Presacral Tumors: Diagnosis and Management

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ABSTRACT

Presacral tumors are uncommon lesions that can be difficult to diagnose because of their nonspecific presenting signs and symptoms. Cross-sectional imaging is essential in evaluating these lesions to determine the optimal surgical approach and the extent of resection. Surgery is the mainstay of treatment as it establishes the diagnosis and prevents the adverse consequences associated with malignant degeneration and secondary bacterial infection. The outcomes for patients with benign presacral tumors are favorable. Although there have been substantial improvements in the prognosis of patients with malignant presacral tumors, the development of newer adjuvant therapies are likely to further improve the oncologic outcomes of malignant presacral tumors such as chordomas and sarcomas.

KEYWORDS: Presacral tumors, management, diagnosis, surgery, prognosis

Objectives: On completion of this article the reader should be able to summarize the diagnosis, management, and treatment of presacral tumors.

Presacral or retrorectal tumors represent a spectrum of heterogeneous lesions ranging from simple benign cysts to complex malignant masses invading surrounding pelvic structures. The incidence of presacral tumors in the general population is not known as the majority of reports on these lesions are from tertiary referral centers and thus do not represent the true incidence of these tumors.¹ The only large series² not from a referral center was published almost 30 years ago by Uhlig and Johnson and found an average incidence of two presacral tumors per year in the metropolitan Portland area. Despite their infrequent occurrence, a basic comprehension of the etiology, presentation, diagnostic evaluation, and management of these lesions is important as incorrect diagnosis or inappropriate management can result in significant morbidity and adverse outcomes.¹

ANATOMY

The presacral space is a potential space. The mesorectum forms the anterior boundary of the space and

the anterior aspect of the sacrum forms the posterior border. Superiorly, the space extends up to the peritoneal reflection and inferiorly to the retrosacral fascia, which passes forward from the S-4 vertebra to the rectum ~3 to 5 cm proximal to the anorectal junction (Fig. 1). Below the retrosacral fascia lays the supralelevator space, another potential space, bound anteriorly by the mesorectum and inferiorly by the muscles of the pelvic floor. The lateral extent of the presacral space is bound by the ureters, the iliac vessels, the lateral stalks of the rectum, and the sacral nerve roots.³⁻⁵

The presacral space contains loose connective tissue, the middle sacral artery, the superior rectal vessels and branches of the sympathetic and parasympathetic nervous systems. The presacral space is the site of fusion of the embryologic hindgut and neuroectoderm and contains totipotent cells from which various tumors may develop. Furthermore, a variety of tumors found in this region arise from adjacent tissues that extend into the presacral space.³

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Anal Cancer and Retrorectal Tumors; Guest Editor, Jan Rakinic, M.D.

Clin Colon Rectal Surg 2009;22:84-93. Copyright © 2009 by Thieme Medical Publishers, Inc., 333 Seventh Avenue, New York, NY 10001, USA. Tel: +1(212) 584-4662.

DOI 10.1055/s-0029-1223839. ISSN 1531-0043.

Figure 1 Relationship of pelvic structures to the presacral space. From Mayo Clinic, Rochester, NY. Reproduced with kind permission of Springer Science and Business Media.

CLASSIFICATION

As a result of the diverse nature of presacral tumors, a variety of classification systems have been proposed to categorize these lesions. The classification system first described by Uhlig and Johnson has been most frequently used and separates these lesions into the following broad categories: congenital, neurogenic, osseous, inflammatory, and miscellaneous.² Due to the impact of the nature of the lesion (benign versus malignant) on the therapeutic approach, Dozois et al⁵ have modified and updated this classification system to subcategorize tumors into malignant and benign entities within these broad categories (Table 1). Lev-Chelouche et al⁶ have classified these tumors into congenital versus acquired and benign versus malignant, resulting in four distinct groups, each consisting of various histologic subtypes, but with similar clinical presentation, diagnosis, and management. Common characteristics and features of some of the more frequently seen presacral tumors are summarized in Table 2.

CONGENITAL

Congenital lesions are the most common presacral lesions accounting for 55 to 70% of all lesions.¹ These originate from embryologic remnants, are usually benign, and are more common in females.^{4,5} These include developmental cysts, chordomas, and anterior meningoceles.

Developmental Cysts

Developmental cysts account for 60% of all congenital presacral lesions and can originate from any of the three

Table 1 Classification System of Presacral Tumors

Congenital
Benign
Developmental cysts (teratoma, epidermoid, dermoid, mucus-secreting)
Duplication of rectum
Anterior sacral meningocele
Adrenal rest tumor
Malignant
Chordoma
Teratocarcinoma
Neurogenic
Benign
Neurofibroma
Neurilemoma (schwannoma)
Ganglioneuroma
Malignant
Neuroblastoma
Ganglioneuroblastoma
Ependymoma
Malignant peripheral nerve sheath tumors (malignant schwannoma, neurofibrosarcoma, neurogenic sarcoma)
Osseous
Benign
Giant-cell tumor
Osteoblastoma
Aneurysmal bone cyst
Malignant
Osteogenic sarcoma
Ewing's sarcoma
Myeloma
Chondrosarcoma
Miscellaneous
Benign
Lipoma
Fibroma
Leiomyoma
Hemangioma
Endothelioma
Desmoid (locally aggressive)
Malignant
Liposarcoma
Fibrosarcoma/malignant fibrous histiocytoma
Leiomyosarcoma
Hemangiopericytoma
Metastatic carcinoma
Other
Ectopic kidney
Hematoma
Abscess

Modified from Uhlig and Johnson.² Original permission from Lippincott Williams & Wilkins.

Table 2 Characteristics of Common Presacral Tumors

Tumor Type	Benign or Malignant	Cells of Origin	Tissue/Cell Type
Dermoid cyst	Benign	Ectoderm	Stratified squamous $\pm$ skin appendages
Epidermoid cyst	Benign	Ectoderm	Stratified squamous
Duplication cyst	Most are benign	Hindgut	Squamous or columnar
Tailgut cyst	Rare malignant degeneration	Primitive gut remnants	Squamous, columnar, or transitional
Teratoma	Malignant potential	Totipotential cells	Tissue from each germ layer
Chordoma	Most common malignant presacral tumor	Notochord	Spinal column
Neurilemmoma	Benign	Schwann cells from neural crest	Nerve Sheath
Chondrosarcoma	Malignant	Cartilage	Chondrocytes
Ewing sarcoma	Malignant	Mesoderm/ectoderm	Most commonly bone

embryonic germ layers.¹ High secondary infection rates have been reported with these types of cysts particularly when they are misdiagnosed as a perirectal abscess and are operatively manipulated.⁷ Depending on the cell layer of origin, developmental cysts can be divided into the following types: epidermoid cysts, dermoid cysts, enterogenous cysts, tailgut cysts, and teratomas.

EPIDERMOID AND DERMOID CYSTS

These cysts result from the abnormal closure of the ectodermal tube. Epidermoid cysts are benign unilocular lesions composed of stratified squamous cells. Dermoid cysts can be differentiated from epidermoid cysts because they are not only composed of stratified squamous epithelium, but can also have skin appendages like sweat glands, hair follicles, or sebaceous cysts.^{3,5} Both types may be associated with a postanal dimple or sinus when they communicate with the skin.⁵

ENTEROGENOUS CYSTS (DUPLICATION CYSTS OF THE RECTUM)

These are thought to develop due to sequestration of the developing hindgut.⁵ Morphologically, they tend to have one dominant lesion with several smaller satellite lesions.⁵ They are diagnosed based on three histologic criteria: (1) continuity or contiguity with the rectum, (2) a well defined muscular wall with a myenteric plexus, and (3) a mucosal lining. This lining is usually similar to rectal mucosa, but can sometimes contain ectopic tissue such as gastric mucosa, pancreatic tissue or urothelial mucosa because they originate from the endoderm.⁸ Though usually benign, malignant degeneration has been reported.³⁻⁵

TAILGUT CYSTS

Also known as cystic hamartomas, their histologic appearance is similar to that of the intestinal tract and they can be composed of squamous, columnar, or transitional epithelium.³ They are usually circumscribed, unencapsulated and multilocular. The presence of columnar and transitional epithelium differentiates these lesions from epidermoid and dermoid cysts, which only contain squamous epithelium. The absence of a well-defined

muscular wall with a myenteric plexus distinguishes them from duplication cysts.⁵ Although rare, malignant degeneration has been reported, with adenocarcinomas being the most frequent histologic subtype.⁴

TERATOMAS

Presacral teratomas are the most common teratomas seen in infancy and are rare beyond the second decade of life.³ They arise from totipotential cells and therefore can be composed of several tissue types including respiratory, nervous, and gastrointestinal epithelium.⁵ They may be solid or cystic and tend to adhere to the coccyx.¹ Germ cell elements, when present, have a malignant potential and degeneration to squamous cell carcinoma from the ectodermal tissue or to rhabdomyosarcoma from the mesenchymal cells can occur if they are left untreated.⁵

Sacroccygeal Chordomas

These are the most common malignant presacral tumors.³ They are considered to originate from the notochord, which is the primitive flexible vertebral column that extends from the base of the occiput to the caudal limit in the embryo.⁵ Although chordomas may occur anywhere along the spinal column, they are most commonly found at the base of the skull and the sacroccygeal region, with more than half occurring in the sacrum.⁵ There is a male predisposition and they rarely occur before the third decade of life.⁵ They are slow growing and tend to invade adjacent structures and metastasize in 20% of cases, most commonly to the lung, liver, and bone.⁹ They can present with specific symptoms such as incontinence or impotence, vague symptoms including pelvic, buttock, and positional lower back pain, or they can be asymptomatic.¹⁰

Anterior Sacral Meningocele

These lesions develop due to a herniation of the dural sac through a defect in the sacrum and can be seen in combination with presacral lipomas or cysts.⁵ Other associated congenital abnormalities such as spina bifida,

tethered spinal cord, uterine and vaginal duplication or urinary or anal malformations can also occur.⁵ The dural sac is in continuity with the subdural space and contains cerebrospinal fluid (CSF), as a result they can present with headaches associated with defecation due to a compression-induced increase in CSF pressure.⁵ They can also present with recurrent meningitis or symptoms related to a mass effect such as constipation, urinary symptoms, or low back pain.³ If these lesions are inadvertently biopsied, life-threatening meningitis can occur.¹¹

NEUROGENIC TUMORS

These originate from the peripheral nerves and are the second most common presacral tumors after congenital tumors and account for 10% of all presacral lesions.¹ Approximately 85% of neurogenic tumors are benign. Benign lesions include neurilemmomas, neurofibromas, and ganglioneuromas, whereas malignant lesions include ganglioneuroblastomas, neurofibrosarcomas, neuroblastomas, ependymomas, schwannomas, and malignant peripheral nerve sheath tumors.^{3,5} These tumors tend to be slow growing and cause minimal or nonspecific symptoms; therefore, at the time of diagnosis they may be of considerable size.⁵

OSSEOUS TUMORS

These include a spectrum of different tumors that originate from various osseous derivatives such as bone, cartilage, fibrous tissue, and marrow. These include chondrosarcomas, osteosarcomas, myelomas, and Ewing sarcoma. These tumors tend to grow rapidly and can be of significant size when diagnosed. When malignant, they most frequently metastasize to the lungs and are associated with a worse outcome.

INFLAMMATORY TUMORS

Uhlig and Johnson's original article classified a group of presacral lesions under this category.² These lesions included foreign body granulomas from barium leaks or surgical sutures and infectious processes originating from the abdomen (e.g., perforated diverticulitis) or the perianal region that have extended into the presacral space.² Most recent series on presacral tumors have not considered this a true category as these lesions do not originate from the presacral space, but instead the lesions are a secondary extension from another location.^{6,9,12}

MISCELLANEOUS TUMORS

These account for 10 to 25% of all presacral tumors.¹ They include masses found elsewhere in the retroper-

itoneum including metastatic disease (usually from the rectum), lymphomas, fibrosarcomas, liposarcomas, malignant fibrous histiocytomas, lymphangiomas, lipomas, leiomyomas, and hemangiomas.^{1,3,5}

DIAGNOSIS AND MANAGEMENT: HISTORY AND PHYSICAL EXAMINATION

Presacral tumors may be found incidentally during a routine physical examination. However, if symptomatic, most symptoms arise as a result of compression or invasion of surrounding pelvic viscera or nerves.^{1,4} Symptomatic patients can have vague complaints of lower back or perineal pain or can have specific symptoms like constipation, urinary or fecal incontinence, or sexual dysfunction. Pain is the most common presenting symptom for malignant lesions and for benign lesions that have become secondarily infected.¹ Patients can also complain of perianal discharge or drainage if the lesion is infected.⁵ Obstructed labor in women from presacral lesions has been reported in the literature and is the premise for removing presacral tumors in women of childbearing age even if they are asymptomatic and appear benign.^{2,13}

A comprehensive neurologic and musculoskeletal examination is important to document preoperative functional status and evaluate for possible neurologic involvement.⁵ On digital rectal examination, a presacral tumor will typically feel like an extrarectal mass displacing the rectum anteriorly with a smooth intact overlying mucosa.^{4,5} It is important to assess the most proximal extent of the lesion as well as the extent and nature of fixation and the relationship to adjacent organs such as the prostate.¹ Other characteristic physical findings reported to be associated with presacral tumors are small midline dimples just posterior to the anus and immediately below the dentate line as well as on the gluteus muscle.¹

The incidence of various presenting symptoms and physical findings is varied in the literature. Glasgow et al¹² reported minimal findings on physical examination with only 35% (12 patients) having palpable abnormalities on digital rectal exam and only 2 out of the 34 patients demonstrating gluteal dimpling. In Buchs et al¹⁴ series of 16 patients with benign presacral lesions, 75% had a palpable mass on digital examination.¹⁴ Jao et al¹⁰ estimated that 97% of patients diagnosed with presacral lesion have a palpable mass on physical examination, whereas dimpling has been reported in 35 to 100% of patients with developmental cysts.¹ This variation in physical findings is likely due to the differences in the nature and location of the presacral tumors reported in different series.

In general, the symptoms caused by presacral tumors are nonspecific and often result in a delay of diagnosis. It is therefore important to have knowledge

Figure 2 Management flowchart for presacral tumors.

of the various presenting symptoms and a high index of suspicion when evaluating these patients.¹ A history of persistent perianal drainage and multiple anorectal procedures without identification of a source of perianal sepsis, a postanal dimple, or a fullness or fixation in the precoccygeal region are subtle findings that can suggest the presence of a presacral tumor.^{4,5} An algorithm depicting the basic steps in the diagnosis and management of presacral tumors is shown in Fig. 2.

DIAGNOSTIC INVESTIGATIONS

Traditionally plain x-ray radiographs were used for the evaluation of presacral tumors, although currently computed tomography (CT) scans and magnetic resonance imaging (MRI) scans have become the diagnostic modalities of choice. Classically bony destruction of the sacrum and/or calcifications were seen on plain radiographs. These findings are most commonly seen with chordomas, but can also occur with sarcomas. The characteristic “scimitar” sign on plain radiographs is associated with

an anterior sacral meningocele, and is described as a rounded concave border of the sacrum without obvious bony destruction.³ Both CT and MRI scans are now used in a complementary manner rather than exclusively in the evaluation of presacral lesions. A CT scan can be used to determine whether a lesion is solid or cystic, evaluate cortical bone destruction, and assess involvement of adjacent viscera.^{1,5} MRI because of its multiplanar ability and superior soft tissue resolution aids in determining the planes of resection, spatial relationship to surrounding structures, and associated cord abnormalities, as well as the extent of bone marrow involvement.⁹

Other diagnostic modalities that can help in the diagnosis and management of presacral lesions include flexible endoscopy, fistulograms, and endorectal ultrasound.^{5,12} Endoscopy can help in evaluating involvement of the rectal mucosa and the proximal extent of the lesion. Fistulograms can help in the diagnosis of a chronically draining sinus that may be originating from a presacral lesion such as a developmental cyst.⁵ Endorectal ultrasound has been used to determine the nature (solid versus cystic) of retrorectal lesions and their relationship to the layers of the rectum.¹⁴ This information is useful in determining the extent of dissection and assessing the need for rectal resection.

The purpose of diagnostic investigations, particularly cross-sectional imaging, is to accurately evaluate the location and nature of the lesion and determine the extent of involvement of the surrounding structures. This, in turn, helps to determine the surgical approach (anterior versus posterior versus combined) and the intraoperative extent of resection (local excision versus en bloc resection).

PREOPERATIVE BIOPSY

The role of preoperative biopsy in the management of presacral tumors has been controversial. Traditionally, authors have considered it a contraindication to biopsy any presacral lesion that is surgically resectable.^{6,10,15-17} Proponents of this approach have cited the risk of infecting the lesion and seeding the biopsy tract with malignant cells in the case of a malignant lesion as reasons for avoiding a biopsy.^{6,10,15} However, some authors have recently suggested that a preoperative biopsy should be a consideration.⁵ The argument being that a proportion of patients with presacral lesions such as Ewing sarcoma, osteogenic sarcomas, neurofibrosarcomas, and desmoids may benefit from neoadjuvant treatment. Therefore, a preoperative biopsy can significantly influence the preoperative management strategy and can alter the extent and nature of the surgical resection.

Preoperative biopsy of a presacral lesion should therefore only be performed if it is likely to change the management and surgical approach.⁵ There is rarely an

indication to biopsy a purely cystic lesion as they are usually benign and there is a significant risk of secondary bacterial infection. Large lesions or lesions invading adjacent structures may warrant a preoperative biopsy to determine the optimal treatment strategy and possible need for neoadjuvant therapy. Preoperative biopsies of presacral lesions should not be done transperitoneally, transretroperitoneally, transrectally, or transvaginally. If lesions are biopsied through these routes there is a high risk of infection, which can make subsequent surgical excision difficult, increasing the risk of perioperative complications and local recurrence. In cases of malignant lesions, it is mandatory to remove the biopsy tract en bloc with the specimen to decrease the risk of recurrence in that tract. Transrectal or transvaginal biopsies would then require removal of these organs, when they otherwise may not have been involved. Therefore, a transperineal or presacral approach is usually ideal as the biopsy tract falls within the margins of surgical resection.

ROLE OF ADJUVANT THERAPY

Most malignant presacral tumors including chordomas and chondrosarcomas are generally considered to be poorly responsive to chemotherapy. However, the tyrosine kinase inhibitor, Imatinib, has recently been shown to be effective in the management of advanced chordomas.¹⁸ The beneficial effect of Imatinib on progression-free survival has also been supported in a multicenter phase II trial.¹⁹ The epidermal growth factor inhibitors, Cetuximab and Gefitinib, have been reported to show a good response in patients with locally recurrent and metastatic chordoma.²⁰ It is likely that with further advances in molecular-targeted therapy more effective agents will be discovered and aid in the management of patients with chordomas in both the adjuvant and neoadjuvant setting. Chemotherapy has a significant role in the management of extremity sarcomas (Ewing and osteosarcomas). Disease-free survival is significantly increased with combined adjuvant and neoadjuvant chemotherapy in the case of high-grade osteosarcomas and Ewing sarcoma of the extremity.⁵ Therefore, it is likely that this benefit can be seen with presacral sarcomas when adjuvant or neoadjuvant chemotherapy is combined with radical oncologic surgery.

The role of radiotherapy in the management of malignant presacral tumors is not clearly defined. However, the current surgical management of malignant presacral lesions such as chordomas and certain sarcomas is limited by technical difficulties in achieving complete surgical resection and local recurrence despite adequate margins. With the availability of sophisticated photon beam techniques including intensity modulated radiation therapy (IMRT) and stereotactic procedures, radiotherapy is likely to play an increasingly important role in the multimodality treatment of these lesions.

SURGICAL MANAGEMENT

Surgery is the mainstay of management of presacral tumors as it establishes a diagnosis, prevents malignant degeneration, and avoids secondary bacterial infection. The goal of surgery is to remove the presacral lesion in its entirety with minimal morbidity to the patient. Incomplete removal of the lesion can result in recurrence of symptoms. It also increases local recurrence and decreases survival rates in the case of malignant presacral lesions. The surgical approach and extent of resection is determined by the location, nature, and size of the lesion while taking into account whether or not the sacrum, pelvic sidewall, or adjacent viscera are involved.⁹ There are three common approaches for resection of presacral tumors:

Anterior approach (transabdominal)
 Posterior approach (perineal)
 Combined abdominoperineal approach

Small low-lying lesions below the level of the S-3 vertebrae can be removed through a posterior approach. Tumors extending above the level of the S-3 vertebrae can be removed either through an anterior transabdominal incision or through a combined anterior and posterior approach depending on the need for concurrent sacrococcygeal resection (Fig. 3). The extent of resection is determined by the nature of the presacral lesion. Benign lesions can be removed with limited dissection as long as the lesion itself can be completely resected. Malignant lesions require a radical resection particularly if they are adherent to adjacent structures in which case en bloc resection of the lesion and involved structures becomes necessary. The adjacent organs that can be involved include the pelvic vessels, the sacral nerves, the sacrum, and the rectum. Most of these

Figure 3 Relationship of tumor to sacral level and proposed approach. From Mayo Clinic, Rochester, NY. Reproduced with kind permission of Springer Science and Business Media.

structures can be technically resected; nevertheless, there can be significant morbidity involved. Although unilateral resection of all the sacral nerve roots will not compromise fecal and urinary function, if both S-3 nerve roots are resected, the external anal sphincter will not function and the patient will become incontinent.⁵ A majority of the sacrum can be resected if necessary as long as more than half of the S1 vertebra is preserved. However, an extensive sacrectomy can be associated with significant perioperative morbidity and functional disability.^{10,21,22} Resection of the rectum may or may not require a permanent colostomy, which in itself can impair patient quality of life. Therefore, it is important to weigh the benefits of extended resections against the risks associated with them in terms of patient outcomes.

PREOPERATIVE CONSIDERATIONS

Surgical management of small presacral lesions can be relatively straightforward as long as the surgeon understands the anatomy of the region and is familiar with the different technical approaches to remove these lesions. Large presacral lesions, particularly if they are malignant and/or if they involve the surrounding structures, can be technically challenging and require a multispecialty approach with careful preoperative planning. Allied specialties usually involved in these procedures include orthopedics, neurosurgery, vascular surgery, plastic surgery, and medical and radiation oncology.

The following are a few preoperative considerations that need to be taken into account when surgically managing these lesions.^{5,9}

1. Accurate preoperative imaging (and restaging after neoadjuvant therapy) to determine the relationship of the tumor to the adjacent structures and the margins of resection.
2. Adequate nutrition with supplemental parental or enteral nutrition if necessary.
3. Placement of an inferior vena cava filter as there is a significant risk for deep venous thrombosis and pulmonary embolism.
4. Equipment and personnel to handle massive transfusion requirements intraoperatively.
5. Appropriate positioning of the patients according to the surgical approach while adequately protecting pressure points and avoiding pressure injury to the nerves and soft tissue.
6. Ureteric stents to aid in the intraoperative identification of the ureters particularly in patients who have large tumors or have received neoadjuvant therapy.

THE POSTERIOR APPROACH

The posterior approach is preferred for small, benign lesions that do not extend above the level of the S-3

vertebrae. The posterior approach is comprised of many techniques including the transsphincteric, transsacral, transrectal, transanorectal, and transsacrococcygeal approaches. Each of these techniques has their own merit and can be used depending on the nature and location of the tumor and the surgeon's experience.

For a transsacrococcygeal approach the patient is placed in the prone jack-knife position and the buttocks are taped apart (Fig. 4A). A longitudinal incision is made over the lower part of the sacrum and the coccyx down to the anoderm while carefully avoiding damage to the anal sphincter complex (Fig. 4B). Transaction of the anococcygeal ligament facilitates exposure of the tumor and separation of the lesion from the mesorectum. The coccyx may be disarticulated if needed to improve exposure, with or without detachment of the gluteus maximus and a concurrent distal sacrectomy (S-4 and S-5).¹⁶ The lesion then can be dissected from the surrounding tissues including the rectal wall which is usually uninvolved. In cases of benign lesions there is usually a fat plane between the lesion and the mesorectum which facilitates this dissection. In the case of very small lesions, particularly if they are cystic, the

surgeon may double glove his or her nondominant hand and, with the index finger in the anal canal and the lower one-third of the rectum, push the lesion out toward the incision. This maneuver allows dissection of the lesion off the rectal wall and prevents iatrogenic injury to the rectal wall (Fig. 4C). Prior infection of the lesion may make this dissection difficult, particularly if the plane between the lesion and rectum is obliterated. In this case if the lesion is adherent to the rectum and cannot be safely separated a portion of the rectal wall can be excised along with the lesion and the defect is repaired in two layers. Routine resection of the coccyx used to be recommended particularly in the case of teratomas. Currently, this is not advocated unless the coccyx is directly invaded by a malignant lesion or a lesion of uncertain malignant potential.¹

Very low lying, small and benign presacral tumors can also be managed through an intersphincteric approach.¹⁴ The patient is placed in either the prone jack-knife or the lithotomy position. A V-shaped or radial incision is made posterior to the anus and the intersphincteric plane is entered. The anal canal and the internal sphincter are bluntly separated from the external sphincter up to the level of the puborectalis muscle. The dissection is then continued in a cephalad direction into the presacral space and the lesion can be dissected out using a combination of sharp and blunt dissection.

Figure 4 (A) Positioning for posterior approach. (B) Coccygectomy. (C) Index finger in anal canal to push tumor outward to facilitate dissection. From Mayo Clinic, Rochester, NY. Reproduced with kind permission of Springer Science and Business Media.

ANTERIOR APPROACH

The anterior approach is usually performed for lesions that have their lowest extent above the level of the S-4 vertebrae and do not have evidence of sacral involvement. After a midline incision is made, the rectum is dissected off the anterior aspect of the lesion and then the posterior aspect of the lesion is dissected off the presacral fascia. If the middle sacral artery is the main arterial supply of the lesion, then it has to be ligated prior to removing the lesion. With advances in minimally invasive techniques, a laparoscopic approach to removing these tumors has been shown to be a safe and feasible option.^{4,17,23,24} In particular, the better visualization offered by laparoscopy is considered to be an advantage over the traditional approach through a laparotomy.

COMBINED ABDOMINOPERINEAL APPROACH

If the presacral tumor extends above and below the level of the S-3 vertebrae, an anteroposterior approach is preferred.^{5,9,16} This approach is ideal for an anterior sacral meningocele, as the communicating stalk can be identified from a posterior approach and then ligated anteriorly through an abdominal incision.

In a combined approach the patient is usually placed in the synchronous (modified dorsal lithotomy)

position or a "sloppy lateral" position to facilitate the combined anterior-posterior exposure.⁵ The abdomen is entered through a low midline incision and the peritoneal cavity is explored to rule out peritoneal or metastatic disease in cases of malignant lesions. After mobilizing the sigmoid colon, the presacral space is entered below the sacral promontory and the mesorectum is dissected off the presacral fascia down to the upper extent of the lesion. The lateral stalks of the rectum are separated from the tumor if they are attached. If the tumor can be separated from the posterior aspect of the mesorectum, the lesion is dissected free in a plane anterior to the mass between its capsule and the mesorectum. Posteriorly, if there is a plane between the lesion and the sacrum, this is also developed. Isolated lesions that do not invade adjacent organs or structures can be dissected free circumferentially in this manner and removed. If the tumor is large it may be adherent to the rectum and an attempt to dissect between the lesion and the mesorectum may result in an undetected iatrogenic injury to the rectum. In this situation, it is then necessary to remove the lesion en bloc with the rectum. If the lesion is benign or considered to have a low risk of recurrence, reestablishment of intestinal continuity can be undertaken with or without a protective diverting stoma. If the tumor extends high on the sacrum with evidence of invasion of both S-3 roots and/ or S-2 roots, then en bloc resection of the rectum, sacrum, and the nerve roots becomes necessary. To maintain fecal and urinary continence at least one S-3 nerve root needs to be preserved. Therefore, if both S-3 nerves are resected, reestablishment of intestinal continuity is not feasible as the patient will be incontinent and a sigmoid end colostomy should be constructed.

Resection of large complex presacral lesions may result in significant blood loss from radiated pelvic blood vessels or the sacrectomy itself. In these situations ligation of the middle sacral vessels as well as the internal iliac vessels and its branches helps reduce blood loss. Preservation of the anterior division of the internal iliac artery which gives off the inferior gluteal artery, reduces the risk of perineal necrosis.⁵ Another intraoperative maneuver that can be helpful is to mobilize the ureters along with the periureteral tissues and secure them laterally with absorbable suture away from the planned margin of the sacrectomy.⁵ With extended resections, particularly if the pelvis has been radiated, a well-vascularized musculocutaneous flap derived from the rectus abdominis can be used to close the perineum. After closure of the abdominal incision and construction of the stoma, the patient is moved into the prone position.⁵ A midline incision is made over the sacrum and coccyx down to the anus and the anococcygeal ligament is transected while the levators are retracted bilaterally. If the rectum is uninvolved, its posterior aspect is separated from the tumor. The gluteus maximus muscles are dissected bilaterally and the sacrospinous and sacrotuberous ligaments as well as the

piriformis muscles are divided to expose the sciatic nerves. An osteotomy is then performed at the level of S-3 or higher if necessary after exposing and preserving at least one of the S-3 nerve roots. The lesion can then be removed en bloc with the attached sacrum, coccyx, and involved sacral roots, with or without the rectum. With resection of the sacral nerves it is important to recognize dural tears and repair them to prevent a CSF leak and recurrent infections. The perineal wound is closed over drains with a musculocutaneous flap used if indicated.⁵

RESULTS OF SURGICAL TREATMENT

Malignant Lesions

The results of surgical treatment for malignant presacral neoplasms depend on the biologic nature of the lesion and the extent of resection and therefore varies among studies. In Galsgow et al's¹² report, all seven patients with malignant presacral tumors developed recurrence of their disease despite adequate resection and had a median survival of 61 months (range 37 to 108 months). Woodfield et al⁹ found no recurrences or mortality among three patients with malignant presacral tumors other than chordomas with a median follow-up of 26 months (range 10 to 61 months). Lev-Chelouche et al⁶ reported an 80% complete resection rate in 12 patients with presacral tumors other than chordomas with a 67% local recurrence and 50% survival.

The prognosis after surgical management of patients with chordomas, which is the most common malignant presacral tumor, has ranged from 15% in older reports to 84% in more contemporary series.^{5,25} The most significant prognostic factor for patients with chordoma is the surgical margins. The risk of local recurrence in the case of a marginal resection is ~70% and has an adverse impact on survival.²⁶ However, despite adequate surgical resection a significant proportion of patients develop locally recurrent disease indicating the need for improved adjuvant therapies. In a report published in 1985, Jao et al¹⁰ reported a 5-year survival rate of 75% for chordomas; the same group has recently found a 5- and 10-year survival rate of 80% and 50%, respectively, for these patients.⁵ Lev-Chelouche et al⁶ reported a complete resection rate of 66% with a 44% local recurrence and 89% survival for patients with chordomas in their series. In Woodfield et al's⁹ experience local recurrence developed in two out of the four chordomas that were resected with no mortality, although they did not specify the margins. Bergh et al²⁵ reported a 10-year survival rate of 84% and a 44% recurrence rate for chordomas in their experience. The majority of the published reports on chordomas are from single institutions and to an extent carry a referral bias regarding its incidence and prognosis. McMaster et al²⁷ evaluated 400 cases of chordomas reported to nine

population-based registries within the National Cancer Institute's Surveillance, Epidemiology and End Result (NSEER) program over a 22-year period from 1973 to 1995. The 5- and 10-year survival rate for sacral chordomas in the NSEER database was found to be 74% and 32%, respectively, and more likely represents the population-based incidence and outcome of these lesions.

The overall survival for benign lesions is almost 100%; however, recurrence rates can vary depending on the extent of resection, as incompletely resected tumors are likely to reoccur.¹ In Buchs et al¹⁴ report of 16 patients with a median follow up of 5 years with benign presacral tumors that underwent complete macroscopic resection by a posterior approach, only one patient with a tailgut cyst developed a recurrence. Glasgow et al¹² reported none of the patients with benign presacral tumors developed recurrence after a median follow-up of 22 months. Lev-Chelouche et al⁶ reported a 100% survival and no recurrences after complete resection in their experience with 21 benign presacral tumors.

CONCLUSION

Because of their rarity, the diagnosis of presacral tumors can be challenging. Accurate and reliable diagnostic imaging is essential to identify the optimal surgical approach. Surgical management should be determined by the nature and location of the lesion and the extent of involvement of surrounding structures while minimizing the morbidity to the patient.

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