Presacral Tumors

21

Scott R. Kelley and Eric J. Dozois

Key Concepts

- The presacral (retrorectal) space is the location of a wide range of rare tumors with incidence as low as 1 in 40,000–60,000 hospital admissions. Discovery in asymptomatic patients is increasing due to expanded use of cross-sectional imaging.
- Advances in cross-sectional imaging and understanding of tumor biology have led to better outcomes for these challenging patients.
- MRI is the best imaging study to assist in diagnosis and operative planning.
- Most benign lesions have malignant potential and observation alone in some patients is acceptable when a dedicated surveillance protocol is in place.
- When performed appropriately and selectively, a CT-guided biopsy of the lesion may assist in management of solid and heterogeneous cystic lesions.
- The surgical principles that should guide a surgeon who manages these lesions are a function-sparing approach for benign lesions and an en bloc approach for malignant lesions.

Introduction

The presacral (retrorectal) space is a potential space and the location of a wide range of rare tumors. Reports from referral centers have indicated the incidence may be as low as 1 in 40,000–60,000 hospital admissions [1–4]. Detection is frequently delayed since patients are often asymptomatic until tumors reach considerable size. Advances in cross-sectional imaging and understanding of tumor biology have led to better outcomes for these challenging patients. Although most surgeons will encounter a patient with a presacral tumor in their career, few will have the opportunity to treat a large volume of these complex lesions. The care of these patients can be greatly optimized by an experienced multidisciplinary team (MDT).

Anatomic Considerations

Evaluation and management of presacral tumors require a thorough understanding of the anatomic relationships of the pelvic viscera, the bony confines of the pelvis, and the neuromuscular structures. Anteriorly the presacral space is bordered by the mesorectum, posteriorly by the anterior table of the sacrum, inferiorly by the levator muscles, and laterally by the lumbosacral plexus, ureters, and iliac vessels (Fig. 21.1).

Several important vascular and neural structures are located where presacral tumors occur. Injury to these may have important physiological, neurologic, and musculoskeletal consequences. Knowledge of anatomy of the thigh and lower extremity is also necessary in complex cases utilizing muscle or other soft tissue flap coverage. When sacrectomy is required, a multidisciplinary surgical team familiar with the anatomy of the sacrotuberous and sacrospinous ligaments, sciatic nerve, piriformis muscle, the thecal (dural) sac, and sacral nerve roots is necessary (Fig. 21.2a and b). Knowledge of sacral nerve root function is important in order to be able to counsel patients on potential functional sequelae that can influence their quality of life. Todd and colleagues evaluated bowel and bladder function in a group of patients following sacral resection. They found that if bilateral S2–S5 nerve roots were removed patients had complete loss of bladder and bowel function. If bilateral S3–S5 were removed, 40% had normal bowel function and 25% had nor-

Supplementary Information The online version of this chapter (https://doi.org/10.1007/978-3-030-66049-9_21) contains supplementary material, which is available to authorized users.

S. R. Kelley (✉) · E. J. Dozois
Mayo Clinic, Division of Colon and Rectal Surgery, Rochester, MN, USA
e-mail: kelley.scott@mayo.edu

S. R. Steele et al. (eds.), *The ASCRS Textbook of Colon and Rectal Surgery*, https://doi.org/10.1007/978-3-030-66049-9_21

mal bladder function. If bilateral S4–S5 were taken, 100% had normal bowel function and 69% had normal bladder function. If unilateral S1–S5 were taken, 87% had normal bowel function and 89% had normal bladder function [5]. If the S1 nerve root or sciatic nerve is resected foot drop can occur, severely impairing ambulatory function [6]. In addition to functional consequences, when high sacrectomy is performed, pelvic stability can be compromised if more than half of the S1 vertebral body is resected. Moreover, preoperative radiotherapy can increase the risk of stress fractures destabilizing the pelvic ring. When spinopelvic stability is a concern, patients undergo sacropelvic reconstruction with metallic fixation, bone grafting, or 3D printed titanium prostheses [7].

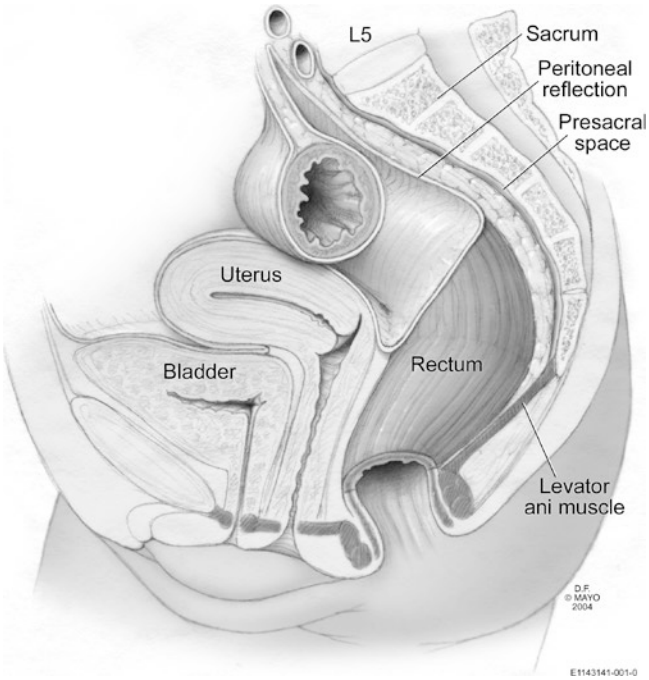

Fig. 21.1 Relationship of pelvic structures to presacral space. (Reused with permission of Mayo Foundation for Medical Education and Research, all rights reserved)

Clinical Presentations

Presacral tumors are often discovered incidentally during routine pelvic/rectal examination, or on imaging for other purposes [8]. If symptoms are present, pain is typically vague, of long duration, and in the pelvis, perineum, and/or low back. The vague nature of pain can make diagnosis difficult, and at times patients are referred to a psychiatrist when no obvious etiology is found on routine physical examination. Typically, pain is heightened by sitting and improved by standing or walking. Pain is more often associated with malignant lesions and can be an ominous sign [2]. Constipation, urinary and fecal incontinence, and sexual dysfunction are typically seen with sacral nerve involvement from advanced tumors. Leg and gluteal symptoms are often associated with extension and mass effect.

Occasionally patients complain of longstanding perineal/sacrococcygeal discharge and their symptoms may be confused with perianal fistulas or pilonidal disease. Singer and colleagues reported on seven patients with presacral cysts (six females, one male). All patients had previously been misdiagnosed and treated for pilonidal cysts, perirectal abscesses, fistula in ano, psychogenic disorder, proctalgia fugax, and posttraumatic or postpartum pain before the correct diagnosis was made. Patients underwent an average of

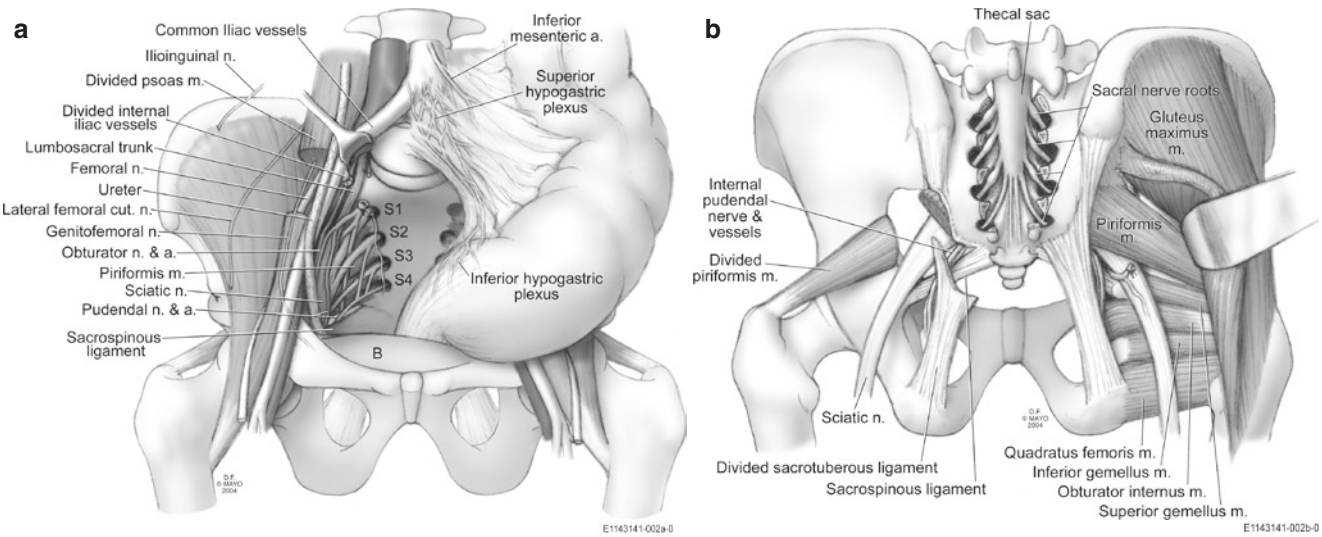

Fig. 21.2 (**a**) Anterior view of the pelvic anatomy. (**b**) Posterior view of the pelvic anatomy. (Reused with permission of Mayo Foundation for Medical Education and Research, all rights reserved)

4.1 prior operative procedures. All patients were successfully treated with resection through a parasacrococcygeal approach after the correct diagnosis was made [9]. Multiple unsuccessful attempts at treatment of anal fistula or pilonidal disease should alert the surgeon to the possibility of a presacral cystic lesion.

Physical Examination

Physical examination should focus on the perineum and rectum. In all but a very small percentage of patients' digital rectal examination will reveal the presence of an extra-rectal mass displacing the rectum anteriorly [2]. It also allows one to determine fixation to the rectal wall and relation to surrounding structures such as the prostate, vagina, and coccyx/sacrum. Evaluation for a post-anal dimple should also be performed. A rigid or flexible proctosigmoidoscopy should be completed to evaluate the mucosa and potentially the upper and lower extent of the tumor. The most common endoscopic appearance is normal mucosa with extrarectal mass effect. The presence of abnormal and/or inflamed mucosa is often suggestive of infection/prior infection or erosion into the rectal wall. Neurologic evaluation of musculoskeletal reflexes and sacral nerve function should be performed if clinically indicated.

Imaging Studies

Anterior/posterior and lateral radiographs are of limited utility, but if obtained can identify osseous expansion, destruction, and/or calcifications of soft tissue occupying masses. In patients with an anterior meningocele, the characteristic "scimitar sign" can often be seen on sacral views. Endorectal ultrasound can be performed at the same time as flexible sigmoidoscopy to assess for invasion of the rectal wall or anal sphincter complex.

Computed tomography (CT) and magnetic resonance imaging (MRI) have dramatically changed the way presacral tumors are evaluated. Both CT and MRI can distinguish between cystic, solid, or mixed (cystic and solid) tumors, and can determine if other pelvic structures (rectum, bladder, ureters, etc.) are involved. Each modality can also define the anatomic extent of the mass, facilitate an accurate diagnosis, and establish the optimal surgical approach (anterior, posterior, or combination). Given the high soft tissue resolution, MRI has become the gold standard imaging modality for evaluating presacral tumors [10, 11]. Magnetic resonance imaging is more sensitive than CT for determining associated cord abnormalities such as sacral nerve root involvement, foraminal encroachment, and dural sac compression [12]. The improved resolution of MRI more clearly defines bony involvement, pelvic sidewall invasion, arterial and venous anatomy, and invasion of surrounding structures (rectum, bladder, ureters, etc.) [11, 13]. Contrast enhanced MRI with gadolinium can also detect meningoceles, thus avoiding the risks associated with myelogram. CT or MR angiography/venography may provide additional information regarding vascular involvement. Preoperative diagnostic accuracy of MRI has been reported to be as high as 100%, with 50–88% sensitivity and 92–97% specificity for differentiating benign from malignant [14, 15]. Low T1 and high T2 signal, gadolinium enhancement, irregular or infiltrative margins, and lesions with heterogeneous and/or solid components are more often associated with malignancy [10, 14, 16].

Hosseini-Nik and colleagues developed an algorithmic approach for MR imaging of presacral tumors. They subdivided fat-containing masses into solid, cystic, or complex lesions. Solid lesions were further subdivided into well and ill defined. Non-fat containing masses were classified as solid or complex, and cystic. Cystic lesions were sub classified as unilocular or multilocular, and solid lesions were differentiated based on the presence or absence of sacral destruction. In addition, they also outlined an optimal MR imaging protocol. They recommend imaging of the pelvis utilizing 1.5 T or 3 T systems with multi-channel phased array torso coils. Multiplanar 2D or high spatial-resolution 3D T2-weighted (T2-W) pulse sequences, together with obliquely oriented 2D T2-W sequences along the long axis of the sacrum helps assess the relationship of the mass to the rectum, sacrum, sacral foramina, and nerve roots. Frequency-selective or inversion recovery fat-suppressed T2-weighted pulse sequences improves the dynamic range for T2-weighting and tissue contrast and also confirms the presence of macroscopic fat. Routine T1-weighted (T1-W) sequences with and without fat saturation examines for macroscopic fat and multiphasic contrast-enhanced acquisitions must be acquired for appropriate characterization. In-phase (IP) and out of phase (OP) T1 gradient-echo imaging is helpful in identifying intracellular lipid [11].

For patients with a presacral cystic lesion thought to be the source of a chronically draining sinus, a fistulogram may help clarify the diagnosis. Our study of choice in this situation is an MRI using fistula-protocol sequences similar to evaluations done in patients with perianal Crohn's disease or suspected occult cryptoglandular fistula-in-ano.

Preoperative Biopsy

Preoperative biopsies have been an ongoing topic of debate [8, 17–20]. Advances in high-resolution imaging have increased the ability to accurately diagnose presacral tumors without tissue [10, 14, 21]. The need for biopsy is predicated on whether the result will change preoperative or operative management. For example, the surgical approach and necessary margins differ significantly for neurofibroma as com-

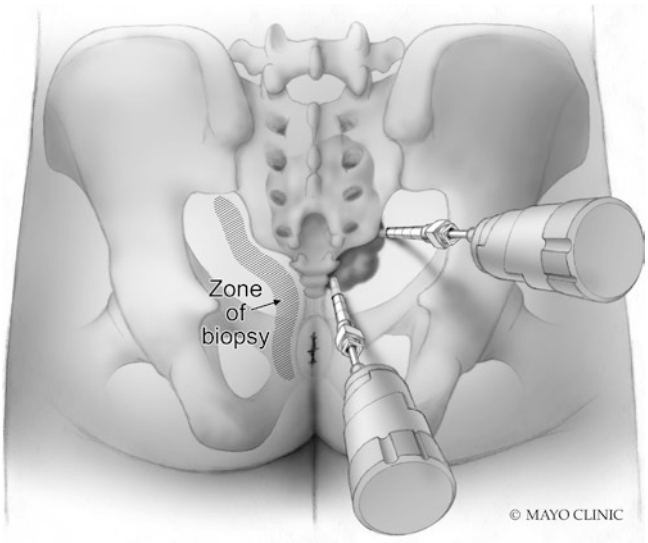

Fig. 21.3 Preoperative biopsy technique using CT guidance. (Reused with permission of Mayo Foundation for Medical Education and Research, all rights reserved)

pared to neurofibrosarcoma. If biopsy is necessary, the intended operative approach should be discussed with the interventional radiologist performing the procedure. Either transperineal or parasacral approaches may be considered depending on the anticipated field of resection (parasacral for planned sacral resection). Needle biopsies should be performed within the field of the proposed area of resection so the needle tract can be resected en bloc with the specimen to decrease the risk of seeding and local recurrence (Fig. 21.3). The external needle entry site can be spot tattooed to aid in future identification. Transperitoneal, transrectal, and transvaginal biopsies should be avoided. If histopathology reveals malignancy, en bloc complete or partial excision of the rectum or vagina with the presacral mass becomes necessary if the biopsy tract traversed one of these organs. In addition, biopsies of tumors in this area can result in bowel perforation, bleeding, and fistulas. Biopsy of cystic lesions increases the risk of secondary infection, and recurrence after resection. Inadvertent biopsy of a meningocele may lead to disastrous sequel such as meningitis and death.

Although historically surgeons have recommended complete excision of any biopsy tract, recently there has been discussion on leaving the biopsy tract in situ. Messick and colleagues reported on 87 presacral tumors, of which preoperative tissue biopsies were obtained in 24/87 (28%). Only 4/24 (17%) underwent excision of the biopsy site to evaluate the tissue (all negative for malignancy). The remaining 20 did not undergo surgical excision of the biopsy site and were followed clinically (or by radiographic imaging) with no reported tumor recurrences in the tract site [4]. Further investigation and evidence is needed before any definitive recommendations can be made on avoidance of biopsy site excision for malignant tumors.

Some patients benefit from neoadjuvant chemotherapy, radiation, hormonal, or immunotherapy, and a tissue diagnosis is often required to make that determination. Large pelvic desmoids can be removed more easily after reducing their size with neoadjuvant radiotherapy. Preoperative chemotherapy and radiotherapy improves outcomes with osseous tumors such as Ewing's sarcoma, osteogenic sarcoma, and neurofibrosarcoma. Hormonal therapy has shown benefit in reducing the size of giant aggressive angiomyxomas, and immunotherapy is showing promise for treating advanced chordomas [22–24]. Thus, when performed safely, a preoperative biopsy can optimize overall management [4, 19].

Classification

The presacral space is primarily composed of connective tissue, nerves, fat, and blood vessels. Totipotential cells that differentiate into three germ cell layers (endoderm, ectoderm, mesoderm) make up the complex embryologic potential space, which can lead to the development of a multitude of tumor types. The original classification described by Uhlig and Johnson divided tumors into congenital, neurogenic, osseous, and miscellaneous [29]. We have modified and updated the classification scheme to subcategorize tumors as malignant or benign, as this greatly impacts therapeutic approaches (Table 21.1).

Lesions found in the presacral space can be broadly classified as congenital or acquired and benign or malignant. Congenital lesions result from abnormalities in embryological processes (fusion of hindgut and proctodeum, degeneration of the notochord, etc.), whereas acquired tumors develop from remnant embryonic or other differentiated tissues found in the presacral space. In general, two-thirds are congenital, of which two-thirds are developmental cysts, with the next most common masses being neurogenic tumors [30]. Around 45–50% are malignant or have malignant change within them [2, 31]. Understanding the various subtypes, disease behavior, and malignant potential is essential to tailor treatment [16].

Epidermoid and Dermoid Cysts

Epidermoid and dermoid cysts (Fig. 21.4) are more common in females, tend to be well circumscribed, have a thin outer layer, result from defects during closure of the ectodermal layer, and are typically benign. They occasionally communicate with the skin surface creating a characteristic postanal dimple and are histologically composed of keratinized stratified squamous epithelium. The cysts are often misdiagnosed

Table 21.1 Mayo classification of presacral tumors

	Benign	Malignant
Congenital	Adrenal rest tumor	Chordoma
	Anterior sacral meningocele	Germ cell tumor
	Developmental cysts (dermoid, epidermoid [aka epidermal], enterogenous [aka rectal duplication], tailgut [aka cystic hamartomas/mucous secreting], teratoma)	Malignant developmental cysts
		Teratocarcinoma
Neurogenic	Ganglioneuroma	Ependymoma
	Neurofibroma	Ganglioneuroblastoma
	Schwannoma (aka neurilemoma)	Malignant schwannoma
		Neuroblastoma
		Peripheral nerve sheath tumors (aka neurofibrosarcoma)
		Primitive neuroectodermal
Osseous	Aneurysmal bone cyst	Chondrosarcoma
	Giant cell tumor	Ewing's sarcoma
	Osteoblastoma	Giant cell tumor
	Osteoma	Myeloma
	Simple bone cyst	Osteogenic sarcoma
		Plasmacytoma
		Reticulum cell sarcoma
		Spindle cell sarcoma
Miscellaneous	Aggressive angiomyxoma	Angiosarcoma
	Benign GIST	Carcinomasarcoma
	Benign hemagiopericytoma	Degenerated hamartoma
	Desmoid (aka fibromatosis)	Epithelioid sarcoma
	Ectopic kidney	Fibrosarcoma
	Endothelioma	Fibromyxoid sarcoma
	Fibroma	Histiosarcoma
	Hamartoma	Hydatid cyst
	Hemangioma	Leiomyosarcoma
	Leiomyoma	Liposarcoma
	Lipoma	Lymphoma
	Lipofibroma	Malignant desmoid
	Myelolipoma	Malignant hemagiopericytoma
	Pecoma	Malignant GIST
	Solitary fibrous tumor	Malignant solitary fibrous tumor
	Tuberculosis	Metastatic carcinoma
		Myeloliposarcoma
		Neuroendocrine tumors
		Rhabdomyosarcoma
		Small cell tumor
		Spindle cell tumor
		Squamous cell carcinoma
		Undifferentiated sarcoma

GIST gastrointestinal stromal tumor

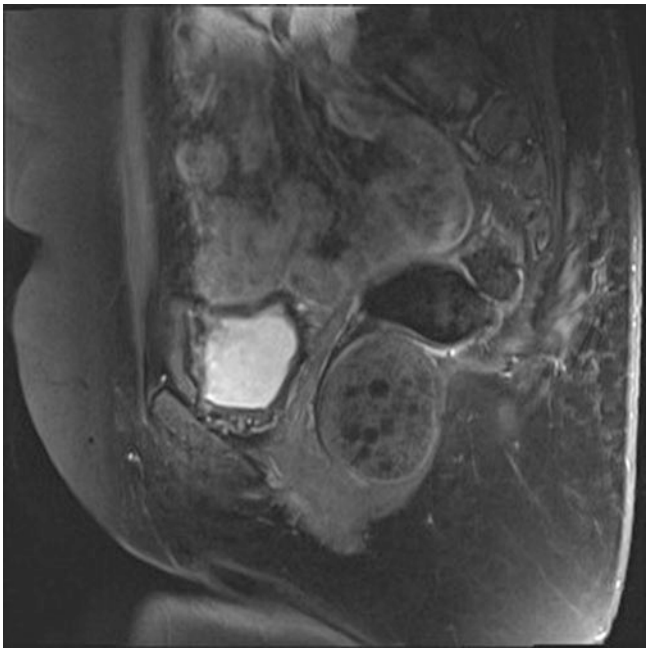

Fig. 21.4 MRI of dermoid cyst. (Reused with permission of Mayo Foundation for Medical Education and Research, all rights reserved)

as perirectal abscesses and can become infected with manipulation. Recurrently infected cysts have been associated with the development of squamous cell carcinoma [1]. Dermoid cysts may contain skin appendages (sweat glands, hair follicles, sebaceous cysts) whereas epidermoid cysts do not [9].

Tailgut Cysts

Tailgut cysts (cystic hamartomas/mucous secreting cysts) are congenital lesions arising from remnants of normally regressing postanal primitive hindgut and are more common in females. The cysts are lined with columnar epithelium and can morphologically resemble the adult or fetal intestinal tract [32]. They do not communicate with the rectal lumen and are often multiloculated or biloculated, and well defined and homogenous (Fig. 21.5). The presence of glandular or transitional epithelium differentiates them from epidermoid and dermoid cysts. Malignant transformation has been reported in some series [27, 33–35].

Enterogenous Cysts

Enterogenous cysts (rectal duplication cysts) are more common in women, often in communication with the rectum, and are thought to derive from the developing hindgut. Since they originate from endodermal tissue they can be lined with squamous, cuboidal, columnar, or transitional

epithelium. Unlike epidermoid, dermoid, and tailgut cysts they have a well-defined muscular wall with a myenteric plexus. The lesions tend to be multi-lobular with one dominant and smaller satellite cysts. In order to be classified as a rectal duplication cysts three anatomic criteria must be met: the cyst must be attached to the alimentary tract; it must be lined by a mucous membrane similar to that part of the gastrointestinal tract; and it must possess a smooth muscular coat. Enterogenous cysts are generally benign, but there are case reports describing malignant transformation [25, 36].

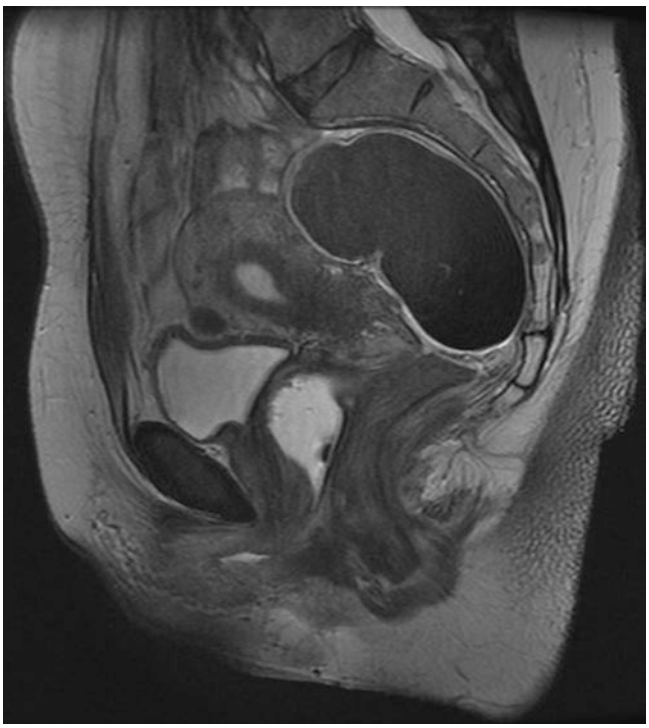

Fig. 21.5 MRI of tailgut cyst. (Reused with permission of Mayo Foundation for Medical Education and Research, all rights reserved)

Teratomas

Sacrococcygeal teratomas are neoplasms that may include all three germ layers (totipotential cells), can contain both solid and cystic components, and are more common in the pediatric age group and females. These tumors can contain tissues from almost any organ system including digestive, nervous, respiratory, and skeletal [37]. Histologically, tumors are referred to as either mature or immature, which reflects the degree of cellular differentiation. The more recognizable the elements (hair, bone, teeth) the more likely the tumor is to be benign, although all should be viewed as potentially malignant. Cystic components are typically benign whereas solid components are more often associated with malignant degeneration. The tumors can reach considerable size and diagnosis is often delayed (Fig. 21.6a–c). Teratomas are often associated with anomalies of the vertebra, urinary tract, and anorectum [38]. The rate of malignancy correlates strongly with age, being much less common beyond the second decade [20, 39]. In infants only 7% of girls and 10% of boys presented with malignancy prior to 2 months, whereas the rates can be as high as 48% and 67%, respectively, after 2 months of age [40]. Malignant degeneration can occur in adults, and incomplete or intralesional resection increases the likelihood of malignant degeneration [41–43]. Because of the diverse germ cell layers these lesions can transform into squamous cell carcinoma (ectodermal origin), rhabdomyosarcoma (mesenchymal origin), or anaplastic (indeterminate cell of origin) tumors [3].

Chordomas

Sacrococcygeal chordomas are the most common malignant tumor of the presacral space. The lesions are more common in male patients and rarely encountered in those less than 30 years of age [2, 44]. They are thought to arise from vesti-

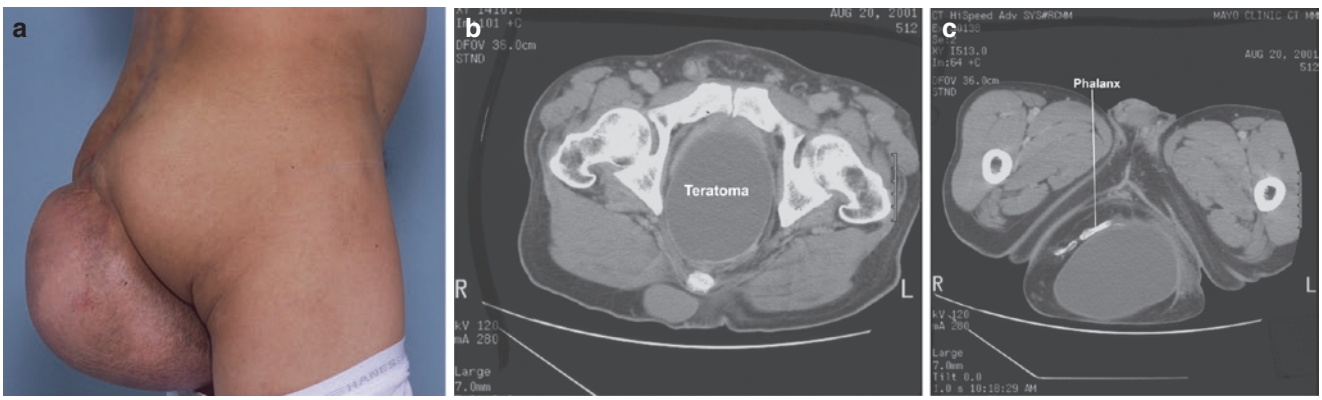

Fig. 21.6 (**a**) Massive cystic teratoma with sacral appendage. (**b**) CT of intrapelvic portion. (**c**) CT of extrapelvic portion with fully developed phalanx. (Reused with permission of Mayo Foundation for Medical Education and Research, all rights reserved)

gial notochord tissue, which embryologically extends from the base of the occiput to the caudal limit in the embryo. The lesions can occur almost anywhere on the spinal cord but are most commonly found in the pheno-occipital region at the base of the skull and the sacrococcygeal region in the pelvis [45, 46]. Symptoms are often vague and include low back, pelvic, and buttock pain, which is aggravated by sitting and alleviated by standing. As a result of the vague symptomology diagnosis is often delayed and the tumors can reach considerable size and result in constipation, fecal and urinary incontinence, and sexual dysfunction. Centrally chordomas contain extracellular mucin and can be soft, firm, or gelatinous. The tumors often contain areas of hemorrhage and can invade or destroy bone and soft tissues and distend into adjacent regions. Local and distant recurrence rates have been documented as 43% and 22%, respectively, and 5- and 10-year survival rates are 67% and 40% [47]. Resection with negative margins is the treatment of choice [47–50].

Meningoceles

Anterior sacral meningoceles arise from protrusions of the thecal sac through a defect in the sacrum, contain cerebrospinal fluid, and can be seen in conjunction with presacral cysts and lipomas. The classic radiographic finding is the "scimitar sign" (sickle-shaped sacrum/hemisacral agenesis), which is a unilateral well-marginated, crescent-shaped defect in the lateral sacrum (Fig. 21.7). Symptoms can include headaches related to postural changes and the Valsalva maneuver (straining/coughing), low back and pelvic pain, constipation/defecatory dysfunction, dyspareunia, and urinary urgency, retention, or incontinence [51]. Anterior sacral meningoceles can be associated with other congenital anomalies including urinary tract and/or anal malformations, uterine and/or vaginal duplication, tethered spinal cord, and spina bifida. These lesions should not be biopsied due to the risk of bacterial contamination of the cerebrospinal fluid and development of iatrogenic meningitis [52]. Surgical treatment consists of obliterating the communication between the subarachnoid space and herniated sac, detethering the spinal cord, and resecting the congenital tumor [53].

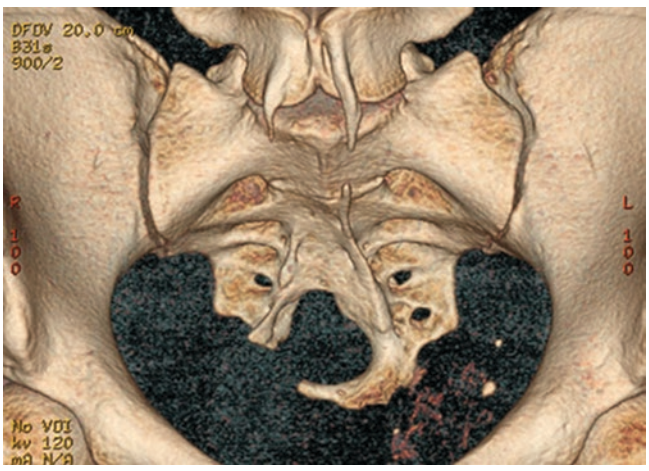

Fig. 21.7 CT pelvis scimitar sign. (Reused with permission of Mayo Foundation for Medical Education and Research, all rights reserved)

Neurogenic Tumors

Neurogenic tumors arise from the peripheral pelvic nerve plexus (Fig. 21.8a and b), make up approximately 10–15% of all presacral masses, and typically affect younger patients (median age 38). Although the vast majority (>90%) are benign, at times differentiating benign from malignant tumors can be difficult without a tissue biopsy. Schwannomas and ependymomas are the two most commonly encountered lesions [2, 26, 48]. Presenting symptoms can include neuropathies and low back and pelvic pain. Benign and malignant tumors have a high local recurrence rate, and survival for malignant tumors is poor. Early detection and aggressive surgical intervention are necessary to improve outcomes. With the use of a nerve-sparing technique a function-preserving resection can be safely completed with an overall improvement in symptoms [54, 55]. The goal is sacral nerve root preservation, but sacrifice may be required for extended resections for malignant tumors.

Osseous Tumors

Osseous lesions that grow into the presacral space make up less than 10% of presacral tumors and can arise from bone, cartilage, fibrous tissue, and marrow. They are more commonly found in males and half are malignant at the time of diagnosis [18]. Osseous tumors can reach considerable size and cause significant local destruction. They have pronounced metastatic potential, with pulmonary being the most common [56]. Although benign, giant cell tumors of the sacrum can metastasize to the lung and transform to a fulminate malignant variant, which has a very poor prognosis [57].

Miscellaneous Lesions

Miscellaneous masses in the presacral space can include heterogeneous pathologies including carcinoids, gastrointestinal stromal tumors, dermoids, angiomyxomas, metastatic deposits, ectopic kidneys, and hematomas [58].

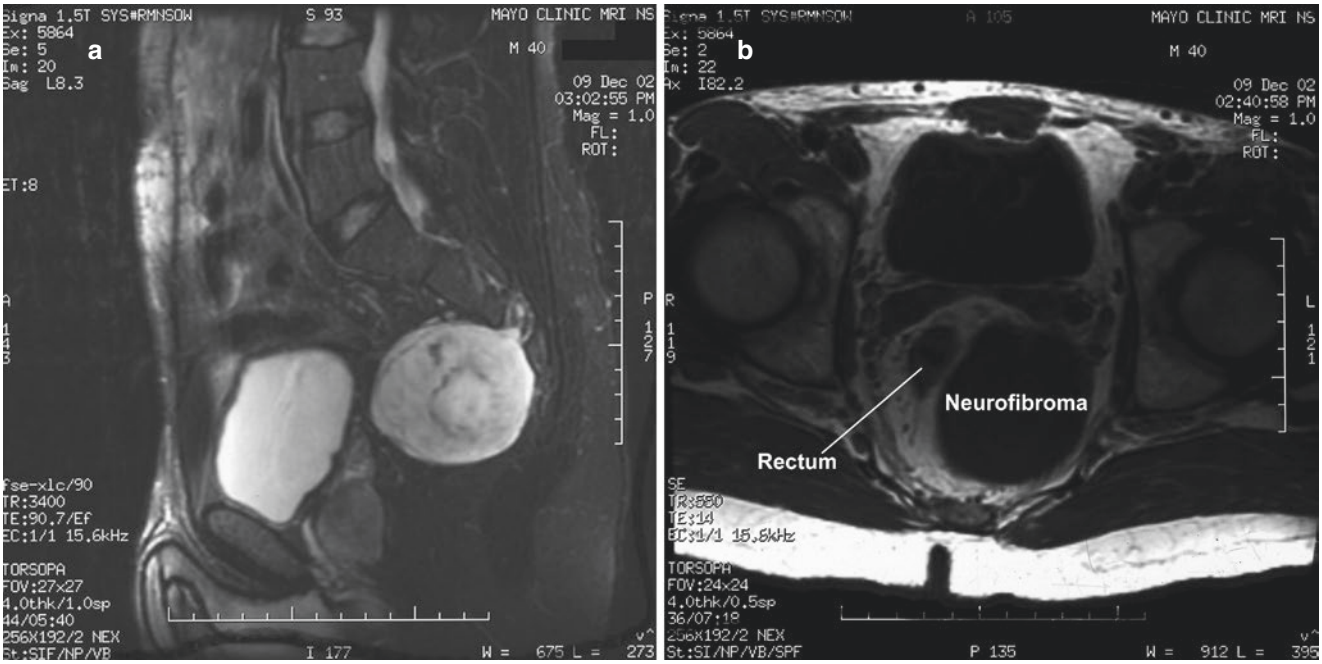

Fig. 21.8 (**a**) T2 weighted pelvic MRI of neurofibroma extending from the third sacral foramina. (**b**) T1 weighted pelvic MRI of neurofibroma. (Reused with permission of Mayo Foundation for Medical Education and Research, all rights reserved)

Currarino Syndrome

Surgeons seeing patients with presacral tumors should be familiar with Currarino syndrome. Currarino syndrome, described in 1981 is a rare congenital malformation associated with three main features: sacral malformation (agenesis or sickle shape), hindgut anomaly, and presacral tumor [59]. It is an autosomal dominant disorder linked to mutations in the HLXB9 gene, although sporadic cases have been described [59, 60]. To date, 43 heterozygous mutations have been reported [61]. As a result, patients can present with variable phenotypes including spinal cord anomalies (tethered cord, thickened filum, syrinx), genitourinary malformations, anorectal and gynecological anomalies, and presacral lesions. More than one presacral lesion can occur in the same patient [62]. The most commonly associated presacral mass is dermoid cyst, although teratomas have been identified in 25–40% of cases. Malignancy in Currarino syndrome is rare and only a small number of adult (19–45 years old) patients with malignant teratoma have been described [39, 63]. An abnormal looking sacrum on imaging is often the tip off that a patient has Currarino syndrome.

Management

The recommendation for treating presacral tumors has historically been surgical resection. Operative indications include known malignancy, concern for future malignant transformation, alleviation of symptoms, and a consistent increase in size (which may make future resection more risky). Small tumors can be addressed independently by colon and rectal surgeons specifically trained to manage these lesions. Larger lesions, or those associated with neuromusculoskeletal structures, are best managed by a multidisciplinary team that can bring specialty expertise to decision-making and assist in a safe surgical approach. For locally adherent malignant tumors, en bloc removal of adjacent organs, soft tissue, and bone is the goal of oncologic resection. At our institution, we have established a decision-making algorithm to guide the management of presacral tumors (Fig. 21.9a and b). The principles that should guide the surgical team include a function-sparing approach for benign lesions and an en bloc oncologic approach for malignant lesions.

There is recent literature supporting nonoperative surveillance (serial imaging) for small cystic lesions without symptoms or suspicious radiologic features [28], although the proof for advisability of this approach needs further investigation. Hopper and colleagues followed six cystic lesions with serial imaging for a median of 20 months (range, 5–66). Interval imaging ranged from every 6 months to every 2 years. At last follow-up, four (67%) were noted to be stable in size [28]. In our own practice, we consider an observational approach for small (<5 cm), asymptomatic neurogenic tumors.

Multidisciplinary Team

An experienced multidisciplinary team is critical for optimal outcomes in patients with complex presacral tumors

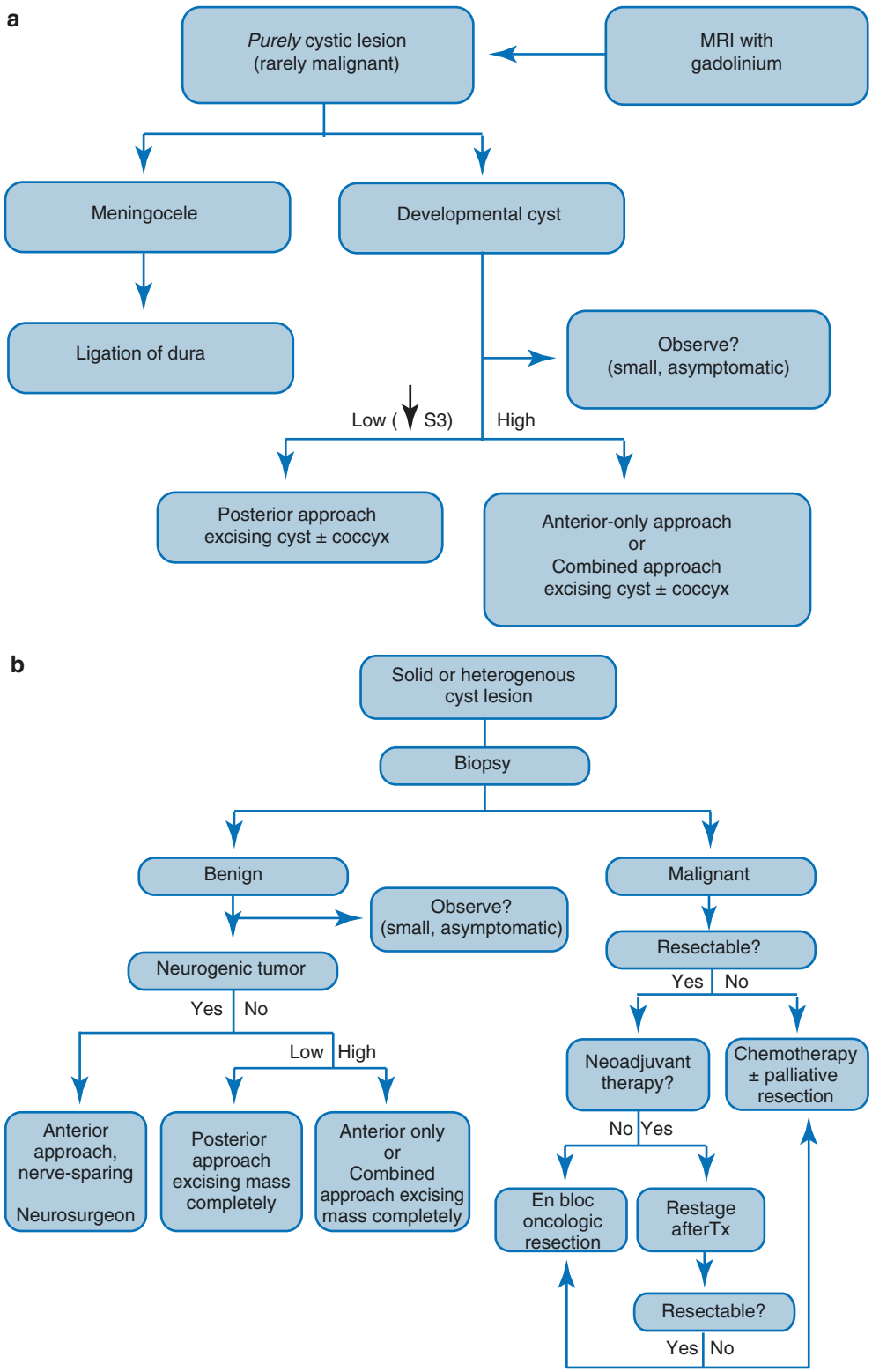

Fig. 21.9 (**a**) Proposed treatment algorithm for purely cystic lesions. (**b**) Proposed treatment algorithm for solid or heterogenous cystic lesions. (Reused with permission of Mayo Foundation for Medical Education and Research, all rights reserved)

[64]. The team may consist of surgeons from colorectal, orthopedic oncology, spine, neurosurgery, urology, vascular, and plastic surgery, as well as medical oncology, radiation oncology, musculoskeletal radiologists, and anesthesiologists with special expertise in complex presacral tumors [26, 65]. A formal discussion at multidisciplinary team conferences is essential for perioperative planning and treatment.

Neoadjuvant Therapy

Although many malignant presacral tumors, such as chondrosarcomas and chordomas, are poorly responsive to radiotherapy and chemotherapy, there are a number which are responsive. The addition of neoadjuvant therapy can decrease tumor size, increase resectability, and potentially decrease the rate of local recurrence. Compared to postoperative administration, the irradiation treatment field in the preoperative setting is smaller and results in less morbidity. Ewing's and osteogenic sarcomas are often associated with metastasis and neoadjuvant chemotherapy is a cornerstone of therapy. Providing chemotherapy in a neoadjuvant setting allows for treatment of micrometastatic disease prior to surgery, as well as decreases delays in chemotherapy treatment that may occur should the patient suffer postoperative complications.

The use of neoadjuvant therapy for presacral sarcomas has been extrapolated from protocols for treating soft tissue sarcomas. Radiation therapy has been shown to decrease local recurrence following resection of both retroperitoneal and extremity sarcomas [66, 67]. Others have shown chemotherapy, with and without radiation, trends toward an improved survival and decreases local and distant relapses for extremity and retroperitoneal sarcomas [26, 68–70].

Due to small case series and heterogeneity in patient populations, it is unclear if patients with malignant cysts benefit from neoadjuvant chemoradiotherapy. We have used this approach in patients with malignant cysts that have either squamous cell carcinomas or adenocarcinomas within them, with the rationale that chemoradiotherapy works well in patients with adenocarcinomas of the rectum and squamous cancers of the anus.

Preoperative Considerations

Optimizing patients for surgery is of paramount importance. When possible, anemia should be improved, protein calorie malnutrition enhanced, and debility reduced. For patients presenting in a debilitated state, social work consultation prior to surgery is important for postoperative rehabilitation and care planning. To decrease intraoperative bleeding, preoperative selective coil embolization of feeding vessels may be useful in some patients with large vascular tumors like hemangiopericytomas (Fig. 21.10a–c) [71].

For complex tumors, the multidisciplinary team should review films and operative planning together before surgery. Significant bleeding can occur with complex resections, and the blood bank should be alerted ahead of time to make sure adequate product is available. An operating room team (anesthesiologists, nurses, assistants, etc.) comfortable with complex pelvic surgery is needed for these procedures.

Surgical Approach

The location, involvement of other pelvic structures, and surgeon experience dictates the operative approach. For tumors superior to the S3–4 sacral bodies, a purely abdominal (anterior) approach should be pursued, while lesions entirely inferior to the S3–4 sacral bodies can be approached through a posterior sacral incision. For tumors extending both proximal and distal to the S3–4 sacral bodies, a combined anterior and posterior approach is often utilized (Fig. 21.11) [10, 21, 65].

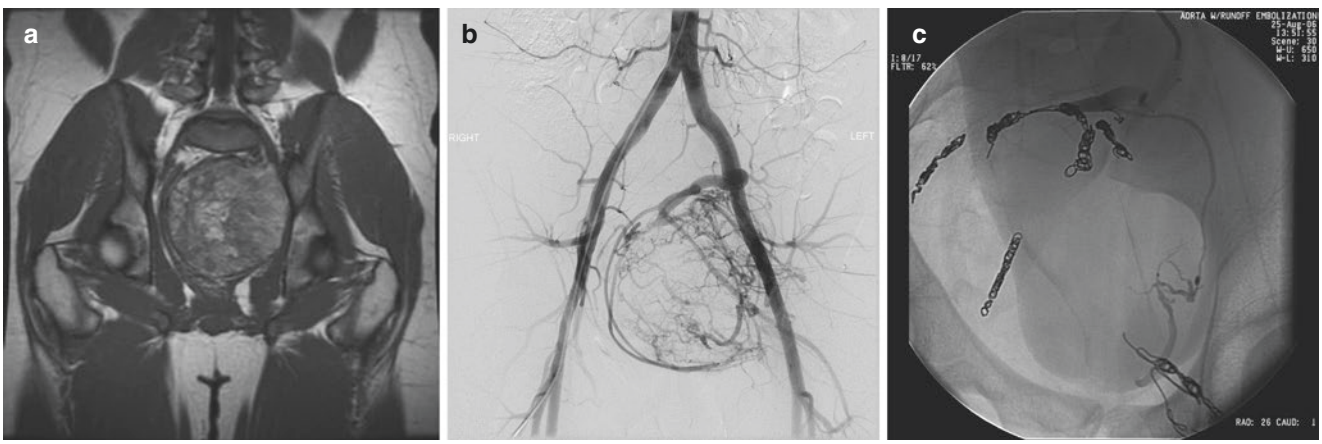

Fig. 21.10 (**a**) MRI of hemangiopericytoma. (**b**) Angiogram of hemangiopericytoma. (**c**) Post-coil embolization of hemangiopericytoma. (Reused with permission of Mayo Foundation for Medical Education and Research, all rights reserved)

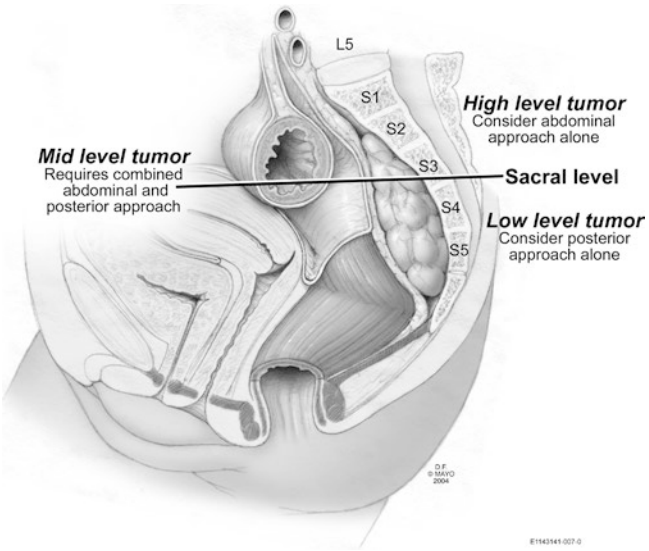

Fig. 21.11 Relationship of tumor to sacral level and proposed approach. (Reused with permission of Mayo Foundation for Medical Education and Research, all rights reserved)

Posterior Approach

The patient is placed in a prone jackknife position and the buttocks are taped apart (Fig. 21.12a). Depending on surgeon experience and preference, an incision (midline, parasacral, paracoccygeal, transverse, and curvilinear to the left of the lower sacrum/coccyx and into the intergluteal fold) is created. The dissection is carried down to the distal sacrum/coccyx and through the anococcygeal ligament, taking care to avoid damage to the sphincter complex. The centrally decussating muscle fibers of the levator muscle (levator plate region) are removed from the tip of the coccyx allowing entry into the presacral space. A pseudocapsule is often encountered and helps facilitate a safe dissection from surrounding tissues including the rectum. A coccygectomy can be performed to facilitate exposure and resection of larger tumors or ones tethered to the coccyx (Fig. 21.12b). The surgeon may double-glove the non-dominant hand, and with the index finger in the anal canal and lower rectum, push the lesion outward to assist with dissection of the tumor from the posterior wall of the rectum (Fig. 21.12c). If necessary, the lower sacrum and/or coccyx can also be excised en bloc with the tumor. If there is concern for local invasion a portion of the rectal wall may need to be excised and the defect closed in layers, otherwise the rectum should be left intact. Rectal integrity can be evaluated with a rigid or flexible endoscope and an air leak test performed by submerging the open operative field with irrigation.

There has been ongoing debate regarding the utility of coccygectomy for every low presacral tumor. Some authors advocate that coccygectomy improves exposure and decreases the risk of recurrence, as the coccyx may harbor a nidus of totipotential cellular remnants that may later evolve into a recurrent cyst [31, 38, 42]. However, multiple studies demonstrating low recurrence rates without coccygectomy support the idea that routine coccygectomy is unnecessary and potentially adds morbidity to patients. Singer et al. did not perform a coccygectomy in six of the seven patients (86%) with benign lesions in their study and saw no difference in recurrence [9]. Mathis and colleagues performed a coccygectomy in 7/28 patients, all of whom underwent resection of presacral tailgut cysts, and reported only a single recurrence (4%) [27]. Messick et al. performed a coccygectomy in 51% of the patients in their series (44/87) and did not appreciate a difference in recurrence for those who did (14%) and did not (20%) undergo resection of the coccyx [4]. We favor coccygectomy in patients with sacrococcygeal teratomas which are uniformly adherent to the coccyx and likely have the highest risk of recurrence if any cyst components are left behind.

Abdominal and Combined Anterior and Posterior Approach

For tumors completely above S3–S4 with no sacral involvement a transabdominal approach can be utilized. If the upper pole of the tumor extends above S3 a combined anterior and posterior approach is usually indicated. Patients can be placed in a variety of positions including supine, synchronous (modified dorsal lithotomy), and modified "sloppy" lateral (Fig. 21.13a–c). For larger tumors bilateral ureteral stents can be placed after induction.

Depending on tumor characteristics (benign/malignant, local invasion) and surgeon experience and comfort either an open or minimally invasive approach is utilized. The abdomen is carefully explored to rule out disseminated disease and other pathology. The lateral attachments of the sigmoid colon are mobilized and the presacral space entered. Ureters and superior hypogastric nerves are identified and preserved. The mesorectum is dissected off the presacral fascia to the level of the upper extension of the tumor. The rectum is mobilized to facilitate identification and exposure of the pathologic area of interest. If the tumor can be safely separated circumferentially from the rectum, presacral fascia, and lateral pelvic sidewalls the dissection can proceed until the mass is removed. Bulky tumors make visualization difficult and may preclude safe dissection between the lesion and rectum. In this event the rectum can be excised en bloc with the tumor, and intestinal continuity reestablished following removal. If the tumor invades both S2 or S3 nerve roots excision of the rectum en bloc with the mass and creation of a permanent colostomy is appropriate since the patient will be rendered incontinent. During mobilization of malignant tumors, no structures attached to the specimen should be

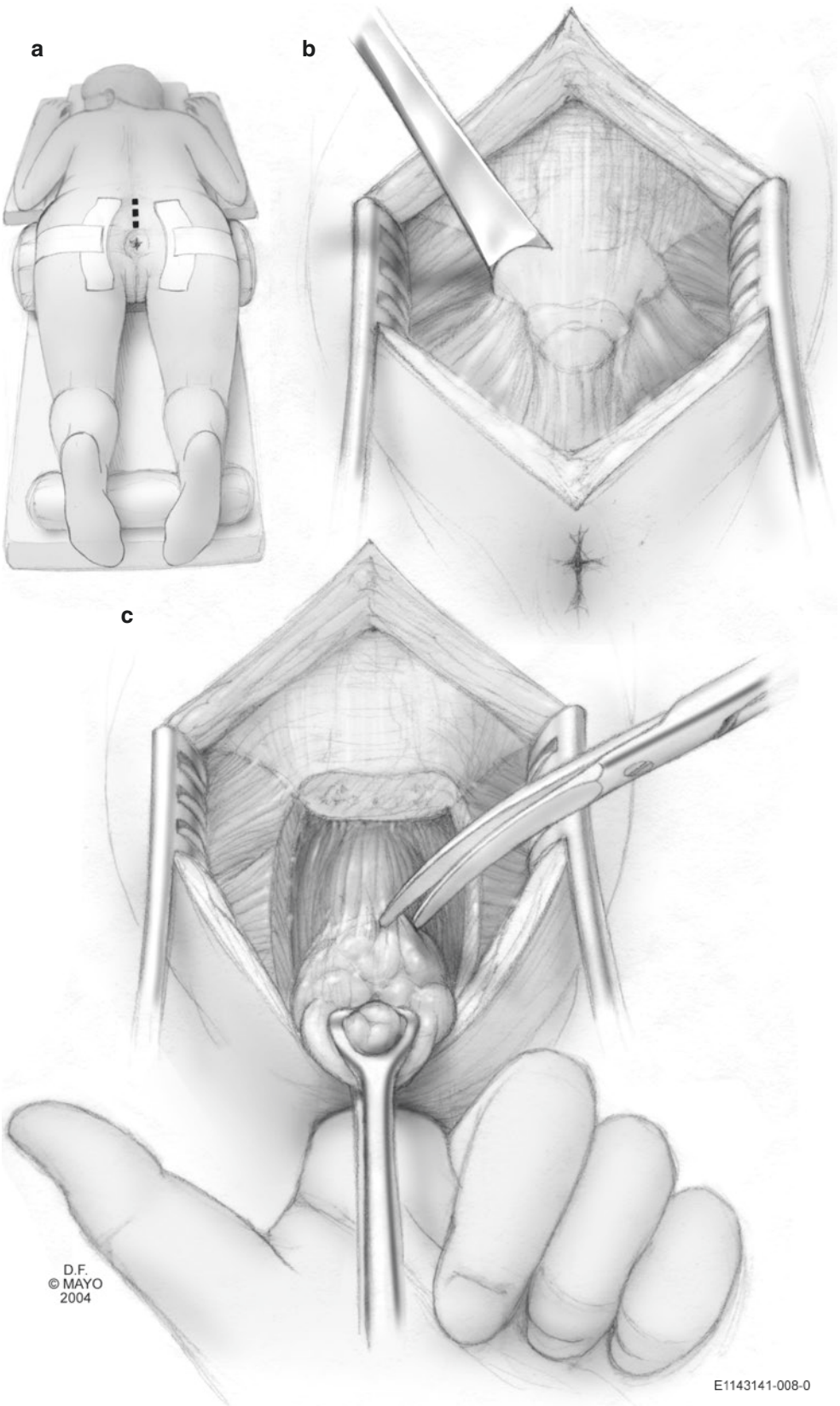

Fig. 21.12 (**a**) Positioning for posterior approach. (**b**) Coccygectomy. (**c**) Index finger in anal canal to "push" tumor outward facilitating dissection. (Reused with permission of Mayo Foundation for Medical Education and Research, all rights reserved)

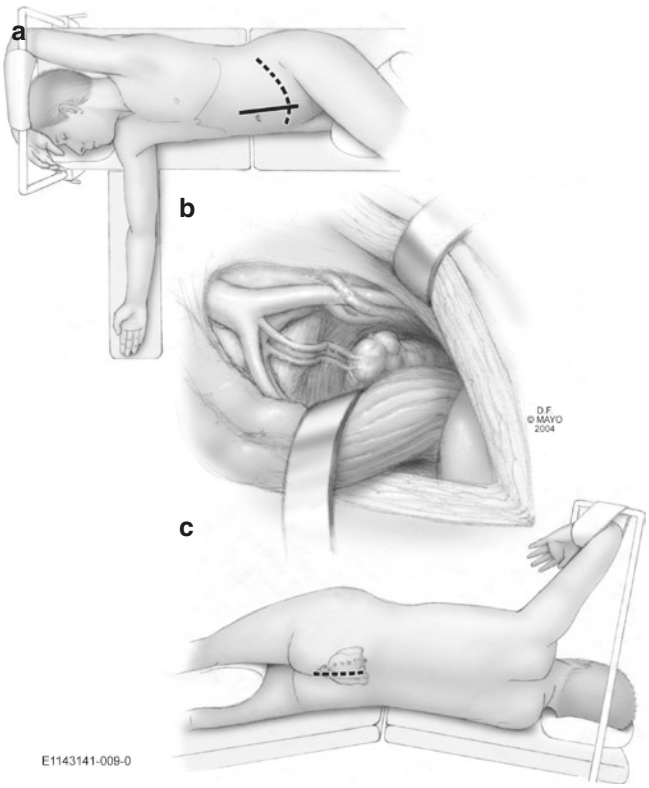

Fig. 21.13 (**a**) Modified lateral position for anterior exposure vis a midline (solid line) or ilioinguinal (dotted line) incision. (**b**) Anterior exposure of vessels and tumor. (**c**) Posterior approach to the sacrum. (Reused with permission of Mayo Foundation for Medical Education and Research, all rights reserved)

separated (ureter, bone, vasculature, nerve, etc.), instead they should be removed en bloc with the tumor.

Substantial blood loss can occur during resection of large presacral tumors, especially in those requiring en bloc sacrectomy. Middle sacral vessels are often significantly enlarged. Selective ligation of the middle sacral artery and in some cases, the internal iliac vessels and their branches, can reduce blood loss (Fig. 21.14). Preoperative catheter based venous and/or arterial embolization can be considered when significant bleeding is anticipated. Preservation of the anterior division of the internal iliac artery and internal gluteal branches reduces the risk of perineal and gluteal necrosis. Multidisciplinary planning with vascular surgery is prudent for cases where significant vascular dissection is anticipated, especially for patients with prior irradiation or anticipated distorted vascular anatomy.

For expected large pelvic or postsacral defects, a plastic surgeon should be involved for tissue interposition/reconstruction. Multiple options are available such as vertical rectus abdominis myocutaneous (VRAM) flap, transverse rectus abdominis (TRAM) flap, omental pedicle flap, gracilis flaps, and gluteus myocutaneous flap (local or V–Y advancement) closures.

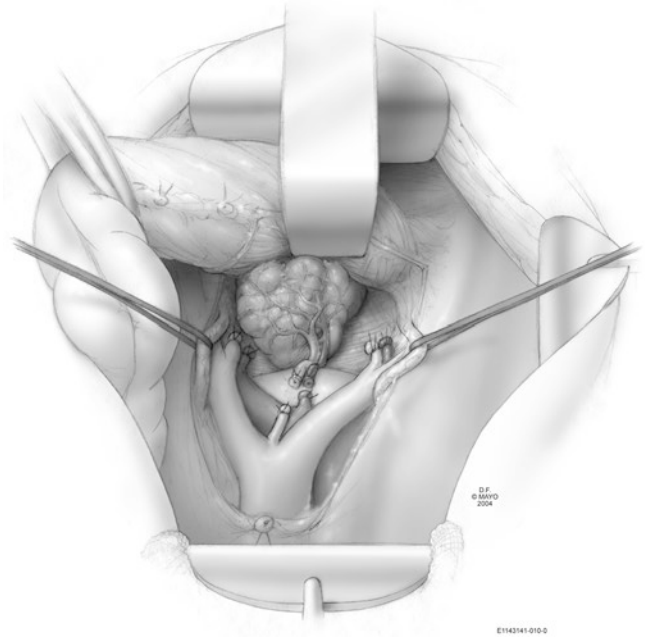

Fig. 21.14 Ligation of middle sacral and internal iliac vessel. (Reused with permission of Mayo Foundation for Medical Education and Research, all rights reserved)

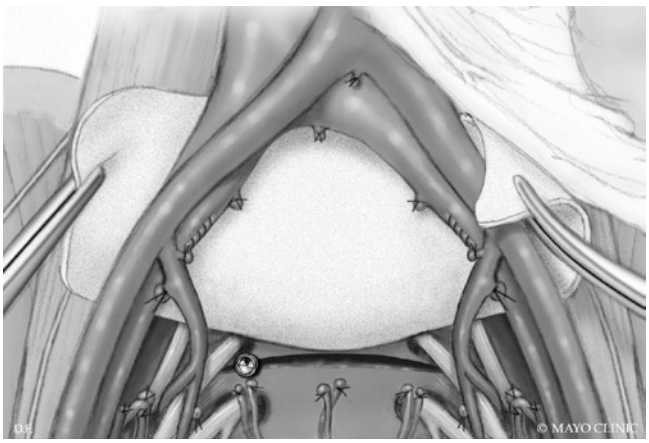

Fig. 21.15 Placement of silastic mesh to protect pelvic vasculature during posterior osteotomies. (Reused with permission of Mayo Foundation for Medical Education and Research, all rights reserved)

For extended sacral involvement it is often necessary to change patient positioning. After the anterior dissection is completed, the abdomen closed, and colostomy matured, the patient is placed in a prone position for the posterior dissection. To reduce injury to vital structures (arteries/veins/ureters) when performing the posterior sacral transection a protective barrier (thick piece of silastic mesh or plastic sheeting, laparotomy pads, etc.) can be placed directly anterior to the sacrum (Fig. 21.15). The mesh will also help protect a pedicled flap that has been placed in the pelvis in preparation for later extraction for perineal reconstruction once the sacrum has been removed. After placing in a prone

position, a midline incision is made over the sacrum and coccyx down to the anus. The anococcygeal ligament is ligated and levator muscles retracted bilaterally. Orthopedic surgery can then continue with dissection of the gluteus maximus muscles bilaterally, transection of the sacrotuberous and sacrospinous ligaments (Fig. 21.16a), and division of the piriformis muscles to expose the sciatic nerves (Fig. 21.16b). An osteotomy is then performed at the desired level exposing and preserving uninvolved sacral nerve roots. For sacral resection in the region of S2–S3 or higher, the thecal sac should be closed with an absorbable suture to decrease issues with cerebrospinal fluid leak or life threatening intra-dural infection (Fig. 21.16c). The tumor is then removed en bloc with the sacrum, coccyx, and involved sacral nerve roots, with or without the rectum. If both S3 nerve roots are sacrificed a permanent colostomy is often necessary.

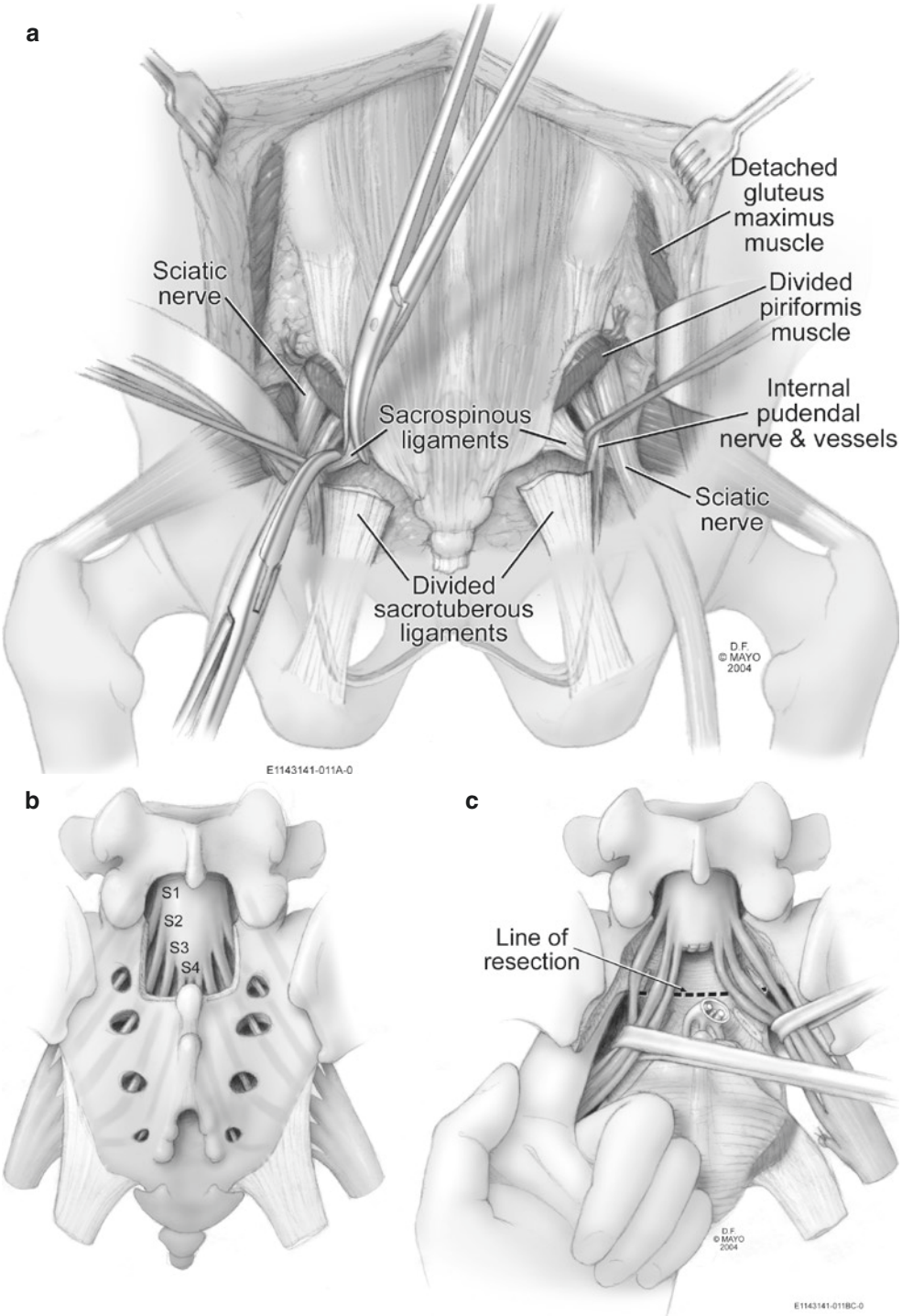

Fig. 21.16 (**a**) Posterior approach exposure of sciatic nerve. (**b**) Sacral nerve roots. (**c**) Ligation of thecal sac. (Reused with permission of Mayo Foundation for Medical Education and Research, all rights reserved)

Minimally Invasive Approaches

Minimally invasive surgery (MIS) has the potential to minimize morbidity and enhance recovery. Laparoscopic and robotic techniques are being more commonly described as safe and feasible means for removing presacral tumors in selected patients (Figs. 21.17 and 21.18) [23, 72–78]. Conditions in which a MIS only approach may not be feasible are very large tumors or malignant tumors that involve the pelvic sidewall, sacrum, or multiple viscera. The overall goals of the surgery must be kept in mind when making a decision on the approach and include complete tumor resection, avoiding disruption of the tumor, and avoidance of injury to surrounding anatomical structures.

Mullaney and colleagues at Mayo Clinic recently performed a systematic review of the literature to determine the feasibility and surgical outcomes of presacral tumors approached using MIS techniques [79]. A total of 82 patients were found that met inclusion criteria. The majority of patients were female (n = 65; 79.2%), with a mean age of 41.7 years (range, 18–89 years). Seventy-three patients (89.0%) underwent laparoscopic or combined laparoscopic-perineal resection, and nine (10.8%) a robotic approach. The conversion rate was 5.5%. The overall 30-day morbidity rate was 15.7%, including one intraoperative rectal injury (1.2%). Ninety-five percent (n = 78) of the tumors were benign. Median length of stay was 4 days for both laparoscopic and robotic groups. No tumor recurrence was noted during follow-up [median 28 months (range, 5–71 months)]. They compared their data from select patients to historical controls from a systematic review of 1064 patients having an open operation. Patients who undergo a minimally invasive approach had a similar mean operating time (155 ± 63 vs. 175 ± 126 min), shorter hospital length of stay (4 vs. 9 days) and comparable 30-day postoperative complications (16% vs. 12.2%) [58]. Selection bias is obviously inherent to the study design, and thus one technique cannot be considered to be superior to another. However, these data suggest that MIS approaches to presacral tumors are reasonably safe and efficacious in select patients when undergoing operation by highly experienced surgeons.

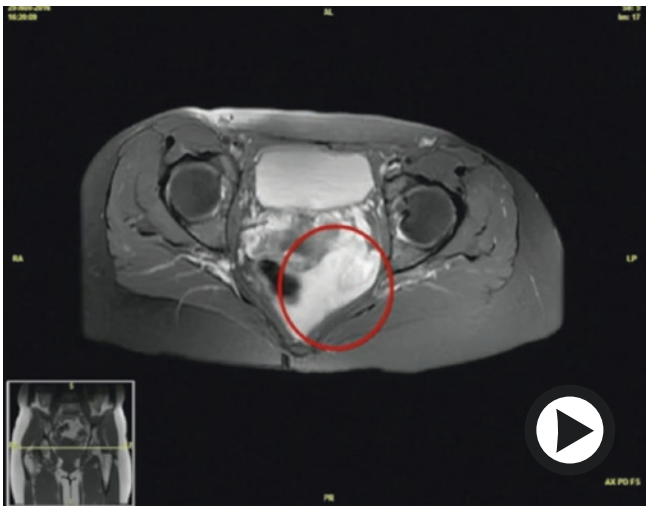

Fig. 21.17 Robotic excision of a giant aggressive angiomyxoma traversing through the levator muscle into the ischioanal space. (Reused with permission of Mayo Foundation for Medical Education and Research, all rights reserved). (https://doi.org/10.1007/000-33b)

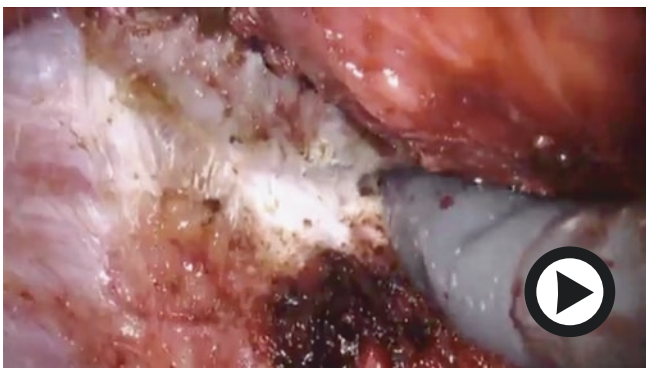

Fig. 21.18 Robotic excision of a presacral cyst below S3 with transvaginal extraction. (Reused with permission of Mayo Foundation for Medical Education and Research, all rights reserved). (https://doi.org/10.1007/000-33a)

Outcomes

Due to the heterogeneity and rarity of presacral tumors, it is difficult to draw any firm conclusions regarding outcomes following treatment from the published literature. Most reported series come from tertiary/quaternary referral centers, with cases accumulated over many years, or decades. As one might expect, there is great variability in follow-up regimens. This fact, and the absence of time-to-event (Kaplan–Meier) calculation of recurrence rates in many series, renders it impossible for the reader to gain more than a general impression of outcomes.

The largest series published since 1975, when Uhlig and Johnson updated the presacral tumor classification system, are outlined in Tables 21.2 and 21.3 [2, 4, 8, 10, 14, 17, 18, 20, 26, 27, 29, 46, 47, 49, 50, 65, 80–82]. For ease of interpretability the tables are separated into benign (Table 21.2) and malignant (Table 21.3), and provide a high-level overview of the numbers and types of tumors presented. Series of both benign and malignant tumors present data ranging from 8 to 48 years. Recurrence rates for benign masses range from 0% to 35%, with the highest recurrences noted for neurogenic tumors. Recurrence rates for malignant lesions range from 0% to 48%, and it is uniformly noted that a R0 resection with wide surgical margins is associated with lower rates of local recurrence.

Table 21.2 Benign tumors

Date	Author	Institution	Cases	Classification	Tumor types	(*n*)
1975	Uhlig et al. [29]	Portland Surgical Center	38	Congenital	Mucus secreting cyst	16
					Indeterminate cyst	7
					Teratoma	2
					Adrenal rest tumor	1
					Epidermoid cyst	1
				Neurogenic	Ganglioneuroma	2
					Neurolemmoma	1
					Neurofibroma	1
				Osseous	Osteoma	1
					Simple bone cyst	1
				Inflammatory	Abscess	2
					Foreign body granuloma	1
				Miscellaneous	Lymphangioma	1
					Desmoid	1
1985	Jao et al. [2]	Mayo Clinic	69	Congenital	Mucus-secreting cyst	16
					Epidermoid cyst	15
					Teratoma	15
					Meningocele	2
				Neurogenic	Neurilemoma	7
					Neurofibroma	3
				Osseous	Giant cell tumor	5
					Aneurysmal bone cyst	1
					Osteochondroma	1
				Miscellaneous	Lipoma	3
					Leiomyoma	1
1993	Bohm et al. [80]	Cleveland Clinic	20	Congenital	Teratoma	9
					Tailgut cyst	6
					Epidermoid cyst	5
1995	Wang et al. [17]	Chang Gung Hosp.	23	Congenital	Epidermal cyst	10
					Teratoma	3
					Dermal cyst	2
				Neurogenic	Neurilemoma	2
				Osseous	Giant cell tumor	4
				Miscellaneous	Leiomyoma	1
					Granuloma	1
2003	Lev-Chelouche et al. [18]	Tel Aviv Univ.	21	Congenital	Tailgut cyst	12
				Neurogenic	Schwanoma	3
				Miscellaneous	Leiomyoma	3
					Fibroma	2
					Angiomyxoma	1
2005	Glasgow et al. [8]	Washington Univ.	27	Congenital	Teratoma	8
					Dermoid/epidermoid cyst	5
					Rectal duplication cyst	2
				Neurogenic	Schwannoma/neurofibroma	5
				Miscellaneous	Leiomyoma	3
				Other	Not described	4
2009	Dozois et al. [26]	Mayo Clinic	46	Neurogenic	Schwannoma	28
					Neurofibroma	17
					Ganglioneuroma	1
2010	Mathis et al. [27]	Mayo Clinic	31	Congenital	Tailgut cyst	31

Table 21.2 (continued)

Date	Author	Institution	Cases	Classification	Tumor types	(*n*)
2012	Macafee et al. [10]	General Infirmary	39	Congenital	Tailgut cyst	13
					Epidermoid cyst	3
					Teratoma	2
				Neurogenic	Schwannoma	11
					Ganglioneuroma	NR
				Miscellaneous	Myelolipoma	NR
					Lipoma	NR
					Mucinous cyst	NR
					Mucin secreting tumor	NR
					Solitary fibrous tumor	NR
2013	Chereau et al. [14]	Hôpital Saint-Antoine	38	Congenital	Tailgut cyst	28
					Dermoid/epidermoid cyst	7
					Teratoma	2
					Rectal duplication cyst	1
2013	Messick et al. [4]	Cleveland Clinic	65	Congenital	Tailgut cyst	28
					Epidermoid cyst	10
					Teratoma	9
					Dermoid	4
					Rectal duplication cyst	2
				Neurogenic	Schwannoma	7
					Ganglioneuroma	1
					Neurofibroma	1
				Miscellaneous	Pecoma	1
					Myelolipoma	1
					Hemangiopericytoma	1
2014	Simpson et al. [20]	Mayo Clinic	21	Congenital	Teratoma	21
2016	Maddah et al. [82]	Mashhad Univ.	23	Congenital	Dermoid/epidermoid cyst	8
					Tailgut cyst	3
					Anterior meningocele	1
					Teratoma	1
					Duplication cyst	1
				Neurogenic	Schwannoma	2
				Osseous	Intra-osseous ganglion cyst	1
				Miscellaneous	Fibromatosis	2
					Hydatid cyst	2
					Lipofibroma	1
					Unknown	1

n number, *NR* not recorded

Table 21.3 Malignant tumors

Date	Author	Institution	Cases	Classification	Tumor types	(*n*)
1975	Uhlig et al. [29]	Portland Surgical Center	25	Congenital Neurogenic	Chordoma	6
					Teratocarcinoma	2
					Neurofibrosarcoma	1
					Ependymoma	1
				Osseous	Osteogenic sarcoma	1
				Miscellaneous	Local & Metastatic cancers	9
					Liposarcoma	2
					Hemangioendothelial sarcoma	1
					Undetermined tumor	1
					Plasma cell myeloma	1

(continued)

Table 21.3 (continued)

Date	Author	Institution	Cases	Classification	Tumor types	(*n*)
1981	Cody et al. [81]	MSKCC	39	Congenital	Chordoma	15
					Epidermoid carcinoma	1
				Neurogenic	Neuroblastoma	4
					Schwannoma	1
					Ganglioneuroblastoma	1
				Osseous	Chrondrosarcoma	3
					Reticulum cell sarcoma	2
					Ewing's sarcoma	1
					Plasmacytoma	1
				Miscellaneous	Unclassified tumor	3
					Hemangiopericytoma	3
					Adenocarcinoma	3
					Carcinoid	1
1985	Jao et al. [2]	Mayo Clinic	51	Congenital	Chordoma	30
					Teratocarcinoma	3
				Neurogenic	Neurofibrosarcoma	2
					Ependymoma	1
					Neuroblastoma	1
				Osseous	Ewing's sarcoma	3
					Osteogenic sarcoma	1
				Miscellaneous	Lymphoma	6
					Myeloma	2
					Fibrosarcoma	1
					Undifferentiated sarcoma	1
1993	Bohm et al. [80]	Cleveland Clinic	4	Congenital	Chordoma	4
1995	Wang et al. [17]	Chang Gung Hosp.	22	Congenital	Chordoma	5
					Teratocardinoma	1
				Neurogenic	Neurofibrosarcoma	1
					Ganglioneuroblastoma	1
				Miscellaneous	Leiomyosarcoma	7
					Undifferentiated sarcoma	2
					Fibrosarcoma	1
					Liposarcoma	1
					Lymphoma	1
					Histiocytoma	1
					Unknown	1
2001	McMaster et al. [49]	NCI	117	Congenital	Chordoma	117
2003	Lev-Chelouche et al. [18]	Tel Aviv Univ.	21	Congenital	Chordoma	9
				Neurogenic	Malignant schwannoma	1
				Osseous	Chrondrosarcoma	2
					Osteosarcoma	1
				Miscellaneous	Desmoid	2
					Angiosarcoma	2
					Fibrosarcoma	1
					Epithelioid sarcoma	1
					Squamous cell carcinoma	1
					Lymphoma	1
2005	Fuchs et al. [50]	Mayo Clinic	52	Congenital	Chordoma	52
2005	Glasgow et al. [8]	Washington Univ.	7	Congenital	Chordoma	3
					Teratocarcinoma	1
				Neurogenic	Malignant schwannoma	1
				Miscellaneous	Radiation induced sarcoma	1
					Leiomyosarcoma	1
2009	Dozois et al. [26]	Mayo Clinic	43	Neurogenic	Neurofibrosarcoma	35
					Ependymoma	6
					Ganglioneuroblastoma	1
					Neuroblastoma	1

Table 21.3 (continued)

Date	Author	Institution	Cases	Classification	Tumor types	(*n*)
2011	Dozois et al. [65]	Mayo Clinic	37	Neurogenic	Neurofibrosarcoma	8
				Osseous	Chrondrosarcoma	7
					Osteosarcoma	3
				Miscellaneous	Undifferentiated sarcoma	6
					Liposarcoma	6
					Leiomyosarcoma	4
					Fibromyxoid sarcoma	1
					GIST	1
					Solitary fibrous tumor	1
2012	Macafee et al. [10]	General Infirmary	17	Congenital	Chordoma	9
				Miscellaneous	Multicystic Adenocarcinoma	2
					Rhabdomyosarcoma	1
					Leiomyosarcoma	1
					Angiomyxoma	1
					Liposarcoma	1
					GIST	1
					NET	1
2013	Chereau et al. [14]	Hôpital Saint-Antoine	9	Congenital	Chordoma	1
				Miscellaneous	Degenerated hamartoma	6
					Unknown sarcoma	2
2013	Messick et al. [4]	Cleveland Clinic	23	Congenital	Chordoma	7
					Teratoma	3
				Osseous	Ewing's sarcoma	1
					Chrondrosarcoma	1
				Miscellaneous	B-cell lymphoma	2
					GIST	2
					Neuroendocrine tumor	2
					Myeloliposarcoma	1
					Histiosarcoma	1
					Squamous cell cancer	1
					Liposarcoma	1
					Fibrosarcoma	1
2014	Simpson et al. [20]	Mayo Clinic	5	Congenital	Teratoma	5
2016	Maddah et al. [82]	Mashhad University	27	Congenital	Chordoma	8
					Germ cell tumor	1
				Neurogenic	Ependymoma	2
					Neuofibrosarcoma	2
					Neuroblastoma	1
					Primitive neuroectodermal	1
				Osseous	Ewing's sarcoma	2
					Chrondrosarcoma	2
					Plasmacytoma	1
					Giant cell tumor	1
				Miscellaneous	Locally invasive cancer	2
					Liposarcoma	2
					Carcinosarcoma	1
					Spindle cell tumor	1
2018	Pan et al. [46]	Xiangya Hospital	451	Congenital	Chordoma	451
2019	Kerekes et al. [47]	Johns Hopkins, Duke, and the Netherlands	1235	Congenital	Chordoma	1235

MSKCC Memorial Sloan Kettering Cancer Center, *n* number, *NCI* National Cancer Institute

The largest body of literature regarding outcomes following treatment of presacral masses is focused on chordomas. McMaster et al. from the National Cancer Institute used data from the Surveillance, Epidemiology and End Results (SEER) database, over a 22-year period (1973–1995). Of 400 cases 33% were spinal, 32% cranial, 29% sacral, and 6% were extra-axial. Fuchs et al. at Mayo Clinic reported on 52 patients who underwent surgical treatment for sacrococcygeal chordoma between 1980 and 2001 (21 years). They found the most important predictor of survival was a wide margin. All patients with a wide margin survived, and the survival rate was significantly different from that for patients who had either marginal or intra lesional excision. Lung metastasis developed in 16 (31%), and all but three of those patients also had a local recurrence [50]. Pan et al. from Xiangya Hospital, China used the SEER database to identify all patients diagnosed with primary spinal chordoma from 1973 to 2014. A total of 808 patients were identified and the overall rate of distant metastatic cases was 8%. Three hundred fifty-seven spinal chordomas (44%) were located in the vertebral column, while 451 (56%) were located in the sacrum or pelvis. Multivariate models showed age ≥60 years, distant metastasis, and non-surgical therapies were independently associated with reduced survival. Tumor site (vertebrae vs. sacrum/pelvis) was not associated with survival for primary spinal chordoma [46]. Kerekes and colleagues from Johns Hopkins, Duke, and the Netherlands completed a systematic review and pooled cohort analysis (1980–2016) of local and distant recurrence in patients undergoing resection of sacral chordomas. They found 57 studies and 1235 cases for review, and noted wide surgical margin was associated with a lower rate of local recurrence; and wide surgical margin, female sex, and patient age ≥65 years was associated with lower rates of distant recurrence [47].

Follow-Up and Observation-Only Patients

There are limited data on which to base any firm recommendations regarding follow-up. In our practice we typically recommend an annual visit with digital rectal examination to assess for recurrence in patients who had benign lesions resected. A pelvic MRI is obtained 1-year post-resection, and then again at 5 years. In the interim, if a mass is palpated, pelvic imaging is performed. For malignant tumors, patients typically undergo an annual physical examination, pelvic MRI, and CT of the chest and abdomen for 5 years. Collaboration with colleagues in medical and radiation oncology is critical as part of postoperative surveillance and need for adjuvant therapy. Recurrences, when they occur, are considered for re-resection if a complete resection is possible.

Patients with small, benign, asymptomatic tumors can safely be approached in a nonoperative fashion and followed if the patient is comfortable with this plan. For cystic lesions, we recommend pelvic MRI every 5 years for a period of 10 years to assess the natural history of the lesion. If little change to the size or morphology of the lesion is noted, longer intervals between imaging can be considered. Decision-making is on a case-by-case basis. Patients should be counseled that if any change in symptoms occur, it should prompt a clinical and radiographic evaluation. For patients with benign solid tumors such as schwannomas, we recommend a similar follow-up.

Conclusions

Presacral tumors represent a rare group of both benign and malignant lesions. Most benign lesions have malignant potential and must be followed carefully if nonoperative treatment is chosen. MRI is the best overall imaging study to assist in diagnosis and operative planning. When performed appropriately and selectively, a biopsy of the lesion may assist in management of solid and heterogeneous cystic lesions. The surgical principles that should guide a surgeon who manages these lesions are a function-sparing approach for benign lesions and an en bloc approach for malignant lesions. Observation alone in some patients is acceptable when a dedicated surveillance protocol is in place. As the discovery of these tumors increases, more surgeons will be asked to evaluate these patients. Given the broad differential and significant implications of mismanagement, presacral tumors should be evaluated and treated by surgeons at centers that have a large experience in managing these complex tumors.

References

1. Spencer RJ, Jackman RJ. Surgical management of precoccygeal cysts. Surg Gynecol Obstet. 1962;115:449–52.
2. Jao SW, Beart RW Jr, Spencer RJ, Reiman HM, Ilstrup DM. Retrorectal tumors. Mayo Clinic experience, 1960–1979. Dis Colon Rectum. 1985;28(9):644–52.
3. Hobson KG, Ghaemmaghami V, Roe JP, Goodnight JE, Khatri VP. Tumors of the retrorectal space. Dis Colon Rectum. 2005;48(10):1964–74.
4. 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.
5. Todd LT Jr, Yaszemski MJ, Currier BL, Fuchs B, Kim CW, Sim FH. Bowel and bladder function after major sacral resection. Clin Orthop Relat Res. 2002;397:36–9.
6. Zoccali C, Skoch J, Patel AS, Walter CM, Maykowski P, Baaj AA. Residual neurological function after sacral root resection during en-bloc sacrectomy: a systematic review. Eur Spine J. 2016;25(12):3925–31.

7. Wei R, Guo W, Ji T, Zhang Y, Liang H. One-step reconstruction with a 3D-printed, custom-made prosthesis after total en bloc sacrectomy: a technical note. Eur Spine J. 2017;26(7):1902–9.
8. Glasgow SC, Birnbaum EH, Lowney JK, Fleshman JW, Kodner IJ, Mutch DG, Lewin S, Mutch MG, Dietz DW. Retrorectal tumors: a diagnostic and therapeutic challenge. Dis Colon Rectum. 2005;48(8):1581–7.
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. Macafee DA, Sagar PM, El-Khoury T, Hyland R. Retrorectal tumours: optimization of surgical approach and outcome. Color Dis. 2012;14(11):1411–7.
11. 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.
12. Lee KS, Gower DJ, McWhorter JM, Albertson DA. The role of MR imaging in the diagnosis and treatment of anterior sacral meningocele. Report of two cases. J Neurosurg. 1988;69(4):628–31.
13. Sung MS, Lee GK, Kang HS, Kwon ST, Park JG, Suh JS, Cho GH, Lee SM, Chung MH, Resnick D. Sacrococcygeal chordoma: MR imaging in 30 patients. Skelet Radiol. 2005;34(2):87–94.
14. Chereau N, Lefevre JH, Meurette G, Mourra N, Shields C, Parc Y, Tiret E. Surgical resection of retrorectal tumours in adults: long-term results in 47 patients. Color Dis. 2013;15(8):e476–82.
15. Sagar AJ, Koshy A, Hyland R, Rotimi O, Sagar PM. Preoperative assessment of retrorectal tumours. Br J Surg. 2014;101(5):573–7.
16. Toh JW, Morgan M. Management approach and surgical strategies for retrorectal tumours: a systematic review. Color Dis. 2016;18(4):337–50.
17. Wang JY, Hsu CH, Changchien CR, Chen JS, Hsu KC, You YT, Tang R, Chiang JM. Presacral tumor: a review of forty-five cases. Am Surg. 1995;61(4):310–5.
18. Lev-Chelouche D, Gutman M, Goldman G, Even-Sapir E, Meller I, Issakov J, Klausner JM, Rabau M. Presacral tumors: a practical classification and treatment of a unique and heterogeneous group of diseases. Surgery. 2003;133(5):473–8.
19. Merchea A, Larson DW, Hubner M, Wenger DE, Rose PS, Dozois EJ. The value of preoperative biopsy in the management of solid presacral tumors. Dis Colon Rectum. 2013;56(6):756–60.
20. Simpson PJ, Wise KB, Merchea A, Cheville JC, Moir C, Larson DW, Dozois EJ. Surgical outcomes in adults with benign and malignant sacrococcygeal teratoma: a single-institution experience of 26 cases. Dis Colon Rectum. 2014;57(7):851–7.
21. 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.
22. Bai HM, Yang JX, Huang HF, Cao DY, Chen J, Yang N, Lang JH, Shen K. Individualized managing strategies of aggressive angiomyxoma of female genital tract and pelvis. Eur J Surg Oncol. 2013;39(10):1101–8.
23. Kelley SR. Robotic extralevator excision of a retrorectal giant aggressive angiomyxoma. Obstet Gynecol Sci. 2018;61(6):693–7.
24. Frezza AM, Botta L, Trama A, Dei Tos AP, Stacchiotti S. Chordoma: update on disease, epidemiology, biology and medical therapies. Curr Opin Oncol. 2019;31(2):114–20.
25. Springall RG, Griffiths JD. Malignant change in rectal duplication. J R Soc Med. 1990;83(3):185–7.
26. Dozois EJ, Wall JC, Spinner RJ, Jacofsky DJ, Yaszemski MJ, Sim FH, Moran SL, Cima RR, Larson DR, Haddock MG, Okuno SH, Larson DW. Neurogenic tumors of the pelvis: clinicopathologic features and surgical outcomes using a multidisciplinary team. Ann Surg Oncol. 2009;16(4):1010–6.
27. Mathis KL, Dozois EJ, Grewal MS, Metzger P, Larson DW, Devine RM. Malignant risk and surgical outcomes of presacral tailgut cysts. Br J Surg. 2010;97(4):575–9.
28. Hopper L, Eglinton TW, Wakeman C, Dobbs BR, Dixon L, Frizelle FA. Progress in the management of retrorectal tumours. Color Dis. 2016;18(4):410–7.
29. Uhlig BE, Johnson RL. Presacral tumors and cysts in adults. Dis Colon Rectum. 1975;18(7):581–9.
30. Dunn KB. Retrorectal tumors. Surg Clin North Am. 2010;90(1):163–71.
31. Wolpert A, Beer-Gabel M, Lifschitz O, Zbar AP. The management of presacral masses in the adult. Tech Coloproctol. 2002;6(1):43–9.
32. Yang DM, Park CH, Jin W, Chang SK, Kim JE, Choi SJ, Jung DH. Tailgut cyst: MRI evaluation. Am J Roentgenol. 2005;184(5):1519–23.
33. Caropreso PR, Wengert PA Jr, Milford HE. Tailgut cyst – a rare retrorectal tuor. Report of a case and review. Dis Colon Rectum. 1975;18(7):597–600.
34. Hjermstad BM, Helwig EB. Tailgut cysts. Report of 53 cases. Am J Clin Pathol. 1988;89(2):139–47.
35. Prasad AR, Amin MB, Randolph TL, Lee CS, Ma CK. Retrorectal cystic hamartoma: report of 5 cases with malignancy arising in 2. Arch Pathol Lab Med. 2000;124(5):725–9.
36. Michael D, Cohen CR, Northover JM. Adenocarcinoma within a rectal duplication cyst: case report and literature review. Ann R Coll Surg Engl. 1999;81(3):205–6.
37. Pantoja E, Rodriguez-Ibanez I. Sacrococcygeal dermoids and teratomas: historical review. Am J Surg. 1976;132(3):377–83.
38. Izant RJ Jr, Filston HC. Sacrococcygeal teratomas. Analysis of forty-three cases. Am J Surg. 1975;130(5):617–21.
39. Dirix M, van Becelaere T, Berkenbosch L, van Baren R, Wijnen RM, Wijnen MH, van der Zee DC, Heij HA, Derikx JP, van Heurn LW. Malignant transformation in sacrococcygeal teratoma and in presacral teratoma associated with Currarino syndrome: a comparative study. J Pediatr Surg. 2015;50(3):462–4.
40. Altman RP, Randolph JG, Lilly JR. Sacrococcygeal teratoma: American Academy of Pediatrics Surgical Section Survey-1973. J Pediatr Surg. 1974;9(3):389–98.
41. Waldhausen JA, Kolman JW, Vellios F, Battersby JS. Sacrococcygeal teratoma. Surgery. 1963;54:933–49.
42. Miles RM, Stewart GS Jr. Sacrococcygeal teratomas in adults. Ann Surg. 1974;179(5):676–83.
43. Hickey RC, Martin RG. Sacrococcygeal teratomas. Ann N Y Acad Sci. 1964;114:951–7.
44. Gray SW, Singhabhandhu B, Smith RA, Skandalakis JE. Sacrococcygeal chordoma: report of a case and review of the literature. Surgery. 1975;78(5):573–82.
45. Dahlin DC. Bone tumors. General aspects and data on 6,221 cases. 3rd ed. Springfield: Charles C. Thomas; 1978. p. 329. ISBN: 0398036926.
46. Pan Y, Lu L, Chen J, Zhong Y, Dai Z. Analysis of prognostic factors for survival in patients with primary spinal chordoma using the SEER Registry from 1973 to 2014. J Orthop Surg Res. 2018;13(1):76.
47. Kerekes D, Goodwin CR, Ahmed AK, Verlaan JJ, Bettegowda C, Abu-Bonsrah N, Sciubba DM. Local and distant recurrence in resected sacral chordomas: a systematic review and pooled cohort analysis. Global Spine J. 2019;9(2):191–201.
48. Stewart RJ, Humphreys WG, Parks TG. The presentation and management of presacral tumours. Br J Surg. 1986;73(2):153–5.
49. 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.
50. Fuchs B, Dickey ID, Yaszemski MJ, Inwards CY, Sim FH. Operative management of sacral chordoma. J Bone Joint Surg Am. 2005;87(10):2211–6.
51. Manson F, Comalli-Dillon K, Moriaux A. Anterior sacral meningocele: management in gynecological practice. Ultrasound Obstet Gynecol. 2007;30(6):893–6.

52. Berstock JR, Bunni J, Thorpe PL. Caution: laparoscopic biopsy of a large pelvic mass may result in disaster. J Minim Invasive Gynecol. 2012;19(5):651–3.
53. Cheng C, Tao B, Bai S, Gao G, Li S, Shang A. Anterior sacral meningocele: a new classification and treatment using the dorsal transsacral approach. Spine. 2020;45(7):444–51.
54. Hébert-Blouin MN, Sullivan PS, Merchea A, Léonard D, Spinner RJ, Dozois EJ. Neurological outcome following resection of benign presacral neurogenic tumors using a nerve-sparing technique. Dis Colon Rectum. 2013;56(10):1185–93.
55. Alderete J, Novais EN, Dozois EJ, Rose PS, Sim FF. Morbidity and functional status of patients with pelvic neurogenic tumors after wide excision. Clin Orthop Relat Res. 2010;468(11):2948–53.
56. Freier DT, Stanley JC, Thompson NW. Retrorectal tumors in adults. Surg Gynecol Obstet. 1971;132(4):681–6.
57. Randall RL. Giant cell tumor of the sacrum. Neurosurg Focus. 2003;15(2):E13. Review.
58. Baek SK, Hwang GS, Vinci A, Jafari MD, Jafari F, Moghadamyeghaneh Z, Pigazzi A. Retrorectal tumors: a comprehensive literature review. World J Surg. 2016;40(8):2001–15.
59. Currarino G, Coln D, Votteler T. Triad of anorectal, sacral, and presacral anomalies. AJR Am J Roentgenol. 1981;137(2):395–8.
60. AbouZeid AA, Mohammad SA, Abolfotoh M, Radwan AB, Ismail MME, Hassan TA. The Currarino triad: what pediatric surgeons need to know. J Pediatr Surg. 2017;52(8):1260–8.
61. Crétolle C, Pelet A, Sanlaville D, Zérah M, Amiel J, Jaubert F, Révillon Y, Baala L, Munnich A, Nihoul-Fékété C, Lyonnet S. Spectrum of HLXB9 gene mutations in Currarino syndrome and genotype-phenotype correlation. Hum Mutat. 2008;29(7):903–10.
62. Leonard D, Pendlimari R, Larson DW, Dozois EJ. Currarino syndrome: typical images of a rare condition. Tech Coloproctol. 2010;14(3):289–90.
63. Pendlimari R, Leonard D, Dozois EJ. Rare malignant neuroendocrine transformation of a presacral teratoma in patient with Currarino syndrome. Int J Color Dis. 2010;25(11):1383–4.
64. MacCarty CS, Waugh JM, Coventry MB, Cope WF Jr. Surgical treatment of sacral and presacral tumors other than sacrococcygeal chordoma. J Neurosurg. 1965;22(5):458–64.
65. Dozois EJ, Jacofsky DJ, Billings BJ, Privitera A, Cima RR, Rose PS, Sim FH, Okuno SH, Haddock MG, Harmsen WS, Inwards CY, Larson DW. Surgical approach and oncologic outcomes following multidisciplinary management of retrorectal sarcomas. Ann Surg Oncol. 2011;18(4):983–8.
66. Yang JC, Chang AE, Baker AR, Sindelar WF, Danforth DN, Topalian SL, DeLaney T, Glatstein E, Steinberg SM, Merino MJ, Rosenberg SA. Randomized prospective study of the benefit of adjuvant radiation therapy in the treatment of soft tissue sarcomas of the extremity. J Clin Oncol. 1998;16(1):197–203.
67. Feng M, Murphy J, Griffith KA, Baker LH, Sondak VK, Lucas DR, McGinn CJ, Ray ME. Long-term outcomes after radiotherapy for retroperitoneal and deep truncal sarcoma. Int J Radiat Oncol Biol Phys. 2007;69(1):103–10.
68. Rosenberg SA, Tepper J, Glatstein E, Costa J, Baker A, Brennan M, DeMoss EV, Seipp C, Sindelar WF, Sugarbaker P, Wesley R. The treatment of soft-tissue sarcomas of the extremities: prospective randomized evaluations of (1) limb-sparing surgery plus radiation therapy compared with amputation and (2) the role of adjuvant chemotherapy. Ann Surg. 1982;196(3):305–15.
69. Adjuvant chemotherapy for localised resectable soft-tissue sarcoma of adults: meta-analysis of individual data. Sarcoma Meta-analysis Collaboration. Lancet. 1997;350(9092):1647–54.
70. Raut CP, Pisters PW. Retroperitoneal sarcomas: combined-modality treatment approaches. J Surg Oncol. 2006;94(1):81–7.
71. Dozois EJ, Malireddy KK, Bower TC, Stanson AW, Sim FH. Management of a retrorectal lipomatous hemangiopericytoma by preoperative vascular embolization and a multidisciplinary surgical team: report of a case. Dis Colon Rectum. 2009;52(5):1017–20.
72. Witherspoon P, Armitage J, Gatt M, Sagar PM. Laparoscopic excision of retrorectal schwannoma. Dis Colon Rectum. 2010;53(1):101–3.
73. Duclos J, Maggiori L, Zappa M, Ferron M, Panis Y. Laparoscopic resection of retrorectal tumors: a feasibility study in 12 consecutive patients. Surg Endosc. 2014;28(4):1223–9.
74. Fong SS, Codd R, Sagar PM. Laparoscopic excision of retrorectal tumours. Color Dis. 2014;16(11):O400–3.
75. Zhou JL, Wu B, Xiao Y, Lin GL, Wang WZ, Zhang GN, Qiu HZ. A laparoscopic approach to benign retrorectal tumors. Tech Coloproctol. 2014;18(9):825–33.
76. Manabe T, Koba R, Nagayoshi K, Sadakari Y, Fujita H, Nagai S, Ueki T, Nagai E, Nakamura M. Laparoscopic excision of neurogenic retrorectal tumors. Asian J Endosc Surg. 2017;10(2):223–6.
77. Cataneo J, Cataldo T, Poylin V. Robotic excision of retrorectal mass. J Gastrointest Surg. 2018;22(10):1811–3.
78. Hove MG, Gil JM, Rodríguez TS, Lomas ÁF, Casajús JP, López-Farré A, Guijarro JJ. Laparoscopic approach to tailgut cyst (retrorectal cystic hamartoma). J Minim Access Surg. 2019;15(3):262–4.
79. Mullaney TG, Lightner AL, Johnston M, Kelley SR, Larson DW, Dozois EJ. A systematic review of minimally invasive surgery for retrorectal tumors. Tech Coloproctol. 2018;22(4):255–63.
80. 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 Color Dis. 1993;8(3):134–8.
81. Cody HS 3rd, Marcove RC, Quan SH. Malignant retrorectal tumors: 28 years' experience at Memorial Sloan-Kettering Cancer Center. Dis Colon Rectum. 1981;24(7):501–6.
82. Maddah G, Abdollahi A, Etemadrezaie H, Ganjeifar B, Gohari B, Abdollahi M, Hassanpour M. Problems in diagnosis and treatment of retrorectal tumors: our experience in 50 patients. Acta Med Iran. 2016;54(10):644–50.