Retrorectal Tumors

Sean C. Glasgow, M.D.¹ and David W. Dietz, M.D.^{1,2}

ABSTRACT

Primary neoplasms of the retrorectal (presacral) space are very rare. These lesions may be congenital or acquired, benign or malignant. They often arise with subtle clinical symptoms, or they may be found incidentally during evaluation for other conditions. Preoperative imaging can provide useful information for operative planning; however, it does not eliminate the need for surgery. Approaches for resection include posterior only and combined abdominoperineal, depending on the characteristics of the lesion. Successful treatment of these enigmatic lesions requires extensive knowledge of pelvic anatomy and expertise in pelvic surgery.

KEYWORDS: Retrorectal tumors, presacral space, chordoma, teratoma

Objectives: Upon completion of this article, the reader should be able to summarize the differential diagnosis, evaluation, and treatment options for lesions of the retrorectal space.

Primary tumors of the retrorectal (or presacral) space are rare lesions. Constituting only 1 in 40,000 hospital admissions in a large series from the Mayo Clinic, they produce symptoms that are often underappreciated by clinicians and the tumors are commonly misdiagnosed, even after extensive radiological work-up.¹ Historical reports have estimated the incidence to be 1 in about 62,000 admissions.² The Cleveland Clinic reported on 24 sacral or retrorectal tumors over 12 years, and we published results for 34 patients with primary retrorectal tumors treated over 22 years.^{3,4} The majority of retrorectal tumors are benign; malignant tumors are notoriously difficult to excise for cure, although they can be slow growing and rarely metastasize until late in their course. The diagnosis of these lesions requires a high index of suspicion, and treatment should be undertaken only by surgeons with expertise in complex pelvic operations.⁵ Although retrorectal tumors can be observed in children (typically teratomas, meningoceles, or large

congenital duplication cysts), this review focuses only on lesions diagnosed in adulthood.

HISTORICAL PERSPECTIVE

Largely because of their clinical rarity and occult nature, the description of retrorectal tumors in modern medicine has occurred relatively recently. In 1885, a report was made of a suppurative dermoid cyst found during autopsy of a young woman.⁶ The lesion was broadly attached to the sacrum and had caused pelvic outlet obstruction during labor, leading to the patient's death. In 1891, Page reported the successful excision of a presacral dermoid cyst through a transverse incision between the anus and coccyx.⁷ Subsequently, others reported benign and malignant sacrococcygeal teratomas.^{8,9}

The German surgeon Kraske is credited with the development of the transcoccygeal approach for the resection of rectal cancers in 1886.¹⁰ He found that

¹Section of Colon and Rectal Surgery, Department of Surgery, Washington University School of Medicine and Barnes-Jewish Hospital, St. Louis, Missouri; ²Siteman Cancer Center, St. Louis, Missouri.

Address for correspondence and reprint requests: David W. Dietz, M.D., Section of Colon and Rectal Surgery, Department of Surgery, Washington University School of Medicine, 660 S. Euclid Ave., Box 8109, St. Louis, MO 63110. E-mail: mdietzd@wustl.edu.

Uncommon Colorectal Neoplasms; Guest Editor, Matthew G. Mutch, M.D.

Clin Colon Rectal Surg 2006;19:61-68. Copyright © 2006 by Thieme Medical Publishers, Inc., 333 Seventh Avenue, New York, NY 10001, USA. Tel: +1(212) 584-4662.

DOI 10.1055/s-2006-942346. ISSN 1531-0043.

removal of the coccyx and the sacrococcygeal ligament allowed greater access to the rectum with limited morbidity; however, there is no report that he used this approach for the treatment of a true retrorectal primary tumor. The Kraske procedure and a strictly transperineal route were the most commonly employed operative approaches to the presacral space during the early 20th century.

The first successful combined anteroposterior resection for a presacral tumor was performed in 1945 by Bowers at Walter Reed Army Medical Center.¹¹ An initial posterior approach for a presumed chordoma was unsuccessful because of extensive vascular collateralization. The patient later was returned to the operating room for a laparotomy, during which both internal iliac arteries were ligated and divided, and the common iliac arteries were temporarily occluded with tapes. He was then placed prone and a sacrectomy to the S1 level proceeded without difficulty, after which flow was reestablished through the iliac vessels and the abdomen closed. Despite bowel incontinence and urinary retention, the patient reportedly made a full recovery and returned to work.

EMBRYOLOGY AND ANATOMY

The retrorectal space exists near the confluence of the embryologic hindgut, bony pelvis, and neuroectoderm of the spinal cord. Lesions may arise *de novo* from any of these structures or congenital variants may be present. Anatomically, the retrorectal space is the pelvic continuation of the retroperitoneum. In adults, this space is filled with loose areolar tissue, fat, and connective tissues. Its boundaries are the rectum anteriorly and the sacrum and investing sacral fascia posteriorly. Some authors include the bony sacrum and its nerve roots as part of the retrorectal space because some retrorectal lesions arise for these structures (e.g., chordoma, neurogenic tumors). Although the rectosacral (Waldeyer's) fascia separates the true retrorectal space from the more inferior/caudal horseshoe-shaped supralelevator space, lesions in both regions are typically considered together, with the levator and coccygeal musculature constituting the inferior boundary. The ureters, internal iliac arteries, and lateral rectal stalks (at the lower levels) mark the lateral extent of the retrorectal space, and it extends cranially to the peritoneal reflection.

PATHOLOGY

Various classification schemes have been reported for retrorectal tumors. Most commonly, these tumors are broadly categorized as *inflammatory*, *congenital*, *neurogenic*, *osseous*, and *miscellaneous* (Table 1). Others have proposed *benign* versus *malignant* and *congenital* versus *acquired*.¹² With improved preoperative imaging and the widespread use of colonoscopy, most modern series

Table 1 Differential Diagnosis of Retrorectal Tumors

Congenital

- Developmental cyst (epidermoid, dermoid)
- Cystic hamartoma (rectal duplication)
- Teratoma
- Teratocarcinoma
- Chordoma
- Adrenal rest tumor
- Anterior sacral meningocele

Inflammatory

- Perineal or pelvirectal abscess
- Endometriosis
- Foreign body granuloma
- Infectious granulomas
- Diverticulitis
- Crohn's disease

Neurogenic

- Neurofibroma
- Neurilemoma
- Ependymoma
- Ganglioneuroma
- Neurofibrosarcoma

Osseous

- Osteoma
- Osteogenic sarcoma
- Ewing's tumor
- Chondromyxosarcoma
- Giant cell tumor
- Aneurysmal bone cyst

Miscellaneous

- Metastatic carcinoma
- Lipoma/liposarcoma
- Hemangioendothelial sarcoma
- Extra-abdominal desmoid
- Plasma cell myeloma
- Fibrosarcoma
- Leiomyoma/leiomyosarcoma
- Hemangioma
- Endothelioma
- Pelvic ectopic kidney

Compiled from references 4, 13, 24, 26.

exclude metastatic and/or locally advanced colorectal and genitourinary carcinomas from the category of retrorectal tumors. Likewise, inflammatory conditions that mimic primary tumors can usually (although not always) be diagnosed prior to surgical exploration. The most common or unique retrorectal tumors are discussed in the following.

Developmental Cysts

These lesions result from abnormal closure of the ectodermal tube (dermoid and epidermoid cysts) or sequestration of the developing hindgut (enterogenous cysts

and cystic hamartomas).¹³ Developmental cysts are always lined with epithelium: either squamous epithelium with (dermoid) or without (epidermoid) skin appendages or cuboidal, transitional, or columnar epithelium (enterogenous cysts and cystic hamartomas). Most often they are multilocular, and disorganized smooth muscle fibers can be identified surrounding their thin walls.

Dermoid and epidermoid cysts are most commonly diagnosed in females in the fourth and fifth decades of life. They are typically benign, and although they do not usually communicate with the rectum, up to 30% become infected.¹⁴ When not infected, these cysts tend to be filled with a viscid green-yellow material.¹³ Differentiating an infected retrorectal cyst from a complex perirectal abscess may be difficult, leading to repeated drainage procedures prior to arrival at the correct diagnosis. Patients with cysts occasionally have a characteristic postanal dimple, representing a vestigial communication to the embryologic ectoderm.

Enterogenous cysts, also termed rectal duplication cysts, arise from the hindgut and are endodermal in origin. As such, they are typically lined with columnar epithelium and secrete a clear to greenish mucoid material. These lesions often have a single large, dominant cyst with several smaller "daughter" cysts. Although they are usually benign, malignant transformation of rectal duplication cysts has been reported.¹⁵ Cystic hamartomas are also known as tailgut cysts or mucus-secreting cysts. They are most often lined with transitional epithelium and are believed to derive from the hindgut. Although smooth muscle bundles may be present, these cysts lack the myenteric and submucosal nerve endings found in enterogenous cysts. They are typically multicystic and contain thick mucus. Similar to the other developmental cysts, cystic hamartomas are more common in women and are almost always benign.

Teratoma and Teratocarcinoma

By definition, teratomas contain cellular elements derived from all three germ layers. They originate from totipotent cells that are abnormally sequestered during in utero development. Although the sacrococcygeal area is the most common site for teratomas in newborns, *de novo* lesions in this region are rare in adults. Teratomas are classified as mature, immature, or malignant (teratocarcinoma). Mature teratomas contain recognizable epithelial and/or mesenchymal cells, and immature tumors consist of primitive endoderm, mesoderm, or ectoderm. Malignant teratomas contain neoplastic cells of germ cell origin (e.g., choriocarcinoma, germinoma), as opposed to teratomas with malignant transformation, which contain malignant cells of somatic origin that have presumably degenerated. Only 4% of teratomas diagnosed at birth are malignant, whereas those found later in childhood tend to be

very aggressive and associated with a poor prognosis.¹⁶ Approximately 30% of adult teratomas harbor malignancy at the time of resection.

Chordoma

Chordomas arise from remnants of the embryologic notochord, a primitive streak running from the skull base to the pelvis and represented by the nucleus pulposus in normal adults. These are rare bony tumors, but about 50% of all chordomas are located in the pelvis and retrorectal space. They are the most common malignant tumor of this region. Chordomas are lobulated, gelatinous tumors that tend to be slow growing but locally destructive. Intratumoral hemorrhage can create a cystic appearance, and a pseudocapsule is commonly present. The pathognomonic feature on microscopy is the presence of "physaliferous" cells, which are vacuolated and filled with mucous droplets. Although they may occur at any age, chordomas typically arise in the sixth and seventh decades and are much more common in men.^{1,4,17} Despite the rare tendency to metastasize, chordomas are difficult to treat for cure. Symptoms are typically delayed until the tumor reaches a large size, making resection with uninvolved margins extremely difficult.

Anterior Sacral Meningocele

These are rare tumors, with only 150 cases reported in the world medical literature.¹⁸ This congenital herniation of the dural sac through the sacrum is commonly associated with other developmental abnormalities, such as spina bifida, tethered spinal cord, and genitourinary tract malformations. The sac communicates with the spinal canal and is filled with cerebrospinal fluid. Symptoms result either from local compression of other organs (e.g., the rectum, producing constipation) or from straining maneuvers causing sudden increases in the intracranial pressure (e.g., onset of headache with bowel movements). Treatment involves ligation of the neck of the dural sac prior to resection and typically can be accomplished through a posterior approach. Large anterior meningocele, such as that pictured in Figs. 1A–C, require an anterior approach with resection of the tumor followed by closure of the dural sac.

Neurogenic and Osseous Tumors

Tumors arising from pelvic nerve roots or peripheral nerves or primary bony tumors can occur in the retrorectal space. Taken together, neurogenic and osseous tumors constitute 20 to 30% of primary retrorectal tumors.⁴ Two thirds of neurogenic tumors in this region are benign; however, even benign tumors that

Figure 1 A young woman with a massive anterior meningocele. (A) Sagittal T2-weighted MR image of the tumor demonstrating communication with the dural sac. (B) Intraoperative photograph showing the tumor protruding from the abdomen after lower midline incision. (C) After resection of the tumor, the defect in the dura is visible prior to suture closure.

arise from the spinal cord can produce devastating neurological morbidity. The most common malignant neurogenic lesions are neuroblastomas, schwannomas, and ependymomas; benign tumors include neurilemmomas and ganglioneuromas. Osseous tumors can derive from cartilage, bone, or fibrous tissue. They behave similarly to bony tumors in other anatomic locations, with sarcomas having a predilection for hematogenous spread to the lungs. Diagnosis is typically made at an advanced stage, when significant destruction of the pelvis has occurred.¹⁹

Miscellaneous

Carcinoid tumors of the retrorectal space are rare but have been described. Most represent direct extension or metastatic spread from rectal carcinoids.²⁰ Retrorectal carcinoid tumors may also arise from the glandular cells present in rectal duplication or tailgut cysts.²¹ These lesions are potentially malignant and should be treated by local excision, typically using a perineal approach. Some authors have suggested the use of urinary 5-hydroxyindolacetic acid (5-HIAA) as a tumor marker to assess for complete resection and as a biochemical marker of recurrence, as for carcinoid tumors in other gastrointestinal locations.²² The reliability of this approach, however, is questionable.

CLINICAL PRESENTATION

Retrorectal tumors are frequently asymptomatic, and they are often found incidentally during evaluation for unrelated physical complaints. Furthermore, even patients with symptoms directly referable to a retrorectal tumor may be initially misdiagnosed as having fistulae in ano; pilonidal cysts; perianal abscesses; psychogenic, post-traumatic, or postpartum pain; or proctalgia fugax. In fact, Singer et al found that patients underwent an average of 4.1 surgical procedures before being correctly diagnosed with primary retrorectal pathology.⁵

Symptoms

Retrorectal tumors may produce minimal or vague symptoms for long periods of time. In fact, benign lesions are most often asymptomatic.⁴ Patients may initially complain of constipation, paradoxical diarrhea, or frequent urge to defecate, as the tumor causes compression of the adjacent rectum. In older series, retrorectal tumors in women were occasionally diagnosed during labor as the cause for dystocia. Pain (either pelvic discomfort or sciatic symptoms) is more common with malignant retrorectal tumors. Two large series have demonstrated that pain is present in 86 to 88% of patients with malignancy versus 22 to 39% of those with benign lesions.^{1,4} Other factors predictive of malignancy include male gender and age

older than 60 years. Chordomas are classically associated with constant dull aching in the low back, pelvis, and buttocks. This pain is typically worsened by sitting and relieved with standing or walking. Patients with anterior sacral meningoceles may complain of headaches after straining during defecation or intercourse, and their symptoms may be reproduced with Valsalva.²³ The diagnosis of retrorectal cyst should be suspected in any patient with a history of repeated attempts at drainage for a fistula in ano or perirectal abscess, especially when no obvious internal rectal opening can be identified or mucoid material (rather than pus) is drained.¹³ Malignant tumors that invade the sacral plexus or nerve roots can lead to bowel and bladder incontinence or urinary retention.

Physical Examination and Proctoscopy

Most large series describing retrorectal tumors report that these lesions are almost always palpable on digital rectal examination.^{1,19,24} The presence of a postanal skin dimple is suggestive of a developmental cyst, although it is unclear how commonly these conditions coexist and in our experience only 7% of patients with a benign tumor have any external examination findings.⁴ Determination of the cranial extent of the lesion is important for operative planning, as is establishing its relationship to the coccyx and sacrum and whether the tumor seems fixed or freely mobile within the pelvis. Most retrorectal tumors feel soft and compressible. Teratomas may contain abnormal calcifications that are palpable. A tender mass is suggestive of either an infected developmental cyst or a primary perirectal abscess with supralelevator extension.

All patients with suspected retrorectal tumors should undergo complete colonoscopy and women should have detailed bimanual pelvic examinations to exclude the much more common conditions of colorectal or ovarian/uterine adenocarcinomas, respectively. The

rectal mucosa overlying a retrorectal tumor appears normal. Small lesions may not be detectable on proctoscopy, but larger lesions demonstrate extraluminal compression of the rectum.

IMAGING EVALUATION

Traditionally, evaluation of suspected retrorectal tumors began with plain radiographs of the pelvis. These are typically normal and provide little additional information. The exceptions are malignant tumors that may produce bone destruction of the sacrum, as well as the rare benign bone-based lesions such as aneurismal bone cysts, giant cell tumors, and osteochondromas.¹³ Teeth or small bone fragments may be visible in presacral teratomas. The "scimitar sign" is characteristic for anterior sacral meningocele; the sacrum has a concave deformity suggestive of the curved sword of the same name.²⁵ Transrectal ultrasonography (TRUS) appears to have utility in establishing the diagnosis of a retrorectal tumor. We found TRUS to have a sensitivity of 100% when combined with rigid proctoscopy.⁴ Others have also described the use of ultrasonography to characterize these lesions.⁵ Although the specificity of TRUS is unknown, it is probably safe to assume that a normal TRUS study performed by an experienced surgeon effectively rules out a retrorectal tumor.

Tomographic imaging, with either computed tomography (CT) or magnetic resonance imaging (MRI), has become the standard for the preoperative evaluation of retrorectal tumors. In particular, MRI with an endorectal coil can provide detailed images depicting the relationship of the tumor with sacral nerve roots, the coccyx, and the musculature of the pelvic floor (Figs. 2A, B). MRI also appears to be slightly more specific for establishing histology of the retrorectal tumor.⁴ Either modality demonstrates whether the lesion is cystic or solid as well as the extent of the lesion. Interestingly, even though the tumor may be readily

Figure 2 Representative (A) axial and (B) sagittal T2-weighted MR images of a retrorectal cystic hamartoma.

visible on preoperative imaging, radiologists have difficulty in differentiating benign from malignant lesions.⁴ Sixty percent of solid tumors are malignant, whereas 90% of cystic lesions are benign. Thus, although certain characteristics on imaging may be suggestive of a benign pathology, imaging alone cannot obviate the need for surgical resection.

SURGICAL APPROACH

Indications for Surgery

In appropriate surgical candidates, all retrorectal tumors should be resected, even if asymptomatic. Ludwig and Reynolds have previously summarized the reasons for operative extirpation as follows¹³:

1. Cystic lesions may become spontaneously infected, making subsequent resection difficult and increasing the odds of recurrence, leading to the need for reoperative intervention.
2. A presacral mass in a woman of childbearing age can cause dystocia and prevent vaginal delivery.
3. Even benign-appearing cystic lesions may harbor occult malignancy.
4. Teratomas may degenerate to malignancy.
5. Resolution of symptoms may improve quality of life, and resection provides ease of mind for patients with retrorectal tumors of uncertain pathology.

The wisdom of a multidisciplinary approach involving colorectal surgeons, neurosurgeons, and possibly orthopedists cannot be overemphasized, especially with tumors involving the sacrum and sacral nerve roots. In general, preservation of at least unilateral S3 nerve roots typically allows normal bowel and bladder function following sacral resection.^{2,17} Resection of higher sacral nerve roots leads to bowel and bladder incontinence and impotence. The surgical approach should be based on preoperative imaging and expected extent of resection. Regardless of the approach, all patients should undergo preoperative bowel preparation and receive prophylactic intravenous antibiotics.

The Role for Biopsy

There is almost no role for biopsy of a retrorectal tumor prior to surgical resection, and such practice should be discouraged among other practitioners who may diagnose these lesions (e.g., gastroenterologists performing endoscopic ultrasonography). Transrectal biopsy can lead to superinfection of cystic lesions, complicating subsequent resection. It may also produce rapidly fatal meningitis in patients with anterior sacral meningocele. Furthermore, there are reports of needle-tract seeding following percutaneous CT-guided biopsies of chordo-

mas.¹³ Retrorectal tumors found incidentally during laparotomy for other conditions should not be biopsied, as biopsy results rarely alter the need for subsequent resection. Perhaps the only indication for biopsy is to make a diagnosis in the patient who is not a candidate for surgery but may benefit from palliative radiation or chemotherapy.

Abdominal- or Perineal-Only Approaches

The perineal-only approach is typically reserved for smaller retrorectal tumors that lie mostly caudal to the S4 level. The upper extents of such lesions are typically palpable on digital rectal examination. An abdominal-only approach may be utilized for high retrorectal tumors that do not involve the sacrum and lie above S4; however, most large lesions are best treated with a combined abdominoperineal approach as discussed subsequently.

For the perineal approach, the patient is placed in the prone jack-knife position with the buttocks retracted laterally with tape. The rectum is prepared by irrigation with iodine solution. Either a transperineal or parasacral incision may be utilized. After the incision is made and muscular attachments to the coccyx and lower sacrum are divided, the coccyx can be dislocated and resected. This maneuver is strictly to facilitate further exposure and resection. Whether coccygectomy decreases recurrence rates is unclear, with various authors offering differing opinions.^{1,19,26,27} Teratomas and dermoid/epidermoid cysts often arise from the coccyx and associated cells, and en bloc coccygeal resection with these tumors seems prudent.

Once the coccyx has been disarticulated, the presacral space is readily accessible and dissection of the tumor from the posterior wall of the rectum can proceed. Alternatively, further sacral disarticulation can be performed as needed. Bimanual palpation or intraoperative rigid proctoscopy can be useful to ensure that the rectal mucosa is not violated. Extensive involvement of the rectal wall can necessitate en bloc proctectomy. In our experience, approximately one third of retrorectal tumors require concurrent rectal resection.⁴ Tumors requiring proctectomy are typically best managed using a combined abdominoperineal approach. Following tumor excision, a closed suction drain is placed in the retrorectal space and the muscles are reapproximated. For large tumors, closure of the pelvic floor with a gluteal flap (often in conjunction with a plastic surgeon) can aid in filling the residual tissue defect.

Combined Abdominoperineal Approach

As mentioned previously, large tumors or those with extension above S4 require a combined abdominoperineal approach. Not surprisingly, smaller tumors amenable to a perineal-only approach tend to result in less

blood loss and fewer transfusions than those in which a combined approach is indicated.⁴ However, there is no difference in overall operative morbidity. For the combined approach, the procedure begins with the patient in either the supine or modified lithotomy position and the abdomen is opened using a low midline incision. The sigmoid colon is mobilized medially and the inferior mesenteric artery identified. Both ureters and the hypogastric nerves are also identified, swept laterally, and preserved. The retrorectal space is entered by incising the peritoneum in the posterior midline at the sacral promontory. The areolar plane between the mesorectal fascia and presacral fascia is then sharply dissected, proceeding caudally until the tumor is encountered. The middle sacral vessels can be ligated, as can branches from the hypogastric arteries, to gain vascular control of the tumor and reduce bleeding during the deeper portions of the pelvic dissection and the subsequent perineal phase of the operation. The upper aspect of the tumor is then carefully dissected from the surrounding structures, typically the presacral fascia posteriorly, the levators laterally, and the posterior wall of the rectum anteriorly. If the tumor is adherent to the rectum, this attachment should not be violated. En bloc proctectomy should be performed, with most patients able to undergo restoration of intestinal continuity by coloanal or low-colorectal anastomosis.

Once vision becomes limited in the deep pelvis and accurate planes of dissection can no longer be defined, the abdomen is closed and the patient is either repositioned in the prone jack-knife position (our preference in cases in which proctectomy is not needed) or left in the modified lithotomy position (proctectomy needed) for the perineal phase of the operation, which then proceeds as previously described. Great care should be taken to avoid injury to the sphincter complex or its innervation. If en bloc proctectomy has been necessary and the resection completed in the prone jack-knife position, the patient can be returned to the modified lithotomy position for creation of the anastomosis. In cases in which proctectomy is anticipated, we usually perform the entire procedure in the modified lithotomy position as this saves considerable operative time spent repositioning the patient.

ADJUVANT THERAPY

The rarity and heterogeneity of retrorectal tumors make it difficult to demonstrate the efficacy of adjuvant treatment. Benign lesions and teratomas with malignant degeneration should be considered cured following complete surgical extirpation. Conversely, achieving curative resections of malignant lesions, especially chordomas, can be very difficult. Aggressive postoperative radiation therapy has been reported for sacrococcygeal chordomas with favorable results, with some data suggesting that

the recurrence-free interval in irradiated patients is longer than that in those treated only with resection.^{3,28} These data notwithstanding, in our experience median overall survival for patients with malignant retrorectal tumors was 61 months, with a median time to recurrence of 24 months. These lesions tend to be slow growing but locally aggressive and produce significant pain and disability. Radiation therapy may have a role in palliation of unresectable presacral tumors. Given the rarity of true retrorectal malignancies, it is unlikely that a definitive study examining the role of adjuvant therapy will be undertaken, and the utility of external radiation will remain anecdotal.

CONCLUSIONS

Retrorectal tumors remain a difficult diagnostic and therapeutic challenge despite the use of newer imaging modalities and improvements in perioperative care. Although they are useful for planning resection, neither CT nor MRI can provide a definitive diagnosis. TRUS is highly sensitive for retrorectal tumors when used in combination with history and physical examination. Specifically, failure to visualize a lesion in the retrorectal space using TRUS obviates the need for further costly imaging to rule out a retrorectal tumor. Complete resection with negative margins is the standard for benign retrorectal tumors, but it is more difficult to resect malignant tumors for cure, and they tend to recur locally despite the use of adjuvant radiation. A multidisciplinary approach, including colorectal surgeons, neurosurgeons, and radiation oncologists, is likely to improve the rate of successful treatment for these enigmatic lesions.⁴

DISCLAIMER

The authors of this article and their spouses have no financial interest in the topics discussed in this article.

REFERENCES

1. Jao S-W, Beart RW, Spencer RJ, Reiman HM, Ilstrup DM. Retrorectal tumors: Mayo Clinic experience, 1960–1979. *Dis Colon Rectum* 1985;28:644–652
2. McCune WS. Management of sacrococcygeal tumors. *Ann Surg* 1964;159:911–918
3. Bohm B, Milsom JW, Fazio VW, Lavery IC, Church JM, Oakley JR. Our approach to the management of congenital presacral tumors in adults. *Int J Colorectal Dis* 1993;8:134–138
4. Glasgow SC, Birnbaum EH, Lowney JK, et al. Retrorectal tumors: a diagnostic and therapeutic challenge. *Dis Colon Rectum* 2005;48:1581–1587
5. Singer MA, Cintron JR, Martz JE, Schoetz DJ, Abcarian H. Retrorectal cyst: a rare tumor frequently misdiagnosed. *J Am Coll Surg* 2003;196:880–886

6. Barnes F. Pelvic tumor obstructing labor. *Br Med J* 1885;1: 420–422
7. Page F. Large extraperitoneal dermoid cyst successfully removed through an incision across the perineum, midway between the anus and the coccyx. *Br Med J* 1891;1:406–409
8. Lisco H. Malignant tumors developing in sacrococcygeal teratomata. *Ann Surg* 1942;115:378–389
9. Neal MP, Carlisle JB. Congenital sacrococcygeal tumors, a case report of teratoma. *South Med J* 1943;36:677
10. Kraske P. Zur exstirpation hochsitzender Mastdarmkrebs. *Arch Klin Chir* 1886;33:563
11. Bowers RF. Giant cell tumor of the sacrum—case report. *Ann Surg* 1948;128:1164–1172
12. Lev-Chelouche D, Gutman M, Goldman G, et al. Presacral tumors: a practical classification and treatment of a unique and heterogenous group of diseases. *Surgery* 2003;133:473–478
13. Ludwig KA, Reynolds HL. Retrorectal tumors. *Clin Colon Rectal Surg* 2002;15:285–293
14. Abel ME, Nelson R, Prasad ML, Pearl RK, Orsay CP, Abcarian H. Parasacrococcygeal approach for the resection of retrorectal developmental cysts. *Dis Colon Rectum* 1985;28: 855–858
15. Springall RG, Griffiths JD. Malignant change in retrorectal duplication. *J R Soc Med* 1990;83:185–186
16. Pantoja E, Rodriguez-Ibanez I. Sacrococcygeal dermoids and teratomas: historical review. *Am J Surg* 1976;132:377–383
17. Baratti D, Gronchi A, Pennacchioli E, et al. Chordoma: natural history and results in 28 patients treated at a single institution. *Ann Surg Oncol* 2003;10:291–296
18. Anderson FM, Burke BL. Anterior sacral meningocele: a presentation of three cases. *JAMA* 1977;237:39–42
19. Cody HS III, Marcove RC, Quan SH. Malignant retrorectal tumors: 28 years' experience at Memorial Sloan-Kettering Cancer Center. *Dis Colon Rectum* 1981;24:501–506
20. Horenstein MG, Erlandson RA, Gonzalez-Cueto DM, Rosai J. Presacral carcinoid tumors: report of three cases and review of the literature. *Am J Surg Pathol* 1998;22:251–255
21. Mourra N, Caplin S, Parc R, Flejou J-F. Presacral neuroendocrine carcinoma developed in a tailgut cyst. *Dis Colon Rectum* 2003;46:411–413
22. Edelstein PS, Wong WD, La Valleur J, Rothenberger DA. Carcinoid tumor: an extremely unusual presacral lesion. *Dis Colon Rectum* 1996;39:938–942
23. Oren M, Lorber B, Lee SH, et al. Anterior sacral meningocele: report of five cases and review of the literature. *Dis Colon Rectum* 1977;20:492–505
24. Uhlig BE, Johnson RL. Presacral tumors and cysts in adults. *Dis Colon Rectum* 1975;18:581–596
25. Kovalcik PJ, Burke JB. Anterior sacral meningocele and the scimitar sign: report of a case. *Dis Colon Rectum* 1988;31: 806–807
26. Freier DT, Stanley JC, Thompson NW. Retrorectal tumors in adults. *Surg Gynecol Obstet* 1971;132:681–686
27. Ng EW, Porcu P, Loehrer PJ. Sacrococcygeal teratoma in adults: case reports and a review of the literature. *Cancer* 1999; 86:1198–1202
28. Amendola B, Amendola M, Oliver E, McClatchey K. Chordoma: role of radiation therapy. *Radiology* 1986;158: 839–843