Case Report

Retrorectal/Presacral Epidermoid Cyst: Report of A Case

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Abstract

Although epidermal cysts are frequently observed throughout the body, they are rarely found in the retrorectal/presacral regions. Retrorectal epidermal cysts are congenital lesions of ectodermal origin and generally occur in women during the reproductive period. In this case report, a 47 year old female patient with 25 years of complaint of a mass extending from the perianal region to the retro-coccygeal region is discussed. A total mass excision with paracoccygeal incision was performed under spinal anesthesia. The histopathologic examination demonstrated an epidermoid cyst. Correct diagnosis and appropriate treatment when first detected significantly decrease the recurrence and complication risks in all retrorectal tumors. Any mass should be completely removed in the treatment.

Key Words: Rectal, presacral, perianal, epidermoid, cyst

Introduction

Tumors with notochordal, cloacal and neurogenic origins can appear in the retrorectal region. Cysts in this region generally have embryologic origins. Retrorectal epidermal cysts are congenital lesions of ectodermal origin and are generally seen in women during the reproductive period [1]. Although epidermal cysts are seen frequently throughout the body, they are rarely found in the retrorectal/presacral regions. These benign perianal masses are mostly seen in women. They have a tendency to grow over time and may become infected and inflamed [2].

Case Report

In this case report, we present a 47 year old female patient with approximately 25 years of complaint of a mass extending from the perianal region to the retro-coccygeal region. The patient reported that the swelling in this region periodically discharged and became smaller, followed by regrowth. On physical examination of the patient, a 5-6 cm, painless, well-circumscribed, mobile mass with the lower edge extending from the perianal region to the retro-coccygeal region and the upper edge extending from the right paracoccygeal region to the subcutaneous tissue was palpated. The mucosa was normal on rectal exam, and exterior pressure was detected in the posterior region. The personal history and family history were unremarkable. A pre-operative image of the mass is shown in Figure 1.

Superficial tissue ultrasonography (USG) detected a well circumscribed, hypoechoic, avascular cystic lesion 56 mm x 45 mm in size with dense content extending from the subcutaneous tissue to the perianal region in the coccygeal region. Surgical excision of the mass was planned after the patient was informed and her consent was obtained. The preoperative hematologic, biochemical and coagulation examinations of the patient were normal. A total mass excision with a paracoccygeal incision was performed under spinal anesthesia. The peroperative and postoperative images of the masses are shown in Figure 2 and Figure 3.

In the microscopic and histopathological examination of the 6.5×4.5×3 cm, grey-brown soft viscous tissue sample, ceratine lamellas were found in the cyst lumens covered with squamous epithelium (Epidermoid Cyst).
The classification system described by Uhlig and Johnson and modified by Lovelady and Dockerty is commonly used for the grading of retrorectal tumors. According to this classification, retrorectal tumors are divided into 5 categories: congenital, inflammatory, neurogenic, osteo and other [3, 4]. Presacral or retrorectal developmental cysts are tailgut cysts, epidermoid cysts, dermoid cysts, teratomas and rectal intussusceptions. Some authors do not include teratomas and rectal intussusceptions in the category of developmental cysts. Retrorectal developmental cysts are rarely seen, and 26-50% of affected patients are asymptomatic [5]. Symptoms differ according to the tumor size, location and the presence of infection or malignity [1]. Although distinct symptoms are present in symptomatic patients, these cysts can be confused with anorectal abscesses, complicated fistulas and pilonidal sinuses. The most frequent symptoms are anal pain and perianal discomfort, difficulty in defecation, tenesmus, changes in bowel habits, bleeding, thin stool, incontinence and a sacrococcygeal mass [5]. In this case report, the only complaint was of a mass in the sacrococcygeal region. Retrorectal epidermal cysts are benign inclusion cysts of skin that are mostly spherical or oval-shaped unilocular lesions. Generally, their walls are covered with multilayered squamous epithelia and do not contain specialized skin components such as hair follicles and sweat glands [1].

Retrorectal developmental cysts detected in 15 cases were reported in the study conducted by Baek et al. [5] between 2001-2009. It was reported that 14 of these cases were symptomatic and 1 was detected incidentally on CT. The mean age in this study was 41, and the patients were all female. The mean cyst diameter was reported as 4.8 cm, and an epidermoid cyst was histopathologically detected in only 5 patients. In the same study, it was stated that 47 presacral, retrorectal or coccygeal developmental cyst were reported in Korea between 1990-2009; the mean age was 40.3; the M/F ratio was 1:6.4, and 40 of these were verified histopathologically, with an epidermal cyst detected in 13. The study conducted by Whittaker et al. [6] between 1922-1936 reported 22 retrococcygeal tumors. Of these, 10 were benign (9 dermoid cysts and 1 fibroma), and the remaining tumors were malignant. An epidermoid cyst was not detected in any of the cases in the study. A study conducted by Jao et al. [3] over a period of 20 years reported 120 patients with retrorectal tumors. Of these tumors, 54 were malignant and 66 were benign. Chordomas were seen more frequently in males, and congenital cysts were seen more frequently in females. A study conducted over a period of 30 years by Uhlig et al. [4] reported 63 retrorectal tumor cases. The literature confirms that epidermal cysts in the retrorectal region are very rarely seen.

The most important elements of the diagnosis of the disease are the medical history and physical examination [5]. Spencer and Jackman [7] concluded that a congenital developmental cyst should be suspected in patients with a
recurrent retrorectal abscess, repeated fistulectomy, presence of a fistula in the anus or the perianal area or rectum without identification of a primary focus in the dentate line of the anus, posterior anal funnel-shaped skin dimple and a palpable fixed or distended lesion in the precoccygeal area. Tumors in the retrorectal region can generally be easily detected with palpation. The correct diagnosis and appropriate treatment are very important for tumors in this region because an incorrect or insufficient first surgical treatment can complicate the correct diagnosis, compromise the chances of a complete surgical excision and increase the recurrence risk, as well as causing serious complications such as fecal incontinence [5]. Great caution should be applied during the diagnosis and treatment of the disease. In the differential diagnosis of perianal cysts, hemorrhoids, fistula, apse, pilonidal sinus/cyst, perianal dermatosis, anal canal cysts, benign/malign teratoma, epidermoid/dermoid cysts and anal skin cancer should be considered [2, 8]. USG, endorectal ultrasonography (ERUS), computed tomography (CT) and magnetic resonance imaging (MRI) can be used in the diagnosis according to the clinical presentation [8]. If the mass is thought to be malignant, the tumor size, structure, rectum invasion depth and lymphatic metastasis should be evaluated with ERUS [9]. CT and MRI may show the size and shape of the mass and assist in surgical decision-making [5]. If the tumors are potentially resectable, a preoperative biopsy should not be performed because it can lead to tumor dissemination, abscess, fecal fistula or meningitis (3). Laboratory tests have no diagnostic value for retrorectal cysts [2]. An increase in tumor markers (AFP, CEA) does not always indicate malignancy. Developmental cysts as well as malignant lesions can also increase in size, and these markers are typically used after general surgery [5].

Complete surgical resection should be performed for the treatment of retrorectal tumors [6, 8]. Depending on the tumor size and location, an anterior, posterior (Kraske) or antero-posterior surgical approach is used. A coccygectomy might be performed to provide a sufficient surgical area in posterior approach [5]. In the paper published by Baek et al. [5], it was reported that coccygectomy was performed in 75% of retro-coccygeal cysts detected in patients in Korea. If the tumor is larger than 5 cm, the upper margin is higher then the 3rd sacral level, and the lower margin is not lower than the 4th sacral level, an anterior transabdominal approach is recommended. The anteroposterior approach should be used if the tumor is larger than 5 cm and there is a suspicion of severe adhesion or malignancy, or if the tumor fills the sacrum [5]. The successful treatment of these tumors requires an extensive knowledge of pelvic anatomy and expertise in pelvic surgery [10]. Although the mass described in our report was larger than 5 cm, it was completely excised with a posterior approach because it extended from the right paracoccygeal region to the subcutaneous region. There was no requirement for preoperative coccygectomy because the cyst was uniformly bounded and easily separated from the surrounding tissues.

Postoperative recurrences generally occur locally. It has been reported that total excision is difficult, the prognosis is poor, and radiotherapy is useful for pain palliation in malignant tumors [6].
In conclusion, although epidermal cysts are frequently seen throughout the body, they are rarely found in the retrorectal/presacral regions. The correct diagnosis and appropriate treatment when first detected significantly decrease the risk of recurrence and complications in all retrorectal tumors. The mass should be completely removed in the treatment.

**Conflict of interest statement:** The authors declare that they have no conflict of interest to the publication of this article.

**References**