Problems in Diagnosis and Treatment of Retrorectal Tumors: Our Experience in 50 Patients

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Abstract- Retrorectal tumors are rare lesions in adults, which remains a difficult diagnostic and management problem. The purpose of this study was to evaluate the results of surgical management of retrorectal tumors in our institution. In a retrospective study, a consecutive series of patients who underwent surgical excision of a retrorectal tumor were identified from a database. Medical records, radiology, pathology reports and surgical approach were checked retrospectively. The data was analyzed using SPSS statistical software (version 18). From 50 patients, 24 were male, and 26 were female with the mean age of 41.7 years. The origin of mass was congenital in 46% (23 cases) and neurogenic in 14% (7 patients), bone origin in 12% (6 cases) and miscellaneous in 24% (12 cases). In total, 56.7% (21 cases) were malignant. Surgical approaches included laparotomy in 11 cases, the sacral approach in 17 cases, the anterior-posterior approach in 14 cases and one case through abdomino-sacral approach. The mean follow-up was 56.7 (10-277) month. Ten patients died due to extensive metastases with a mean survival of 46.6 (1-158) months. Primary urethrorectal tumors are very rare. Successful treatment of these tumors requires careful clinical evaluation and expertise in pelvic surgery.

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Introduction

Retrorectal space is a potential space which is surrounded by rectum in the anterior, sacrum and coccyx in the posterior, peritoneum fold in the above and coccygeal, *levator ani* and *puborectalis* muscles in the below, and iliac, and ureter vessels are external verge. Because of the connectivity of nervous and skeletal and intestine system during fetal life, a diverse group of embryonal tumors is created in this area.

Because of the rarity of these tumors, the surgeons have few experiences about sacral and pre-sacral tumors (1). Their prevalence in large referral centers such as the Mayo Clinic has estimated approximately 1 in 40,000 hospital admissions (2). These tumors can present at any age and present a broad range of histopathological types (3). Detection of these tumors may pose questions regarding the differential diagnoses, diagnostic procedures, possible surgical approaches and next prognosis (4).

Differential diagnoses of the tumors outside of the rectum include benign and malignant lesions derived from soft and bony tissues of this area.

We examine 50 cases of retrorectal tumors with different pathological types during a period of 24 years (1989 till 2013) which were treated by the authors in surgery departments of Ghaem and Omid hospitals, Mashhad University of Medical Sciences. Clinical protests, surgical approaches, and clinical outcomes were evaluated in each patient and the encountered diagnostic and therapeutic problems were highlighted.

Materials and Methods

In a retrospective review from February 1989 to November 2013, a total of 50 patients with retrorectal tumors were studied in Ghaem and Omid hospitals, Mashhad University of Medical Sciences. Medical

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records, preoperative presentations, radiology and pathology reports, surgical approaches were checked retrospectively. The patients were followed up for postoperative complications, mortality, and recurrence. Finally, the data was analyzed using SPSS statistical software (version 18).

Surgical approaches

Abdomino-sacral approach

In abdomino-sacral approach, the patient was placed in decubitus lateral position and abdomen was opened through long incisions from the suprapubic area toward left groin. Sacral artery and vein were identified, and vessels were edged out with vascular loops, entered to the sacral space just below the sacrum promontorium and rectum posterior was separated from the sacrum.

Posterior incision started from the sacrococcygeal junction, and continued up to the first sacral vertebra in the midline and then went upward and out on the iliac crest about 2 inches; lateral was curved to the superior posterior iliac spine.

The anococcygeal raphe, the origin of gluteal muscles, and sacrotuberous and sacrospinous ligaments were separated, and pudendal nerve ligaments were maintained. Sacrum was unroofed and after maintaining at least one S3, the osteotomy was performed at the level of S3 or higher, if necessary, all the sac nerve were cut in this method, and all lesions were removed as enblock and sometimes with the rectum.

Combined anterior and posterior approach

This surgical approach was started from anterior, and the patient was placed in supine position, and the internal iliac artery and vein were separated in the two sides. Arteries and veins of the external and middle sacral were cut and closed. In a posterior approach, the patient was placed in prone position. The posterior incision was started from sacrococcygeal junction and expanded to above in the midline. Other actions were similar to the abdomino-sacral technique.

Results

Of 50 patients, 24 were male, and 26 were female with the mean age was 41.7 (range, 16 to 74) years and included different pathological types (Table 1).

Table 1. Different pathological types of retrorectal tumors in 50 patients

	Benign		Malignant	
Congenital (24 cases)	**Dermoid and epidermoid cyst**	7	Chondroma	8
	Epidermal cyst	1		
	Tailgut cyst	3	Primitive neuroectodermal	1
	Anterior meningocele	1		
	Infected teratoma	1	Germ cell tumor	1
	Alimentary duplication cyst	1		
Osseous (6 cases)	**Ewing sarcoma**			2
	Chondrosarcoma			2
	Malignant giant cell tumor			1
	Plasmacytoma			1
Neurogenic (7 cases)	**Schwannoma**			2
	Neuroblastoma			1
	Neurofibrosarcoma			1
	Ependymoma			2
	Malignant peripheral nerve sheet tumor			1
Miscellaneous (13 cases)	**Malignant spindle cell tumor**			1
	Fibromatosis			2
	Recurrent rectal cancer invading sacrum			2
	Lipofibroma			1
	Liposarcoma			2
	Hydatid disease			2
	Unknown origin			1
	Carcinosarcoma			1
	Intra-osseous ganglion cyst			1

The origin of the mass was congenital in 46% (23 cases) and neurogenic in 14% (7 patients), bone origin in 12% (6 cases) and miscellaneous in 24% (12 cases).

In total, 56.7% (21 cases) were malignant. Of 19 women, 7 cases (36.8%) and of 18 men, 14 cases (77.7%) had malignant tumors. Ten cases had a benign cystic lesion and of 27 cases of the solid lesion, 21 cases (81.4%) were malignant.

In two patients of the studied group, the final diagnosis was not obtained by a pathologist. The first case was a 57-year-old man with a history of dysuria and hesitancy (urination along with pressure on the bladder and straining) from 13 years ago, and no diagnosis was confirmed during this period until he referred with fever and severe pain in the perineum. Computed tomography (CT) scan showed a collection on the left side of the pelvic. He underwent trans-sacral drainage, and necrotic and infected tissues were discharged. In CT scan, the abscess is under control with the dimensions of 6×9 cm on the left side of the pelvic and perianal fistula, and compression effect on the rectum. But because of patient's history of ischemic heart disease and high cardiac risk during surgery, he did not undergo a definitive surgery. Therefore, a definitive diagnosis with pathological confirmation was not available

The second patient was a 23-year-old woman who has had left sciatic pain from two years ago. In CT scan, there was a large cystic lesion in the left half of the pelvis with bone involvement from S1 to S5 that had undergone resection through anterior-posterior approach. But, in the pathologic evaluation of the resected tissue, pseudocyst with necrotic content with no malignancy evidence was reported and definite origin of the tumor remained unknown.

Presenting symptoms were so various and included: sciatic pain in 10 patients (bilateral sciatic pain in 3 cases, right sciatic pain in 4 cases, and left sciatic pain in 3 cases), abdominal pain of lower abdomen in 7 patients, difficult defecation in 5 patients, prolonged perineal secretion in 3 patients, the asymptomatic vaginal mass which was found in normal examination of the vagina in 3 cases, lower back pain in 4 patients, abdominal mass in 1 patient, dyspareunia in 1 patient, headache resulting in an increase in arterial pressure in 1 patient, dystocia in 1 patient, infertility in 1 patient, paraplegia in 1 case, dysuria in one patient, perineal mass in 2 cases, sacral mass in 2 cases and incidentally discovered in one patient following fetus abortion.

The mean time of symptoms prior to diagnosis was 37.42 months. Fifteen cases (30%) had 1-4 surgical errors because of incorrect diagnoses (Table 2).

Table 2. Surgical errors in 15 patients due to diagnosis of other diseases

Supposed diagnosis	No	Procedure	Definitive diagnosis
Ovarian cyst	5	Laparotomy	Meningocele, tailgut cyst, neurofibrosarcoma, dermoid cyst
Ovarian tumor	1	Laparotomy	Schwannoma
Unsuccessful laparotomy	1	Unsuccessful laparotomy	Germ cell tumor(seminoma)
Anal fissure	1	Anal sphincterotomy	Ewing's sarcoma
Fistula in ani	2	Fistulectomy (4 patients)	Infected benign teratoma, fibrolipoma
hemorrhoid	1	Hemorrhoidectomy	Primitive neuroectodermal
Unknown pelvic cyst	1	Drainage of cyst to rectum (twice)	Tailgut cyst
Vaginal tumor	1	Biopsy of vaginal tumor	Fibromatosis
Hypertrophy of prostate	1	Prostatectomy(endoscopic)	Chondrosarcoma
Herniated lumbar disc	2	Removal of disc	Chordoma
Uteral Prolapse	1	Repair of uteral prolapse	Carcinosarcoma with s_5 involvement

Preoperative diagnosis of retrorectal masses was performed based on physical examination which was confirmed by ultrasonography, radiography of the sacrum (Figure 1), CT scan and magnetic resonance imaging (MRI). A CT Myelogram was performed only in 1 patient with tailgut cyst suspected to myelomeningocele.

Various surgical approaches have been used including laparotomy in 11 cases, the sacral approach in 17 cases, the anterior-posterior approach in 14 cases, and one case was operated on through abdomino-sacral approach. As aforementioned, one patient did not undergo surgery due to cardiac disease (Table 3).

The mean follow up time was 56.7 (10-277 months. One case was excluded after 2 months of follow-up, and 10 patients died due to extensive metastases with a mean survival of 46.6 (1-158) months.

Two patients died after surgery. The first case died after completion of surgery because of unstable hemodynamic status. This patient was a 58-year-old man that had been undergone surgery through the anterior and posterior approach for chondrosarcoma

resection. Another case was a 55-year-old man with sacral chondrosarcoma. Sacrectomy was performed through the anterior and posterior approach, and abdomino-perineal excision was done, and a sigmoid colostomy was inserted for him.

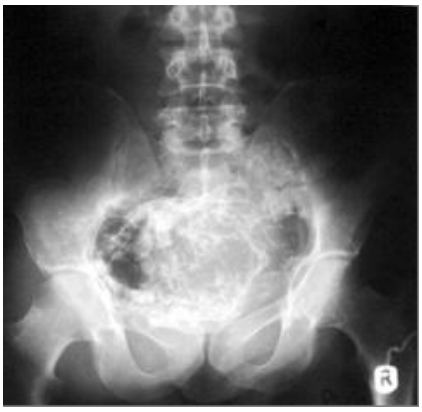

Figure 1. An X-ray image of the pelvis of a 55-year-old man with chondrosarcoma shows a large mass with extensive calcifications affected almost all the sacrum

The patient on the third day after surgery complicated to focal peritonitis due to colon perforation at the back of colostomy location. Perforation occurred due to the erosion of hard and sharp stool in the colon. Unfortunately, the patient died due to septicemia 10 days after surgery. Thirty-seven patients are still alive. Complications were observed in 8 patients (Table 4).

Table 3. Surgical approaches in 50 patients

Procedure	No	Procedure	No
Anterior and posterior approach	14	Transsacral biopsy	9
Laparotomy excision	10	Sacral approach resection	8
Laparotomy biopsy	6	Abdominosacral resection	1
Laparotomy colostomy	1		

Table 4. Postoperative complications in 50 patients with retrorectal tumor

Complication	Procedure	Definitive diagnosis	Treatment	Outcome
Meningitis	Laparotomy	Meningocele	Conservative	Alive 271 months
Persisted urinary retention+1`infertility	Fistulectomy for anal fistula +sacral resection + failed attempts	Infected benign teratoma	Augmented cystoplasty + self-urinary catheterization	Alive 191 months
Impotence	Anterior and posterior approach + rectal resection and end colostomy	Giant cell tumor	Conservative	Alive 121 months
Urinary incontinence	Transacral resection	Ependymoma	Conservative	Alive 30 months
Vaginal stricture	Anterior and posterior approach	Rectal tumor invading scrum and vagina	Conservative	Died 14 months
Fecal peritonitis	Anterior and posterior approach	Chonrosarcoma	Laparotomy	Died postoperative
Rectosacroperineal fistula	anterior and posterior approach	Ependymoma	Sigmoid colostomy	Alive 214 months
Paraplegia and rectosacral fistula	Transsacral drainage	Lumbosacral hydatid cyst	Conservative	Alive 288 months

Discussion

Tumors of retrorectal space are uncommon. Because of the rarity of these tumors and their various symptoms, they are often misdiagnosed or incorrectly treated. In our study, 30% of patients had 1-4 previous wrong surgery with an incorrect diagnosis which indicates the low plexity of these conditions.

Preoperative diagnosis and extent of the disease should be defined as clearly as possible before the surgery (5). Some believe that sacrum CT scan is the technique of choice in the initial evaluation of these lesions. CT scan determines the area of soft tissue involvement. However, MRI is better than CT in the detection of osteopenia and bone destruction (Figure 2).

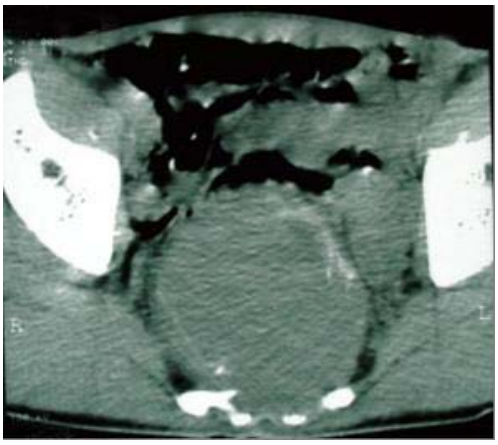

Figure 2. The pelvic computed tomography scan of a 32-year-old man with recurrent cell giant tumor shows bone destruction in the lower part of the sacrum (S2-S5).

Many believe that MRI has the highest sensitivity and specificity and is the investigation of choice for these lesions (7,8). MRI can show the tumor longitudinal extension and exact extension in the craniocaudal direction of the disease that has of main importance for the surgery. Myelography along with CT scan is the best way to determine the exact anatomy of the anterior sacral meningocele and allows surgeons to make an ideal decision (Figure 3).

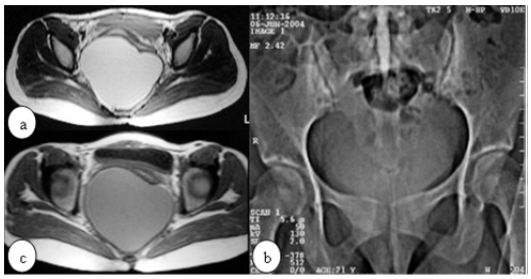

Figure 3. Magnetic resonance imaging and computed tomography (CT) myelography of tailgut cyst in a 21-year-old woman; T1 (a) and T2 (b) images show the cystic appearance large, and CT myelography (c) don't show any relationship with the dura sac.

Some researchers believe that solid tumors should be sampled before the surgery. Indeed, some pre-sacral tumors may benefit from chemotherapy or radiation therapy before the surgery. This is performable particularly in bone tumors such as Ewing's sarcoma. However, trans-sacral biopsy of any lesion should be avoided because such an action for cystic lesions may lead to cyst infection (2).

The surgical approach for such lesions is complex and often need a multi-direction approach and a set of surgical specialties including general surgery, orthopedics, and neurosurgery (3,6). We made use of collaboration of a neurosurgeon based on need. Patient and the surgeon may face a dilemma: the choice between radical resection with high potential morbidity or partial excision with higher recurrence rates (10).

Successful treatment includes complete resection, preservation of neurological function and patient survival which the latter rarely can be achieved (1). Inadequate resection due to poor exposure of the area at the time of the first surgery leads to local recurrence and subsequent destruction of the sacrum (11).

The level of sacrum resection should be upper the tumor and avoided from wound contamination with tumor cells, and if caudal sac of the spinal cord was cut or had leakage, it should be repaired (12). On the other hand, resection with extension to upper parts of the sacrum is often associated with urinary and fecal incontinence and infertility. Every patient who has undergone sacrum resection should be aware of possible complications (13,14). However, if the surgeon was not ready for this sacrifice, finally, the tumor performs this sacrifice.

Bilateral loss of nerve roots below the S1 level during sacrectomy leads to urinary retention and incontinence (16). Some researchers showed that at least one or both S3 nerves must be preserved to maintain normal urinary and fecal continence (17,18). In patients with unilateral loss of the sacral nerve, no considerable disorder was caused by anorectal and urinary function (3,17).

In our series, 2 cases that underwent sacral resection were complicated to bladder dysfunction. Both patients had an S2-S5 sacral resection. Only one case of resection was complicated, and then augmentation cystoplasty and self-catheterization were performed. The second patient has still a Foley catheter.

In our study, in one patient that sacrum resection from S3 to S5 was performed along with rectum resection, there was no problem about bladder function, but the patient had infertility after the surgery which it appears to be due to sympathetic dysfunction.

Partial resection of the sacrum from caudal to the middle vertebrae of S1 does not make the pelvic unstable (19). Thus the stability of the spine needs to maintain at least half of the S1 vertebrae and related lumbosacral ligaments (11). Complete sacrectomy need to connect both sides of the spine and ileum to rebuild the pelvis.

Selection of the appropriate surgical approach can be a mystery (5). The trans-sacral approach had a lot of popularity for many years. But pre-sacral access is limited, and artery control is insufficient (3). Trans-

perineal Kraske approach is simple and has minimal morbidity. But this approach has some problems to remove large tumors. Moreover, it is associated with high recurrence rate (20). There is the possibility of resecting the rectum and ureters in trans-abdominal approach and also, we have the possibility to control iliac vessels. Thus, it is the choice of approach in pre-sacral masses.

We can resect the masses with less than 5 cm in diameter through the trans-coccygeal or perineal.

Sacrum low tumors involved S3 and lower can be resected through the posterior approach by sacrectomy of a full half. While, to achieve wide margin in sacral tumors which have often a large size when diagnosed, a combined posterior and anterior resection or abdomino-sacral approach is required.

Cystic lesions are most common and often consist 2.3 of rectrorectal lesions. There was a dominance of cystic tumors in our study. In the adult population, 10% of cystic lesions are malignant, whereas, in solid lesions, 60% are malignant (23). In our group, all cystic lesions were benign, and 81% of the solid lesions were malignant, and cystic lesions were more common in females (female: male ratio=7: 3).

Because of various clinical protests, the examiner should consider the possibility of retrorectal tumors in numerous occasions, and clinical examination should be performed by focusing on the posterior part of the rectum.

Treatment success in posterior rectal tumors is complete removal of the lesion with preservation of neurological function and the patient's life, which, unfortunately, this triad is less achieved. Tumors of the lower part of the sacrum which involve sacral third vertebra and the lower must be completely resected by the posterior approach, but for the tumors of the upper sacrum, anterior and posterior resection or laparotomy is necessary to achieve adequate margins. Besides, to preserve the continence of normal excretion, at least one or both the third sacral spinal nerves must be preserved.

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