Retrorectal tumors: A case series

Retrorektal tümörler: Vaka serisi

B. Bülent MENTEŞ, Osman KURUKAHVECİOĞLU, Bahadır EGE, Ahmet KARAMERCAN, Sezai LEVENTOĞLU, Ömer YAZICIOĞLU, Mehmet OĞUZ

Department of Surgery, Gazi University, Faculty of Medicine, Ankara

Background/aims: *Retrorectal masses are rarely encountered in surgical practice, and they arise from congenital remnants, and from osteogenic, neurogenic, inflammatory, or metastatic processes. The majority of these masses are benign but many can be malignant, so they need to be treated with aggressive surgical management.* ***Methods:*** *In this case series, eight patients with retrorectal masses of various etiologies are described, with particular emphasis on diagnosis and surgical treatment.* ***Results:*** *In our series, we noted one duplication cyst, one tailgut cyst, one epidermoid cyst, one teratoma, one gastrointestinal stromal tumor, one epithelial malignant tumor, one inflammatory mass, and one retrorectal mass of as yet unknown origin. In three patients, complete excision via posterior sagittal approach was performed, one underwent abdominoperineal en-block proctectomy, two were only biopsied for neoadjuvant therapy, and two are waiting for operation.* ***Conclusions:*** *Surgery is the main treatment of choice, and the surgical strategy should be decided according to the localization and nature of the retrorectal mass.*

Key words: Retrorectal mass, retrorectal space, tailgut cyst, rectal duplication cyst, rectum

Amaç: *Retrorektal kitleler cerrahi pratikte nadir görülen olgulardır ve konjenital kalıntılar, osteojenik, nörojenik, inflamatuvar veya metastatik kaynaklı olarak ortaya çıkarlar. Bu kitlelerin çoğunun benign olmasına rağmen bir kısmı da malign olabilir. Bu nedenle retrorektal kitleler agresif cerrahi yaklaşımlarla tedavi edilmelidirler.* ***Yöntem:*** *Bu vaka serisinde etyolojisi birbirinden farklı sekiz hastanın tanı ve cerrahi tedavisindeki özellikler sunulmaktadır.* ***Bulgular:*** *Serimiz; duplikasyon kisti, tailgut kisti, epidermoid kist, teratom, gastrointestinal stromal tumor, epiteliyal malign tumor, inflamatuvar kitle ve bir adet histolojisi tanımlanmamış kitle lezyonundan oluşmaktadır. Hastaların üçüne posterior sagittal yaklaşımla eksizyon ve birine de abdominoperineal rezeksiyon uygulandı. İki hastaya neoadjuvan tedavi için biyopsi yapılırken iki hasta da henüz ameliyat edilmedi.* ***Sonuç:*** *Cerrahi temel tedavi seçeneğidir ve seçilecek cerrahi tedaviye, retrorektal kitlenin yerine ve türüne göre karar verilmelidir.*

Anahtar kelimeler: Retrorektal kitle, retrorektal alan, tailgut kisti, rektal duplikasyon kisti, rektum

INTRODUCTION

Retrorectal masses are rarely encountered in surgical practice. The incidence of these tumors is reported as 1/40,000-63,000 admissions to large referral centers (1, 2). Most general surgeons treat only one such patient during the course of their careers (3). The majority of these masses are benign but many can be malignant, so they need to be treated with aggressive surgical management. Furthermore, benign tumors can transform to malignant lesions, or can include malignant parts.

The retrorectal space is also known as the presacral space. The borders of this region are the fascia propria of the rectum anteriorly, presacral fascia overlying the sacrum posteriorly, the peritoneal reflection superiorly, Waldeyer's fascia inferiorly, and the lateral stalks of the rectum, the ureters and the iliac vessels (3). Retrorectal tumors generally originate from the embryologic remnants of this space. Suspicion is the main factor in the diagnosis of these tumors. The location and borders of the tumor and possible invasion to adjacent tissues can be easily detected using computed tomography (CT), magnetic resonance imaging (MRI), or transrectal ultrasound (TRUS). However, misdiagnosis or inadequate surgery can lead to serious complications. Their rarity and the diagnostic therapeutic dilemmas contribute to the controversies regarding the treatment of these tumors. Retro-

Address for correspondence: Osman KURUKAHVECİOĞLU
Gazi Üniversitesi Tıp Fakültesi
Genel Cerrahi Anabilim Dalı
06500 Beşevler, Ankara, Turkey
Tel: +90 312 202 57 19
E-mail: okurukahveci@yahoo.com

Manuscript received: 20.10.2006 **Accepted:** 08.10.2007

rectal tumors can be classified into five categories (congenital, neurogenic, inflammatory, osseous and miscellaneous).

In the literature, retrorectal tumors have been reported as case reports of one or two cases. The largest series were reported from the Mayo Clinic, Memorial Sloan-Kettering and the Cleveland Clinic (3). However, some of these series include chordomas as the most common malignant cause of retrorectal mass. Chordomas are well-described and relatively frequent invasive tumors that arise from the fetal notochord within the vertebral bodies. This tumor is not typically placed in the retrorectal space, but it also extends to the sacrum and the postsacral region. Therefore, surgical treatment consists of wide excision of the sacrum/parasacral tissues usually by the neurosurgeon, while other retrorectal lesions are generally treated by the colorectal surgeon.

In this case series, eight patients with retrorectal masses of various etiologies are described with particular emphasis on diagnosis and clinical approach.

CASE SERIES

This case series includes eight patients with retrorectal tumors. The female/male ratio was 5/3, and the mean age was 48.50 ± 7.42 (38-59). The demographic and clinical features are listed in Table 1. Rectal pain and perirectal mass sensation were the main symptoms. Four patients were asymptomatic, whereas three suffered from rectal pain and two of these three patients also described mass sensation. Patient 6 suffered from intermittent vaginal purulent discharge. This retrorectal cyst neighbored the left posterior border of the vagina. An inflammatory cyst was detected by CT scan (Figure 1).

Some of the typical CT or MR views of the patients are shown in Figure 1. All of the retrorectal masses in this case series except for those of patients 3 and 8 were localized under the level of the fourth sacral element.

Patients 1, 2 and 4 were operated via a posterior sagittal approach. In brief, a posterior paramedian incision was made, and after dividing the subcutaneous fatty tissue, the lumbosacral fascia was exposed. The levator muscles and the anococcygeal ligament lie just under the lumbosacral fascia (Figure 2). Transection of this ligament allows mobilization of the coccyx. In some cases, excision of the coccyx provides further exposure of the retrorectal space. In our series, the coccyx was resected only in patient 4 (Figure 2). Great care must be taken when resecting the tumor from the posterior wall of the rectum to avoid injury. The diagnoses of patients 1, 2 and 4 were rectal duplication cyst, tailgut cyst and epidermoid cyst, respectively.

In three patients (nos. 3, 7, 8), transanal biopsies were performed. In patient 7, transanal core needle biopsy revealed teratoma. This patient eventually underwent abdominoperineal rectal resection en bloc with the tumor, which invaded the distal rectum/proximal anal canal. Patients 3 and 8 had

Table 1. Demographic and clinic features of the patients with retrorectal mass

No	Age	Sex	Symptoms	Presentation/ Preoperative diagnosis	Operation	Diameter of tumor (cm)	Histopathology
1	38	F	Asymptomatic	Retrorectal mass Duplication cyst?	Total resection with posterior sagittal approach	4 x 4	Rectal duplication cyst
2	50	F	Asymptomatic	Retrorectal mass Tailgut cyst?	Total resection with posterior sagittal approach	6 x 7	Tailgut cyst
3	53	M	Rectal pain, perirectal mass sensation	Retrorectal mass	Transanal biopsy	8 x 7	Malignant epithelial tumor
4	41	F	Rectal pain	Retrorectal mass	Total resection with posterior sagittal approach	2 x 3	Epidermoid cyst
5	50	F	Asymptomatic	Retrorectal mass	Unsuccessful transabdominal approach/ waiting for second operation	3 x 3	Unknown
6	42	F	Vaginal discharge	Retrorectal inflammatory mass – vaginal discharge	Not operated, waiting for operation	5 x 3	Inflammatory cyst (unconfirmed)
7	59	M	Asymptomatic	Retrorectal mass	Core needle biopsy + abdominoperineal resection (en bloc with anorectum)	3 x 4	Teratoma
8	55	M	Rectal pain, perirectal mass sensation	Retrorectal mass	Transanal biopsy	14 x 9	Gastrointestinal stromal tumor

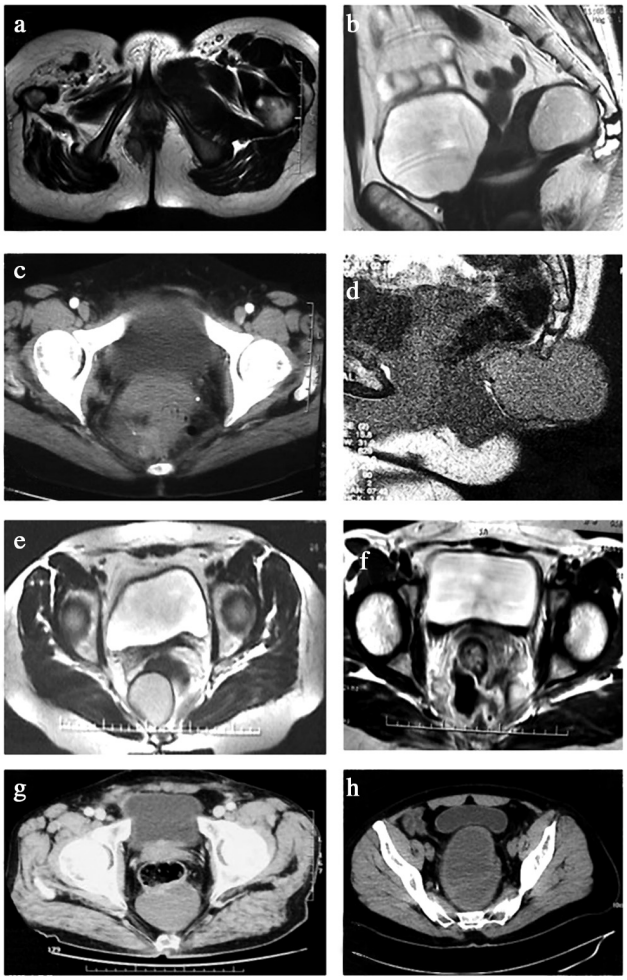

Figure 1. CT or MR views of the eight patients: **a)** No. 1. rectal duplication **b)** No. 2. tailgut cyst **c)** No. 3. malignant epithelial tumor **d)** No. 4. epidermoid cyst **e)** No. 5. retrorectal mass not operated **f);** No. 6. inflammatory mass **g)** No. 7. teratoma **h)** No. 8. gastrointestinal stromal tumor.

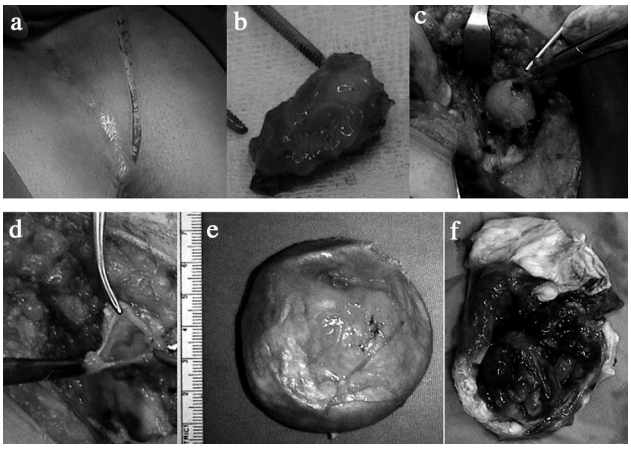

Figure 2. Operative views and surgical specimens of some of the cases: **a)** Paramedian posterior sagittal incision **b)** Coccyx resection (whenever necessary) **c)** Exposure of the retrorectal mass **d)** Opening of the recta duplication cyst in order to check possible connection to the rectum **e)** Complete excision of the tailgut cyst **f)** Complete excision of the epidermoid cyst.

large retrorectal masses which appeared unresectable in CT and MR images (Figure 1). Transanal biopsies were performed, and these patients will be evaluated for radical surgery after neoadjuvant therapy.

Patient 5 had undergone an operation with transabdominal approach in another hospital, but the mass could not be resected. The retrorectal mass was located below the level of the fourth sacral vertebral corpus in the CT and MR images (Figure 1). A posterior sagittal approach was proposed, but she wanted to wait for several months before the second operation for social reasons. Patient 6 suffered from intermittent vaginal purulent discharge and an inflammatory cyst was detected by CT scan, but she refused surgery.

DISCUSSION

Because most retrorectal tumors are asymptomatic, they are incidentally discovered during routine gynecologic examinations. Accordingly, retrorectal tumors are most frequently seen in females (4). However, malignant forms are more frequently encountered in males. In our series, five of the eight patients were female, and the average age was 48. Three of the eight patients had malignant tumors, and all of them were male. In accordance with previous series, half of our patients were asymptomatic, and three suffered from rectal pain. Two of these three patients also had perirectal mass sensation. One patient was admitted with vaginal discharge. Occasional symptoms of retrorectal tumors are perirectal pain, mass sensation, constipation, painless rectal bleeding, change in caliber of stool, or urinary frequency (5). Generally, malignant or inflammatory tumors present with pain (1, 6).

Obstruction is infrequent, but in pregnant females, this can be life-threatening during labor (7). Chronic fistulas can be the only symptom of inflammatory retrorectal tumors, as exemplified in patient 6 in this series.

Retrorectal tumors can be classified as congenital, neurogenic, inflammatory, osseous and miscellaneous. Congenital lesions account for 55 to 70% of all retrorectal tumors (1, 8, 9), and they harbor embryologic remnants. Congenital lesions include chordomas (remnants of notochord), teratomas, anterior sacral meningoceles and developmental cysts (dermoid, epidermoid, enteric duplication, or tailgut cysts) (5). Tailgut cysts develop from pre-

cursors of the gastrointestinal tract, and they contain mucus-secreting columnar epithelium, as exemplified in patient 2. Teratomas arise from totipotential cells and if they contain germ-cell elements, they may have malignant potential. If teratomas are left untreated, 5 to 10% of them undergo malignant degeneration (4). In patient 7, a malignant retrorectal teratoma invaded the anorectum, and an en bloc anorectal resection was indicated. Chordomas are the second most common type of retrorectal tumors, and most of them are malignant. However, they tend to grow erratically, and most are not confined to the retrorectal space. Rectal duplication cysts are remnants of duplicated rectum and contain colonic mucosa and muscle and serosa. Inflammatory lesions are not common as congenital lesions. If perineal abscesses spread to the supralevator space, they can be evaluated as retrorectal tumors. Many retrorectal tumors develop inflammation after biopsy is performed.

The differential diagnosis of a retrorectal mass can be narrowed using a combination of diagnostic tools. Rectal examination is the essential method. Due to their location, retrorectal tumors are palpable on rectal examination as extrinsic masses, and they are usually soft and compressible (1, 10). These features of retrorectal tumors may lead to misdiagnosis unless the examiner is careful and suspicious (8). Colonoscopy can rule out any rectal mucosal changes in cases of rectal bleeding. Extrinsic masses can be detected with barium enema, but it provides no additional information (1, 10). TRUS has been used to characterize the lesion as solid or cystic, and it occasionally shows internal echoes due to mucoid or inflammatory debris (5).

The margins, characteristics and location of the tumor, as well as possible invasions to the adjacent tissues, can be easily evaluated with CT and MRI. However, radiologic diagnosis with CT or MRI is easily in agreement with the final histology (4). Biopsy of retrorectal tumors is controversial. It is generally feared that biopsy before radical resection may cause recurrence (1). However, others have reported no complications after biopsy (6, 11, 12). Furthermore, the tissue obtained is often not enough to make a definitive histological diagnosis (10, 13-15). Complete excision is necessary to prevent recurrence, infection, and possible malignant transformation. Therefore, preoperative biopsy should only be performed for unresectable lesions for neoadjuvant treatment. Two patients in this series underwent transanal biopsy for the planning of neoadjuvant therapy. In another patient with teratoma (patient 7), preoperative histologic confirmation was needed to justify en bloc resection of the anorectum.

Total resection is the best choice of treatment for the lesions that appear resectable. There are three main surgical approaches for retrorectal tumors. Posterior or transcoccygeal resection can be performed for small tumors. If the tumor does not extend above the level of the fourth sacral element, posterior approach is the appropriate method. Anterior or abdominal approach is preferred for lesions with lowest extension above the fourth level of the sacral element. The last choice is the combination of these two methods. If the lesion begins under the fourth level of the sacral element and extends above it, a combination approach may be necessary (3). It deserves emphasis that one of the patients in this series had undergone an unsuccessful transabdominal attempt for resection of a lower-located retrorectal mass in another center.

Complete resection with negative margins is the standard surgical approach for benign retrorectal tumors. However, 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 will likely improve the rate of successful treatment of these enigmatic lesions.

REFERENCES

1. Jao S-W, Beart RW, Spencer RJ, et al. Retrorectal tumors: Mayo Clinic experience, 1960-1979. Dis Colon Rectum 1985; 28: 644-52.
2. McCune WS. Management of sacrococcygeal tumors. Ann Surg 1964; 159: 911-8.
3. Hobson KG, Ghaemmaghami V, Roe JP, et al. Tumors of the retrorectal space. Dis Colon Rectum 2005; 48: 1964-74.
4. Glasgow SC, Birnbaum EH, Lowney JK, et al. Retrorectal tumors: a diagnostic and therapeutic challenge. Dis Colon Rectum 2005; 48: 1581-7.
5. Killingworth C, Gadacz TR. Tailgut cyst (retrorectal cystic hamartoma): report of a case and review of the literature. Am Surg 2005; 71: 666-73.
6. Cody HS 3rd, Marcove RC, Quan SH. Malignant retrorectal tumors: 28 years' experience at Memorial Sloan-Kettering Cancer Center. Dis Colon Rectum 1981; 24: 501-6.
7. Sobrado CW, Mester M, Simonsen OS, et al. Retrorectal tumors complicating pregnancy. Report of two cases. Dis Colon Rectum 1996; 39: 1176-9.

8. Uhlig BE, Johnson RL. Presacral tumors and cysts in adults. Dis Colon Rectum 1975; 18: 581-9.
9. Stewart RJ, Humphreys WG, Parks TG. The presentation and management of presacral tumors. Br J Surg 1986; 73: 153-5.
10. Hannon J, Subramony C, Scott-Conner CE. Benign retrorectal tumors in adults: the choice of operative approach. Am Surg 1994; 60: 267-72.
11. Singer MA, Cintron JR, Martz JE, et al. Retrorectal cyst: a rare tumor frequently misdiagnosed. J Am Coll Surg 2003; 196: 880-6.
12. Bohm B, Milsom JW, Fazio VW, et al. Our approach to the management of congenital presacral tumors in adults. Int J Colorectal Dis 1993; 8: 134-8.
13. Hjermstad BM, Helwig EB. Tailgut cysts. Report of 53 cases. Am J Clin Pathol 1988; 89: 139-47.
14. Sidoni A, Bucciarelli E. Ciliated cyst of the perineal skin. Am J Dermatopathol 1997; 19: 93-6.
15. Prasad AR, Amin MB, Randolph TL, et al. Retrorectal cystic hamartoma: report of 5 cases with malignancy arising in 2. Arch Pathol Lab Med 2000; 124: 725-9.