22
Presacral Tumors

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Key Concepts

- Unless contraindicated, presacral tumors should be surgically excised because of the risk of malignancy.
- MRI should be performed to characterize the lesions and to plan surgery.
- Lesions that are below sacral level S4 can be excised through a posterior/perineal approach.
- Complete, non-piecemeal excision is critical to avoiding recurrence or infection.

Introduction

Retrorectal masses are a group of lesions that encompass a wide spectrum of disease processes, ranging from congenital lesions (with varied malignant potential) to inflammatory disease processes and overt malignancy [1, 2]. In general, retrorectal tumors are extremely rare, with the incidence of the tumors varying in the reported literature [1–3]. The Mayo Clinic has reported that retrorectal tumors represent 1 in 40,000 hospital admissions [4]. Diagnosis of these lesions is usually incidental on physical exam or on imaging studies, as symptomatology is usually vague [4]. Imaging remains the key to preoperative characterization of these lesions in addition to preoperative planning. Although the majority of patients will have undergone computed tomography (CT scan), magnetic resonance imaging (MRI) is an essential element in the preoperative evaluation. Although the role of preoperative biopsy has been a source of debate, because of the fear of recurrence at or seeding of biopsy tracts, there is a good single institutional data to support its selective use [5].

Anatomic Considerations

The presacral or retrorectal space is not a true space but rather a potential space (see Chap. 1). It is a unique area in that it represents a developmentally critical location where several types of embryological distinct cell lines converge for the final steps prior to the completion of ontogeny. It is these changes that produce the variety of benign and malignant and solid and cystic growths that can occur in this space [1]. The retrorectal space is the area posterior to the rectum, but, more specifically, its superior extent is the pelvic peritoneal reflection, its lateral limits are the ureters and iliac vessels, posteriorly it is defined by the sacrum, and anteriorly it is defined as the posterior wall of the rectum. The inferior border is the levator complex and the coccygeal muscles (Figure 22-1) [3].

The retrorectal space presents a multitude of challenges to the surgeon, and this subset of procedures is not recommended for those uninitiated in pelvic surgery. The sacral nerve rootlets are located in this retrorectal space, and thus injury to and sacrifice of these structures can have substantial implications on rectoanal and sexual function. In cases requiring the unilateral sacrifice of all of the sacral nerve rootlets, the patient will likely retain normal anorectal and sexual function. Bilateral sacrifice of the third sacral nerve rootlet will usually result in fecal incontinence [6, 7].

Classification

Histology/Pathology

The classification of presacral masses encompasses a wide variety of etiologies and tissue types (Table 22-1). The classification of these retrorectal lesions, first elaborated by Uhlig and Johnson in 1975, divides these lesions broadly into congenital, acquired, neurogenic, osseous, and “others” [3]. Understanding the various subtypes, disease behavior, and malignant potential is essential to tailor treatment regimens.

Congenital Lesions

Congenital lesions represent two-thirds of all retrorectal lesions, which are thought to arise from various combinations of the three embryonic cell layers. These congenital

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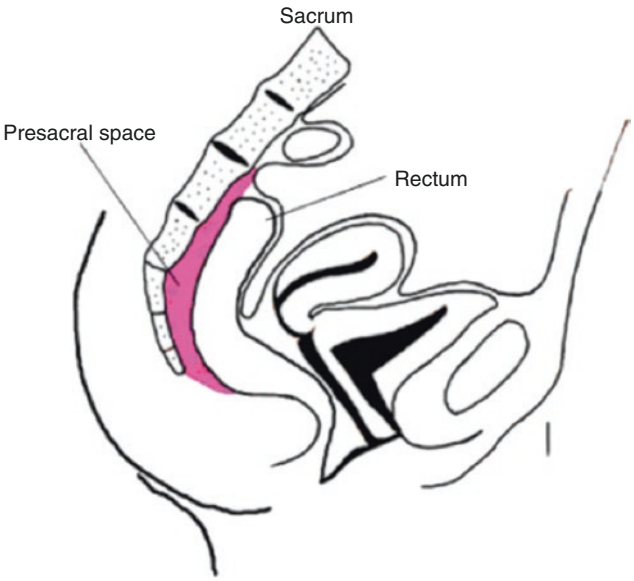

FIGURE 22-1. Location of the presacral space (Reprinted with permission from Ghosh J, Eglinton T, Frizelle FA, Watson AJ. Presacral tumours in adults. Surgeon. 2007 Feb;5(1):31–8 © 2007, Elsevier Ltd. [30]).

lesions can be cystic or solid [8]. In general, these lesions are more common in females than males [4, 8].

Dermoid and Epidermoid Cysts

Dermoid and epidermoid cysts are lined with squamous epithelial cells and may contain various skin appendages such as hair or nails (Figure 22-2). These lesions are thought to arise from the ectodermal layer in embryonic development. Patients can have a postanal dimple or sinus that can be mistaken for an abscess and errantly drained [9, 10]. This also accounts for the high rate of infection of these cysts.

Enterogenous

Unlike dermoid and epidermoid cysts, enterogenous cysts are multilocular. Enterogenous cysts arise from the endoderm of the primitive hindgut. These lesions can also undergo malignant degeneration.

Tailgut Cysts

Tailgut cysts are also referred to as retrorectal cystic hamartomas which arise from the persistence of the hindgut. Rectal duplication cysts contain all of the layers of the intestinal tract (Figure 22-3). Rectal duplication cysts can also undergo malignant change [11].

TABLE 22-1. Classification of retrorectal tumors

Congenital
Developmental cyst
Epidermoid cyst
Dermoid cyst
Teratoma
Teratocarcinoma
Chordoma
Anterior meningocele
Rectal duplication
Adrenal rest tumors
Neurogenic tumors
Neurofibroma
Neurilemmoma
Ependymoma
Ganglioneuroma
Neurofibrosarcoma
Malignant peripheral nerve sheath tumors
Osseous
Osteoma
Osteogenic sarcoma
Sacral bone cyst
Ewing's tumor
Giant-cell tumor
Chondrosarcoma
Chondromyxosarcoma
Miscellaneous
Metastatic or recurrent disease
Lipoma
Fibroma
Leiomyoma
Hemangioma
Desmoid
Liposarcoma
Leiomyosarcoma
Fibrosarcoma
Endothelioma
Granuloma
Perineal abscess
Fistula

Teratomas

Teratomas also contain cells from all three germ layers, but, more importantly, these lesions are true neoplasms. They can contain both solid and cystic components. Up to 10 % of these lesions contain cancer, and thus aggressive extirpation should be pursued. Because of the diverse germ cell layers, these lesions can become squamous cell carcinomas, rhabdomyosarcomas, or anaplastic tumors [1]. These tumors can contain tissues from almost any organ system including digestive and respiratory or bony tissue. Similar to other congenital lesions, teratomas are more common in females. They are also more common in children than adults. Factors that are associated with malignant degeneration and/or recurrence are incomplete resection and resections where the coccyx is not removed [1, 12].

FIGURE 22-2. CT image of an epidermoid cyst.

FIGURE 22-3. CT image of rectal duplication cyst.

Chordomas

The most common malignant tumor of the presacral space is the sacrococcygeal chordoma. These tumors arise from what is believed to be vestigial notochord tissue. These lesions are more common in male patients under 40 with an incidence of about 0.08 per 100,000. These lesions can occur almost anywhere on the spinal cord but are most commonly found in the presacral area. The patients present with vague symptomatology including low back pain. The 5- and 10-year survival rates are 67 and 40 %, respectively, and though surgery remains a mainstay of treatment, it is associated with a high recurrence rate [13].

Anterior Sacral Meningocele

These lesions arise from protrusions of the dural sac through a defect in the sacrum. The classic radiologic finding of the "scimitar sign" can often be seen on plain films. Patients often have vague symptomatology including headaches related to postural changes and Valsalva [4, 14]. Magnetic resonance imaging usually easily characterizes these lesions, and percutaneous biopsy should be avoided for fear of bacterial contamination of the cerebrospinal fluid and iatrogenic meningitis.

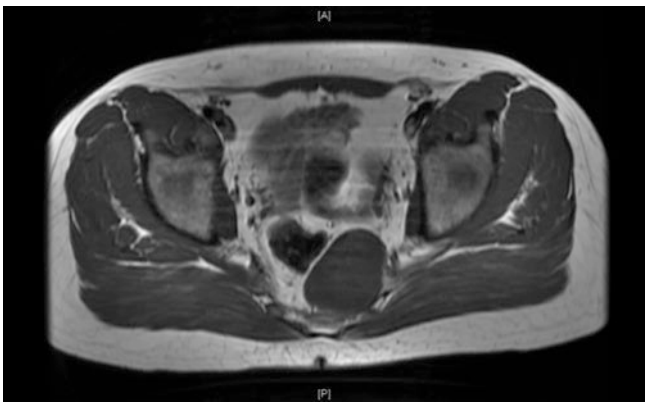

FIGURE 22-4. MRI image of a ganglioneuroblastoma.

Neurogenic Tumors

Neurogenic tumors represent about 10 % of all retrorectal tumors (Figure 22-4). They arise from peripheral nerves and include neurofibromas, schwannoma, ganglioneuroma, neuroblastomas, ganglioneuroblastoma, and ependymoma. Ependymomas are the most common of these tumors [4, 15]. Differentiation between benign and malignant variants can be difficult, and these tumors can produce significant neuropathy as a presenting symptom.

Osseous Lesions

Osseous lesions include giant-cell tumors, osteoblastoma, aneurysmal bone cysts, osteogenic sarcoma, Ewing's sarcoma, myeloma, and chondrosarcomas. These lesions represent 10 % of all retrorectal tumors. These may be the most aggressive of all the retrorectal tumors and can be very locally destructive and have pronounced metastatic potential [1, 16].

Diagnosis

History and Physical

Because of the location of these tumors in the presacral space, the symptomatology tends to be vague and nonspecific. Many of these tumors are diagnosed incidentally on rectal examination, and in fact 97 % of presacral lesions are palpable on digital rectal examination [4]. Many patients will have lower back pain or pelvic pain; however, in general,

there is not a plethora of common findings. Patients with congenital cysts/tumors may have a postanal sinus; however, the most likely etiology of a postanal sinus is perianal fistulous disease. Therefore the lesions may be diagnosed after several unsuccessful attempts at treatment of a perianal fistula that usually culminates in cross-sectional imaging as the true manner of identification. Patients with advanced tumors can have constipation, sexual dysfunction, urinary incontinence, and other leg and gluteal symptoms related to local extension and mass effect. Neurologic exams with attention to these symptoms in addition to gluteal and lower extremity dysfunction allow for preoperative documentation of these defects and aid in assessing the locally invasive nature of the lesion.

Imaging Studies

The preoperative assessment of a retrorectal tumor should include intraluminal evaluation of the rectum via flexible sigmoidoscopy. Understanding the extent of the mass of the tumor on the rectum and the ability to assess the mucosal integrity of the rectum are both important elements of the preoperative preparation. Flexible sigmoidoscopy allows for a better assessment of the upper and lower extents of these tumors, in addition to the relationship of the lesion to the sphincter complex. Endorectal ultrasound (ERUS) can be utilized to assess the relationship of tumors to the muscular layers of the rectum and the anal sphincters; despite the fact that majority of the lesions are well circumscribed, the subset of tumors that are not can be quite locally advanced and destructive. ERUS can also allow a very preliminary assessment of sacral bony destruction by tumors.

Plain films have limited utility but can sometimes demonstrate osseous destruction of the sacrum or calcifications within the tumor itself. In patients with anterior sacral meningocele, the classic "scimitar sign" can often be seen on plain films, but usually cross-sectional imaging is a requirement for confirmation. Magnetic resonance imaging (MRI) with gadolinium is the imaging modality of choice for retrorectal tumors. MRI is critical in the management of these tumors by facilitating accurate diagnosis, determining the anatomic extent of the lesion, and selecting the optimal surgical approach. Information that can be extracted from an MRI is much more granular in comparison to other modalities, including key elements such as location, size, morphology, margins, and interface [17]. MRI determination of the location of lesions in relation to the sacral vertebral bodies allows for planning of abdominal versus posterior versus combined surgical approaches. Characterization of the lesion as solid or cystic is easily achievable via MRI, but subtle nodularity or septation of these lesions allows for further characterization of these lesions into their various subtypes (Figure 22-5). Threatened margins can be more easily identified via MRI

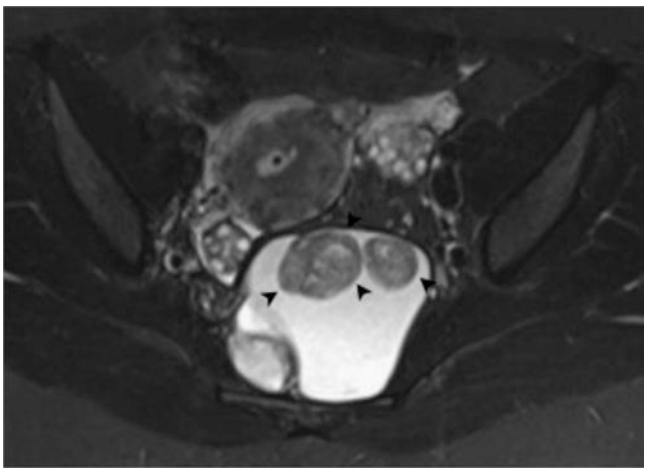

FIGURE 22-5. MRI of presacral cyst. T2-weighted imaging of an epidermoid cyst shows a bilobulated cystic lesion with pools of keratin debris (*arrows*) inside the larger cyst (Reprinted Loock MT, Fornès P, Soyer P, Rousset P, Azizi L, Hoeffel C. MR imaging features of nongynaecologic cystic lesions of the pelvis. Clin Imaging 2013;37(2):211–8 © 2013 Elsevier Ltd, with Permission from Elsevier. [31]).

such as bony erosion, or invasion of tumors and pelvic side wall invasion are more clearly definable. Arterial and venous anatomy is seen in much greater detail. What MRI excels at in comparison to CT scan is defining invasion of the muscular walls of the rectum, particularly in cases of sacrococcygeal chordoma [18]. These details, in total, make multimodality and multispecialty planning for operative interventions requiring en bloc resection of the rectum, partial sacrectomy, and arterial reconstruction or endovascular techniques much easier.

Preoperative Biopsy

Biopsy of presacral tumors presents a twofold question. First, is biopsy associated with a higher rate of local recurrence? Second, does biopsy have proven utility in the management of presacral tumors, i.e., does it changes the management? In general, biopsy of cystic lesions should only be undertaken in situations where there is some question of the characterization of the lesion *after* a high-quality MRI interpreted by an experienced radiologist. To be clear, it is universally acknowledged that biopsy of presacral lesions via the transrectal or transvaginal route is strongly discouraged, as it is possible to infect a sterile cystic lesion. In addition, biopsy via these routes necessitates either partial or complete proctectomy or vaginectomy to remove the biopsy tract in continuity with the presacral tumor in order to prevent recurrence. Biopsy of a meningocele via any route should be avoided for fear of an infection of the cerebrospinal fluid and resultant meningitis.

Early work from several authors discouraged biopsy of these tumors for fear of local recurrence [19–21]. More recent data suggests that percutaneous biopsy of retrorectal tumors can be performed without an increased risk of recurrence. In a single institutional series of 87 patients, Messick et al. performed biopsy of 24 patients (28 %) prior to surgical extirpation with no postoperative tumor recurrences. In this same series, only 4 of the 24 patients underwent excision of their biopsy site, also without any reported recurrences [5]. In our current practice, we do not biopsy all solid presacral lesion and even fewer mixed solid or cystic lesions. There is a role for biopsy in unresectable, sizeable, or aggressive tumors such as Ewing's sarcoma or osteogenic sarcoma where preoperative radiation or chemotherapy could be of value for systemic or local control or to improve the likelihood of resectability. It is our current practice to excise the biopsy tract and site at the time of definitive surgery.

Management

Role of Preoperative Neoadjuvant Therapy

Retrorectal tumors can exhibit a diverse set of behaviors and can be quite large and locally advanced by the time they are diagnosed. In addition, the subset of pelvic sarcomas has fairly significant systemic metastatic potential. With this in mind, there is a definite role for neoadjuvant chemotherapy for some of these tumors. In cases of large locally advanced presacral tumors, where resectability is at issue, neoadjuvant radiotherapy may render some benefit in decreasing tumor size and increasing resectability.

Surgical Treatment

Unless the lesion is unresectable or there is evidence of systemic metastasis, presacral tumors should be resected, as 30–40 % of the lesions will be malignant and benign lesions can undergo malignant change. Furthermore, approximately up to 10 % of cystic lesions will become chronically infected and can complicate any planned operative intervention [2–5].

Preoperative Planning

The key to preoperative planning is understanding the extent of the resection field. In patients that have direct invasion of the muscular wall of the rectum, proctectomy must be anticipated. In cases of bony invasion, partial sacrectomy is planned. Pelvic sidewall involvement may necessitate intraoperative radiotherapy and vascular or ureteric reconstruction. The assembly of a multispecialty team of colorectal, urologic, neurosurgical, orthopedic, vascular, and plastic surgeon is a prerequisite for many of these undertakings.

FIGURE 22-6. Transverse incision marked, as well as the sacrum and coccyx, for a posterior approach.

Surgical Approach

The location, the morphology, and the impingement or involvement of other pelvic structures dictate the operative approach. In general, a well-circumscribed presacral lesion whose uppermost extent can be palpated on digital rectal examination can usually be approached via a posterior approach. Several single institutional series also seem to share consensus where the S4 level is the line of division between abdominal and posterior approaches [3, 5, 22–25]. In lesions above the S4 level of the spine, a purely abdominal approach can be considered, while lesions below S4 can be approached posteriorly. Lesions spanning both above and below are best approached via a combined abdominal and posterior approach.

Posterior Approach

Patients are given a mechanical, cathartic bowel preparation the night before in preparation for this procedure. After intubation, the patient is placed in the prone jackknife position atop a large bolster. The rectum is irrigated with a dilute solution of betadine and saline; after this the buttocks are taped apart. While the incision for this procedure is usually described as a midline incision from the lower portion of the sacrum down to the anus, while yet others describe a transverse incision (Figure 22-6), our practice is different. The technique used by our group involves making a curvilinear incision; the incision is placed just to the left of the lower portion of the sacrum and carried in a curvilinear caudad direction around the lateral aspect of the coccyx toward the midline and the intergluteal fold just below the tip of the coccyx. Once the intergluteal fold is reached (below the tip of the coccyx), the incision is extended downward in the midline to a point approximately 2–3 cm short of the anal orifice (Figure 22-7). The reason for this type of incision is that the

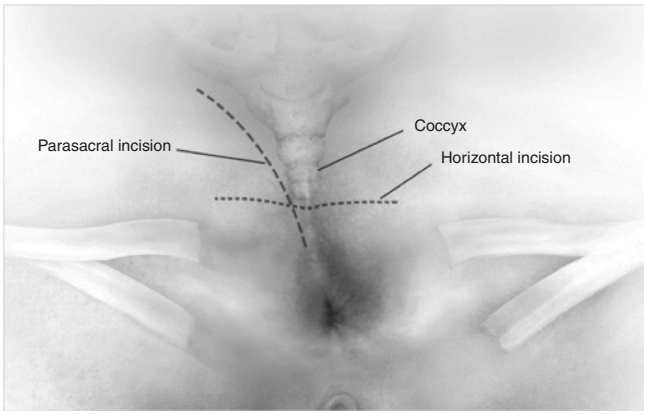

FIGURE 22-7. Posterior approach for the removal of a presacral tumor and placement of incision. The patient is in prone jackknife position, and the incision can either be horizontal on the anococcygeal ligament or curvilinear to the left of the lower sacrum/coccyx and into the intergluteal fold (With permission from Ludwig KA, Kalady MF. Transsacral approaches for presacral cyst: rectal tumor. Operative Techniques in General Surgery 2005;7:3-126-136 © 2005 Elsevier Ltd. [32]).

curvilinear incision allows for easier access to the lateral aspect of the coccyx, which is routinely removed.

Once the skin incision is completed, the dissection is deepened until the coccyx and the anococcygeal ligament are visualized. The anococcygeal ligament is divided, and extreme care is taken to identify the posterior aspect of the sphincter complex in order to preserve it. After this, the coccyx is freed along both sides of its lateral aspects and then the coccyx is removed (Figures 22-8 and 22-9). It is our practice to routinely remove the coccyx for two reasons. The first is that many of the congenital cysts are tethered to and originate at the coccyx, and it is thought that preserving the coccyx results in a higher recurrence rate [3, 26]. The second reason we routinely remove the coccyx is that removal allows for better visualization of the retrorectum and the mass, which creates a somewhat wider operative field, which facilitates removal of these tumors. This technique allows for intact removal of the lesion and reduces the likelihood of inadvertent perforation of the lesion, which is linked to a higher rate of recurrence and infection.

The lesion can be usually "shelled out" by dissecting it off of the sacrum and then slowly rolling the most proximal aspect of the tumor toward the incision from a cephalad to a caudad direction and then slowly dissecting it off the rectum (Figures 22-10, 22-11, and 22-12). There is quite often a feeding vessel that is encountered on the proximal aspect of many of these lesions that needs to be controlled; this can be safely and easily accomplished with a long handled bipolar energy source. After the removal of the tumor (Figure 22-13), the operative field is submerged beneath irrigant, and a proctoscope is used to insufflate the rectum to check for an air leak and assure that the rectum has not been violated. The soft tissue and the incision are closed in multiple layers over a closed suction drain (Figure 22-14).

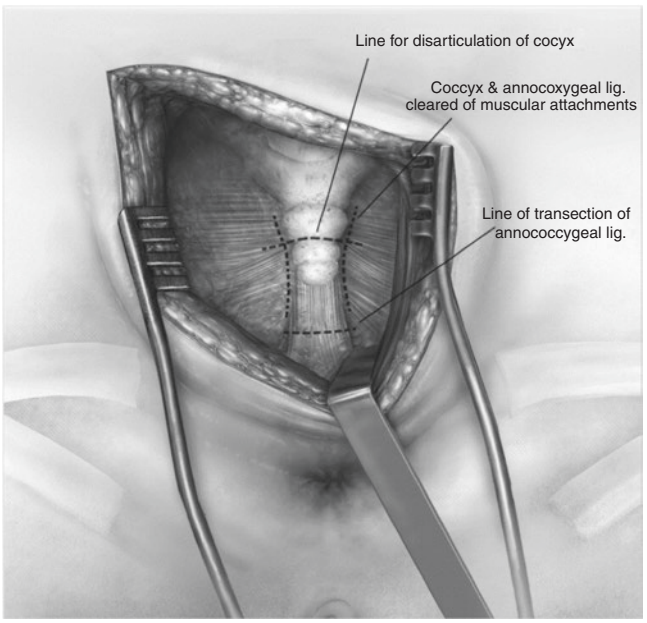

FIGURE 22-8. The anococcygeal ligament is divided, and the coccyx is subsequently cleared of its lateral attachments and removed; this facilitates dissection along the sacrum (With permission from Ludwig KA, Kalady MF. Transsacral approaches for presacral cyst: rectal tumor. Operative Techniques in General Surgery 2005;7:3-126-136 © 2005 Elsevier Ltd. [32]).

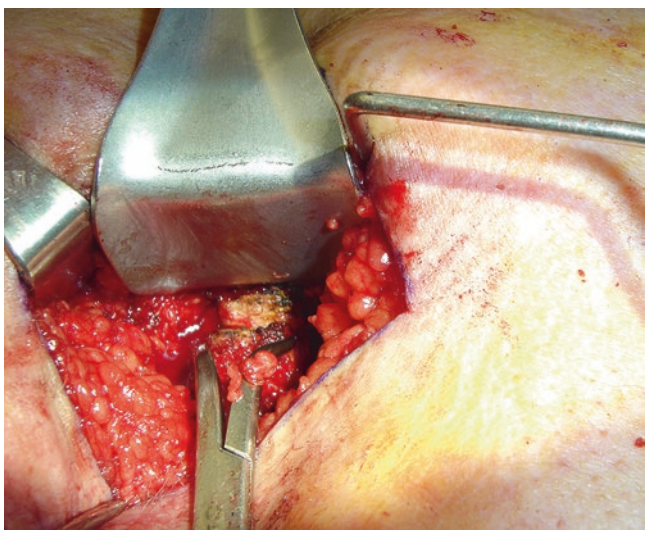

FIGURE 22-9. The tip of the coccyx is removed en bloc with the specimen.

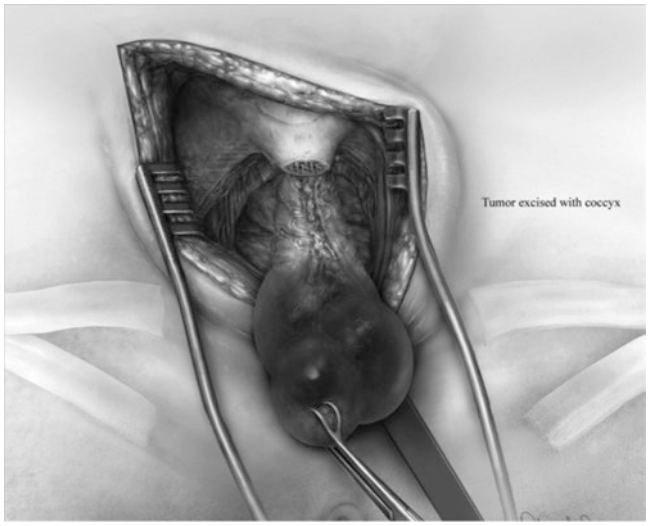

FIGURE 22-10. Now with access to the presacral space, the surgeon can carefully dissect the cyst off of the sacrum and "roll" it toward himself from cephalad to caudad (With permission from Ludwig KA, Kalady MF. Transsacral approaches for presacral cyst: rectal tumor. Operative Techniques in General Surgery 2005;7:3-126-136 © 2005 Elsevier Ltd. [32]).

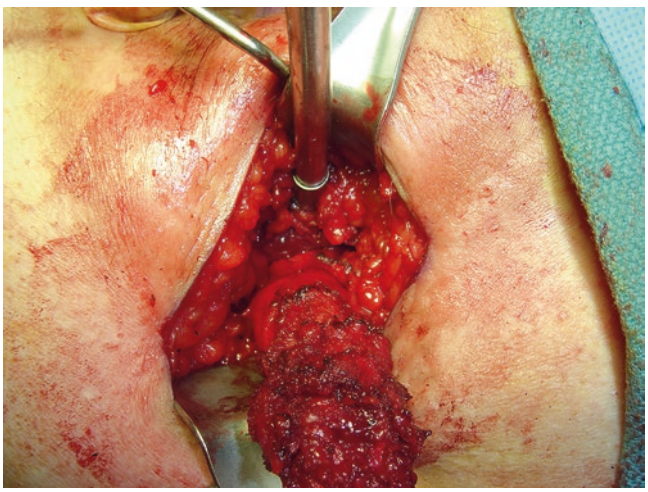

FIGURE 22-11. The presacral mass is mobilized off the rectal wall.

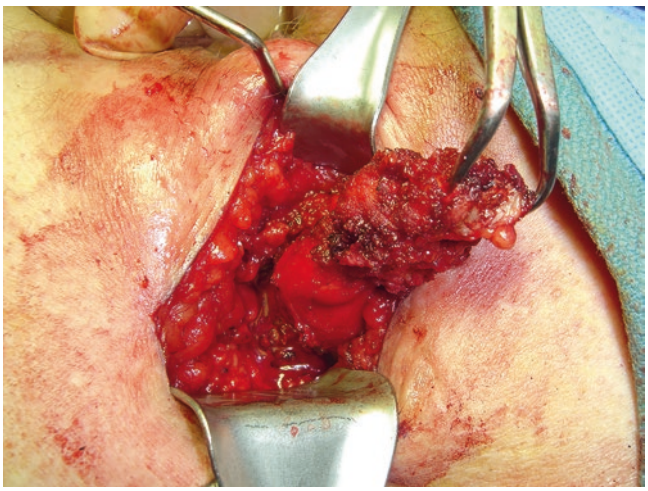

FIGURE 22-12. Side view of the coccyx tip and mass en bloc dissection from the rectal wall.

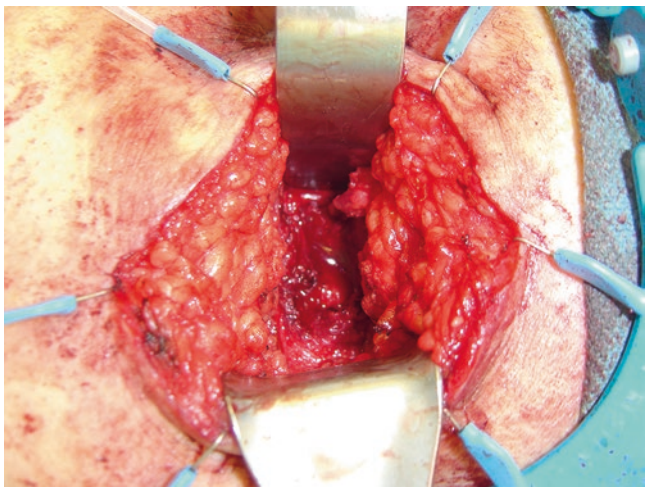

FIGURE 22-13. With the specimen out, a large cavity is present, and the posterior rectal wall can be visualized.

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 patients should be placed in lithotomy so that if a perineal or posterior approach is needed, access to the area has been anticipated and facilitated. Ureteric stents can be placed in bulky, high tumors.

A standard midline incision is utilized, and a thorough examination of all quadrants of the abdomen should be performed to assure that there are no metastases. The sigmoid colon is mobilized along the white line of Toldt, and the presacral space is entered at the level of the sacral promontory in the same fashion as a total mesorectal excision. The left and right hypogastric nerves are identified and preserved. The rectum is pulled forward. The lesion can then be dissected away from the mesorectum with preservation of the rectum.

When the lesion is large (Figure 22-15) and the space is small or visualization posteriorly is less than ideal, there are several maneuvers that can aid with visualization and facilitate posterior dissection. The lateral stalks can be taken down to the level of the levators, and the rectum can be mobilized anteriorly to the pelvic floor; in addition the superior rectal artery can be divided at the level of the sacral promontory to take tension of the mesentery, and the root of sigmoid and left colon mesentery can be detached from the retroperitoneum and aorta all the way to the root of the inferior mesenteric

FIGURE 22-14. The incision is closed in multiple layers over a suction drain.

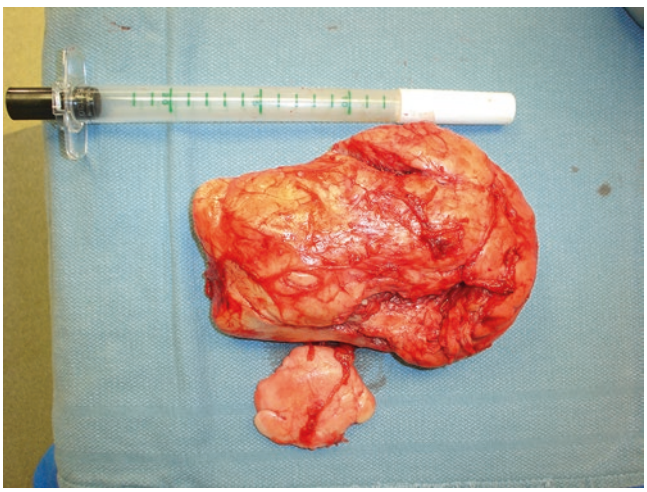

FIGURE 22-15. A large presacral mass.

artery. These maneuvers, in combination, allow the rectum to be pulled up and out of the pelvis to allow easier visualization of the dissection planes and better retraction. There is often a feeding vessel to the tumor in the midline, and ligating the middle sacral vessels can often help stem potential blood loss. The tumor is then dissected anteriorly off of the rectum and posteriorly off of the sacrum and laterally off of the sidewalls. In situations where tumor is densely adherent to the posterior rectum, a proctectomy should be performed for en bloc removal with the tumor. Most of the time, a stapled low colorectal anastomosis can be performed, but on occasion a hand-sewn coloanal anastomosis may be necessary.

If the internal iliac artery or vein needs to be sacrificed, communication with the anesthesiologist in advance of ligation is ideal, as the sacrifice of these vessels can sometimes be associated with large-volume bleeding misadventures and blood products should be on hand. If the involvement of these vessels is identified preoperatively, catheter-based venous or arterial embolization can be considered in advance of surgery.

In situations where the lowermost portion of the tumor cannot be reached from the abdominal approach, there are two options: the first is to place the patient in high lithotomy and proceed via a posterior approach, and the second is to close the abdomen and place the patient in prone jackknife position. The visualization and performance of the posterior approach with the patient placed in high lithotomy are challenging, and it is our preference to close the abdomen and subsequently flip the patient to the prone jackknife position. The visualization is superior, and the incidence of cyst perforation is much lower. In addition, partial sacrectomy of the lower sacrum including nerve rootlets can be accomplished via this approach when necessary.

In patients where the tumor is quite large and the anticipated pelvic or perineal defect is quite large, there are several options for tissue interposition or reconstruction. A transabdominal rectus abdominis myocutaneous (TRAM) flap can be transposed into the pelvis to fill fairly impressive defects. For more modest defects, less morbid options may be V-Y fasciocutaneous flap closure and unilateral or bilateral gracilis transposition.

Closed suction drainage of the pelvis and the perineum should be performed in these patients.

Outcomes

Malignant Lesions

In a single institutional report, Messick et al. reported on 87 patients who had excision of retrorectal tumors; the overall recurrence rate was 16 %, with the recurrence rate of malignant tumors being 30 %. In this particular series, all of the recurrences in the malignant cohort were distant, and the median survival was 47.5 months [5]. In series where the tumors are extracted piecemeal or the tumors are violated, the recurrence rate can be as high as 65 % or higher [21, 27].

Although retrorectal sarcomas tend to be locally advanced, half of all patients have reasonable long-term survival. Dozois et al. reported a median survival of 4.7 years with survival at 2 and 5 years reported at 75 and 55 %, respectively [24]. In other data acquired from the Surveillance, Epidemiology, and End Results (SEER) program, McMaster and colleagues reported on sacral chordomas, which represent 29 % of all chordomas. In this study, the 5- and 10-year survival rates for sacral chordomas were 74 and 32 %, respectively [13]. Another series of 39 patients with malignant retrorectal tumors by Cody and associates reported a 5- and 10-year survival of 50 and 37 %, respectively. In this series, 38 % of these tumors were chordomas and 15 % were neurogenic tumors [2].

Finally, Wang and colleagues reported a series of 45 patients with presacral tumors, in which 48 % of the patient had malignant tumors. Incomplete resections were associated with inferior outcomes. The 5-year survival rate for malignant tumors was 41 % [28].

Benign/Cystic Lesions

The Cleveland Clinic series of tumors located strictly below S4 reported that 95 % were approached via a posterior approach only, and the local recurrence rate for the benign cohort was 11 %. Coccygectomy was performed in 51 % of patients; however, there was no difference in the recurrence rates between patients that underwent coccygectomy and those that did not [5]. Glasgow et al. published a series of 34 patients with retrorectal tumors where 26 patients had benign tumors. At a mean follow-up of 22 months, none of the patients in the benign group had recurred [29]. Another series by Jao and associates reported on a series of presacral lesions, of which 66 were benign retrorectal tumors. Of note, there was a 15:1 ratio of females to males. The overwhelming majority of the lesions were resected through a posterior approach, with 10 of 66 patients experiencing a recurrence [4].

Conclusion

Presacral tumors represent a diverse set of tumors with a strong predominance of the congenital cysts. The symptomatology of these tumors is often vague, and early diagnosis is an unusual event. Many of these tumors are found on digital rectal examination, and many are found incidentally on imaging or in the workup of nonspecific symptomatology. The tumors can have solid, cystic, or mixed features. Surgical extirpation is recommended for almost all tumors, as they a third can contain a malignancy and they can undergo malignant degeneration. MRI is essential in preoperative planning, as in a multidisciplinary team. Biopsy of lesions should only be reserved for lesions that are thought to be unresectable or metastatic. The majority of lesions that are below the S4 level can be approached via a posterior approach. Larger or more locally advanced lesions may require both an abdominal and perineal approach with en bloc resections of a portion of the sacrum or rectum. Lesions that are resected completely without disruption have a better prognosis than those that are not.

References

1. Hobson KG, Ghaemmaghami V, Roe JP, Goodnight JE, Khatri VP. Tumors of the retrorectal space. Dis Colon Rectum. 2005;48(10):1964–74.
2. Cody 3rd HS, Marcove RC, Quan SH. Malignant retrorectal tumors: 28 years' experience at Memorial Sloan-Kettering Cancer Center. Dis Colon Rectum. 1981;24(7):501–6.
3. Uhlig BE, Johnson RL. Presacral tumors and cysts in adults. Dis Colon Rectum. 1975;18(7):581–9.
4. Jao SW, Beart Jr RW, Spencer RJ, Reiman HM, Ilstrup DM. Retrorectal tumors. Mayo Clinic experience, 1960-1979. Dis Colon Rectum. 1985;28(9):644–52.
5. Messick CA, Hull T, Rosselli G, Kiran RP. Lesions originating within the retrorectal space: a diverse group requiring individualized evaluation and surgery. J Gastrointest Surg. 2013;17(12):2143–52.
6. Gunterberg B, Kewenter J, Petersen I, Stener B. Anorectal function after major resections of the sacrum with bilateral or unilateral sacrifice of sacral nerves. Br J Surg. 1976;63(7):546–54.
7. Gunterberg B, Petersen I. Sexual function after major resections of the sacrum with bilateral or unilateral sacrifice of sacral nerves. Fertil Steril. 1976;27(10):1146–53.
8. Bullard Dunn K. Retrorectal tumors. Surg Clin North Am. 2010;90(1):163–71. Table of Contents.
9. Singer MA, Cintron JR, Martz JE, Schoetz DJ, Abcarian H. Retrorectal cyst: a rare tumor frequently misdiagnosed. J Am Coll Surg. 2003;196(6):880–6.
10. Abel ME, Nelson R, Prasad ML, Pearl RK, Orsay CP, Abcarian H. Parasacrococcygeal approach for the resection of retrorectal developmental cysts. Dis Colon Rectum. 1985;28(11):855–8.
11. Springall RG, Griffiths JD. Malignant change in rectal duplication. J R Soc Med. 1990;83(3):185–7.
12. Hickey RC, Martin RG. Sacrococcygeal teratomas. Ann N Y Acad Sci. 1964;114:951–7.
13. McMaster ML, Goldstein AM, Bromley CM, Ishibe N, Parry DM. Chordoma: incidence and survival patterns in the United States, 1973-1995. Cancer Causes Control. 2001;12(1):1–11.
14. Williams B. Cerebrospinal fluid pressure changes in response to coughing. Brain. 1976;99(2):331–46.
15. Stewart RJ, Humphreys WG, Parks TG. The presentation and management of presacral tumours. Br J Surg. 1986;73(2):153–5.
16. Freier DT, Stanley JC, Thompson NW. Retrorectal tumors in adults. Surg Gynecol Obstet. 1971;132(4):681–6.
17. Hosseini-Nik H, Hosseinzadeh K, Bhayana R, Jhaveri KS. MR imaging of the retrorectal-presacral tumors: an algorithmic approach. Abdom Imaging. 2015;40(7):2630–44.
18. Sung MS, Lee GK, Kang HS, Kwon ST, Park JG, Suh JS, et al. Sacrococcygeal chordoma: MR imaging in 30 patients. Skeletal Radiol. 2005;34(2):87–94.
19. Luken 3rd MG, Michelsen WJ, Whelan MA, Andrews DL. The diagnosis of sacral lesions. Surg Neurol. 1981;15(5):377–83.
20. Bohm B, Milsom JW, Fazio VW, Lavery IC, Church JM, Oakley JR. Our approach to the management of congenital presacral tumors in adults. Int J Colorectal Dis. 1993;8(3):134–8.
21. Lev-Chelouche D, Gutman M, Goldman G, Even-Sapir E, Meller I, Issakov J, et al. Presacral tumors: a practical classification and treatment of a unique and heterogeneous group of diseases. Surgery. 2003;133(5):473–8.
22. Macafee DA, Sagar PM, El-Khoury T, Hyland R. Retrorectal tumours: optimization of surgical approach and outcome. Colorectal Dis. 2012;14(11):1411–7.
23. Sagar AJ, Tan WS, Codd R, Fong SS, Sagar PM. Surgical strategies in the management of recurrent retrorectal tumours. Tech Coloproctol. 2014;18(11):1023–7.
24. Dozois EJ, Jacofsky DJ, Billings BJ, Privitera A, Cima RR, Rose PS, et al. Surgical approach and oncologic outcomes

following multidisciplinary management of retrorectal sarcomas. Ann Surg Oncol. 2011;18(4):983–8.
25. Merchea A, Dozois EJ. Lesions originating within the retrorectal space. J Gastrointest Surg. 2014;18(12):2232–3.
26. Aktug T, Hakguder G, Sarioglu S, Akgur FM, Olguner M, Pabuccuoglu U. Sacrococcygeal extraspinal ependymomas: the role of coccygectomy. J Pediatr Surg. 2000;35(3):515–8.
27. Kaiser TE, Pritchard DJ, Unni KK. Clinicopathologic study of sacrococcygeal chordoma. Cancer. 1984;53(11):2574–8.
28. Wang JY, Hsu CH, Changchien CR, Chen JS, Hsu KC, You YT, et al. Presacral tumor: a review of forty-five cases. Am Surg. 1995;61(4):310–5.
29. Glasgow SC, Birnbaum EH, Lowney JK, Fleshman JW, Kodner IJ, Mutch DG, et al. Retrorectal tumors: a diagnostic and therapeutic challenge. Dis Colon Rectum. 2005;48(8): 1581–7.
30. Ghosh J, Eglinton T, Frizelle FA, Watson AJ. Presacral tumours in adults. Surgeon. 2007;5(1):31–8.
31. Loock MT, Fornès P, Soyer P, Rousset P, Azizi L, Hoeffel C. MR imaging features of nongynaecologic cystic lesions of the pelvis. Clin Imaging. 2013;37(2):211–8.
32. Ludwig KA, Kalady MF. Transacral approaches for prescral cyst: rectal tumor. Oper Tech Gen Surg. 2005;7: 3-126-136.