Pelvic plexus tumors

Kurtus Dafford, M.D.,1 Daniel Kim, M.D.,2 Natasha Reid, M.D.,3 and David Kline, M.D.4

1Department of Neurosurgery, Tulane University Medical Center, New Orleans; 2Department of Neurosurgery, Ochsner Clinic Foundation, New Orleans, Louisiana; 3Department of Obstetrics and Gynecology, Virginia Commonwealth University, Richmond, Virginia; and 4Department of Neurosurgery, Louisiana State University Health Sciences Center, New Orleans, Louisiana

Object. Pelvic plexus tumors are unusual entities. These lesions often reach significant size prior to clinical presentation due to vague lower extremity or other symptoms refractory to exhaustive workup and the potential space of the retroperitoneum.

Methods. The authors retrospectively reviewed the charts of 44 patients who presented to Louisiana State University Health Sciences Center with pelvic plexus tumors. All such patients were included in the present study if there was at least 12 months of follow-up. Clinical evaluations were examined, including typical clinical presentation, imaging studies, and other preoperative evaluations. These tumors include 38 peripheral neural sheath tumors (86%), two malignant neural sheath tumors (5%), and four nonneural sheath tumors (9%). The authors also examined the relation of pelvic neural sheath tumors to neurofibromatosis Type 1 (NF1).

Results. Histopathological examination confirmed 18 solitary neurofibromas (41%), 12 NF1-associated neurofibromas (27%), eight schwannomas (18%), two malignant nerve sheath tumors (5%), and four other nonneural sheath tumors (9%). The other nonneural sheath tumors consisted of one each of desmoid, ganglioma, lipoma, and an unspecified calcified mass.

Conclusions. The optimal treatment for retroperitoneal lesions remains operative excision with adjunctive therapy specific to the lesion encountered.

Key Words • neurofibromatosis • pelvic plexus • peripheral nerve sheath tumor • retrospective study

Retroperitoneal soft tissue tumors are unusual and often present as a pelvic or abdominal mass.20,23 These lesions can be divided into peripheral neural sheath tumors and peripheral nonneural sheath tumors. The peripheral neural sheath tumors in the present study arose from the nerve sheath of the pelvic plexus, as the name implies; the peripheral nonneural sheath tumors included ones that arose from any of the surrounding structures in the retroperitoneal space. These two categories can be further subdivided into their respective benign and malignant entities.10

The retroperitoneal space is not an actual area but rather a potential space. Tumors arising in the retroperitoneal space can reach significant size before becoming clinically evident. Often, such masses present as an abdominal or pelvic mass or mimic other musculoskeletal syndromes,4 20,22,23 further delaying their recognition and treatment. Clinical suspicion for these tumors can point the physician to obtaining adequate radiological imaging to aid in the diagnosis. Pelvic lesions pose a particular challenge to the general surgeon, orthopedic surgeon, and neurosurgeon in terms of diagnosis, treatment, surgical approach, and long-term patient outcome. Many pelvic lesions fall into the category of peripheral neural sheath tumors, and association with other known disease entities should be considered; this is especially true of patients with NF1, also known as von Recklinghausen disease.

In this paper, we will take a retrospective look at the charts of patients who presented with pelvic plexus tumors to the neurosurgical department at Louisiana State University Health Sciences Center from 1975 to 2006. The symptoms on presentation, surgical interventions, relation to NF1, and outcomes will be discussed. This information should help guide other clinicians in considering pelvic plexus lesions in the differential diagnosis when vague lower extremity, bladder symptoms, or a painless mass in the abdomen or pelvis are noted on clinical examination.

Clinical Material and Methods

This is a retrospective examination of 44 patients who presented with pelvic plexus tumors to the Louisiana State
University Health Sciences Center from 1975 to 2006 and had at least 12 months of follow-up. The senior authors, Drs. Daniel Kim and David Kline, performed all resections. The clinical presentation in these patients consisted of the presence of a pelvic mass, pelvic pain, paresthesias, or weakness. Pelvic pain or paresthesia was the presenting symptom in 38 (86%), and a pelvic mass was discerned in 26 (59%) of 44 patients. Weakness of either quadriceps with knee extension or foot dorsiflexion was present in 11 (25%). Urinary urgency was a nonspecific symptom in some patients. Patients underwent preoperative imaging studies to include magnetic resonance imaging and/or CT to delineate the extent of these lesions. Preoperative EMG studies were performed to assess baseline function in patients.

Results

Pathological examination confirmed 18 solitary neurofibromas (41%), 12 NF1-associated neurofibromas (27%), eight schwannomas (18%), two malignant nerve sheath tumors (5%), and four other nonneural sheath tumors (9%). The other nonneural sheath tumors consisted of one each of desmoid, ganglioma, lipoma, and a calcified mass not otherwise specified on pathological review.

The primary goal of surgical intervention was the gross-total resection of the lesion. A subtotal resection was documented if positive margins were found on pathological examination or if incomplete resection was carried out during the primary procedure. Total resection was achieved in 14 (78%) of the solitary neurofibromas, six (50%) of the NF1-associated neurofibromas, seven (88%) of the schwannomas, and all four nonneural sheath tumors. Neither malignant peripheral nerve sheath tumor could be completely resected. Local resection with inclusion of plexus elements was performed in both patients. One patient with a malignant neural sheath tumor died 4 years after the procedure, and in the other no recurrence was noted at the last clinic visit, but the patient has subsequently been lost to follow-up.

After surgical intervention, moderate pain reduction was noted in 34 (77%) patients. This was evaluated based on a decreased requirement for narcotic pain medications postoperatively. The follow-up period ranged from 8 to 48 months, with a mean of 14 months. Weakness was the most common postoperative finding in patients. This was especially true in the patients with NF1-associated neurofibromas, among whom 11 (92%) had some demonstrable weakness postoperatively. However, 50% returned to preoperative strength levels within 12 months of surgical intervention.

Discussion

Ackermann is credited with the first description of the retroperitoneal space. He described its location as between the lumbar and iliac regions and between the peritoneum and the posterior parietal wall of the abdominal cavity. The floor is formed by the fascia covering the quadratus lumborum and psoas major muscles. The roof is formed by the parietal peritoneum, which in turn covers the pancreas, the third and fourth portions of the duodenum, the kidneys, the adrenal glands, and, laterally, the ascending and descending colon. The space is composed of loose areolar and fat tissue through which pass the ureters, various nerves and ganglia, renal vessels, gonadal vessels, aorta, and inferior vena cava. Although our focus was on pelvic tumors, this potential space must be kept in mind during the clinical evaluation of patients with pelvic tumors. Tumor growth may proceed unhindered into this space, allowing tumors to become quite large and involved with the surrounding structures before clinical presentation. Knowledge of this space is also needed for surgical planning.

Benign neural sheath tumors have often been referred to collectively as “neurofibromas.” However, as further case reports became available, the distinction between neurofibromas and schwannomas (also referred to as neurilemomas) became evident. Pathological studies provide some distinction between these two entities. A distinct feature is the higher collagen or elastin content found in neurofibromas. Both neurofibromas and schwannomas are believed to be derived from cells of Schwann cell origin, and special staining techniques assist in the diagnosis. Both tumor populations stain positive for S100 protein, but staining may be absent in some neurofibromas. Schwannomas are readily identified by the presence of Antoni Type A and B areas. The Antoni A portion is a relatively cellular area with spindle-shaped cells. When these cells form palisades, they become known as Verocay bodies. Antoni type B tissue is an arrangement of spindle cells in a clear mucinous matrix. Neurofibromas are reported to demonstrate a more myxomatous stroma than that seen in schwannomas, which causes heavier staining for mucopolysaccharide with Alcian blue in the neurofibroma. In addition, neurofibromas have been shown to stain heavier for reticulum, and with Bodian staining may show more axons than schwannomas.

Malignant degeneration is a concern, especially of larger neurofibromas and especially in patients with NF1. The natural history of schwannomas is not well documented, but there seems to be consensus that these lesions tend not to have the same potential for malignant degeneration that is found in neurofibromas. Neurofibromas, on the other hand, show up to a 15% chance of malignant degeneration in patients with von Recklinghausen disease, although the incidence cited in some reports ranges from 5 to 42%. The authors of many reports on these tumors advocate wide resection margins in patients in whom nerve function is already compromised. In the absence of NF1, the risk of malignant degeneration of these tumors is considered quite low.

Neurofibromatosis Type 1, or von Recklinghausen disease, is an autosomal-dominant disease with patients fulfilling two of many criteria for diagnosis. It is thought to be a common neurological disorder, with an incidence of one in 2500 to 4700 births reported. Diagnostic criteria include two or more of the following: six or more café-au-lait spots, each greater than 5 mm in greatest diameter in the prepubescent individual or greater than 15 mm in greatest diameter in the postpubertal patient; more than two neurofibromas of any type or one plexiform neurofibroma; freckling in the axillary or intertriginous areas; optic glioma; more than two Lisch nodules; distinctive osseous abnormalities; or a first-degree relative with NF1 according to the above criteria. Lisch nodules appear as
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Malignant peripheral neural sheath tumors comprise a group of previously heterogeneously named lesions classified as malignant schwannomas, malignant neurilemmas, and neurofibrosarcomas. There are also pathological difficulties with these tumors share a cellular origin with other spindle-cell sarcomas like monomorphic synovial sarcoma, leiomyosarcoma, and fibrosarcoma. As previously noted, there is a high incidence of these lesions in patients with von Recklinghausen disease or NF1. An interesting clinical feature of these tumors is also their multifocality and the development of secondary tumors with the same morphological characteristics. These lesions are all considered aggressive in nature, and en bloc resection is the goal of therapy.\(^1\,2\,8\,13\,21\)

The diagnosis of malignant peripheral nerve sheath tumors has relied on the combination of a gross fusiform tumor in relation to nerve, microscopic features of spindle cells with fascicular patterns and varying degrees of mitosis, the presence of areas of necrosis or tumor calcification, the presence of associated benign Schwann cells, and positive immunohistochemical staining for S100 protein and neuron-specific enolase.\(^9\)

Desmoid tumors are rare soft tissue tumors that arise from the musculoaponeurotic and facial tissue and manifest as locally invasive bands of fibrous tissue. These tumors will become adherent to surrounding structures, making resection with clear margins difficult when found in association with contiguous neural or vascular structures. Total excision of this lesion is the treatment of choice, with lower recurrence rates reported for microscopically negative margins. Recurrence rates of 5 to 50% in the setting of negative margins have been reported in the literature, compared with a 90% recurrence rate in tumors with microscopically positive margins.\(^10\,31\)

Adjunctive therapy has included radiation therapy or interstitial brachytherapy with a dose of 45 to 65 Gy.\(^7\,13\) Tamoxifen has been used as a chemotherapeutic agent with variable results in the reduction of recurrence rates in patients with these tumors.\(^13\)

Lipomas, although usually benign, can create neurological compromise when they cause neural compression by either enveloping the nerve or being in close approximation in a confined space. They have a fine capsule and are composed of adipose tissue. Surgical excision of these lesions becomes difficult if there has been a previously attempted resection or there is close approximation with plexus elements. Less common are lipomas arising from an intraneural component with an extraneural mass. These lesions can be associated with a lipohamartomatous condition, in which the nerve contains a significant fatty-fibrous mass. In this setting, the lesion can sometimes be debulked by resection of the involved fascicles with intraoperative monitoring for nerve function.\(^7\,10\,11\)

Gangliomas, or ganglioneuromas, are lesions composed of equal portions of neural and glial cells. They are rarely found in the peripheral nervous system. Very little information is available in the literature on this entity in the peripheral nervous setting.\(^10\,11\)

Malignant nonneural sheath tumors usually involve the neural elements by direct extension. These lesions can arise directly from the surrounding structures, or they may represent metastasis to the nerve. Resection with clear margins is the recommended treatment, with adjunctive therapy directed at the underlying pathological entity.\(^5\,10,23\)

Imaging studies of pelvic lesions have included CT and magnetic resonance imaging. Nishino and associates\(^15\) describe the diagnostic challenges associated with the localization of tumors in the retroperitoneal space. A familiarity with retroperitoneal structures, method of tumor spread, specific tumor components, and tumor vascularity help to narrow the differential diagnosis when using these two modalities. Bass and colleagues\(^7\) have reported CT findings of plexiform neurofibromas of the retroperitoneum, which may help differentiate benign from malignant lesions. In their study, benign lesions were typically bilateral, symmetric, low attenuating masses in the paraspinal or presacral location (Fig. 1). Asymmetry in size or attenuation of the lesion was suggestive of underlying malignancy.\(^1,8\) This information is useful in guiding the surgeon toward or away from performing biopsy sampling of the lesion. Plexiform neurofibromas carry an inherent risk of sensorimotor loss on biopsy sampling because the...
nerve fibers are interspersed with the lesion. Biopsy sampling of these lesions usually requires obtaining multiple samples to ensure a true representation of diseased tissue, and at times results can still be inconclusive. The risk of incurring neurological deficits after biopsy sampling must be weighed against the information provided by the procedure. In patients in whom pain is the only presenting complaint, the senior authors recommend observation of the lesion over relatively frequent intervals to further guide indications for resection with increasing size or further neurological impairment.

High signal intensity with a low intensity band surrounding a thickened perineurium have been demonstrated on T2-weighted magnetic resonance imaging. Hypointense septations have also been reported on T2-weighted imaging of neurofibromas and plexiform neurofibromas (Fig. 2). Electromyographic studies obtained preoperatively provide some guidance for the intended surgical approach. In patients with compromised neural function or silent EMG studies, the tumor may be resected along with the associated neural elements without further compromise of function.

Surgical Exposure

Surgical approaches to the pelvic plexus are defined best in the general surgical and gynecological literature. Most surgeons would opt for a transabdominal approach to true pelvic lesions with the patient in the supine position. More laterally located lesions, especially those involving the lumbar plexus and thus not in the true pelvis, can be exposed via a retroperitoneal approach. Occasionally, lumbosacral plexus tumors may extend through the sciatic notch, and then a second subgluteal approach may be necessary for complete excision. This would allow for the greatest exposure of lesions involving the pelvic plexus. However, if the lesion involves or extends into the ischiorectal fossa, a posterior approach to these lesions (described by Miller et al.23) may be more effective, as this approach allows access to lesions originating below S-2 without introducing further neural compromise. The most common postsurgical complication of the posterior approach was noted to be wound dehiscence, which was treated with Sitz baths.

There is no substitute in the treatment of pelvic tumors for adequate surgical exposure, and help from other specialties is paramount in this regard. In benign neural sheath tumors, the dominant tumor in this series, tissue coverings including the capsule were incised and removed to expose the tumor proper. However, in cases of suspected or known malignant tumors, we recommend that the capsule be left intact for en bloc resection of the lesion (Fig. 3). Unfortunately, malignancy could not be determined in some cases until after the capsule had been opened and frozen sections obtained and analyzed. We then worked around the entire periphery of the tumor, often using fibrillar or a similar agent to keep capsule, coverings, and associated fascicles separate from the tumor proper. Heavy sutures were then placed in the upper points of the tumor to permit it to be shifted slightly so that its deeper aspects and proximal and distal poles could be dissected from capsule and surrounding tissues. Care was taken with bipolar cautery of the entering and leaving vessels, and in a few cases they had to be tied off or occluded by vascular clips. In this fashion, all but the very large tumors could usually be enucleated, sparing the pelvic plexus. The very large tumors had to be debulked with the cavitational ultrasonic surgical aspirator (Valley Lab) or, in the case of more fibrous ones, by sharper dissection using scissors, scalpel, and/or the bovie. Strict and patient attention was paid subsequently to hemostasis of the tumor bed by use of bipolar cautery and placement of fibrillar and/or Gelfoam, and occasionally fibrin or similar hemostatic aids.
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Conclusions

Pelvic plexus tumors are uncommon. Vague lower extremity symptoms and/or findings in a patient with no other obvious pathological entity should raise the clinical suspicion of a retroperitoneal or pelvic lesion. Occasionally these patients will present with a palpable abdominal or pelvic mass and no neurological compromise. These lesions are often quite large on presentation because of the potential space of the retroperitoneum. Magnetic resonance imaging and CT studies are essential for delineating the extent of tumor or tumoral invasion into adjacent structures in these patients. Clues may also be garnered from these studies as to whether these lesions are malignant or benign. Preoperative EMG studies may aid the surgeon in defining the suitability for complete resection without further neural compromise. En bloc resection of any lesion of the retroperitoneum is advocated to control local recurrence or the compromise. En bloc resection of any lesion of the retroperitoneum. Magnetic resonance imaging and CT studies are essential for delineating the extent of tumor or tumoral invasion into adjacent structures in these patients. Clues may also be garnered from these studies as to whether these lesions are malignant or benign. Preoperative EMG studies may aid the surgeon in defining the suitability for complete resection without further neural compromise. En bloc resection of any lesion of the retroperitoneum is advocated to control local recurrence or the compromise. En bloc resection of any lesion of the retroperitoneum is advocated to control local recurrence or the compromise.

References


Address reprint requests to: Daniel Kim, M.D, F.A.C.S., Department of Neurosurgery, Ochsner Clinic Foundation, 1514 Jefferson Highway, New Orleans, Louisiana 70121. email: neurokimdaniel@yahoo.com.