Case report

Retrorectal Dermoid Cyst: a Rare Clinical Entity

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ABSTRACT: The retrorectal space represents the anatomical site at which level we identify the embryologic reminiscents in which it can develop liquid tumors - *cysts* or solid tumors - *neoplasia*. These tumors are rare but pose a diagnostic and therapeutic interest. This paper presents the case of a young 18 years-old diagnosed incidentally at a gynecological examination, with a palpable tumor developed, at the retrorectal space. Imaging examinations - transvaginal ultrasound and abdominal - pelvic computer tomographic exam - have supported the presence of a cystic tumor with a maximum diameter of 7.8 cm., in the space retrorectal. The lesion presented surgical indication, so it needed a posterior approach with resection of the coccyx enough for the control and safety of the operation. Histopathological examination revealed a dermoid cyst. Five years after surgery the patient is presented in good general condition, asymptomatic without clinical and imaging signs of local-regional recurrence.

***KEYWORDS**: retrorectal cyst, dermoid cyst, retrorectal lesions, retrorectal space.*

Introduction

Development cysts (embryonic) are the most frequently encountered retrorectal lesions. The disease most often interests young women (1). Speciality literature describes epidermoid , dermoid, enteric or neuroenteric cysts, according to their origin and pathologic outcome (2). Although retrorectal cysts are asymptomatic, sometimes patients claim specific symptoms secondary of mass effect caused by the lesion volume (constipation, rectal fullness, dysuria, pelvic pain), or by some complications. Clinical, palpation can feel an intra - anal formation, intact or centered by a depression, indicating an ano-rectal fistula apparently trivial but it can also hide an unpleasant surprise.

Differential diagnosis inflicts with retrorectal tumors and complete surgical excision is the optimal treatment that brings healing, confirms the diagnosis and avoids evolutionary complications.

Case presentation

This paper presents the case of an urban 18 years old female patient, admitted in the surgery department of the Clinical Hospital CF 2 Bucharest, on suspicion of a retrorectal tumor detected following a gynecological examination.

Family history, personal and pathological information found nothing significant. Gynecological examination supported by transvaginal ultrasound detected a tumor in retrorectal space. Clinical examination revealed a normostenic patient, normal weight with a good general condition, conscious, cooperative. Visible mucous membranes and skin are normal colored. Woman hair, subcutaneous adipose tissue represented evenly. Impalpable peripheral lymph nodes. Normal torax, RR-16 /min, auscultation - bilateral vesicular murmur.

Rhythmic and clear heart sounds. BP : 110/60 mm Hg. Pulse-70 bpm. Abdomen is symmetrical, mobile with respiratory movements, smooth, painless. Bowel without features. Renal lodges are supple, Giordano maneuver is negative, physiological urination. Digital rectal exam reveals a normotonic anal sphincter, rectal ampulla with supple walls. In the posterior wall of the rectal ampulla, overlying sphincter area, palpation can feel a round tumor, about 5 cm in diameter, with regular shape, uniform consistency, soft, elastic, mobile, painless.

Normal biological examinations, Alpha fetoprotein also shows a normal value, ie 1.90 ng / ml.

Transvaginal ultrasound revealed the presence of an oval formation of 6/5, 5/8 inches, with regular contours and homogeneous echostructure at the level of retrorectal space.

Thoracic - pleura - pulmono – mediastinal radiography reveals emphasis of bilateral perihilar design.

Abdominal CT scan with contrast substance confirmed a spontaneous hypodense image with fluid densities and own wall, with inside septs that load contrast substance. Formation is fluid, noniodofil, size 7,7 / 5,2 cm, with mass effect on the rectum. Tomographic examination conclusion - sacrococcygeal cystic lesion with development in the rectorectal space (Fig. 1.2).

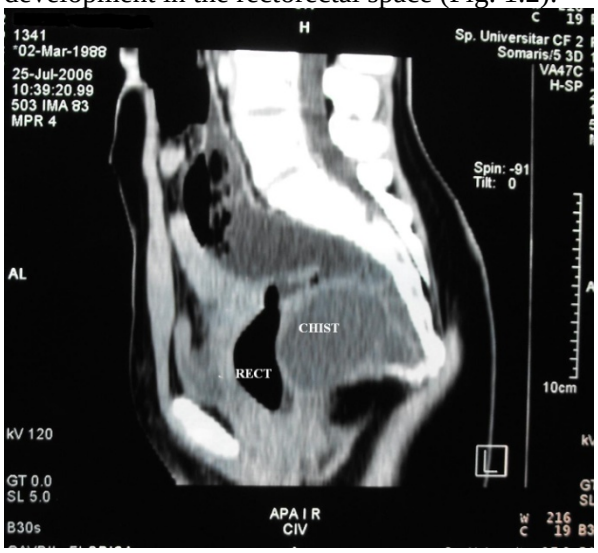

Fig. 1 Pelvic CT sagittal section

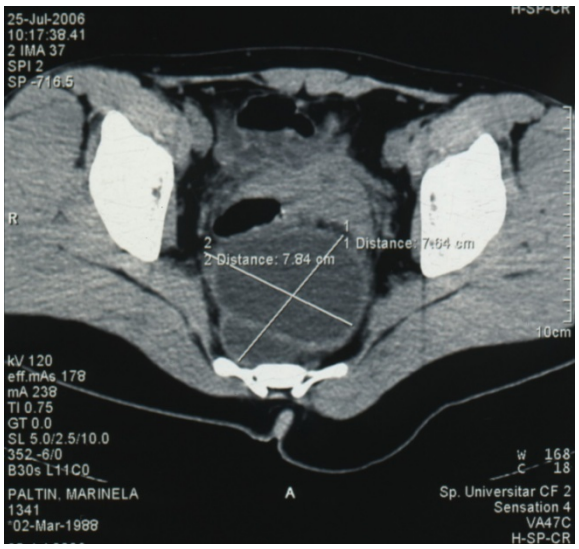

Fig. 2 Pelvic CT cross section

Fine needle aspiration biopsies, left parasacrococcigian space, under transanal digital control, discharges a thick liquid, yellowish, odorless, whose cytology study is inconclusive.

The surgery is performed under general anesthesia. Prone patient with the pelvis slightly ascended by a support. It is performed a left paramedian incision in the sacrococcygeal raphe, about 2 cm cranial to the anus (Fig. 3).

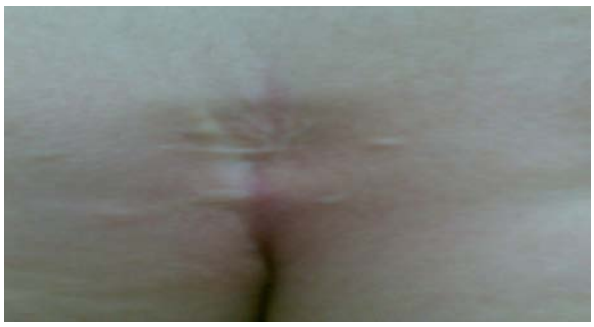

Fig. 3. Left paramedian postincisional scar – cranial to the anus.

Sacro-coccygeal joint from this level is discovered - coccyx excision is performed with attention to cutting the ano - coccygeal raphe in order to keep the integrity of the external sphincter. It reveals a cystic tumor of soft consistency, fluctuant content which starts from the top of the external anal sphincter and extends cranial occupying the entire retrorectal space. Formation shows an intimate relationship with their own rectal fascia, which is confused with. Ablation of the formation is performed by dissection with digital control of the superior rectal wall. The procedure ends with residual cavity drainage and suture the wound in anatomic planes with rare, nonabsorbable wires.

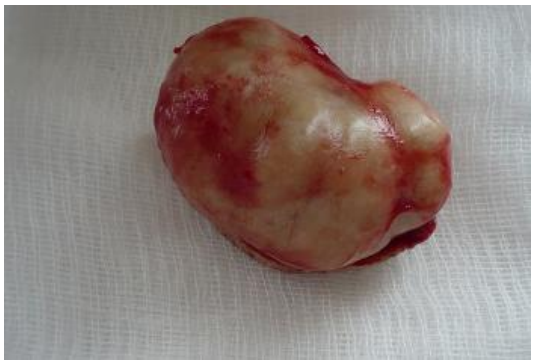

Fig. 4 Macroscopic appearance of the excision piece.

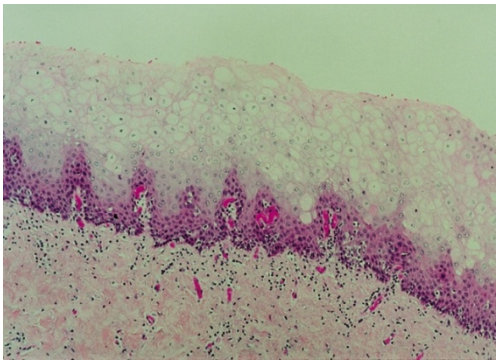

Fig .5 Microscopic appearance of the excision piece.

The specimen is represented by a cystic tumor, approximately 6 cm in diameter, containing a fetid sebum, yellow, in the amount of about 100ml. Pathological examination confirms the sacrococcygeal dermoid cyst (Fig. 4.5)

Postoperative evolution was simple, the drain was suppressed in the sixth day after surgery, the surgical wound healed per primam ; patient was discharged in the - 9th postoperative day and will be observed in ambulatory conditions.

Discussion

Retrorectal cystic lesions in adults are rare and in most cases are congenital. Developmental cysts are the most common congenital entity and includes the histologic type epidermoid cysts, dermoid, enteric and neuroenteric (3). Specialized literature describes two types of enteric cysts: hamartomatos cyst or mucin secreting cyst and cystic rectal duplictions.Anatomical, developmental cysts are described between the anterior sacrum side and posterior wall of the rectum. They occur most often in young women in a female / male ratio of 3:1 (3). Their most common complications are infection, abscessing and a later fistulization. Bleeding or malignancy is found in the case of enteric cysts. Chronic infection affects about 30-50% of cases, especially in epidermoid cysts and enteric ones. In these cases, the clinical picture is dominated by deep pelvic pain, fever and local inflammatory events. Evolution is with peri- anal or intra - anal fistulization with elimination of pus and remission of symptoms. A communication or a intra - anal fistula can exist without being related with the infection. Rectal bleeding is common in rectal duplication when cyst contains ectopic gastric mucosa, although it can occur secondary to cyst irritation or infection. Malignant degeneration, estimated at about 7% and met after 30 years of evolution has been reported for enteric cysts, histopathological types commonly found as adeno-carcinomas or squamous cell carcinomas. (4)

Clinical manifestations

Clinical symptoms are variable and are determined by the volume of the cyst, its mass effect, or the presence or absence of infection. It is estimated that about 50% of these cysts are completely asymptomatic, being discovered accidentally during a gynecological examination or abdominal- pelvic ultrasound made for another reason. Inspection of the posterior perianal region may reveal a funnel - shaped depression on the posterior anal line or even a chronic perianal fistula that opens to the skin, posterior of the anus or intra - anal on the posterior midline. Digital rectal exam may reveal a firm formation, smooth from retrorectal space that bulge into the lumen of the rectum. Symptoms are often attributed to the effects of compression, patients complaining of constipation, rectal fullness and tenesmus. Defecation is difficult, painful with anal and hypogastric pain. May occur, phenomena related to compression in the urinary tract manifested as high urinary frequencies and dysuria. Sometimes patients have sciatic-type pain. Sometimes symptoms is dominated by infectious complications or rectoragia.

Imaging

Simple abdominal radiography may reveal various sacrococcigians bone lesions that often evolve together with developing cysts. Retrorectal space widening on the profile incidence in the lateral study of barium enema may be suggestive of the diagnosis. Ultrasound examination shows a uni-or multi - locular cystic lesion located retrorectal, sometimes with internal echoes caused by mucoid material or present infection. Sometimes a fibrous linking-path from the cyst to the rectal lumen may suggest the presence of fistula. Computerized tomographic examination shows a well-defined oval uni - or multi - locular formation, thin walled and CT signal attenuation. Rarely are associated fine calcifications especially in dermoid and enteric cysts(4). If the cyst is infected, the walls may appear thickened with pericystic inflammatory changes. In chisto - anorectal fistulas CT appearance is of a cystic cavity containing air and liquid.

Differential Diagnosis

Sacrococcigian teratoma

Presacral sacrococcygeal teratoma is the most common lesion seen in pediatric pathology. Most of them (90%) are diagnosed at birth or in the first years of life and are benign. Risk of malignant increases with the years. Anyway, they are rarely diagnosed in adults. Usually there are no sacrococcigien abnormalities in simple radiography. TC or MRI examination show a well-defined heterogeneous formation with mixed content : cystic and solid. Rarely is entirely cystic and then usually is

benign. In about 50% of the sacrococcygeal teratoma cases they contain fat or calcification (5).

Anterior sacral meningocele

Anterior sacral meningocele is a congenital pathology although sporadic familial cases have been reported in the Currarino syndrome. It can be associated with epidermoid cyst. It is defined as a meningeal - cyst that occurs in the presacral space secondary to the agenesis of the anterior side of the sacrum. In half of the cases it is associated with other malformations : spina bifida, horned uterus, non-perforated anus. Simple radiograph shows sacral bone defect. Tomography and magnetic nucelar resonance examination - confirm the bone defect and highlights well-defined cystic tumor, unilocular, developed in the prerectal space. MRI images can show a communication between meningocele and tecal bag.

Anal gland cyst

In terms of pathology there are mucus-secreting cysts lined by a combination of stratified squamous, columnar and transition epithelium. It often communicates with anal crypts. Nuclear tomography and magnetic resonance examination reveal a retrorectal uni- or multi- locular cystic lesion, near the anal sphincter. Cysts may interest the cocis /tailbone, anal glands or ischiorectal space.

Rectal leiomyosarcoma

Rectal leiomyosarcoma is an infrequent tumor in less than 0.1% of cases. Most of them grow in the lower third part of rectum and men are particularly affected. Extension of these tumors is mainly local, by contiguity and vascular. They can invade rectal mucosa leading to ulceration and bleeding. Computerized tomographic examination reveals a heterogeneous tumor, well-defined, multilobular, with cystic necrotic components.

Retroperitoneal psudomixom

It is a rare clinical entity characterized by excessive mucin secretion. Glandular epithelium of the cyst is responsible for mucin secretion. Nuclear tomography and magnetic resonance sections formation show a homogeneous, well-defined, cystic, septated. Frequently mural nodules or calcifications are observed in the cyst.

Pelvic cystic lymphangioma

It is a rare benign tumor, especially common in childhood genetically determined. Although frequent location is cervical, it can be encountered in the retrorectal space. Cysts are lined with endothelial - type epithelium and secrete clear fluid. TC images show a multicystic lesion, well marked with fine septs. Common complications are bleeding and infection inside the cyst.

Treatment

Retrorectal cyst treatment is essentially surgical, removal of the cyst wall represents the main objective. The team will be composed of surgeons familiar with colorectal surgery, supplemented if necessary by an orthopedic surgeon and neurosurgeon. Posterior approach is preferred by most surgeons when the upper pole of the tumor is accessible to digital rectal examination. The patient is positioned prone flexed. The incision will be placed in the left inter-gluteal trench at about 2 cm from the anal orifice, extended cranial to the sacrum. The incision is thorough to the sacrum, coccyx and anooccigeal ligament. The ligament is sectioned in addition to the disjoints of S5 to coccyx (6). Most authors recommend excision of the coccyx for better exposure and a lower rate of recidivism.

Because it is assumed that all cystic lesions originate in the coccyx, cystic lesion should be removed en bloc with it (7). After removing the coccyx enter the retrorectal space through division of the levator anus(3). At this point, depending on the level and volume of the tumor it is questioned if to remove some sacral elements. When it implies the removal of some sacral segments, insertion of gluteal muscles will be divided bilaterally. Piriform muscles will be divided in the same way, and sacro-tuberous ligaments and sacro - spine will be identified and sectioned. The last two sacral nerves, after some authors even S3 – unilateral, may be slaughtered without neurological consequences. (8)

In contrast, bilateral sectioning of S3 is associated with sphincter incontinence. Removing the lower portion of the sacrum does not cause static pelvic instability. Bleeding can be a problem until complete removal of the sacrum. In case of high tumors vascularity is considered, which may require patient repositioning in anterior abdominal approach for hemostatic in uncontrolled bleeding. High lesions can claim a combined anterior and posterior approach. This abdominal - sacral approach provides good exposure and optimal vascular control. After lower median laparotomy will mobilize the left colon, left ureter will be exposed and preserved. Posterior plane between the parietal and visceral fascia is gently

dissected to separate mesorectum from the tumor. This generally requires cutting the rectum wings to mobilize the rectum through the plane of retrosacrate fascia. After this, laparotomy closes in anatomical layers and the patient is repositioned and the operation is continued through posterior approach. After tumor excision, the wound will be drained and sutured in anatomical planes. Specific postoperative complications include neurogenic bladder, parietal infections, sphincter incontinence, rectal fistula or retrorectal abscess.

Conclusions

A wide variety of cystic lesions occur in the retrorectal space and most of them are congenital. Development cysts show an identity of its own which determine radiological and histopathological diagnostic dilemma. Imaging examinations have characteristic signs, but definitive diagnosis remains the anatomical - pathological exam. Surgical treatment aims to establish an accurate diagnosis, lesion removal and prevent complications.

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