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CASE REPORT

Retroperitoneal Mucinous Neoplasm Arising from Colonic Duplication Cyst

María M. Rojas-Rojas[1] · Marcela Mejiah[1] · Martha Mora[2] · Jorge Otero[3] · Fernando Arias-Amézquita[4] · Eduardo Londoño-Schimmer[4] · Paula A. Rodríguez-Urrego[1]

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Introduction

Enteric duplication cysts are uncommon abnormalities with a reported incidence of 1 in 4,500 to 10,000 live births. These lesions are defined as mucosa lined structures, contiguous and attached to the bowel wall, and surrounded by at least one smooth muscle layer. Most of them (80%) are diagnosed within the first 2 years of life and the symptoms depend on the location [1]. They can occur in any portion of the alimentary tract, but the retroperitoneum is a very unusual location; the most common location is the small bowel (duodenum 2–12% and jejuno-ileal > 50%), followed by the esophagus (15%), stomach (4–9%), and large bowel/rectum (7%). Complications can include hemorrhage, intussusception, obstruction, or malignant transformation [2]. Malignant change is an unusual complication of alimentary tract duplications and the exact incidence is unknown. Thirty-three cases of malignant tumors arising specifically from duplications of the large intestine have been reported including adenocarcinoma ($n = 26$), squamous cell carcinoma ($n = 3$), carcinoid tumor ($n = 2$), mucinous cystadenocarcinoma ($n = 1$), and gastrointestinal stromal tumor ($n = 1$) [3–5]. The aim of this report is to describe the first case of a low-grade mucinous neoplasia that originated in a retroperitoneal colonic duplication cyst.

Case Report

A 26-year-old woman with a history of polycystic ovarian syndrome who complained of abdominal cramps was found to have a retroperitoneal mass during follow-up transvaginal ultrasound. Ultrasound found a paraovarian mass measuring 57 × 49 × 43 mm. A contrast computed tomography reported a prevertebral septated cystic lesion with calcifications at the level of L5-S1. Magnetic resonance imaging demonstrated a round mass with lobulated margins and thickened wall and septa located in front of the sacral promontory next to the right common iliac vessels. Laboratory studies were CEA 1.47 ng/mL (0–5 ng/mL), CA125 10.36 U/mL (0–35 U/mL), and AFP 0.98 ng/mL (0–15 ng/mL).

According to these findings, the patient underwent laparoscopic resection revealing a retroperitoneal calcified cystic and solid tumor. The tumor was in close contact with the right ureter and iliac vessels but no relationship with any part of the bowel and/or mesenteric root was observed. There was no evidence of mucin or any peritoneal disease at the time of surgery. The entire mass was resected without rupture and it was sent for histopathological examination.

Pathological Findings A 6-cm rounded and firmed mass with intact surface was received. Sectioning revealed a unilocular cyst filled with mucin. The wall was firm and calcified. Light microscopy showed a colonic type lining and a wall that resemble the gastrointestinal tract with a concentric layer of smooth muscle (muscularis propria). However, the submucosa displayed small submucosal glands (Fig. 1(a)). Although the majority of the mucosa was denuded, it focally showed an area of bizarre architecture with hyperplastic changes somewhat reminiscent of a serrated polyp (Fig. 1(b)). The lesion was submitted entirely, and the rest of the wall was hyalinized

✉ Paula A. Rodríguez-Urrego
paula.rodriguez@fsfb.org.co

[1] Department of Pathology, Hospital Universitario Fundación Santa Fe de Bogotá, Calle 119 # 7-75, Bogotá, Colombia

[2] Nursing Department, Hospital Universitario Fundación Santa Fe de Bogotá, Calle 119 # 7-75, Bogotá, Colombia

[3] Department of Clinical Oncology, Hospital Universitario Fundación Santa Fe de Bogotá, Calle 119 # 7-75, Bogotá, Colombia

[4] Department of Surgery-Subdivision of Peritoneal Diseases, Hospital Universitario Fundación Santa Fe de Bogotá, Calle 119 # 7-75, Bogotá, Colombia

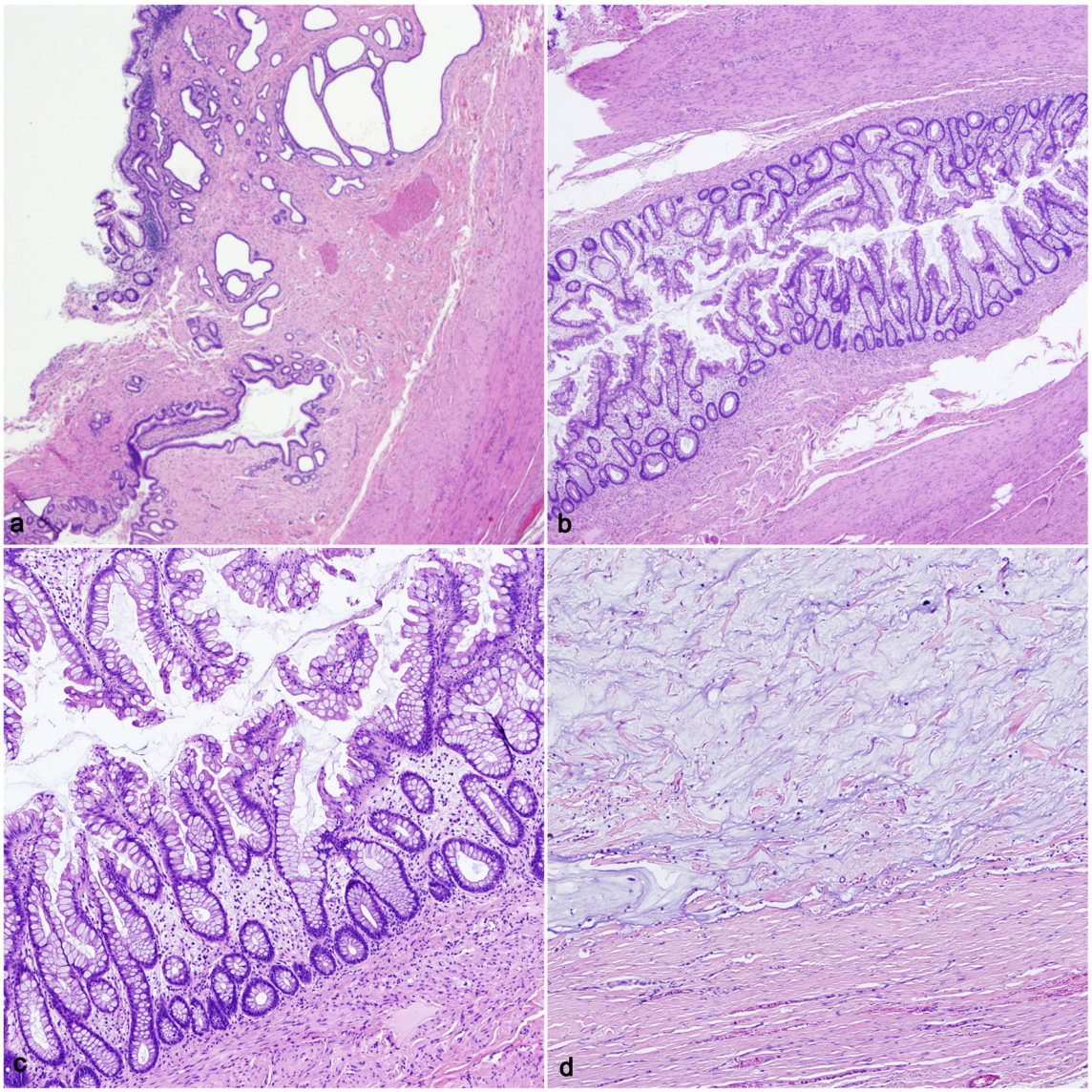

Fig. 1 Histopathological examination. (a) Hematoxylin and eosin (H&E) 40× duplication cyst wall with colonic type lining and submucosal glands. (b) H&E 40× foci of mucinous and serrated mucosal changes without high-grade dysplasia. (c) H&E 100× higher power field of colonic type mucosa with epithelial changes compatible with low-grade mucinous neoplasia, limited to the mucosa. (d) H&E 200× hyalinized wall with mucin dissection

with intramural dystrophic calcifications, ossification, and mucin extravasation with the epithelial changes limited only to the mucosa. These findings were consistent with a low-grade mucinous neoplasm that originated from retroperitoneal colonic duplication cyst.

The patient was followed up and no abnormal serum markers were found. Two months after the surgery, she was taken for an exploratory laparoscopy to visualize the right colon, appendix, and abdominal cavity. A slightly thickened distal third of the appendix and a lymphadenopathy of the mesoappendix were seen. Appendectomy and biopsies of the retroperitoneum were performed. The histopathological exam of the appendix showed benign reactive follicular hyperplasia, and the retroperitoneal biopsies were negative for tumor cells or mucin.

Discussion

Alimentary tract duplications are rare abnormalities and the cause is not completely known. It is thought that they result from embryological development fault of the dorsal foregut and could be associated with other congenital anomalies [6]. The preoperative diagnosis of these lesions is challenging given that symptoms are nonspecific and can include abdominal pain, diarrhea, constipation, nausea, vomiting, and palpable

mass [3]. In the present case, the patient had occasional abdominal cramps and the mass was detected by routine ultrasound.

Enteric duplications found in the retroperitoneum are uncommon. In our case, the mass was in close contact with the right ureter and iliac vessels but without connection to the alimentary tract. According to literature, only 15 cases of retroperitoneal enteric duplication cyst without connection to the alimentary tract had been reported, with none of those cases presenting malignant transformation [7, 8].

Malignant change in duplication cysts is an extremely rare complication. A study of malignancies arising from duplications cysts from any part of the digestive tract analyzed 67 reported cases and found that 46.2% of the tumors originated from duplications of the large intestine. The most common histologic type was adenocarcinoma, followed by squamous cell carcinoma and carcinoid tumor [3]. Malignant change was more common in duplications of the large intestine in comparison with duplications from other part of the digestive tract, and special attention should be paid in these cases.

Only six cases of mucinous neoplasias arising from enteric duplications have been reported. Four cases arise from small intestine duplications, one from an appendiceal duplication and one from a colonic duplication (mucinous cystadenocarcinoma). The case herein, represents the first colonic duplication cyst with malignant transformation of a low-grade mucinous neoplasia located at the retroperitoneum [5, 9–12].

The preoperative diagnosis of malignant transformation is difficult but should be considered when a solid component is found within a duplication accompanied by high serum CEA or CA19-9. In this setting, surgical removal is considered the treatment of choice to relieve symptoms and prevent other type of complications such as bleeding, intestinal obstruction, and infection [3].

The differential diagnosis for retroperitoneal neoplastic lesions includes mucinous cystadenoma, cystic teratoma, cystic lymphangioma, mullerian cyst, epidermoid cyst, cystic change in solid neoplasms, and pseudomyxoma retroperitonei [13]. In our case, the histological examination showed the presence of a cystic lesion with colonic mucosal lining surrounded by a muscular layer, guiding the diagnosis of colonic duplication cyst. The appendix was benign and no mucin deposits were seen at laparoscopy, ruling out a secondary involvement from a primary appendiceal tumor.

Conclusion

Malignant transformation arising in enteric duplication cysts is extremely rare. We report a unique case of a low-grade mucinous neoplasm that originated in a colonic duplication cyst located at the retroperitoneum. In the patient, the histopathological findings and the medical history allowed confirmation of the diagnosis. It is recommended to perform a complete surgical removal and histological examination of duplication cysts, in order to avoid complications and exclude the presence of malignant component.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

Ethical Approval Ethical approval was obtained for publication of this case report.

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