limited by the close confines of the intervertebral foramina and spinal canal. This results in relatively early compression of the spinal cord, cauda equina, or nearby nerve roots and subsequent neurological symptomatology. The intrasacral space is unique in that it has the capacity for regional expansion of neurogenic tumors, resulting in a delayed clinical presentation as the slow-growing mass fills the sacrum. Once this finite volume has been filled, the schwannoma will either: 1) advance cephalad within the spinal canal, often intradurally; 2) track through the sacral foramina; or 3) erode through the walls of the sacrum. Erosion of the sacral bone is presumably secondary to the pressure effect of the slowly enlarging mass and not due to any inherent invasive properties of the schwannoma.

In accordance with previous reports of spinal axis schwannomas, the tumors in our series showed no sex predilection, there was a peak incidence of symptom onset in the fourth decade of age, and pain with or without dysesthesiae was the most prevalent symptomatic feature. Demonstrable sensorimotor deficits were uncommon, whereas diminished deep-tendon re-

Fig. 8. Photomicrographs of giant sacral schwannomas. Upper Left: Typical schwannoma showing Antoni A and B patterns with associated vascular sclerosis. H & E, × 64. Upper Right: Typical schwannoma showing hypocellular proliferation of cytologically benign Schwann cells with associated vascular changes. H & E, × 160. Lower Left: Cellular schwannoma with Antoni A and B pattern variation and degenerative vascular changes. H & E, × 64. Lower Right: Cellular schwannoma with hypercellularity and occasional mitotic figures. H & E, × 400.
flexes and a palpable retrorectal mass on digital examination were consistent findings.

Plain roentgenography, myelography, and CT are considered essential and complementary studies in the preoperative assessment. Computerized tomography has largely supplanted adjunctive radiological studies due to its ability to clearly delineate both the intraspinal and intrapelvic extent of tumor involvement. However, myelography does provide additional information in regard to the tumor's superior aspect, is predictive of intradural extension, and can effectively rule out additional intraspinal lesions.

Preoperative needle biopsy or limited tissue sampling is not recommended. The information gained from a biopsy does not affect the surgical management of a large destructive sacral mass with neurological involvement. Also, the risk of infection following a transrectal or percutaneous biopsy is real, as shown by the two patients biopsied in this series. Both were found at surgery to have developed bacterial infection of the tumor bed.

Choice of surgical approach is dependent upon the degree of sacral destruction, intrapelvic extension, and sacroiliac joint involvement. Schwannomas with a modest presacral component can usually be resected by the posterior transsacral approach alone. If a large presacral portion is present, then an anterior transabdominal operation is performed initially to gain control of the vascular plexus that encompasses the mass and to ensure identification and protection of important intrapelvic structures. The intrasacral tumor remnants can then be resected safely and with easy accessibility via a posterior transsacral operation, either as a separate delayed procedure or as a combined operation performed under the same anesthetic. Radical sacral resection necessitated by diffuse sacral destruction or sacroiliac joint involvement is required to minimize the chance of tumor recurrence. The degree of sacroiliac joint instability following tumor resection is evaluated 9 to 12 weeks postoperatively and will determine the need for bone reconstruction.

All but three tumors in this series displayed classic features of benign schwannoma. Three specimens showed a greater than usual degree of cellularity with occasional mitotic figures. Such "cellular schwannomas" have been considered benign. In our limited series, these features did not appear to affect prognosis.
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Considering the quality of current CT scans and the development of magnetic resonance imaging, it is reasonable to follow these patients with serial scans (Figs. 9 and 10). The need for further surgical treatment, however, will ultimately be based upon the clinical status of the patient. In fact, three patients who have evidence of tumor persistence on CT have not shown any change in tumor size and have not developed new neurological symptoms that would invite surgical intervention. We are, therefore, content to merely observe these patients for the time being.

Conclusions

Our experience suggests that these histologically benign but neurologically devastating tumors should be aggressively resected with the intent of complete extirpation. Although the intraoperative sacrifice of sphincter function or the creation of new sensorimotor deficits may be formidable concepts to a patient with intact neurological function preoperatively, these tumors will ultimately compromise bladder and bowel function and in some instances threaten pelvic arch stability if left untreated or inadequately resected. Mere debulking operations also run the risk of greatly increasing the difficulty and dangers of a later more definitive procedure. Complete tumor removal may require both an anterior and a posterior approach and possibly a reconstruction. Complete tumor removal may require both an anterior and a posterior approach and possibly a reconstruction procedure, but it can be accomplished with minimal risk and the anticipation of an excellent prognosis.

References