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CONTENTS

PRIMARY RETROPERITONEAL MUCINOUS CYSTADENOMA EMERGING FROM THE RETRORECTAL SPACE IN A MALE PATIENT – A CASE REPORT

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Abstract

This paper presents a rare and according to our knowledge a unique case in the world literature of a primary retroperitoneal mucinous cystadenoma (PRMC) with borderline malignancy in a male patient with localization in the retrorectal space, presented in a form of a system of perianal fistulas. The patient is a 67-year-old male, with a 23-year-old history of perianal fistula, which in the past two years has progressed by forming several severe fistulous channels on both sides of the anus and gluteal regions from which there was gross secretion of a mucinous content. He was operated on the assumption that the gross mucoproduction is from the rectal mucosa. In the first part of the operation, which was carried out in a jackknife position with epidural anesthesia, all the perianal and gluteal mucoid masses along with part of the muscle tissue and superficial fascia were removed creating a big defect. When it was obvious that the mucoid masses are emerging deeply from the praesacrococcygeal – retrorectal space, the patient was put in gynecological position with general anesthesia, and cylindrical amputation of the rectum according to Milles – Thompson, with removing of all of the tumorous tissue, was performed. During the final stage, the perineal wound was closed by rotating and sliding skin flaps. Postoperatively, there was a minor skin dehiscence on the perineal wound that healed spontaneously, and now four months after the operation, the patient is well, grateful, and without signs of local recidivism. PRMC is an extremely rare entity of only about 50 cases reported in the world literature and most of them in female patients. Our case will be overall 4-th reported case of PRMC with borderline malignancy (2, 4, 12) and a first case localized in the retrorectal space presenting with system of perianal fistulas.

Key words: primary retroperitoneal mucinous cystadenoma, retrorectal space

ПРИМАРЕН РЕТРОПЕРИТОНЕАЛЕН МУЦИНОЗЕН ЦИСТАДЕНОМ КОЈ ПОТЕКНУВА ОД РЕТРОРЕКТАЛНИОТ ПРОСТОР КАЈ МАЖ – ПРИКАЗ НА СЛУЧАЈ

Апстракт

Презентираме редок и спрема нашите сознанија од достапната светска литература единствен случај на примарен ретроперитонеален муцинозен цистаденом (ПРМЦ) со

“borderline“ малигнитет кај маж локализиран во ретроректалниот простор и презентиран во облик на систем од перианални фистули. Пациентот е 67 годишен маж со 23 годишна анамнеза за перианална фистула која во последните 2 години прогредира со формирање на повеќе изразени фистулозни канали од обете страни на анусот и глутеалните регии од кои се цеди обилна мукоидна содржина. Пациентот е опериан со убедување дека обилната мукопродукција потекнува од ректалната мукоза.Во првиот акт на операцијата во “jackknife” позиција и епидурална анестезија се отсранети сите глутеални и перианални желатинозни маси, а на некои места заради инфламаторни синехии и дел од мускулната фасција со формирање на голем дефект. Кога се виде дека желатинозните маси извираат длабоко од пресакрококцигеалниот-ретроректален простор се пристапи кон вториот акт на операцијата во гинеколошка положба и општа анестезија и се направи цилиндрична ампутација на ректумот по Milles-Thompson со отстранување на целото туморозно ткиво. На крајот, перианалната рана се затвори со ротациони и лизгачки кожни резенки. Иако постоперативно имаше помала кутана дехисценција, сега 4 месеци по операцијата нема знаци за рецидив а перинеалната рана е комплетно затворена. ПРМЦ е исклучтелно редок ентитет од само околу 50 случаи објавени во литературата. Нашиот случај ќе биде четврти случај на ПРМЦ од групата на оние со низок степен на малигнитет (2, 4, 12), а прв објавен случај на ПРМЦ во светската литература локализиран во ретроректалниот простор и презентиран во облик на систем од перианални фистули.

Клучни зборови: примарен ретроперитонеален муцинозен цистаденом, ретроректален простор

INTRODUCTION

Primary Retroperitoneal Mucinous Cystadenoma (PRMC) is a very rare kind of tumor of only about 50 cases reported in the literature, and most of them are in female patients (1). Our case is the 4-th case of PRMC with borderline malignancy reported in male patients worldwide, with unique localization in the rectorectal space, presenting itself with a system of perianal fistulas from which there is a constant gross secretion of a mucinous content. To our knowledge, the development of such a tumor in the retrorectal space of a male patient has never been reported in the literature. Mucinous cystadenomas usually develop in peritoneal cavity from the ovary (14), appendix (15, 16) or pancreas (17). In the retroperitoneum, there is no epithelial tissue, therefore PRMC are extremely rare. They are not connected with any of the above mentioned structures and are with still unclear histogenesis and biological behavior.

CASE REPORT

In this section, we are presenting a 67-year-old male patient with a 23-year-old history for perianal fistula. In the past two years the disease has progressed in a form of several fistulous channels on the both sides of the anus and gluteal regions, from which there was a gross secretion of a mucinous content (Figure 1).

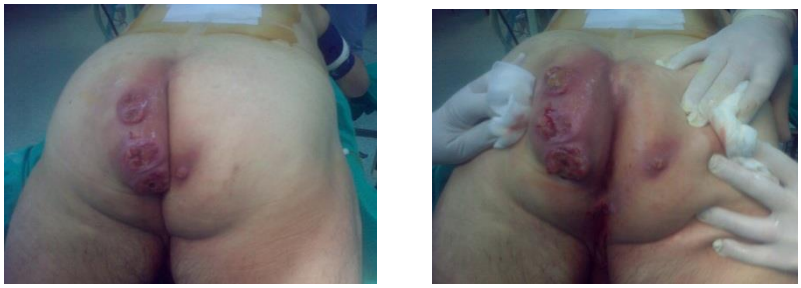

Figure 1 Preoperative condition

On inspection of the perianal and sacrococcygeal region there were four big external openings of fistulous channels from which, on pressure, there was a leakage of an odorous, mucinous secretion (Figure 2).

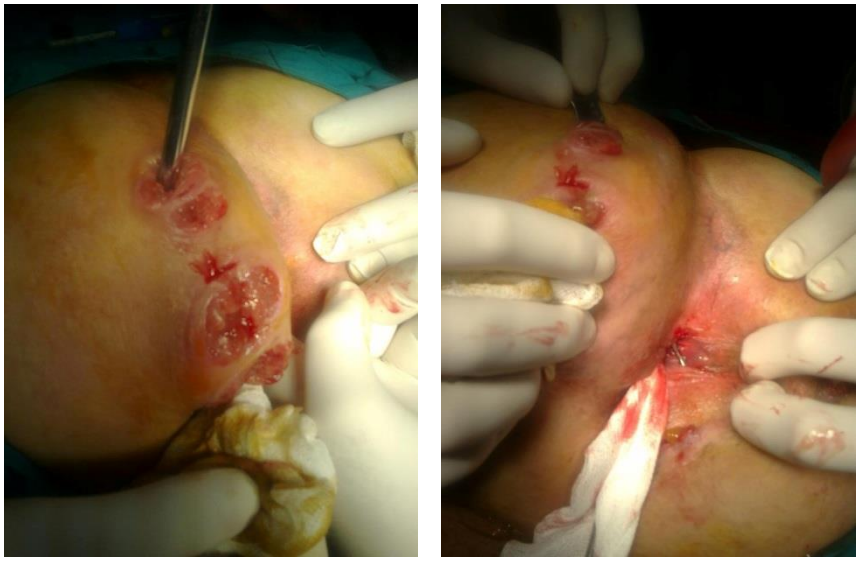

Figure 2 Probing the fistulous channels

On DRE and anoscopy a mammilla was registered, 8-10 cm from the anus, as a sign of communication with the rectum. Unfortunately, it was not practical to perform fistulography at that time. The ultrasound showed no signs of intra-abdominal tumor, as well as the colonoscopy has showed no signs of malignant process on the colon and rectum. On two consecutive patohistological findings from the material collected by the curettage of the fistulous channels there were no signs of malignancy. The patient was scheduled for operation without further investigations on the assumption that the gross mucoproduction arises from the rectal mucosa. Preoperatively, the colon was prepared conventionally, and prophylactic antibiotic regimen was started. The patient was informed and gave his consent for temporary or permanent colostomy. In the operating room he was put in a jackknife position, and, on explorative anoscopy, the internal opening on the posterior rectal wall about 8-10 cm from the anal verge was registered. During the first stage, total excision of the fistulous channels along with the skin and the subcutaneous tissue was performed. At several places and predominantly from the praesacral region mucoid masses emerged, at spots arranged in polycystic structures of cysts with a thin capsule. Furthermore, at several places, there was a severe fibrotic reaction, and in order to remove it, the superficial fascia along with the part of the muscle tissue had to be removed, as well as a part of the external anal

sphincter, which created a big tissue defect in the perianal and sacrococcygeal region (Figure 3).

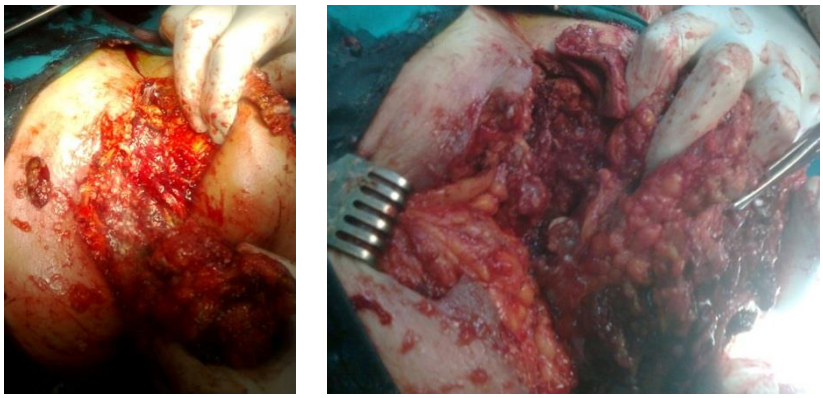

Figure 3 Removing the mucoid masses from perianal end gluteal region

The opening on the rectum proved to be very big; thus, the decision was made to perform an anterior rectal resection according to Dixon or "Ultralow" procedure. For this purpose the external sphincter was reconstructed, epidural anesthesia was combined with general anesthesia, and the patient was put in a gynecologic position. On the laparotomy, there were no mucouide masses in the peritoneal cavity. Throughout the mobilization of the rectum, it was concluded that the majority of the masses in form of polycystic structures with thin fibrous capsules filled with mucinous content were located in the praesacrococcigeal - retrorectal space. In order to fully clean the region of all the tumorous tissue, a cylindric amputation of the rectum, according to Milles-Thompson, with definitive colostomy was carried out. At the end the perineal wound was closed with sliding and rotating skin flaps. The postoperative course although prolonged went well, and the patient was discharged on the 14-th postoperative day with minor dehiscence of the skin perineal wound, that was proposed to be treated with daily washings with antiseptic solutions (Figure 4).

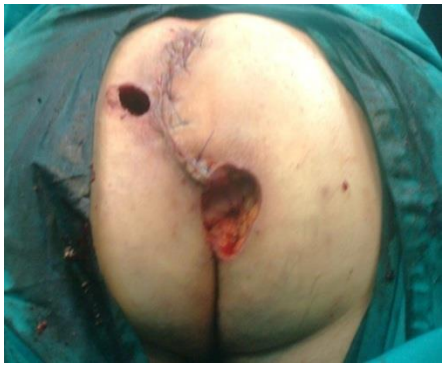

Figure 4 The condition on discharge from hospital

The patohistological finding from the operation was consistent with low malignant mucinous cystadenoma or mucinous cystadenoma with borderline malignancy that probably developed from terathoma or inclusion cyst in the retrorectal space (Figure 5) This was confirmed by additional immunohistochemical analyses where positivity for CK20,

CKAE1/AE3, as well for CEA, was found, and the tumor cells were negative for Vimentin, WT1, PLAP, AFP, CD57 and calretinin (Figure 6 a,b).

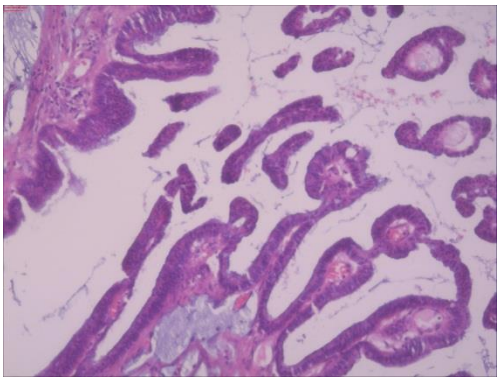

Figure 5 Hematoxylin and eosin staining

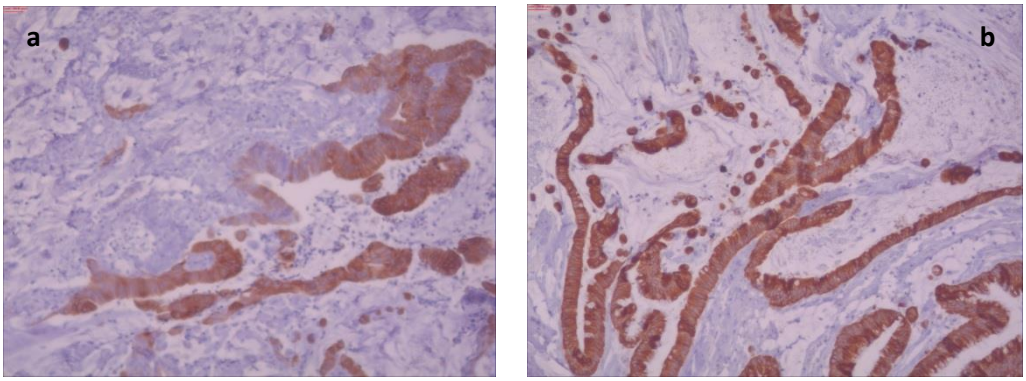

Figure 6 Positive imunostaining for CK20 (a) and for CKAE1/AE3 (b)

Now almost four months from the operation the patient is well, grateful and the perineal wound is completely healed without signs of local recidivism (Figure 5).

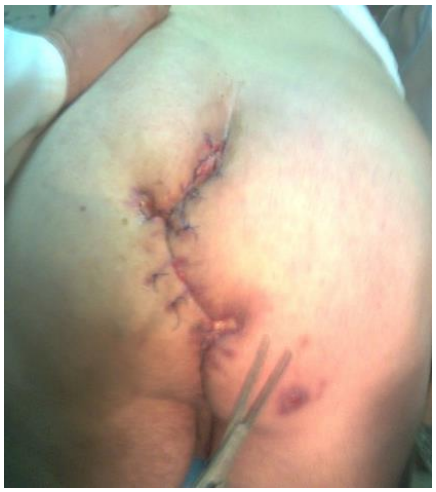

Figure 7 Current condition

DISCUSSION

Mucinous cystadenomas are tumors that usually develop from the ovarium, appendix or pancreatic tissue in a form of unilocular and multilocular cystic lessons filled with gelatinous yellowish fluid and lined with thin walls of cuboidal epithelium. They can become quite large and compress the surrounding structures or sometimes they can burst and spill the content in to the peritoneal cavity that results in pseudomyxoma peritonei (18). If they have a foci of metaplasia, dysplasia or malignant degeneration on the patohistological examination, they are classified as benign, borderline, or malignant.

Their development in the retroperitoneal space as primary tumors is extremely rare. In 2008, G. Bifulco et al. (1) reported a total of 48 cases in the literature. In 2009, A. Benkirane et al. (2) reported only 17 cases of PRMC with borderline malignancy in the English literature. In 2012, P. Navin et al. (3) reported a total of 19 cases of PRMC of e benign type in the English literature. The first case in a male patient was presented by Motoyama et al. (4) in 1994, as a form of PRMC of borderline malignancy localized in the right perinephric region and presenting as a cystic mass. T. P. Thamboo et al. (5), in 2006, reported the first case of primary retroperitoneal mucinous cystadenocarcinoma in a male patient localized anterior to the left psoas muscle and presented as a cystic mass. After that, there were three more cases in male patients overall reported as cystadenocarcinomas (6, 7, 8), three reported as benign cystadenomas (9, 10, 11) and two as PRMC with borderline malignancy (2, 12). All of them were located mostly in the upper parts of the retroperitoneum, presented either as a unilocular or multilocular large cystic mass or as a polycytic structure. Consequently, our case is overall the 4-th case of PRMC with low malignant potential in the male patient in the literature worldwide, and most importantly, it is the first case of a tumor developing in the praesacrococcigeal-retrorectal space and manifesting itself with a system of perianal fistulas from which there is gross secretion of mucinous content.

Table 1

Congenital	Osseous
Developmental cysts (epidermoid, dermoid, and mucus-secreting cysts; teratoma)	Osteoma
Chordoma	Osteogenic sarcoma
Teratocarcinoma	Simple bone cyst
Adrenal rest tumor	Ewing's tumor
Anterior sacral meningocele	Chondromyosarcoma
Duplication of rectum	Aneurismal bone cyst
	Giant cell tumor
Inflammatory	**Miscellaneous**
Foreign body granuloma	Metastatic carcinoma
Perineal abscess	Liposarcoma
Internal fistula	Lipoma
Retrorectal abscess	Fibroma
Chronic infectious granuloma	Fibrosarcoma
	Leiomyoma
	Leiomyosarcom
	…

Retrorectal space lies above the horse-shoe shaped supralevator space, behind the rectum, and it is bounded superiorly by the peritoneal reflection in communication with retroperitoneal space, anteriorly by the fascia propria of the rectum, laterally by the lateral ligaments, ureters and iliac vessels, and inferiorly, by the rectosacral or Waldeyer's fascia. It is a common place for embryologic remnants from which neoplasms and cysts may arise forming a group known as retrorectal tumors (19, 21). There is a worldwide accepted classification of these lesions – Table 1 (20).

The histogenesis of a PRMC, as well as their biological behavior, are not clearly explained. There are four major theories about their origin in the retroperitoneal space. **The first one** is that they develop from ectopic ovarian tissue, which is supported by the fact that the patohistological and immunohistochemical features of the PRMC are the same as their ovarian counterparts. This same theory could not explain the fact that in all of the specimens after surgery ovarian tissue could not be found or the existence in a male population. **The second theory** is that the tumors arise from terathomas in the retroperitoneum with proliferation of the mucinous epithelium. **The third** is that they develop from congenital intestinal duplication, and **the fourth,** the most widely accepted one, is that these tumors are a product of mucinous metaplasia of the mesothelial cells from the peritoneal inclusion cysts, which is most likely our case (13).

In almost all of the cases reported, the diagnosis was not made preoperatively. Moreover, all of the cases have been detected as unilocular or multilocular cystic lesions (using CT, MRI or at least ultrasound) that arise within the retroperitoneum, sometimes of enormous dimensions. The diagnosis was established on the patohistological examination of the extracted specimen. In our case the ultrasound could not detect the process hiding in the retrorectal space, and the operation was made without further investigation, because it was assumed that it is a case of system of perianal fistulas with mucinous secretion coming from the activity of the rectal mucosa. We usually strongly recommend the use of the CT and MRI in such cases, which can clearly show the location of the tumor and the relationship with the surrounding structures, especially the rectum and the anal sphincters. Endorectal ultrasound can also be good in that manner. Fistulography, sygmoidoscopy, and barium enema, if necessary, are used as a proof in our case. Punctional biopsy along with other retrorectal tumors should be contraindicated because spreading of malignant cells may occur.

The operation is an essential modality of treatment whenever there is a suspicion of PRMC, because if not malignant at a certain time, it usually becomes one. The operation should be radical in eliminating all the tumorous tissue and, if possible, not spilling the content of the cysts on the surrounding tissue. In the majority of the cases adjuvant chemotherapy is not recommended if the operation is radical, except in cases where there is aggressive component like anaplastic carcinoma or sarcoma on the patohistological finding, although there isn't much hope for those patients. In the other cases, if it is a benign cystadenoma, the patient is cured, and for the borderline and malignant cases, periodic controls are recommended. In our case, the process was spread throughout the whole perirectal and perianal region with destruction of the posterior rectal wall and posterior part

of the sphincterous apparatus. Despite the intention to preserve the anal sphincters, amputation of the rectum was necessary to clean all the area of the tumor.

CONCLUSION

Although this could be the first reported case of this condition in the literature, the retrorectal space is the perfect place for development of such tumors if we consider its anatomic characteristics and neoplastic processes that can be found there, as well as the proposed theories for the histogenesis of the PRMC. Being aware of this possibility, CT and especially MRI should be essential diagnostic tools in preoperative evaluation in cases of perianal fistulisation with predominantly mucinous secretion that will show the exact location of the tumorous process, the extension, and the relationship with the surrounding structures, especially the rectum and anal sphincters. Since the radical operation is the main modality of successful treatment and a cure for these tumors on this location, the patient should be informed and give his approval for a worse possible outcome for his quality of life postoperatively, for example, amputation of the rectum in order for the surgeon to completely clean all the tumorous tissue.

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