Spinal extradural schwannoma

PAOLO CELLI, M.D., GIUSEPPE TRILLÒ, M.D., AND LUIGI FERRANTE, M.D.

Division of Neurosurgery, St. Andrea Hospital, Department of Neurological Sciences, Second Faculty of Medicine, La Sapienza University Medical School, Rome, Italy

Object. The authors endeavor to define the clinical and surgery-related profile of spinal nerve sheath tumors located in the extradural space outside both the dural sac and, apparently, the nerve roots’ sleeve.

Methods. A series of 24 extradural schwannomas was retrospectively selected after reviewing the notes of spinal nerve sheath tumors surgically treated at La Sapienza University of Rome. Clinical data, tumor-related characteristics, and outcome were analyzed.

Women predominantly harbored these tumors. On admission sensory nerve root dysfunction was infrequently reported, whereas pyramidal tract deficits were often present. The tumor, generally large, was most frequently located in the intermediate thoracic segments and high cervical region; only one was reported in the lumbosacral region. Considerable erosion of vertebral bodies was reported in almost one third of the cases. In four patients eloquent nerve roots, that of C-5 in three and that of S-1 in one, were involved with the tumor. Radical tumor resection, with preservation of the nerve roots, was possible in several cases, whereas in two patients manipulation and resection of the C-5 root produced transient and permanent, respectively, root palsy. At follow-up examination patients for whom walking was impossible before surgery were now able to walk.

Conclusions. Extradural schwannomas can be distinguished from other nerve sheath tumors growing inside the spinal canal by their clinicoradiological features and unlikely nerve root origin. After surgery, recovery from pyramidal tract deficits, even severe, is noteworthy; in the authors’ experience, however, resection of an involved appendicular root is more likely to result in a permanent and significant radicular deficit.

KEY WORDS • spinal tumor • extradural tumor • nerve sheath • schwannoma • nerve root

Extradural schwannomas have not been the focus of investigation in the series of spinal nerve sheath tumors, and have been the subject of relatively little study in general, being more often described in a single case report or small series. Their clinical profile, therefore, is not clearly defined. In recent years we have surgically treated a number of patients with cervical and thoracic extradural schwannomas, in whom the pathological tissue was found to be totally outside the nerve roots and the tumor could be radically removed while conserving nerve root integrity. This prompted us to review the operative records pertaining to patients with spinal nerve sheath tumors who underwent surgery in our department and to assess cases involving the extradural lesions.

Clinical Material and Methods

We searched the cross-index files of the pathology and surgery registers of the Neurosurgical Institute of Rome, University La Sapienza, collected during a 50-year period. Excluding completely or largely extraspinal tumors and those for which there were inadequate operative reports, we obtained data on 24 extradural schwannomas in 24 patients who form the basis of this clinical series (Fig. 1). In all cases the pathological tissue was reported to be external to the dural sac but, apparently, also outside the nerve roots. In fact in some cases no nerve root was seen in significant relation to the tumor. In others the nerve roots appeared on the surface of or were engulfed in and crossed the pathological tissue, which was defined as “adherent” or, especially in the intervertebral foramina, “attached to” and “infiltrating” the sleeve of the roots, whose size and appearance, however, were not clearly pathological.

Patient Population

In no patient were there the clinical stigmata of neurofibromatosis or other diagnosed schwannomas at the time of surgery or during the follow-up period. The tumors were more prevalent in women (71%). The median age of the patient population was 44.5 years. The duration of clinical manifestations ranged from 1 month to 6 years (median 24 months). This duration was no shorter in the 12 patients in whom surgery was performed after the..
advent of CT scanning (after 1977) than in those treated before (median 24 and 10 months, range 2–60 and 1–72 months, respectively).

Clinical data and the correlation between clinical syndrome and spinal level of tumor are reported in Table 1. Local vertebral pain was reported by almost all patients, but symptoms or sensory signs of clearly radicular distribution were present in only five patients: pain in two with thoracic tumors, paresthesias in two with subaxial cervical tumors, and paresthesias in one with a T1–4 tumor.

Fig. 1. Artist’s drawings depicting the more typical configuration and extension of spinal extradural nerve sheath tumors in frontal (upper) and axial (lower) views. These tumors can grow in all directions (longitudinally in the epidural space, anteriorly in one or two excavated VBs, posteriorly eroding the laminae, and anterolaterally in the intervertebral foramen) and frequently extend in the paraspinal space through the enlarged intervertebral foramen or a bone defect. When they are large and extend extraspinally, their more typical configuration is characterized by lobulating contours and irregular margins; moderate to massive involvement of laminae and VBs is a constant finding and destruction of the spine segments on one side of the midline is sometimes striking.

<table>
<thead>
<tr>
<th>Variable</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>sex</td>
<td></td>
</tr>
<tr>
<td>F</td>
<td>17</td>
</tr>
<tr>
<td>M</td>
<td>7</td>
</tr>
<tr>
<td>age (yrs)</td>
<td></td>
</tr>
<tr>
<td>median</td>
<td>44.5</td>
</tr>
<tr>
<td>mean</td>
<td>38.6</td>
</tr>
<tr>
<td>range</td>
<td>13–65</td>
</tr>
<tr>
<td>duration (mos) of preop symptoms</td>
<td></td>
</tr>
<tr>
<td>≥6</td>
<td>29%</td>
</tr>
<tr>
<td>≤12</td>
<td>67%</td>
</tr>
<tr>
<td>60–72</td>
<td>17%</td>
</tr>
<tr>
<td>median</td>
<td>24</td>
</tr>
<tr>
<td>range</td>
<td>1–72</td>
</tr>
<tr>
<td>nerve root dysfunction†</td>
<td></td>
</tr>
<tr>
<td>sensory nerve</td>
<td>5</td>
</tr>
<tr>
<td>motor nerve</td>
<td>1</td>
</tr>
<tr>
<td>LE CST</td>
<td>20</td>
</tr>
<tr>
<td>tumor level‡</td>
<td></td>
</tr>
<tr>
<td>C1–2</td>
<td>7</td>
</tr>
<tr>
<td>T4–7</td>
<td>3</td>
</tr>
<tr>
<td>T5–8</td>
<td>1</td>
</tr>
<tr>
<td>T9–12</td>
<td>3</td>
</tr>
<tr>
<td>L5–S2</td>
<td>1</td>
</tr>
<tr>
<td>tumor location</td>
<td></td>
</tr>
<tr>
<td>IS</td>
<td>4</td>
</tr>
<tr>
<td>IS-for</td>
<td>15</td>
</tr>
<tr>
<td>IS-for-PV</td>
<td>5</td>
</tr>
<tr>
<td>plain x-ray finding</td>
<td></td>
</tr>
<tr>
<td>bone abnormality</td>
<td>17§</td>
</tr>
<tr>
<td>resection</td>
<td></td>
</tr>
<tr>
<td>total</td>
<td>23</td>
</tr>
<tr>
<td>subtotal</td>
<td>1</td>
</tr>
<tr>
<td>schwannoma type</td>
<td></td>
</tr>
<tr>
<td>benign</td>
<td>23</td>
</tr>
<tr>
<td>melanotic</td>
<td>1</td>
</tr>
<tr>
<td>spinal stabilization</td>
<td>1</td>
</tr>
<tr>
<td>transient op complication**</td>
<td></td>
</tr>
<tr>
<td>neurological</td>
<td>2</td>
</tr>
<tr>
<td>nonneurological</td>
<td>3</td>
</tr>
</tbody>
</table>

* Clinical data for patients undergoing a second surgery reflect those obtained at the first procedure (performed at outside institutions). Abbreviations: for = foraminal; IS = intraspinal; LE CST = lower-extremity corticospinal tract; PV = paravertebral.
† Sensory nerve root dysfunction included pain in two cases, paresthesia in two, and hypesthesia in one; the motor nerve root deficit was a foot flexion–extension disorder; and in the 20 cases involving a lower-extremity corticospinal tract deficit nine patients were nonambulatory.
‡ Clinical syndromes related to tumor level included the following. Atlantoaxial level tumors: dysesthesia/hypesthesia/mild amyotrophy of the hands and pyramidal tract deficit of the legs (four cases [two nonambulatory], Brown–Séquard syndrome (two cases), and torticollis only (one case); C4–7 tumors: nerve root paresthesia and Brown–Séquard syndrome (one case), nerve root paresthesia alone (one case), and ascending pathway pain contralateral to the lesion as well as hypesthesia (one case); T1–12 tumors: Brown–Séquard syndrome or variation (12 cases [root pain in two, and six nonambulatory] and spastic–flaccid paraparesis (one nonambulatory case); and L5–S2 tumors: moderate foot flexion–extension deficit (one case).
§ Includes six cases of severe VB erosion.
‖ After combined posterior–anterolateral removal of a C4–6 schwannoma, stabilization of the spine was performed by placing an iliac bone graft and plate system.
** Neurological complications included deterioration of long tract pathways (to Grade III [see Table 2]) soon after excision of a C1–2 schwannoma (one case [patient recovered preoperative {Grade IV} status]) and sensorimotor C-5 nerve root palsy due to manipulation (one case [recovery was complete after 3 months]). Nonneurological complications included CSF leakage related to sectioning of involved roots (three cases [resolution after bedrest and CSF drainage]).
tumors, and touch and pinprick hysthesias in one patient with a lower cervical growth. On admission (Table 1) corticospinal tract dysfunction was present in 20 patients, nine of whom were unable to walk and suffered sphincter function impairment. Flexion–extension weakness of the feet was demonstrated in one patient who harbored a lumbar-sacral tumor. Of the seven patients with C1–2 tumors upper-extremity symptoms, including various combinations of mild atrophy of the intrinsic hand muscles and sensory complaints, occurred in four. No motor deficit was observed in three patients with tumors at C1–2 (one case) and below C-3 (two cases), who exhibited torticollis, nerve root paresthesias, and root sensory deficit, respectively.

Tumor Locations

The tumor was located in the cervical region in 10 patients (seven at C1–2 and three below C-3), the thoracic region in 13 (one at T1–4, nine at T5–8, and three at T9–12), and the lumbar-sacral region (L5–S2) in one case. The tumor generally extended for more than one vertebral segment and occupied a lateral position in the spinal canal, nearly always with an intraspinal–extraspinal hourglass shape (six of seven C1–2 tumors progressed in the atlantoaxial interlaminar space, whereas all three subaxial cervical tumors, 10 of 13 thoracic tumors, and the one lumbar lesion grew through one or more intervertebral foramina). Five tumors, all cervical, possessed a significant extraspinal paravertebral component. In some cases the tumor extended anteriorly with coarse erosion of the VB, whereas a thoracic growth appeared under the myofascial planes through the eroded laminae.

In all 17 cases in which plain x-ray films or the radiologist’s reports of the spine were available, bone abnormalities were demonstrated. Striking enlargement of the interlaminar space between the atlas and axis was a typical finding, plainly demonstrated in lateral views, in five cases of C1–2 schwannomas. In almost one third of cases (six of 17) erosion of one or two VBs was present (Figs. 2 and 3) with well-defined marginal sclerosis (Fig. 2). Imaging studies—CT scans (after 1977; Figs. 2 and 3) with CT myelograms in some and/or MR images (available after 1988; Fig. 4)—were obtained in 10 of 12 patients who underwent surgery after 1977. The preoperative diagnosis was epidural or nerve sheath tumor, but in two patients in whom CT scans were obtained it was spinal metastasis based on extensive changes of the VBs (Fig. 2).

Operative Procedure

Twenty-two patients underwent surgery for the first time and two underwent a second operation when symptomatic clinical recurrence was demonstrated 15 and 20 years, respectively, after an incomplete excision performed at other institutions.

A posterior or posterolateral approach was used to reach C1–2 tumors, one lower cervical lesion, and thoracic and lumbar-sacral lesions, while two subaxial cervical schwannomas were removed via an anterolateral or combined posterior–anterolateral approach. Excision was apparently total in 23 cases (first operation in 21 and second operation in two with clinical recurrences) and subtotal in one patient to preserve normal-appearing and -functioning C-5 nerve root.

Results

Histopathological Features

Macroscopically, the pathological tissue was described as having clear borders, an irregular nodular surface, and being unencapsulated, but well circumscribed and readily cleaved.

Based on histological features, 23 tumors were diagnosed as fascicular (Type A) and/or loose reticular (Type B) schwannomas; one was a melanotic schwannoma without definitive malignant features (mild pleomorphism and spindletoid figures), and its histopathological, immunohistochemical and electron microscopy features have been previously reported.65 Immunohistochemically, all tumors were positive for S100 staining.

Soon after surgery two patients experienced significant but transient neurological deterioration (Table 1): one patient who underwent surgery for a C1–2 lesion exhibit-
ed an increase in long tract motor dysfunction (to Grade IV), but her function had recovered to its preoperative status (Grade III) at discharge; the other patient, who underwent surgery for a cervical tumor, suffered a palsy, not present before surgery, of the C-5 nerve root, which was manipulated at length intraoperatively but not sectioned (the only case in which the tumor was removed incompletely); recovery was complete within 3 months.

At long-term follow-up examination (median 9 years; Table 2), 15 patients remained alive. Of those no longer living, one patient in whom a thoracic melanotic schwannoma was removed suffered local recurrence and died of lung metastases 11 months after the operation, and six died of causes unknown or unrelated to the spinal tumor. Two were lost to follow up. No other clinical recurrence was observed in the patients who underwent surgery in our department; the only patient in whom excision was subtotal remained in stable clinical and radiological condition 4 years later.

Noteworthy improvements of pyramidal tract deficit were evident in all patients at discharge (except for the patient in whom it worsened soon after surgery and in whom status eventually recovered). Of 18 patients who attended follow-up examinations seven who were nonambulatory at surgery were eventually able to walk, at least with aid (Table 2).

At discharge the patient in whom the lumbosacral tumor was removed experienced slight improvement in foot movements, whereas the four patients treated for high cervical schwannomas experienced resolution or improvement of upper-extremity sensory disturbances.

Surgery-related nerve root palsy, still severe 10 years later, was present in one patient in whom the C-5 nerve root was sacrificed intraoperatively. She also suffered persistent but moderate deep-seated pain in the distribution of the divided root.

Discussion

Spinal Extradural Schwannomas

Apart from rare intramedullary schwannomas, spinal nerve sheath tumors are classified as intradural (50–83%),5,11,13,20,32,38,54,59,60,71,81,88,96,104 intradural–extradural (7–24%),5,11,13,20,32,38,54,59,60,71,81,88,96,104 and purely extradural (2–31%).5,11,13,20,32,38,54,59,60,71,81,88,96,104

In published series of spinal nerve sheath tumors,5,11,32,34,38,39,44,49,54,59,71,81,88,91,96,104 extradural tumors have not been systematically addressed, being given no more than passing attention and being noted mainly in association with descriptions of the more frequent intradural and intradural–extradural tumors. With some exceptions,64 reports of extradural nerve sheath tumors, published sporadically since 1914,7 have involved single or only a few cases, mainly reported as examples of benign epidural lesions7,97 or to illustrate peculiar clinicoradiological features,6,10,18,31,45,50,58,62,75,77,87,92,101,102 pathological characteristics,15,67,85 or surgical approaches and results.33,36,51,61,65,66 In these papers the macroscopic appearance of the tumor and nerve roots, if adherent to or enfolded in the pathological tissue, is hardly ever reported explicitly and the nature of the nerve root involvement is not discussed. The terms used to describe the relationship of the tumor to the roots (attached to, developing on, arising or originating from, close relationship to, and crossed by) and of the roots to the tumor (involved, embedded or engulfed in, running or just passing across, and crossing through) do not seem to have specific and differing meanings. In the present series schwannomas developed in the spinal canal outside the dura, and the nerve roots involved were not definitely abnormal in appearance.

Patient Population, Clinical Manifestations, and Imaging Features

There were no clinical signs of neurofibromatosis69 and all schwannomas were solitary.

The mean patient age was 38.6 years and the majority (71%) were women; in the general category of spinal nerve sheath tumors (mostly intradural and intradural–extradural) the reported mean age of patients was similar (range 38–45 years)5,11,13,20,32,38,54,59,60,91,94 but neither sex was prevalent11–13,32,38,39,54,71,81,91,96,104 or males formed the ma-
Spinal extradural schwannoma

The duration of clinical history in our patients was extremely variable (range 1 month–6 years). It is noteworthy that in three of four patients with longer preoperative histories, the first symptom was not pain but lower-limb hypesthesia. The duration of history did not seem related to the year of surgery (medians 10 months in patients who underwent surgery before 1977 and 24 months in those treated after 1977, when CT scanning, and after 1988, when MR imaging became available), to severity of neurological deficit (severe deficits were found in patients with both long and short histories), or spine alterations on admission studies (even in patients with brief histories there was extensive osseous erosion revealed on x-ray films). The incidence of striking osseous changes, even in patients suffering rapid clinical deterioration, suggests that, irrespective of the duration of clinical history, these tumors are associated with a long period of latency before symptom onset, remaining minimally symptomatic or asymptomatic at length. The presence of a tough dural sac, which tends to withstand gradual compression by an extradural slow-growing mass, may explain why these types of tumors often become large while manifesting few neurological deficits and why, once the resistance of the dural membrane is overcome, the long pathway dysfunction occurs or worsens during a short period. In one of our cases, intact motor function deteriorated to severe paraparesis in a few weeks.

Radicular complaints (pain, paresthesia, or superficial hypesthesia) were infrequent, observed in almost one fifth of our patients; this finding is in contrast to the high incidence of preoperative nerve root symptoms reported in patients in whom overall spinal nerve sheath tumors were resected; in these cases, pain, generally radicular, was the first or an early symptom in 49 to 75% of cases, the most frequent symptom in 72 to 85%, or the only symptom in 30%.

The clinical syndromes produced by extradural schwannomas were nonspecific because of their spinal level and the epidual position of their growth (Table 1). A clinical syndrome with different combinations of hand-based amyotrophy/dysesthesia/paresthesia/tactile agnosia was observed in four of seven our patients with C1–2 schwannomas; these upper-extremity disturbances have been reported by others in patients with similar extradural high cervical schwannomas. This clinical syndrome is known to be produced by compression in the region of the occipital foramen and high cervical segments mainly by intradural extramedullary tumors but also by nontumoral extradural lesions.

On admission, 87% of 23 patients with cervical and thoracic tumors in the present series (which, however, included patients treated before the advent of CT scanning) had corticospinal tract deficits, and 39% (nine of 23) were unable to walk. The severity of pyramidal tract dysfunction seems in some way related to the spine level of the lesion and incidence of nerve root symptoms. Indeed, patients with C1–2 and thoracic schwannomas who did not complain of root disturbances exhibited severe pyramidal tract deficits, whereas all three patients with cervical tumors below C-3 suffered radicular dysfunction, but only one experienced mild hemiparesis.

Radiographically documented bone abnormalities of the spine were observed in all cases of schwannoma in the present series (that is, those for which x-ray films or radiological descriptions were available), and coarse erosion of VBs was present in approximately one third of these. Likely due to their extradural position, the osseous changes in schwannoma are clearly more frequent than the incidence (30–65%) and more extensive than the enlargement of the intervertebral foramen reported for spinal nerve sheath tumors in general.

Before the advent of CT scanning, radiographic documentation of VB erosion allowed an extradural schwannoma to be diagnosed preoperatively as a metastatic growth, especially in the presence of rapid neurological deterioration. Although CT scanning was performed, the tumors in two of our patients were preoperatively misjudged as malignant; however, the re-review of the plain x-ray films clarified the lesion’s benign features (a round radiolucent, cystlike defect sharply defined by a border of sclerotic bone; Fig. 2).

Level Distribution of Lesion Within the Spine

Along the longitudinal axis of the spine, the extradural schwannoma was located in the cervical region (10 cases [41.7%]), the thoracic region (13 cases [54.1%]), and one...
(4.2%) was present in the lumbosacral spine. In particular, the tumors were more frequent at atlantoaxial level (29% of all schwannomas and 70% of the cervical lesions) and in the intermediate (T5–8) thoracic segments (37.5% of all schwannomas and 69.2% of the thoracic lesions). By contrast, spinal nerve sheath tumors in general (mainly intradural and intradural–extradural) are fairly evenly distributed or have predilection for the cervical, lumbar-sacral, or thoracic levels. In particular they have mainly been reported in the low thoracic (~ 50% below T-8) and thoracolumbar regions, whereas their incidence at high cervical sites (C1–2 or the foramen region) is less than 10.4%, and represents 9.1 to 38% of the cervical manifestations. On the other hand, the majority (38%–100%14) of C1–2 nerve sheath tumors were reported to be located extradurally.

In the present series, in the spinal canal the schwannomas were generally positioned lateral to the dural sac, but the anterior spread of neoplastic tissue into the VBs was relatively frequent. A vertebrocolumnar extension into the intervertebral foramina or through the eroded VBs was observed in 83% of tumors, and in five of these (all cervical) the paraspinal component was significant. The high incidence of this hourglass pattern of growth through the intervertebral foramen, explained by the tumor’s extradural position, is clearly greater than the overall 14 to 23% rate reported for spinal nerve sheath tumors.

Surgical Treatment and Long-Term Results

Surgical approaches for dumbbell-shaped spinal schwannomas have been well analyzed by McCormick and Lot and George. In our patients, most of whom underwent surgery a number of years ago, the approach was almost always posterior and involved a laminectomy, which was often enlarged by undertaking a unilateral facetectomy on the side of the tumor. In only two patients with subaxial cervical tumors who underwent surgery more recently was the approach purely anterolateral or combined posteri–anterolateral. In 22 patients who underwent primary surgery in our department and two who underwent repeated operation for recurrence, tumor removal was reported to be macroscopically total. Resection, however, was subtotal in the case of a C4–6 schwannoma to preserve the C-5 root which had a normal appearance and function. In the two patients who underwent repeated operation, clinical recurrence occurred 15 and 20 years, respectively, after incomplete removal of the tumor at other institutions. This long period to recurrence is in line with the not infrequent delayed relapse of spinal schwannomas, even after subtotal removal. No patient treated in our department experienced tumor recurrence, except for one who underwent surgery for a melanotic schwannoma, who died of disseminated malignancy; however, the prognosis of melanotic nerve sheath tumor is unpredictable, and the aggressive and fatal course that occurred in our patient has also been described by Royta, et al. for a similar pigmented, not malignant, spinal extradural schwannoma. Therefore, typically ominous preoperative features, such as striking osseous erosion demonstrated radiographically and, in some cases, the rapid neurological deterioration, do not necessarily predict an unfavorable prognosis.

As shown in Table 2 the comparison of neurological status pre- and postoperatively and neurological status preoperatively and at follow up (median 9 years) in 18 patients showed noteworthy improvement of pyramidal tract deficits. Before discharge improvement was shown in almost all cases and at follow-up examination all seven patients who were nonambulatory preoperatively were now able to walk. One patient, who had been almost completely paraplegic (Grade V) and in whom a preoperative diagnosis of vertebral metastasis (Fig. 2) was established underwent emergency surgery and was able to walk 6 months later. Similar striking functional recovery after removal of an extradural nerve sheath tumor has been reported by others and the reversibility of neurological damage, even severe, has been attributed to the fact that loss of function may be due more to venous stasis than to irreversible mechanical damage or arterial ischemia.

In the present series, no sacrifice of any root was possible in six cases, in which all but one cervical lesion was radically removed. In the other 18 patients, 21 nerve roots were sacrificed; 20 were of little functional significance (C-2 in seven, thoracic in 12, and S-2 in one), and only one, C-5, was of functional importance. Thus, in most cases the decision to cut the roots enveloped by the tumor may have been determined more by their relative functional unimportance than by the impossibility of radical resection of the pathological tissue without damaging the root.

Four schwannomas involved eloquent nerve roots. In one patient with a lumbosacral tumor the S-1 roots were preserved, but an S-2 root was resected. In the other three patients an eloquent and functionally intact C-5 nerve root was embedded in the pathological tissue. In one of these the root was left intact and the tumor was radically removed using meticulous microsurgical dissection of the pathological tissue from the root’s sheath. In another patient the root was manipulated at length but not sectioned, and the operation was limited to incomplete tumor excision; postoperatively there was a root palsy, which abated completely within 3 months. In the last patient the nerve root was sectioned to allow for complete excision of pathological tissue; a corresponding radicular deficit was severe and has remained unchanged for 10 years.

The aforementioned observations suggest two practical considerations. 1) Radical removal of these tumors without sacrificing the underlying nerve root seems possible; likewise McCormick reported gross-total resection of an extradural intraspinal–extraspinal C5–6 schwannoma without needing to sacrifice the root. Encasement and adherence of pathological tissue to the nerve root can make it difficult to determine whether the schwannoma is actually infiltrating or simply enfolding the radix. In the early stage of surgery, however, a cautious, blunt nerve root–sparring dissection should be attempted so that unnecessary sacrifice of eloquent nerve roots can be avoided. 2) When sectioning a functionally important root, the risk of causing severe radicular impairment appears to be real in cases involving these types of extradural schwannomas. In the present series the deficit was permanent in a unique case in which the root was sacrificed, and it was transient in another case in which the root was manipulated at length. On the other hand, it is known that this risk is generally very low when surgery involves removal of overall nerve sheath tumors and it seems to be practically nonexistent when excising intraradicular nerve sheath tumors. It has been
suggested that extradural schwannomas growing outside the nerve root do not significantly, or only later, affect the radicular function; thus, more often, at the time of surgery the affected roots are functional and not compensated. Therefore, during surgery of extradural schwannomas, the intraoperative monitoring of nerve root function is strongly advised.

Origin of Extradural Extraradicular Schwannomas

In our series the operative notes were in line with the description of a tumor that was generally large and not really capsulated, but possessed clear margins and was readily separated from the dura and the bone; it grew outside the dural sac, apparently outside the sleeve of the roots involved (more often the thinner roots of high cervical and thoracic segments), and even if enveloped by the pathological tissue, it seemed normal in size and appearance.

Although the point of origin of extradural schwannomas is not definitely known, one source to consider first are the Schwann cells or “perineurial” or “perineurothelial” cells of nerve fibers inside the nerve root sheath, which are the origin of the more common intradural and/or intraradicular schwannomas. Because of early breakthrough or infiltration of the nerve root sleeve, the tumoral cells may infiltrate the extradural space and grow almost completely outside the nerve root, which therefore does not appear to be significantly enlarged. This reasonable hypothesis, however, does not explain the complete absence of pathological tissue inside the dural sac, where tumors as large as these should easily extend from within the root. Additionally, infiltration and transgression of the root sheath in the early stages of tumor development contradict the fact that this sleeve, outpouching from the dural sac, is in general a remarkably effective barrier against inflammatory or neoplastic invasion. Furthermore, certain findings in our patients do not agree with this hypothesis; for instance, there was no relationship between nerve roots and tumor in several cases; in some cases, cerebrospinal fluid flowed freely from the stump of resected small thoracic roots folded in the tumor, which should exclude the presence of an intraradicular component of tumor; and, finally, there was no tumor residue or recurrence demonstrated at follow-up examination in patients in whom no nerve root was sacrificed.

In light of these findings we believe that an extraradicular origin should be considered. Roytta, et al. reported that extradural schwannomas have a rare but well-established lesion. They are rarely observed in the spine, even though the sacrum seems to be the second most frequent site. These tumors are usually found intradurally and are characterized by a lack of function of the affected roots. The vertebral bone could also be a potential origin of these lesions, because of the frequency and extension of osteolytic lesions of the spine. Intraosseous schwannomas are rare but well-established lesions. They are rarely observed in the spine, even though the sacrum seems to be the second most frequent site. These tumors are usually found intradurally and are characterized by a lack of function of the affected roots. Therefore, the origin of these schwannomas may be explained using the two theories typically proposed when these tumors are observed intraoperatively or because the nerves involved are reasonably considered only contiguous and not in continuity with the tumor. This pattern is not infrequent; in large surgical series involving schwannomas (excluding those primarily arising from spinal and cranial nerves), a nerve of origin was identified in fewer than half of the cases. Therefore, it seems more logical to suggest that the extradural tumors in our cases shared a relationship with these paraspinal schwannomas, both growing close to and around the vertebral column.

The origin of these schwannomas may be explained using the two theories typically proposed when schwannomas are without a parent nerve (because no nerve is observed intraoperatively or because the nerves involved are reasonably considered only contiguous and not in continuity with the tumor). This pattern is not infrequent; in large surgical series involving schwannomas (excluding those primarily arising from spinal and cranial nerves), a nerve of origin was identified in fewer than half of the cases. Therefore, it seems more logical to suggest that the extradural tumors in our cases shared a relationship with these paraspinal schwannomas, both growing close to and around the vertebral column.
nerve ending. Thus, any evident relation of the tumor with the small nerve of origin would be lost. The vertebral column is surrounded by thin sensory and sympathetic nerve plexuses, ventrally supplied mainly by small filaments from sympathetic chain, rami communicantes, and perivascular nerves of segmental arteries and dorsally by main contributions from a branch of the spinal nerve, which, with its spinal and sympathetic components, enters the spinal canal. These nerves extend to the bone, periosteum, ligaments, dura, anulus fibrosis, and accompanying vessels of the epidural space.

Therefore, these thin paraspinal nerves can be considered the most reasonable source of paravertebral schwannomas in which a parent nerve is absent, both in the anterior regions and extradurally in the spinal canal. An alternative hypothesis is that these nonnerve related schwannomas originate from multipotential mesenchymal cells or from variably primitive neuroectoderm cells (from neural crest to mature Schwann cells) that are abnormally displaced during embryonic development. Ectopic rests are more predisposed to oncogenic influences than similar cells in normal anatomical locations, and this predisposition should be particularly true for Schwann cells, which typically multiply when separated from nerve fibers, as in the case after nerve injury.

Conclusions

Because of their preoperative clinical features (high incidence of corticospinal tract deficits and low incidence of radicular disturbances) and radiological characteristics (striking erosion of the vertebral bones), extradural schwannomas should be considered a subgroup of spinal nerve sheath tumors distinct from more common intradural, intradural–intradural, and intradural nerve sheath tumors growing inside the spinal canal. Some surgery-related considerations, such as the nonpathological appearance of the involved nerve roots, the possibility of radical removal of the tumor without the sacrifice of any radix, and the high risk, in our experience, of severe radicular palsy, if the involved roots are resected, indicate that these types of tumors have an unlikely origin in the nerve roots.

Acknowledgment

We are grateful to Mr. Remo Romolo Natali for the artistic drawings.

References

29. Feigin I, Ogata J: Schwann cells and peripheral myelin within
Spinal extradural schwannoma

63. Martin P, Kleyntiens F: [Subdural tumors of the occipital foramen.] Rev Neurol 82:313–334, 1950 (Fr)
73. Orf G: [Clinical and morphological findings in an infiltrating spinal neurinoma.] Arch Psychiatr Nervenkr 213:396–407, 1970 (Ger)