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Retrorectal tumors

Jeffrey A. Neale, MD, FACS, FASCRS[a,b,*]

[a] Department of Colon and Rectal Surgery, Henry Ford Hospital, 2799 West Grand Blvd, Detroit, MI 48202
[b] The Colorectal Institute, Fort Myers, FL

A B S T R A C T

Tumors that arise in the retrorectal (presacral) space are uncommon lesions. Their diagnosis and management remain difficult. These tumors present with nonspecific signs and symptoms, thereby leading to a difficulty in diagnoses. For complete evaluation of the lesion, cross-sectional imaging, most importantly MRI, is required for diagnostic purposes and can distinguish between benign and malignant lesions, as well as the extent of resection. MRI is also utilized to determine the most appropriate surgical approach. Surgical removal leads to favorable outcomes for patients with benign purely cystic retrorectal tumors. Preoperative tissue diagnosis with transperineal and transsacral biopsies of solid or heterogeneous cystic lesions plays a role in determining the necessity of neoadjuvant therapy, which may decrease local recurrence after surgery and avoid an unnecessary delay in systemic therapy. However, to perform a biopsy, one must abide by certain principles during the biopsy.

Introduction

The retrorectal (presacral space) is an area that may become occupied by a group of heterogeneous and rare tumors. These lesions range from benign cysts to complex malignant masses that can invade the surrounding pelvic structures. These tumors are often indolent and produce nonspecific signs and symptoms. As a result of delayed diagnosis, the tumors can reach a substantial size and may involve other organs at the time of their diagnosis. In recent years, the advances in imaging modalities, understanding of tumor biology, and adjuvant therapy have improved the management of these tumors.

The retrorectal space consists of various embryological remnants originating from various tissues. Most tumors located in this space are benign, with malignant neoplasms being more common in pediatric patients and solid lesions more likely being malignant than cystic lesions.

The incidence of the retrorectal tumors in the general population is unknown. However, there are several retrospective series which support that between one and six patients are diagnosed yearly in tertiary referral centers.[1–3] Uhlig and Johnson[4] demonstrated the incidence of two presacral tumors per year in a metropolitan population at a non-tertiary referral center, and Jao et al.[5] found that retrorectal tumors accounted for 1 in 40,000 hospital admissions.

Although they are infrequent, the understanding of anatomy, presentation, ideal imaging modalities, and management of these lesions will avoid the significant morbidity and complications associated with incorrect diagnosis and inappropriate management.[1]

Clinical anatomy and physiology

The retrorectal space lies above the presacral fascia, between the upper two-thirds of the rectum and sacrum. This is the site that contains totipotential cells and is where fusion of the embryologic hindgut and the neuroectoderm occurs. A variety of tumors found in this space are extensions of tumors arising from adjacent tissues.[6] These factors account for the wide diversity of tumors in this region. This potential space extends superiorly to the level of the peritoneal reflection and inferior to the retrosacral fascia (Waldeyer's fascia). Below this fascia, another potential space exists, bounded inferiorly by the muscles of the pelvic floor and anteriorly by the mesorectum. This is known as the supralevator space. Laterally, the space is bounded by the endopelvic fascia (lateral rectal stalks), ureters, sacral nerve roots, and iliac vessels.[6–8] The presacral space contains connective tissue, the superior rectal vessels, middle sacral artery, and components of both sympathetic and parasympathetic nervous systems (Fig. 1). Injury to these vascular and neural structures may have profound effects on rectoanal physiology and cause substantial neurological and musculoskeletal morbidity. Anorectal function will be maintained if all unilateral nerve roots have to be sacrificed or if the upper three sacral nerve roots are left intact bilaterally.

* Correspondence address: The Colorectal Institute, 13770 Plantation Road, Fort Myers, Florida Suite 2, FL.
E-mail address: JeffNeale@thecolorectalinstitute.com

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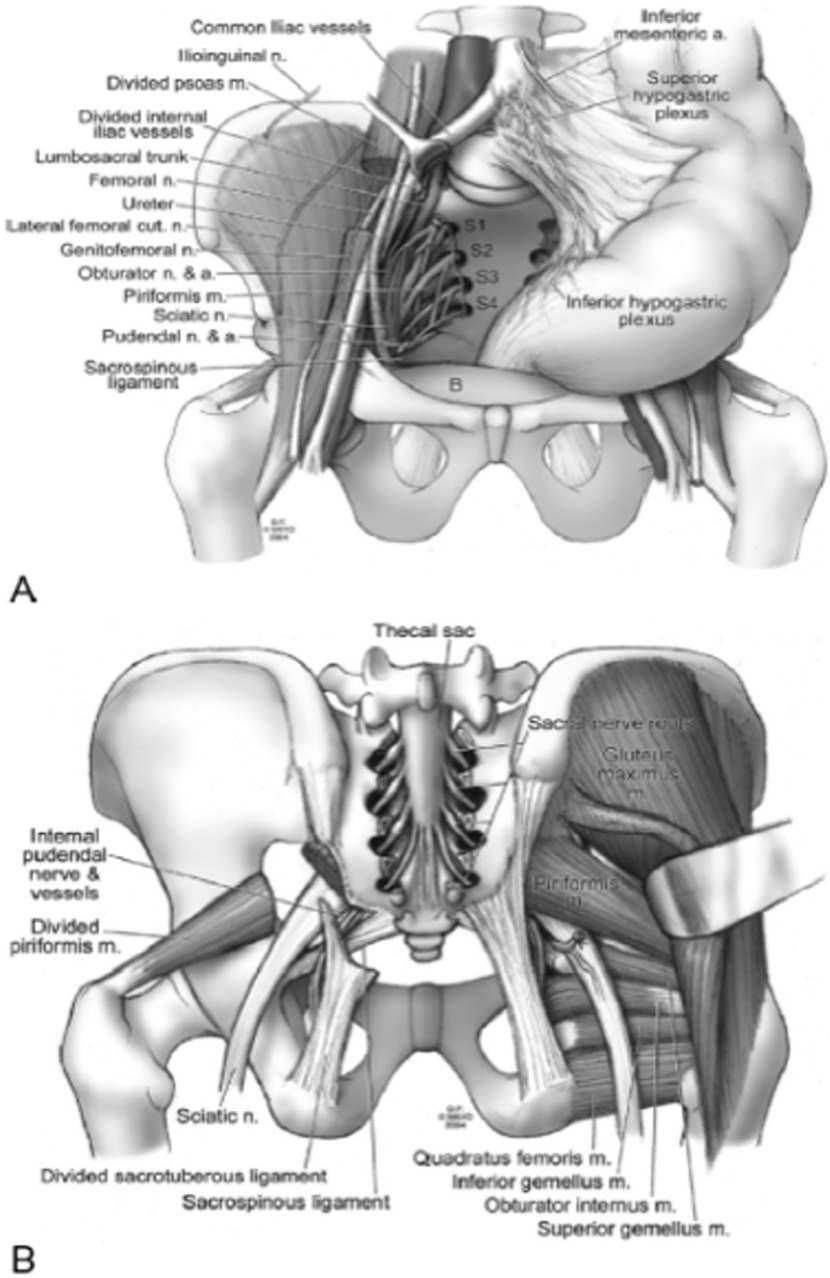

Fig. 1. (A) Anterior and (B) posterior view of the pelvic anatomy.

However, if the S3 nerve roots are sacrificed bilaterally, there will be a lack of contraction by the external sphincter during dilation of the rectum and various degrees of incontinence can occur.[9] If greater than half of the vertebral body of S1 remains intact after sacrectomy, pelvic stability will be maintained. If this area underwent radiation preoperatively, spinopelvic stability may require augmentation with fusion.[5,8]

Classification

There have been a variety of classifications systems proposed to categorize the diverse nature of presacral tumors. Uhlig and Johnson[4] first classified retrorectal tumors into broad categories: congenital, neurogenic, osseous, inflammatory, and miscellaneous. Dozois et al.[8] modified Uhlig and Johnsons classification and subcategorized them into malignant and benign entities, as the therapeutic approach is different for each (Table).

Congenital lesions

Congenital lesions are thought to arise from the remnants of embryonic tissues and can present as either cystic or solid lesions. The cystic congenital lesions include developmental cysts and anterior meningoceles. The solid congenital lesions consist of teratomas, sacrococcygeal chordomas, and adrenal rest tumors. Congenital lesions are the most common retrorectal tumors, accounting for nearly two-thirds of all tumors in the presacral space. They are usually benign and are more common in females.[4,5,10]

Cystic lesions

Developmental cysts

Developmental cysts make up two-thirds of the retrorectal tumors seen in clinical practice. They are almost always benign.