Case Report

DOI: 10.21276/APALM.1453

Prerectal mucinous cystadenoma: A case report and considerations about its origin

Jaime Zuloaga[1], Javier Arias-Diaz[2]*, Maria Jesus Fernandez-Aceñero[3], Cristina Diaz-del-Arco[4] Julio Mayol[1] and Antonio Torres[1]

[1]Dept. of Surgery, Complutense University of Madrid, Instituto de Investigación Sanitaria Hospital Clínico San Carlos, Spain
[2]Dept. of Surgery, Complutense University of Madrid, School of Medicine, Instituto de Investigación Sanitaria Hospital Clínico San Carlos, Spain
[3]Dept. of Pahology, Complutense University of Madrid, Instituto de Investigación Sanitaria Hospital Clínico San Carlos, Spain
[4]Dept. of Pathology, Instituto de Investigación Sanitaria Hospital Clínico San Carlos, Spain

ABSTRACT

Mucinous cystadenoma is a generic denomination usually applied to cystic formations filled with mucinous content and showing signs of epithelial proliferation. They usually arise in ovaries, pancreas and appendix, but there have been occasional reports in many other locations. Most retroperitoneal and mesenteric mucinous cystadenomas have affected women, and a potential müllerian origin has been postulated, although for most of them their histogenesis remains uncertain. Some cases have shown malignant behavior. We herein describe a case of mucinous cystadenoma affecting a young man with an unusual prerectal location. An exhaustive study was performed as a basis for the discussion of possible origins for this tumor. This lesion showed premalignant proliferative changes, suggesting an early stage in a potential malignant transformation.

Keywords: *Mucinous Cystadenoma; Tailgut cyst; Cysts/pathology; Rectal Diseases/pathology; Rectal Diseases/surgery*

Introduction

Mucinous cystadenoma usually arise in ovaries, pancreas and appendix, but they have been reported in many other locations.[1] Most retroperitoneal and mesenteric mucinous cystadenomas have affected women, and a potential müllerian origin has been postulated, although their histogenesis remains uncertain for most of them.[2][3] Some cases have shown malignant behavior.[1]

We herein describe a case of mucinous cystadenoma affecting a young man with an unusual prerectal location. This lesion showed proliferative changes, suggesting an early stage in a potential malignant transformation.

Case Report

A 45-year-old male presented with a history of low rectal bleeding and a palpable rectal mass at 6 cm of the anal margin. At colonoscopy the mass had a rounded surface and a submucosal origin. Transrectal ultrasonography identified a 4x3.5 cm extraluminal tumor, which appeared encapsulated and independent of both the mucosa and muscular layers of the rectum.Pelvic MRI scan showed a lobulated cystic tumor, measuring 5 cm of diameter and located 7 cm above pectinate line, anteriorly to the rectum (Figure 1). The tumor was independent of the rectal mucosa and also was separated from the prostate and seminal vesicles by a fatty dissection plane. The tumor capsule appeared thin and uniform, and no adenomegalies are found. RMI findings were thus concordant with the diagnosis of an epidermoid or duplication cyst.No additional pathological findings were observed in the abdominal US. Routine blood tests were normal, and serum tumor markers CA 15.3, CA19.9 and CEA were within normal range. Feminization traits were discarded, male and female sex hormone levels and those of sex hormone binding globulin were within normal limits.

At laparotomy, a 5-cm mass was found anterior to the rectum. It was noted to be intimately involved with the muscular layers of the rectum wall but it did not appear to involve the prostatic fascia. A low anterior resection with stapled colorectal anastomosis and a loop ileostomy were performed. The patient showed uneventful recovery and he is alive and well eighteen months after discharge.

The lesion was sent as a whole. It was found multilocular when cut, with thin delicate septa within its wall and filled with a sticky viscous material. Serial sectioning revealed no areas of macroscopic papillary growth or thickening in the cyst wall. The rectosigmoid colon was normal.

Microscopic analysis of hematoxylin-eosin sections of the cyst wall revealed that the mass was growing within the adventitia and the muscular coat of the rectum with no mucosal involvement. The cyst loculi were filled

with amorphous bluish material (mucin) and sparse inflammatory cells. The outer cyst wall corresponded to fibrous paucicellular tissue with no muscular layer or adventitial tissue and the muscular coat of the rectum was compressed by the tumor growth. The inner wall was partially denuded of epithelium, but in some areas it showed a well-preserved layer of columnar tall cells (Figure 2a). These cells were relatively uniform and showed mucin production, mainly in the apical cytoplasm. The nuclei were all basally located and showed neither atypia nor mitosis (Figure 2b). Some foci of pseudostratification with micropapillary formations were noted (Figure 2c), but the basal membrane was preserved. No other mucin lakes or epithelial inclusions were noted in other areas of the rectal specimen sent for examination after thorough sampling.

Immunohistochemistry performed on the cyst wall revealed intense positivity for pancytokeratins (low and high molecular weight), cytokeratin 20 (marker of hindgut origin; Figure 3a), and carcinoembryonic antigen (CEA). Markers suggesting mesothelial (calretinin and mesothelin), müllerian (cytokeratin 7), prostatic (p63, chromogranin A, vimentin and prostate specific antigen) or intestinal (CDX-2) origin, were all negative. Estrogen and progesterone receptors were also negative. Proliferative index with Ki67 (Figure 3b) was very low and restricted to the basal layer of cells in the foci of pseudostratification, therefore suggesting a benign nature of the lesion. The cyst did not communicate with the rectal lumen at any point, so we could not confirm diverticular disease as the origin of the lesion. Besides, the cyst wall did not show a muscle coat and it only had the epithelial inner lining and a peripheral narrow band of connective tissue, what seems to discard a duplication cyst of the rectal wall. Final diagnosis was mucinous cystadenoma.

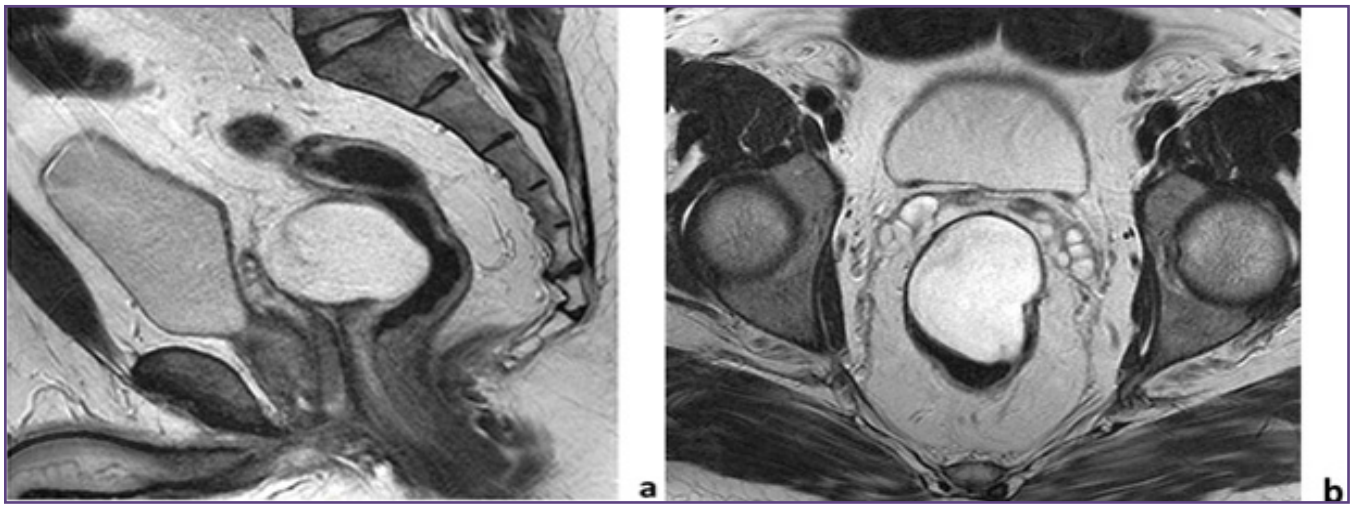

Fig. 1: Pelvic MRI scan showing a lobulated cystic tumor, located anteriorly to the rectum and independent of the rectal mucosa. A fatty dissection plane separates the cyst from the prostate and seminal vesicles. A sagittal plane (a) and an axial one (b) are shown.

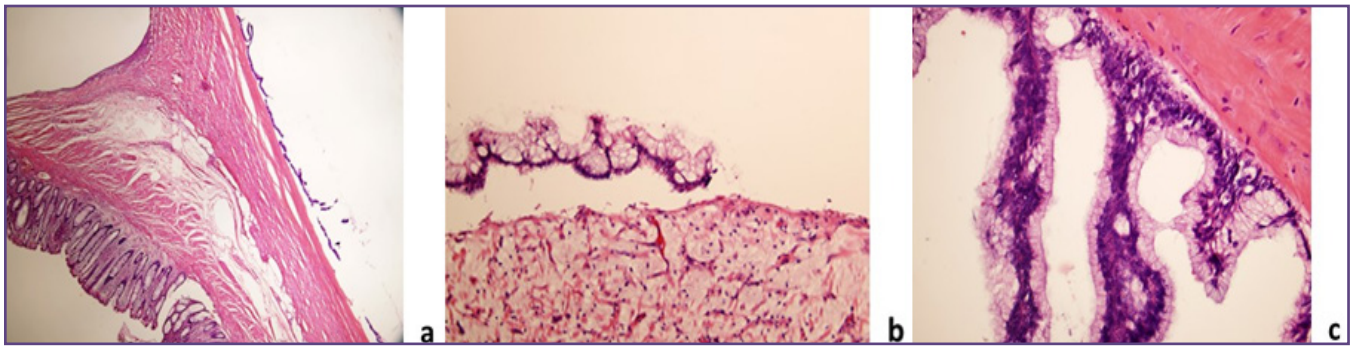

Fig. 2: A. Low-power view of the cyst wall showing the relation to the bowel mucosa (lower left). Note the cyst wall is only composed of fibrous tissue and compresses the muscular coat. B. Medium power view of the cyst wall showing a partially detached monolayer of epithelial cylindrical cells with apical mucin. C. Medium power view of the areas with pseudostratification of nuclei and papillary ingrowths into the cyst lumen (A, H-E x 40; B and C: H-E, x 200).

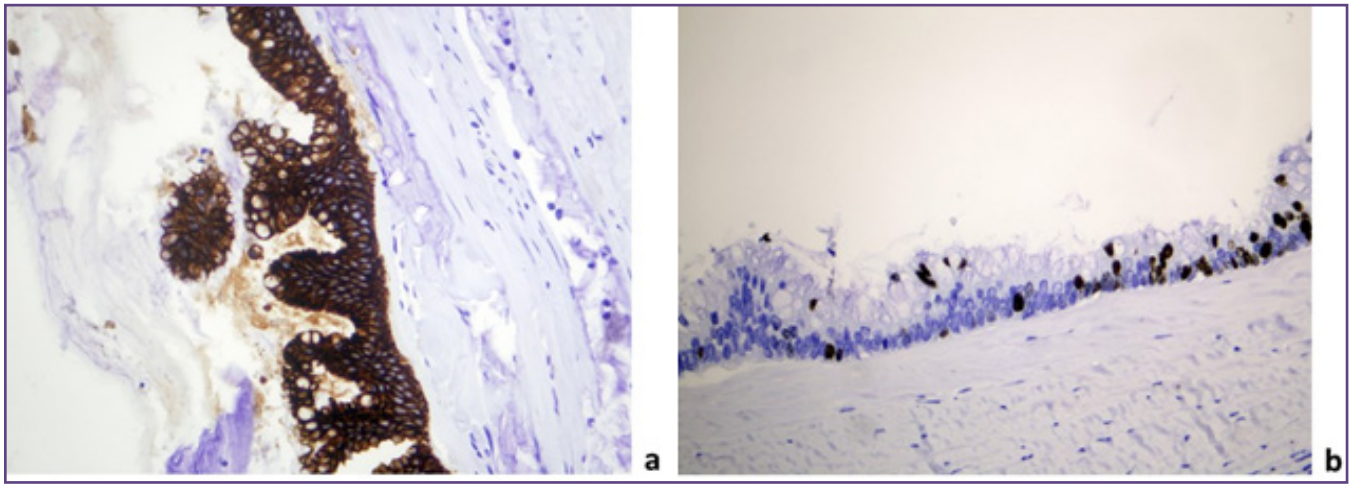

Fig. 3: A. Immunohistochemical staining for cytokeratin 20 confirms hindgut origin of the epithelial lining. B. Immunohistochemical staining for Ki67 confirms low mitotic activity, limited to basal layers. (A. Immunohistochemistry for CK20, x 200; B. Immunohistochemistry for Ki67, x200).

Discussion

The pathologic diagnosis was eventually based on pure morphological grounds. We had a multilocular prerectal cyst lined by mucin secreting epithelium with micropapillary projections and pseudostratification, but without any sign of stromal invasion suggestive of malignant transformation. The histopathological image was in all aspects similar to that seen in appendicular or ovarian mucinous cystadenoma with some features of the so-called borderline lesions in this group.[4] A thorough literature review of this issue revealed rare reported cases of mucinous cystadenoma originating in the retroperitoneum or the mesentery. Nevertheless, very few cases were located in the perirectal anterior area.[5] These reports affected mainly women and discussion regarding origin suggested most a müllerian origin or a mesothelial one, based on the positivity of two cases for calretinin.[6] We consider that the mesothelial origin has been reasonably excluded in the present case, for the epithelial lining was negative for calretinin and mesothelin.

The müllerian remnant in male is the prostatic utricle. In our patient, there was a lack of continuity between the cystic mass and the prostate. In addition, the plasma levels of sex hormones were normal and immunohistochemical markers indicative of a müllerian origin were absent.

The simple cyst in the seminal vesicle is a unilocular cystic lesion located at lateral to the midline in the retrovesical area. In our case, the cyst was also independent of the seminal vesicles, and did not contain sperm fragments, which seems to be a characteristic finding in these cysts.[7]

Another potential source for this lesion could be a teratoma with complete regression of all non-ectodermal components, but even in this case it would be expected to have some mixture of epithelial types, which was not found in our patient. A cyst arising from remnants of the cloacogenic membrane could be another possibility. However, the cytokeratin 20 positivity makes it however very unlikely. The absence of any history of prior colorectal biopsy or procedure makes also unlikely for it to arise in a misplaced or ectopic colorectal mucosa.

We consider that the most plausible explanation for our lesion would be a premalignant epithelial proliferation in a tailgut cyst with an unusual location.[5] Nevertheless, the prerectal location cast some uncertainty over this statement.[8]

Tailgut cysts usually arise in the presacral space and are related to a lack of involution of the tailgut, which usually regresses completely between the 7-8th week of development. It is known that most of these cysts are lined by mucin-secreting epithelium of intestinal type and there are reports of aggressive mucinous adenocarcinomas arising in long-standing cysts of this type.[9] We can guess that the neoplastic growth of the mucin secreting epithelium lining the cyst wall has led to a papillary mucinous cystadenoma, similar to the ones originating in müllerian structures. It might be suggested that this cystoadenomatous pattern can be a premalignant phase of the mucinous adenocarcinomas known to arise in these embryologic remnants.

Given that tailgut is a dorsal growth in the developing embryo, it is puzzling that remnants of such a clearly retrocloacal structure should be present in prerectal location. In fact, the possible tailgut origin of prerectal cysts is opposed by some.[8]

Nevertheless, there have been a few reports of cysts affecting the anterior wall of the rectum that have been eventually labeled to be of tailgut origin.[5][10] In those reports, cysts arising in the prostate utricle or the seminal vesicle, and also duplication enteric cysts, have been considered in the differential diagnosis. The former possibilities have been excluded in the present case with immunohistochemical studies, and the latter can also be excluded based on the lack of a neatly developed muscular and adventitial coat around the cyst wall.

One peculiar aspect of our case is that there was only one kind of epithelium. Tailgut cysts usually show different types of epithelial linings, including squamous and cuboidal ones. Besides, it is fairly frequent in these lesions to have intense inflammatory changes with erosion of the epithelium and substitution by macrophages or lymphocytes.[8] This inflammatory reaction was absent in our case. However, the reactivity for cytokeratin 20 supports this embryologic origin in tailgut structures.

Conclusion

We report a case of mucinous cystadenoma, which we consider related to a congenital tailgut cyst of unusual location and discuss on the histogenesis of this lesion based on immunohistochemical data.

Reference

1. Tapper EB, Shrewsberry AB, Oprea G, Majmudar B. A unique benign mucinous cystadenoma of the retroperitoneum: a case report and review of the literature.Arch Gynecol Obstet. 2010;281:167-169.
2. Subramony C, Habibpour S, Hashimoto LA. Retroperitoneal mucinous cystadenoma.Arch Pathol Lab Med. 2001;125:691-694.
3. Isse K, Harada K, Suzuki Y, et al.Retroperitoneal mucinous cystadenoma: report of two cases and review of the literature. Pathol Int. 2004;54:132-138.
4. Brown J, Frumovitz M. Mucinous tumors of the ovary: current thoughts on diagnosis and management.Curr Oncol Rep. 2014;16:389.
5. Jang SH, Jang KS, Song YS, et al. Unusual prerectal location of a tailgut cyst: a case report. World J Gastroenterol. 2006;12:5081-5083.
6. Demirel D, Gun I, Kucukodaci Z,et al. Primary retroperitoneal mucinous cystadenoma with a sarcoma-like mural nodule: an immunohistochemical study with histogenetic considerations and literature review.Int J Gynecol Pathol. 2013;32:15-25.
7. Aumuller G, Riva A. Morphology and functions of the human seminal vesicle. Andrologia. 1992;24:183-196.
8. Hjermstad BM, Helwig EB. Tailgut cysts. Report of 53 cases.Am J Clin Pathol. 1988;89:139-147.
9. Mathis KL, Dozois EJ, Grewal MS,et al. Malignant risk and surgical outcomes of presacral tailgut cysts. Br JSurg. 2010;97:575-579.
10. Au E, Anderson O, Morgan B, et al. Tailgut cysts: report of two cases.Int J Colorectal Dis. 2009;24:345-350.

***Corresponding author:**
Javier Arias-Diaz, MD PhD, Dpto. de Cirugía, Facultad de Medicina, Universidad Complutense de Madrid, Ciudad Universitaria, Madrid, Spain
Phone: +34 607715705
Email: javardi@gmail.com

Financial or other Competing Interests: None.

Date of Submission : 01.04.2017
Date of Acceptance : 23.05.2017
Date of Publication : 25.08.2017