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Introduction

Retrorectal tumors encompass a heterogeneous group of lesions and neoplasms that demonstrate variable growth patterns. As a result of their anatomic location these rare lesions, manifest with non-specific symptoms that makes their initial diagnosis difficult. Often they are found incidentally on physical exam or imaging in the work up of other disease processes. While these lesions are fairly well documented in the literature they are discussed largely in case studies. The largest series is maintained by the Mayo clinic representing 1 in 40,000 hospital admissions [1, 2]. This chapter will illustrate and characterize the etiology, diagnosis and management of these lesions, specifically focusing on minimally invasive approaches to this disease process.

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Anatomy

The retrorectal space lies between the upper two-thirds of the rectum and the sacrum, above the rectosacral fascia. The boundaries of this potential space include the posterior wall of the rectum (fascia propria) anteriorly and the sacrum posteriorly. This space extends superiorly to the peritoneal reflection and laterally is bound by the lateral stalks of the rectum, the ureters, the sacral nerve roots and the iliac vessels. This retroperitoneal space contains loose connective tissue. The presacral fascia protects the extensive vertebral plexus of presacral vessels that lie deep to it.

The retrorectal space is thought to contain totipotential cells from all three germ cell layers. The wide range of histological variance may be due to the presence of multiple embryologic remnants and heterogeneous tissue types. These lesions range from benign cysts to malignant masses which can invade the surrounding pelvic structures [3].

Classification

While a variety of classifications systems have been proposed for presacral tumors, the current system is a modification of Uhlig and Johnson's system which places retrorectal tumors into broad categories: congenital, neurogenic, osseous, inflammatory, and miscellaneous. In the current

scheme, Dozois et al. subcategorized these lesions into malignant and benign entities [4, 5]. While the majority of the lesions in the presacral space are benign up to one third will be malignant. Malignant tumors occur more frequently in men and are associated with worse outcomes than benign with regard to recurrence, function and post-operative complications [6]. The current classification system aids in the characterization of these lesions but in guiding the appropriate therapeutic approach.

Congenital Lesions

Congenital lesions arise from the remnants of embryonic tissue and may be cystic or solid in nature. Benign cystic congenital lesions include developmental cysts and anterior meningoceles while benign solid lesions consist of teratomas, and adrenal rest tumors. Malignant congenital lesions consist of sacrocoxygyl chordomas, teratocarcinoma and can result from the malignant degeneration of cystic lesions. Congenital lesions are the most common retrorectal tumors, accounting for 60.5% of all tumors in the presacral space. They are usually benign and are more common in females. Malignant lesions typically present at an older age (53.9 ± 11.5 vs. 40.1 ± 12.2 years; $p = 0.05$) and occur more frequently in males [7].

Cystic Lesions

Developmental Cysts

Developmental cysts are the most common congenital lesions and can be further classified based on cell layer of origin and are divided into epidermoid, dermoid, duplication and tail gut cysts.

Epidermoid Cysts and Dermoid Cysts

Epidermoid and dermoid cysts result of closure failure of the ectodermal tube. They are lined with squamous epithelial cells. Epidermoid cysts are composed of stratified squamous cells; they are typically unilocular lesions and do not contain skin appendages while dermoid cysts have stratified squamous epithelium with skin append-

ages (sweat glands, hair follicles, sebaceous cysts). They tend to be well circumscribed, round and have a thin outer layer. Both of these cysts may communicate with the skin and be associated with a postanal dimple or sinus. The postanal dimple or sinus that can be frequently misdiagnosed and managed as a perirectal abscesses, fistula in ano, or pilonidal disease. If errantly drained there is a secondary infection rate of 30% [2, 8].

Duplication Cysts (Enterogenous)

Enterogenous cysts arise from the endoderm of the primitive hindgut. As these lesions originate from endodermal tissue, they are typically lined with squamous, cuboidal, or columnar epithelium. They are called rectal duplication cysts if they are related to the rectum. These tumors usually have a multilobular appearance with one dominant lesion and smaller satellite cysts. Similar to epidermoid and dermoid cysts, they are more common in women and may become infected. Although the vast majority of these lesions are benign, malignant degeneration is possible [9].

Tail Gut Cysts (Cystic Hamartomas)

Tail gut cysts, or cystic hamartomas, arise from remnants of the postanal primitive gut that fail to regress. These multinodular encapsulated, well-circumscribed cysts may contain of squamous, columnar, or transitional epithelium. Morphologically, these cysts are similar to the adult or fetal intestinal tract. Tail gut cysts can be differentiated from epidermoid and dermoid cysts by the presence of glandular or transitional epithelium as well as the presence of a defined muscular wall with a myenteric plexuses. In general, these are benign lesions, however, malignant transformation has been reported in up to 13% in some series [10, 11].

Anterior Sacral Meningocele

Anterior sacral meningoceles are a result of a defect in the thecal sac and may be seen in combination with presacral cysts or lipomas. Only rarely do these cysts contain neural elements; if they are present the lesion is considered a

myelomeningocele. Anterior sacral meningocele may be associated with other congenital anomalies, such as spina bifida, tethered spinal cord, urinary tract or anal malformations as well as uterine or vaginal duplications [12]. They are secondary to protrusions of the dural sac through a unilateral defect in the anterior sacrum. Of note, this defect results in a sacrum that demonstrates a rounded concave border without bony destruction on plain radiograph resulting in the classic radiologic finding of the “scimitar sign” seen on plain films (Fig. 26.1). Patients often have vague symptoms including headaches related to postural changes, Valsalva maneuver, coughing and defecation. This can be attributed to the compression-induced increase in cerebrospinal fluid pressure due to the continuity between the dural sac and subdural space. Biopsy or aspiration is contraindicated as secondary infection may result in life-threatening meningitis. Surgical management requires ligation of the dural defect [12, 13].



Fig. 26.1 Classic radiologic finding of the “scimitar sign” associated with sacral meningocele

Solid Lesions

Teratomas and Teratocarcinoma

Teratomas are neoplasms derived from pluripotential cells and include all three germ cell layers. They include epithelium of the gastrointestinal tract, respiratory tract, and nervous system. These lesions may be solid or cystic and often contain both components (Fig. 26.2). Teratomas have the potential may undergo malignant squamous cell carcinoma arising from the ectodermal tissue or rhabdomyosarcoma arising from the mesenchymal cells. Furthermore, anaplastic tumors in which the germ cell origin is not be distinguishable are also seen. This is of primary concern because up to 10% of teratomas will undergo malignant degeneration if left untreated [3, 13]. Teratomas are more common in females and in the pediatric population and are often associated with anomalies of the vertebra, urinary tract, or ano-rectum. Histologically, these tumors are referred to as either “mature” or “immature” reflecting the degree of cellular differentiation [11]. Malignancy is rare beyond the second decade; however, the neonatal malignancy rate is 4%. In adults, malignant degeneration can occur in 40–50% [11, 14]. These lesions tend to adhere to the coccyx and surgical approach requires en bloc coccygectomy (Fig. 26.3). Incomplete or intralesional resection increases the likelihood of malignant degeneration. These lesions can also become infected and be misdiagnosed as a perirectal abscess or fistula.

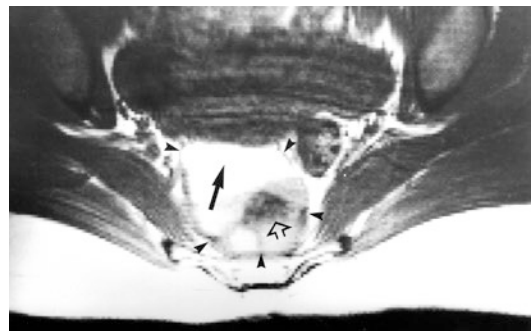


Fig. 26.2 Teratomas may be solid or cystic and often contain both components

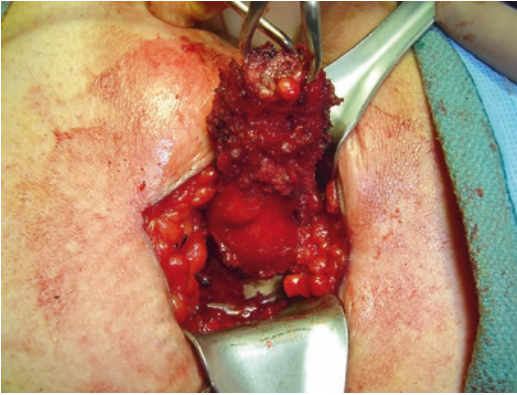


Fig. 26.3 Coccygectomy with retrorectal tumor removal

Due to their location diagnosis is often delayed and these tumors may reach considerable size [15, 16].

Sacroccygeal Chordomas

Chordomas arise from the primitive notochord, which extends from the base of the occiput to the caudal limit of the embryo. They are the most common malignancy in the retrorectal space and may occur anywhere along the embryologic notochord with 30–50% occurring in the sacroccygeal region (Fig. 26.4) [17]. These lesions occur more frequently in men and are rarely encountered in patients younger than 30 years of age. The most common symptoms include pelvic, buttock, and lower back pain aggravated by sitting and alleviated by standing or walking. These slow-growing tumors may be soft, gelatinous, or firm and may invade, distend, or destroy bone and soft tissue (Fig. 26.5). Hemorrhage and necrosis within tumors may lead to secondary calcification and pseudocapsule formation. Teratomas metastasize to lung, liver, and bone in 20% of cases [18]. These tumors often reach substantial size because of delays in diagnosis secondary to the indolent nature of the disease. Patients may be asymptomatic, present with vague complaints including positional buttock, pelvic, or lower back pain; or they may present with specific symptoms secondary to invasion, including impotence and incontinence. Local recurrence rates are high despite radical resection; the 10-year survival rate is only 9–35% [19].

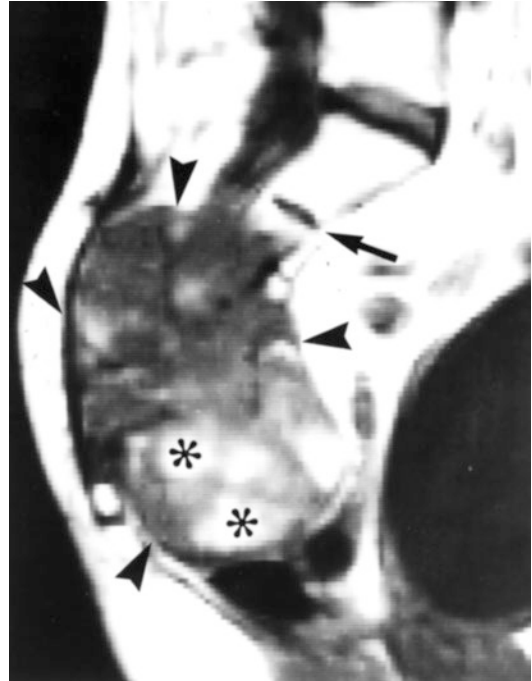


Fig. 26.4 Chordoma



Fig. 26.5 Chordomas are slow-growing tumors may be soft, gelatinous, or firm and may invade, distend, or destroy bone and soft tissue

Neurogenic Tumors

Neurogenic tumors represent 10% of retrorectal tumors and are the second most common presacral lesion after congenital lesions. They typically arise from peripheral nerves and 85% are benign. Neurogenic tumors include neurilemmomas, ganglioneuromas, ganglio-neuroblastomas, neurofibromas, neuroblastomas, ependymomas,

and malignant peripheral nerve sheath tumors (neurofibrosarcoma, malignant schwannomas, and neurogenic sarcomas) [20]. Neurilemmomas were the most common benign neurogenic tumors. Neurofibrosarcoma were the most common malignant neurogenic tumors. Malignant tumors had a higher recurrence rate compared to benign neurogenic tumors (42 vs. 6.7%; $p < 0.05$) [7]. These slow-growing tumors cause non-specific symptoms and may be of considerable size when diagnosed. If symptoms are present, pain distribution and neurologic dysfunction are related to the route of the affected nerve. Preoperative tissue biopsy is of paramount importance as the operative approach is guided by pathology.

Osseous Tumors

Osseous tumors represent approximately 10% retrorectal tumors. They arise from bone, cartilage, fibrous tissue, or marrow. Osseous tumors include chondrosarcoma, osteosarcoma, myeloma, and Ewing's sarcoma. These tumors arise from the bone, cartilage, fibrous tissue, and marrow. All osseous tumors of the presacral space are associated with sacral destruction.

Giant cell tumors are the most common benign osseous tumors. Although benign, giant-cell tumors can metastasize to the lungs, Ewing tumors are the most common malignant osseous tumor [6, 20].

Miscellaneous Tumors

Miscellaneous tumors account for 10–25% of all retrorectal tumors and include lipoma, fibroma, leiomyoma, hemangioma, endothelioma, locally aggressive desmoid tumors, various sarcomas, metastatic adenocarcinoma, hematomas, carcinoid tumors, anomalous pelvic ectopic kidneys and inflammatory tumors. Inflammatory lesions include extension of infection from Crohn disease and perforated diverticulitis. Leiomyoma is the most frequent benign histologic type, followed by a fibroma. The most frequent malignant tumors are metastatic tumors [21].

History and Physical Examination

Due to their indolent growth, presacral tumors are typically found incidentally on imaging, physical exam or during childbirth. The symptoms caused by retrorectal lesions are related to their site, size and, in the case of retrorectal cysts (Fig. 26.6), the presence or absence of infection. Benign lesions tend to be asymptomatic whereas malignant lesions are more likely to produce symptoms. Symptomatic patients typically complain of vague, long-standing pain localized to the low back or perianal area associated with rectal ache, or deep rectal pain. Their pain may be postural, aggravated by sitting and improved by standing or walking. Often the onset of pain relates to local trauma such as a fall on the sacrum or coccyx. If the sacral plexus is involved, patients may experience referred pain in the legs or buttocks.

Patients with retrorectal tumors may present with signs and symptoms infection, pelvic outlet obstruction or central nervous system manifestations. Patients may present with isolated or recurrent fever, chills and rigors. Patients may also complain of perineal discharge and may have midline dimpling just posterior to the anus or the gluteal muscle. This may lead to misdiagnosis of a fistula or pilonidal disease. Large retrorectal tumors may cause symptoms of pelvic outlet obstruction including constipation, sexual dysfunction, rectal or urinary incontinence, or obstructive labor in pregnant patients secondary to obstruction or direct invasion. Large masses may interfere with the passage of stool giving the

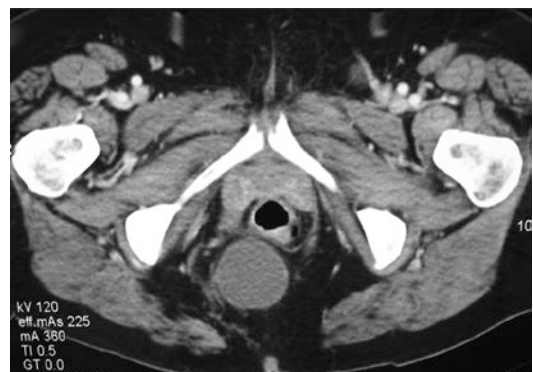


Fig. 26.6 Retrorectal cyst

feeling of incomplete evacuation or disturbances in bladder function secondary to interference with pelvic parasympathetic supply, direct pressure on the bladder or urethra, or obstruction of the pelvic ureters. Both urinary and fecal incontinence may occur due to impingement on the sacral nerve roots or overflow incontinence secondary to outlet obstruction.

Patients should be carefully examined, focusing on the perineum and rectal examination. Identification of a postanal dimple may assist in identifying the presence of a developmental cyst. Approximately, 97% of tumors are diagnosed incidentally on rectal exam [2]. Digital rectal exam frequently reveals the presence of an extrarectal mass displacing the rectum anteriorly with a smooth and intact overlying mucosa. Rectal examination is also critical in assessing the level of the uppermost portion of the lesion, degree and extent of fixation, as well as the relationship to other pelvic organs. Rigid or flexible sigmoidoscopy can be used to assess the overlying mucosa and rule out transmural penetration of the tumor. Location of the mass should be recorded, as well as whether it is lobulated or solitary, and whether it is possible to define its upper limits. In particular, the mass must be assessed for its relationship to the sacrum and the coccyx. This assessment is important as the tumor location will determine the operative approach. A careful neurologic exam focusing on the sacral nerves and musculoskeletal reflexes is mandatory and may also aid in the diagnosis of extensive local tumor invasion. Soiling and a pouting anus may indicate interference with the sacral nerves. Laxity of the anal sphincters and saddle anesthesia of the perineum further support involvement of sacrococcygeal nerves [5].

Imaging

Once a diagnosis of a retrorectal tumor is suspected radiographic imaging should be obtained to assist in the verification of the diagnosis. Plain radiographs, computed tomography (CT), magnetic resonance imaging (MRI), and ultrasonography (US) all play a role in the identification of these lesions.

Plain radiographs of the sacrum are often obscured by overlying viscera containing gas, fecal material, or osseous structures making these images non-specific for retrorectal tumors. However, anterior-posterior and lateral radiographs (AP/LAT) of the sacrum can identify osseous expansion seen in a meningocele or destruction, or calcification of soft tissue masses indicative of locally aggressive tumors including chordomas, sarcomas, giant cell tumor, aneurysmal bone cyst, and neurilemoma. Furthermore, barium enemas may demonstrate anterior displacement of the rectum prompting more specific imaging.

Computed tomography (CT) scans and magnetic resonance imaging (MRI), have emerged as the imaging modalities of choice in diagnosing retrorectal tumors. These modalities complement each other. Computerized tomography can characterize lesions as solid or cystic, determine the spatial relationships between structures in the pelvis and evaluate for cortical bone destruction. MRI with contrast is more sensitive in evaluating soft tissue specifically, spinal imaging where it can demonstrate cord anomalies, thecal sac compression, osseous or nerve root involvement (Fig. 26.7). MRI is extremely important in deter-



Fig. 26.7 MRI with contrast is more sensitive in evaluating soft tissue specifically, spinal imaging where it can demonstrate cord anomalies, thecal sac compression, osseous or nerve root involvement

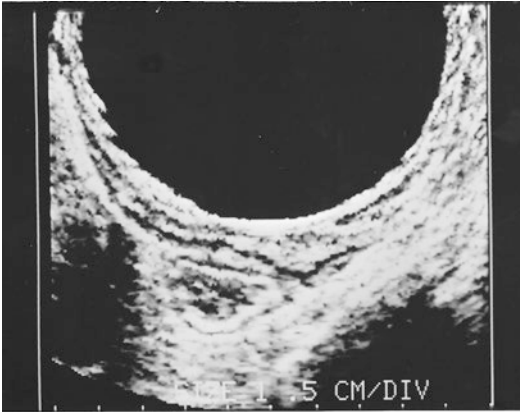


Fig. 26.8 Ultrasound can distinguish masses in the rectal wall from extramural lesions, determine the relationship of tumors to the muscular layers of the rectum and the anal sphincters as well as differentiate between cystic and solid lesions

mining level and extent of resection and can assist in determining the appropriate surgical approach [22].

Endorectal ultrasound, while used less, and less can provide further anatomic delineation of retrorectal tumors. Ultrasound can distinguish masses in the rectal wall from extramural lesions, determine the relationship of tumors to the muscular layers of the rectum and the anal sphincters as well as differentiate between cystic and solid lesions (Fig. 26.8). Tumor involvement of the rectal wall arising outside the rectum requires resection.

Preoperative Biopsy

The role of preoperative biopsy in diagnosis and management of retrorectal tumors is controversial. Historically, preoperative biopsy was contraindication in any potentially resectable presacral tumor for fear of increased local recurrence. In solid tumors, the concern that biopsy may cause seeding of malignant cells in the biopsy tract was frequently cited. These experts advocated that the best biopsy is en bloc operative excision. Preoperative biopsy was only acceptable if the lesion was considered to be inoperable or if clear that surgical excision cannot be undertaken with-

out significant risk to the patient, a preoperative diagnosis using a biopsy would be necessary to prevent inappropriate therapy [21, 23]. These recommendations, however, do not take into account the availability of modern imaging, better knowledge of tumor biology, and opportunities for neoadjuvant therapy.

Data has emerged suggesting that solid or heterogeneous tumors should be biopsied preoperatively. Accurate preoperative diagnosis of these tumors improves outcomes as optimal management of benign and malignant lesions differs considerably. Surgically, a wide-margin is required for oncologic resection in malignant lesions. On the other hand, resection with close-margin is acceptable for benign lesions to spare function and avoid morbidity [24]. Additionally, neoadjuvant therapy assists in optimizing oncologic outcome in specific tumor subtypes including Ewing sarcoma, osteogenic sarcoma, neurofibrosarcomas, and desmoid tumors. Furthermore, preoperative biopsy of presacral tumors has been determined to be safe and more highly concordant with postoperative pathology in comparison with imaging alone.

Dozois et al. recommended several guiding principles when performing biopsies on solid or heterogeneous retrorectal tumors. Prior to performing a biopsy, coagulation studies should be performed to minimize the risk of hematoma formation, which may contaminate the involved areas. The ideal approach is transperineal or parasacral as the biopsy tract must be within the field of the future surgical resection. Ideally, the CT guided biopsy is best performed by a radiologist experienced in the diagnosis and management of pelvic tumors, who is in direct consult with the primary surgeon regarding the biopsy's approach. Furthermore, transperitoneal, transretroperitoneal, transvaginal, and transrectal biopsies are to be avoided—the tract of the biopsy must be removed en bloc. Biopsies performed transrectally or transvaginally may also lead to infection, a more difficult complete excision, or increase the probability of postoperative complications and recurrence. Biopsy obtained via these routes necessitates either partial or complete proctectomy or vaginectomy to remove

the biopsy tract in continuity with the presacral tumor minimize recurrence [5, 10, 26].

Biopsy, however, has its limitations. Specifically, in the presence of a cystic lesion, biopsy may result in infection rendering its future complete excision more difficult and increasing the likelihood of postoperative complications and recurrence. More importantly, inadvertent transectal needling of a meningocele may lead to disastrous sequelae, such as meningitis and even subsequent death.

Management

The Role of Neoadjuvant Therapy

The availability of neoadjuvant tumor irradiation and systemic chemotherapy have altered the management of patients with retrorectal tumors. These tumors exhibit a wide range of behaviors and can be large and locally advanced by the time they are diagnosed. Accurate preoperative diagnosis can facilitate the application of neoadjuvant chemoradiation therapies and improve patient outcomes. Neoadjuvant radiotherapy can decrease in the size of large radiosensitive tumors, including chordomas and intradural myxopapillary ependymomas, which may spare vital structures that would otherwise be resected to obtain wide margins. Regardless wide margins and en bloc resection are often difficult to achieve in large malignant tumors resulting in high local recurrence rates approaching 64%. Neoadjuvant radiotherapy may decrease the size of the field of radiation, preventing the morbidity associated with applying adjuvant radiation therapy to the entire surgical bed, previous tumor site, all contaminated surgical planes, and the sites of all skin incisions in those cases where wide margin and en bloc resection cannot be achieved [10].

Neoadjuvant chemotherapy is essential to the treatment of some retrorectal tumors such as Ewing sarcoma and osteogenic sarcoma. Chemotherapeutic agents such as imatinib have been shown promote progression-free survival in patients with advanced chordomas however, this data is limited and the role

of chemotherapy in this population requires more rigorous analysis [25]. As the role of other agents are defined, their utilization may decrease recurrence rates and improve survival.

Surgical Approach

All presacral tumors should, unless the lesion is unresectable or there is evidence of systemic metastasis, should undergo en bloc resection. Approximately, 30–40% of lesions will be malignant and benign lesions may undergo malignant degeneration [27]. The utilization of a multidisciplinary team in the treatment of large and complex lesions is mandatory. The primary operative goal in the treatment of benign lesions is complete resection without tumor spillage while preserving surrounding tissues. For malignant lesions wide, en bloc removal of adjacent organs, soft tissue, and bone (if locally adherent) is the goal of resection. Only an experienced multidisciplinary surgical team consisting of a colorectal surgeon, orthopedic oncologic surgeon, spine surgeon, urologist, plastic surgeon, vascular surgeon, musculoskeletal radiologist, medical oncologist, radiation oncologist, and specialized anesthesiologist can appropriately evaluate and surgically treat tumors that are large and extend to or destroy the hemipelvis or the upper half of the sacrum [3].

The extent, location, and size of the tumor dictates the optimal approach (Fig. 26.9). The location, nature, and size of the lesion as well as the involvement of adjacent viscera, sacrum, or pelvic sidewalls appropriate surgical approach for retrorectal tumors is ascertained by appropriate imaging (CT and MRI). The extent of surgery is then determined by the of tumor characteristics: as previously stated, benign retrorectal tumors require complete gross resection, whereas malignant tumors will require radical resection, including en bloc resection of adjacent organs if involved. Incomplete resection in both benign and malignant tumors increases local recurrence [5]. The common approaches for resection of retrorectal tumors are the anterior (transabdominal) or combined abdominoperineal, the posterior (perineal) approaches and in rare instances transrectally.

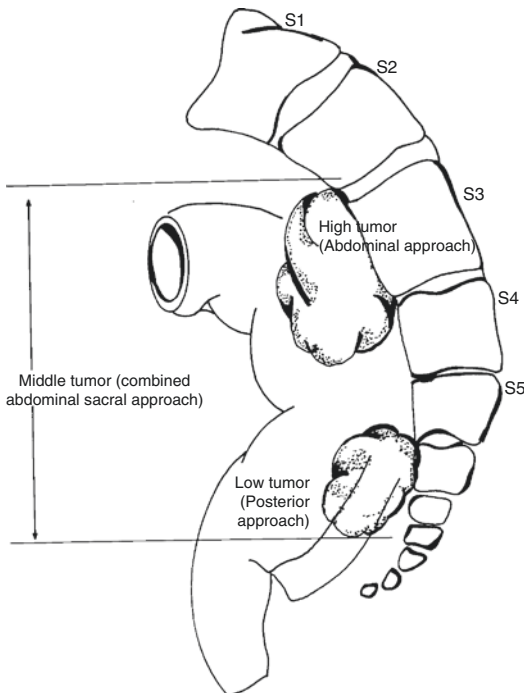


Fig. 26.9 The extent, location, and size of the tumor dictates the optimal approach to surgery for presacral tumors

Combined Abdominal and Perineal Approach

Although there are subsets of tumors that are appropriate for a purely abdominal approach, it is advisable to prepare the patient as if a combined abdominal approach is planned to allow for all contingencies. The anterior portion is performed when the most caudal portion of the lesion is above the level of S3–S4 based on preoperative imaging. Traditionally, these lesions have been approached through a laparotomy; however, advanced laparoscopic and robotic techniques have been described. A particular advantage of the anterior approach is that it allows the surgeon to gain wide exposure to major pelvic structures, including the pelvic viscera, vasculature, and ureters. During the transabdominal approach, the sigmoid colon is mobilized and the rectum is placed on stretch so that the pelvis can be examined. The retrorectal space is entered through the relatively avascular plane anterior to the sacrum. The mesorectum is then dissected from the anterior portion of the lesion. Prior to the removal of

the tumor, the arterial supply to the lesion must be identified and ligated. The middle sacral vessels are often significantly enlarged and should be ligated before mobilization is attempted.

While attempt at preservation of nerve roots and other vital structures is key to meticulous dissection, malignancies involving the rectal wall or that are locally invasive will require en bloc resection. Depending on the extent of the neoplasm, completion of the operation may require the patient to be repositioned in either lithotomy or prone position so that the remainder of the excision can be carried out through a posterior or perineal approach. All biopsy tracts should be excised and should include the skin through which the biopsy was performed. Ideally, a 2 cm margin should be sought [20]. Various bony and nerve structures may be sacrificed, depending on the location of the lesion. The help of a neurosurgeon or orthopedic surgeon is invaluable in these circumstances.

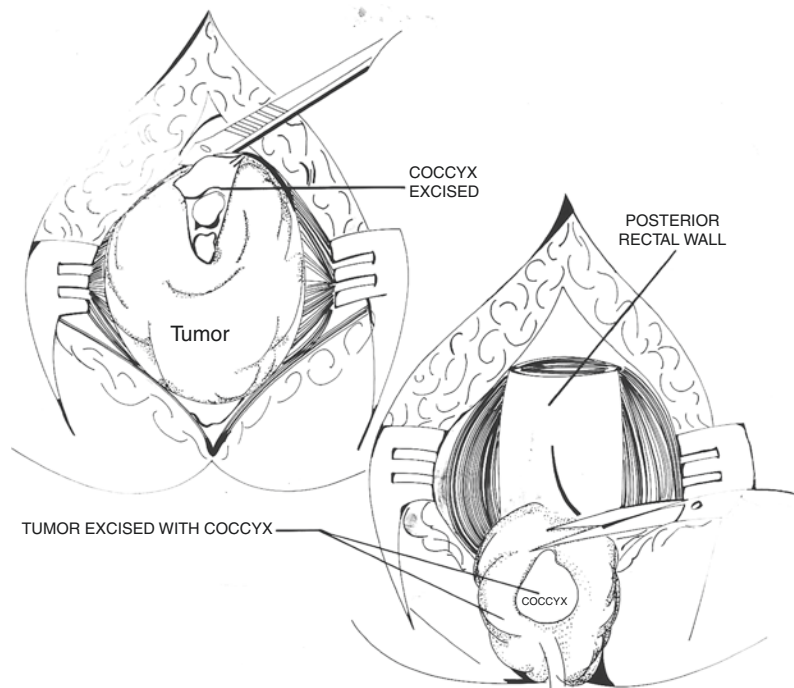
Posterior Approach

The posterior approach is useful for lesions below S3. If the superior border of the tumor is palpable digital examination, the posterior approach should be considered. For lesions that extend more superiorly and on preop MRI show nerve involvement, the posterior approach provides better visualization.

The patient is placed in the prone jackknife position and a midline parasacrococcygeal, curvilinear, or horizontal incision is made and deepened to define the sacrum, coccyx, and anococcygeal ligament. The anococcygeal ligament is detached from the coccyx and displaced, revealing the levator ani muscle with the central decussating fibers passing from the rectum to the coccyx. Resection of the tumor may be facilitated by transection of the anococcygeal ligament and coccyx. The lesion can then be dissected from the surrounding tissues, including the rectal wall, in a plane between the retrorectal fat and the tumor mass itself. If necessary, the lower sacrum, coccyx or both can be excised en bloc with the lesion (Fig. 26.10).

A nerve sparing technique has been described by Dozios et al. [28] from the Mayo clinic. Their technique includes preoperatively localization

Fig. 26.10 If necessary, the lower sacrum, coccyx or both can be excised en bloc with the retrorectal lesion



with MRI is obtained in all patients to determine the precise location, extent, and nerve of origin of the tumor as well as imaging characteristics of the tumor and the involvement of surrounding structure. The use of lower extremity and/or sphincter electrodes, were used for intraoperative neurophysiological monitoring of spontaneous electromyographic (EMG) activity during surgical dissection, manipulation of nerve(s), or tumor resection. The operative approach is determined in the previously discussed fashion. In the case of using an anterior approach the iliac vessels, lower aorta, and inferior vena cava are mobilized, pre-sacral space entered at the level of the promontory, and the avascular plane posterior to the mesorectum is developed caudally providing adequate exposure. Of note the he hypogastric plexuses and associated sympathetic trunks are identified and avoided. The remainder of the dissection is performed by avoiding traction on the surrounding tissues using the EMG as a guide to prevent injury.

Major sacral resection generally is reserved for patients with malignant lesions. In a retrospective analysis of bowel and bladder function in patients having major sacral resection in a single institution during a 10-year period, patients who had unilateral sacrectomy, normal bowel and bladder function was

retained in 87% and 89%, respectively. In patients who had bilateral S2–S5 nerve roots sacrificed, all had abnormal bowel and bladder function. In patients who had bilateral S3–S5 resection, normal bowel and bladder function was retained in 40% and 25%, respectively. In patients who had bilateral S4–S5 resection, with preservation of the S3 nerves bilaterally, normal bowel and bladder function was retained in 100% and 69%, respectively. In patients who had asymmetric sacral resections, with preservation of at least one S3 nerve root, normal bowel and bladder function was retained in 67% and 60%, respectively. These results show that unilateral resection of sacral roots or preservation of at least one S3 root in bilateral resection preserves bowel and bladder function in the majority of patients [18, 28].

Outcomes

Malignant Lesions

Studies identifying the outcomes regarding resection of malignant lesions the are limited as the nature of these diseases due to their rarity and thus reports are limited to case series and observational studies. The biologic nature of the lesion and the extent of resection and therefore varies

among studies; however, the recurrence rates are higher and outcomes poorer among tumors classified as malignant. The risk of local recurrence after a poor oncologic resection approaches 70% with decreased long-term survival prospects. In the Glasgow et al. [1] report, seven of seven patients with malignant presacral tumors developed recurrence of their disease despite adequate resection and had a median survival of 61 months. Lev-Chelouche et al. [21] reported an 80% complete resection rate in 12 patients with presacral tumors other than chordomas with a 67% local recurrence and 50% survival. Wang et al. [29] reported their series of 22 patients with malignant retrorectal tumors that included five chordomas and seven leiomyosarcomas. No preoperative biopsy was obtained and no neoadjuvant therapy was attempted. Despite the use of postoperative chemotherapy and radiotherapy on selected patients the 5-year survival rate was 41%.

Chordomas are the most common malignant presacral tumor. The most significant prognostic factor for patients with chordoma is the surgical margins. Despite adequate surgical resection a significant proportion of patients develop locally recurrent disease indicating the need for improved adjuvant therapies. In a report published in 1985, Jao et al. [2] reported a 5-year survival rate of 75% for chordomas; the same group has recently found a 5- and 10-year survival rate of 80% and 50%, respectively, for these patients. McMaster et al. [17] evaluated 400 cases of chordomas reported to nine population-based registries within the National Cancer Institute's Surveillance, Epidemiology and End Result (NSEER) program over a 22-year period from 1973 to 1995. The 5- and 10-year survival rate for sacral chordomas in the NSEER database was found to be 74% and 32%, respectively, and more likely represents the population-based incidence and outcome of these lesions.

Benign Lesions

Despite limited reports the overall survival for completely resected benign lesions is uniformly associated with low recurrence rates and complete remission. However, incompletely resected tumors are prone to recurrence. In a series by Glasgow et al. [1] none of the 26 patients

with benign presacral tumors developed recurrence after a median follow-up of 22 months. Lev-Chelouche et al. [21] reported a 100% survival and no recurrences after complete resection in their experience with 21 benign presacral tumors.

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