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A Rare Coexistence of Retrorectal and Ovarian Cysts: A Case Report

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Abstract

Introduction: Retrorectal cysts are rare benign lesions which are frequently diagnosed in middle-aged females. According to their origin and histopathologic features, retrorectal cysts are classified as squamous-lined (dermoid or epidermoid) cysts, postanal gut (tailgut) cysts, and rectal duplications (enteric or enterogenous cysts, enterocystomas). Described in this case report is an extremely unusual patient, a woman who simultaneously had a retrorectal cyst and an ovarian serous cystadenoma in addition to a long history of misdiagnosis and multiple unsuccessful surgeries.

Case Presentation: The patient was a 45-year-old female who presented with back pain, rectal fullness, constipation, and urinary symptoms. Upon her first pregnancy, a cystic pelvic mass had been misdiagnosed as an ovarian cyst. During the following 17 years, she had undergone several ineffective operations. The last CT scan and MRI studies revealed two separate noncalcified, unilocular, cystic lesions with well-defined borders in the retrorectal and retroperitoneal spaces. Two cysts were excised completely by a combined abdominoperineal approach. Pathological assessment revealed a dermoid cyst and an ovarian serous cystadenoma. No complications occurred during the 18 months of follow-up.

Conclusions: Coexistence of a retrorectal cyst and a serous cystadenoma is very unusual. Retrorectal cysts are rare entities that remain a difficult diagnostic and therapeutic challenge. Misdiagnosis and multiple unsuccessful surgeries are common. Complete surgical removal is the treatment of choice and requires a multidisciplinary approach in complicated cases.

Keywords: Rectum, Ovary, Cyst, Tumor, Epidermoid, Serous Cystadenoma

1. Introduction

Retrorectal cystic lesions in adults are so rare that most general surgeons treat only one such case during the course of their careers (1). These lesions' incidence rate is one in approximately 40,000 admissions (1-3). Most cases are congenital, and the developmental cysts are the most common congenital entity encountered in the retrorectal space (4, 5).

Although developmental cysts can be found in all age groups, they are frequently diagnosed in middle-aged women in a 3:1 female-to-male ratio (2, 4). They are benign in most cases (2) and have several histologic types. The most important types are squamous-lined (dermoid or epidermoid) cysts, postanal gut (tailgut) cysts, and rectal duplications (enteric or enterogenous cysts, enterocystomas) (6). They can be uni- or multilocular, and their content varies from clear fluid to dense mucus (2). Because of their rarity and nonspecific clinical presentations, the diagnosis of these lesions requires a high index of suspicion (7). Their diagnostic and therapeutic dilemmas contribute to the controversies regarding the treatment of these tumors (1). Due to the risk of malignant transformation, suppuration and pressure symptoms, retrorectal cysts should be removed in an operation in perineal or abdominal access (5). Other than that, misdiagnosis or inadequate surgery can lead to serious complications (1). Described in this case report is an extremely unusual patient, a woman who simultaneously had a retrorectal cyst and an ovarian serous cystadenoma in addition to a long history of misdiagnosis and multiple unsuccessful surgeries.

2. Case Presentation

A 45-year-old female was admitted to our surgical department at the Amir-AL-Momenin Hospital, Semnan, Iran, in May 2012. She presented a 17-year history of lower abdominal and back pain, rectal fullness, pain on defecation, constipation, and symptoms associated with genitourinary obstruction due to a pelvic mass. During her first pregnancy, a cystic pelvic mass had been diagnosed as an ovarian cyst, but the cyst was only drained during the caesarian section. Afterwards, she underwent five additional laparotomies, at first to remove a presumed ovarian cyst and then to treat the actual problem, the retrorectal cyst. The surgeries had been carried out in different

centers and even by unprofessional hands. The cyst was not resected completely, and it recurred after each of these procedures. After these unsuccessful surgeries, no other major procedure was done, and the patient was followed with transabdominal ultrasonography approximately every six months. When the patient's symptoms became unbearable, the cyst was drained through a minimal perineal incision between the coccyx and anus. In the last year, her symptoms did not improve after perineal drainages. It seemed that the cyst was not completely evacuated. A CT scan and MRI studies confirmed two large, separated, noncalcified, unilocular cystic mass lesions with well-defined borders in the retrorectal and retroperitoneal spaces. On a T1-weighted MRI, the lesions were low signal, while on a T2-weighted MRI, the lesions were high signal, indicating fluid content. There were no areas of heterogeneity or irregularity that enhanced with contrast. The retrorectal mass was located in the pelvic cavity behind the rectum and the vaginal canal, compressing and anteriorly displacing the uterus (Figure 1).

We decided to resect the cysts completely, because the case was so complicated after 19 surgeries (laparotomies and perineal drainage). A team of two general surgeons experienced in anorectal surgery, a urologist, a neurosurgeon, and two anesthesiologists operated on the patient for eight hours.

For complete excision, we used a combined abdominoperineal approach. At first, JJ stents were inserted through cystoscopy to find and protect the ureters during dissection, on account of the massive adhesions and anatomic distortion caused by previous operations. Then the laparotomy was done through a midline incision, revealing a large cystic lesion, adherent to the small bowel and ureters. The mass was carefully dissected and isolated. It seemed to have originated from the left ovary, and it contained clear fluid. The cyst was resected by salpingo-oophorectomy.

The second mass in the retrorectal space was completely apart from the first cyst. Although exceedingly difficult because of adhesions and fibrosis from the previous surgeries, the cyst was dissected with special attention to avoid injuring the rectum, ureters, or pelvic nervous plexus. For complete dissection of this cyst, the operation was continued with a perineal approach. A longitudinal incision was made between the anus and the coccygeal bone, the subcutaneous planes were divided, and the lumbosacral fascia was exposed. The anococcygeal ligament was transected. The retrorectal space was exposed by transection of the fibers of the levator ani. After complete dissection of the cyst, it was removed with a tract that had been created between cyst and skin in the site of the last perineal drainage. The pelvic floor was then reconstructed by suturing the fibers of the levator ani.

Upon histopathological examination, the first specimen consisted of the ovary containing a creamy brown unilocular 9 $\times$ 7.5 cm collapsed cyst with a smooth internal surface. The cyst was reported as "ovarian serous cystadenoma." The second specimen was a unilocular 12 $\times$ 6.5 $\times$ 8 cm cyst filled with yellow fluid and sebaceous material; the inner aspect of the cyst had a creamy smooth surface. Microscopically, this cyst was lined by stratified squamous epithelium. Also seen were scattered sweet gland-like structures, areas of erosion, granulation tissue formation, chronic inflammation, and fibrosis. The diagnosis was "dermoid cyst."

Postoperative recovery was complicated by respiratory failure, but eventually the patient improved and was discharged in generally good condition on the twelfth postoperative day. No complications occurred during the 18-month follow-up.

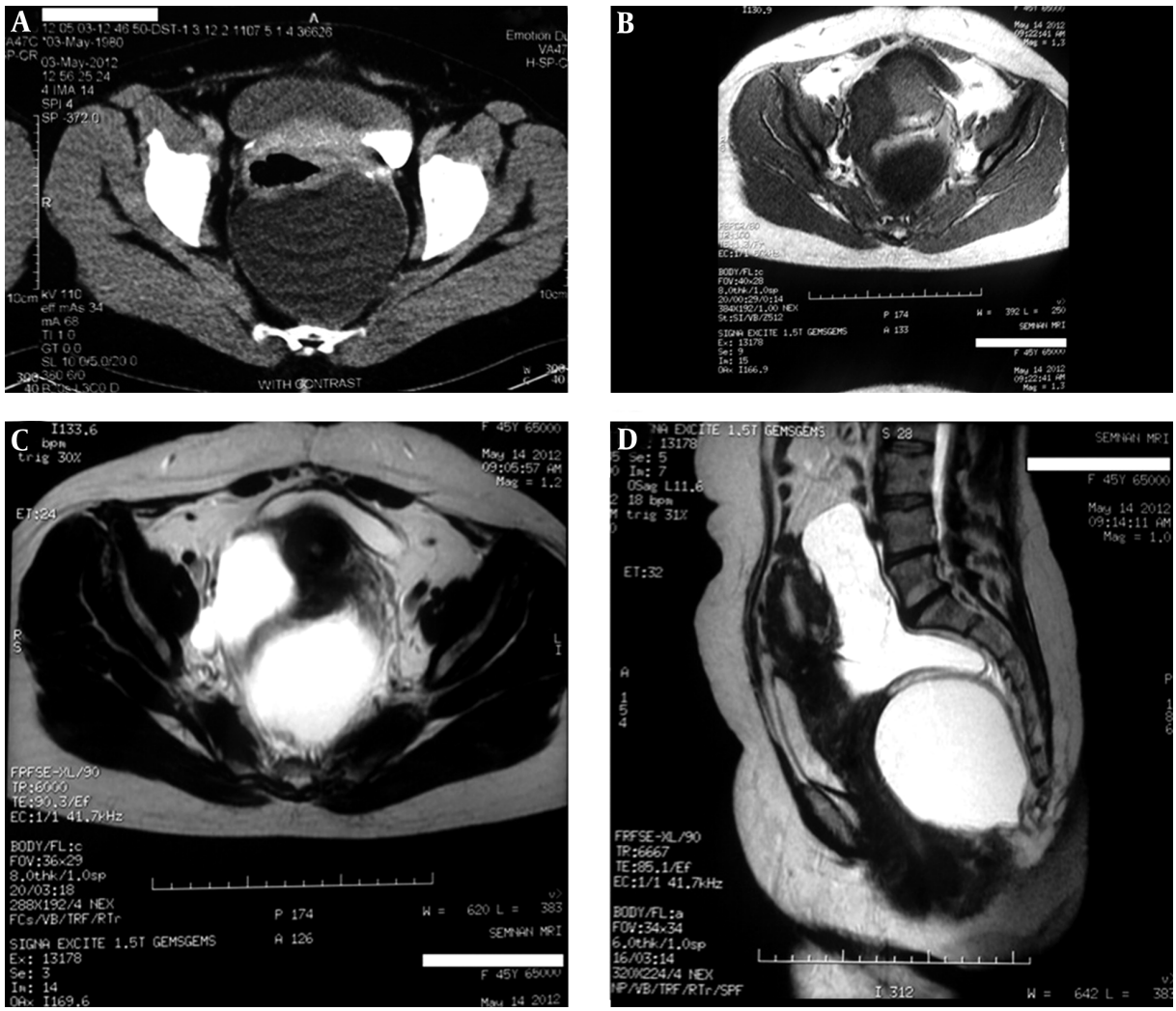
3. Discussion

The case described in the present report had two different problems simultaneously: a retrorectal cyst and an ovarian serous cystadenoma. The coexistence of a retrorectal and serous cystadenoma had been mostly unreported in previous articles. In this case, the ovarian cyst may have been a recently created mass that had been added to an old problem, the retrorectal cyst. This was probably the reason for the patient's persistent symptoms despite four drainages during the preceding year.

Unlike retrorectal cysts, serous cystadenomas are very common benign tumors of the ovary that may occur in any age group (8). Tumors of the ovary are common forms of both benign and malignant neoplasia in women, but about 80% are benign, and these occur mostly in young women between the ages of 20 and 45 years. The benign forms may be entirely asymptomatic, and there are occasionally unexpected findings upon abdominal or pelvic examination or during surgery (9). Treatment of serous cystadenomas can involve a cystectomy or oophorectomy, depending on the amount of ovary involved (8).

Retrorectal dermoid cysts are rare developmental cystic lesions that are thought to arise from caudal embryonic vestiges (4, 10). These benign lesions are lined with stratified squamous epithelium and are filled with dense muddy or fatty material. They can be differentiated from epidermoid cysts with both gross and microscopic analysis, because they contain skin appendages such as hair follicles, sweat glands, and tooth buds (4). Because these cysts are sometimes multiple (4), we had initially misinterpreted the MRI findings of our patient as multiple retrorec-

Figure 1. CT and MRI Images of the Patient



A, contrast-enhanced axial CT scan showing a large, homogeneous mass behind the rectum and the bladder, which are anteriorly displaced; B, axial T1-weighted MR image shows the masses are hypointense; C, axial T2-weighted MR image of the cysts showing masses are hyperintense; D, sagittal T2-weighted MR image shows two large, separated, well-circumscribed cystic masses, in the retrorectal and retroperitoneal spaces, that compress and anteriorly displace the uterus.

tal cysts; but during the operation, we found two different origins for the cysts.

The symptoms of congenital retrorectal masses are often subtle and nonspecific (10). Many of these masses are incidentally discovered during routine gynecologic examinations (1, 6), as we saw for the first time in our case; however, some patients have symptoms secondary to infection in the cyst. They present with a history of recurring retrorectal abscesses and repeated operations for anal fistulae (6). A small group of patients having large cysts experience symptoms resulting directly from the compression of the surrounding structures where the mass causes pain, constipation, narrowed stools, or pollakiuria (4, 10).

Because retrorectal cysts are rare entities and have nonspecific clinical presentations, a large proportion of cases are initially misdiagnosed. These patients may present with a history of multiple unsuccessful drainage procedures (7, 11). It often happens that patients have been treated with an ineffective method for many months before they finally receive professional help (5).

Singer et al. reported seven patients with retrorectal cysts who had been misdiagnosed before referral. These patients had been treated for fistulae in ano, pilonidal cysts, perianal abscesses, psychogenic, lower back, post-

traumatic, or postpartum pain, and proctalgia fugax before the correct diagnosis was made. The patients underwent an average of 4.1 operative procedures (7, 12). Our case had several times been diagnosed and treated for a large ovarian cyst and even after correct diagnosis underwent multiple unsuccessful procedures. In addition to the rarity of the occurrence of retrorectal cysts, the most probable reason for such a situation is physicians' lack of experience in their diagnostics (5). A history of multiple procedures should alert the clinician to the diagnosis of a retrorectal cyst. Once suspected, the correct diagnosis can be made with a physical examination and a CT scan or MRI before a definitive surgical procedure (7). Pelvic CT and especially MRI are the most important diagnostic tools when dealing with retrorectal cysts. Endorectal ultrasound can also be useful. It confirms the diagnosis and allows the precise determination of the size of the cyst and its relation to the rectum wall (5). All of these diagnostic tools provide crucial information for preoperative planning (2, 10).

The cornerstone of the treatment is complete surgical excision (6, 10). Chronic infection is the most frequent complication, occurring in 30%–50% of developmental cysts (3, 4). Because of the high predilection to infection, the preferred treatment for retrorectal cysts is complete excision. Removal is also recommended because of malignant degeneration arising in developmental cysts (6, 13, 14). Dermoidal cysts turn malignant in 10% -15% of cases (5). The approach to retrorectal masses can be abdominal, perineal, or a combination of the two, depending on the location and size of the lesion and its relationship with adjacent structures. If the mass does not extend above the level of the fourth sacral element, the perineal approach is the appropriate method. The abdominal approach is to be reserved for lesions whose lowest extent is above the level of the fourth sacral element, because this approach allows the best exposure of the pelvic structures (1, 15, 16). The combined approach is to be used for very large masses extending both proximally and distally to the fourth sacral element or for frankly malignant lesions with an infiltrative pattern that makes the isolation of the mass impossible by the perineal approach alone (10). We used the combined approach, because the retrorectal mass was extended below the level of the fourth sacral vertebral corpus in the MR images, and the transabdominal approach alone did not provide enough access to the two cysts.

Complete resection with negative margins is the standard surgical approach for retrorectal tumors. So in complicated cases, a multidisciplinary approach involving colorectal surgeons, neurosurgeons, possibly orthopedists, and radiation oncologists (for malignant tumors) is overemphasized in the literature (1, 17, 18). In spite of the benign course of the disease in the present case, we used a team of experts, because we expected probable adhesions, fibrosis, and anatomic distortion after several previous surgeries. The long-term outcome after resection of a retrorectal lesion depends upon the type of tumor and on adequate resection at the initial operation. For benign lesions, after complete surgical removal, the prognosis is good (10, 17).

3.1. Conclusions

The coexistence of a retrorectal cyst and a serous cystadenoma is very unusual. Retrorectal cysts are rare entities that remain a difficult diagnostic and therapeutic challenge. Misdiagnosis and multiple unsuccessful surgeries are common. Complete surgical removal is the treatment of choice and requires a multidisciplinary approach in complicated cases.

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