



Case Report

Enterovesical fistula caused by ileal primary adenocarcinoma

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ABSTRACT

Introduction: Small bowel neoplasms are rare entities, with only 3.2% of gastrointestinal tumors localized in this segment. Adenocarcinoma is the second most common histologic type among small bowel neoplasms. The disease's symptoms are non-specific, with abdominal pain being the most common. Among the complications, fistulas are a far rare condition.

Case report: We present the case of an 86-year-old woman, hypertensive, diabetic, and former smoker, with lower abdominal pain, hematuria, fecaluria and fever associated with weight loss. Upon physical examination presented pale mucous membranes. Magnetic resonance imaging of the abdomen showed parietal thickening in the distal ileum segment with an anterior wall bladder fistula. Absence of metastases.

Subjected to transurethral endoscopic biopsy of the bladder lesion, which anatomopathological study was compatible with invasive mucinous adenocarcinoma. An enterectomy was performed with primary enteroanastomosis, associated with partial cystectomy by video-laparoscopy. Anatomopathological study of the surgical specimens concluded mucinous adenocarcinoma with signet ring cells located in the small bowel and bladder. Immunohistochemical exam has identified findings compatible with mucinous adenocarcinoma of origin in the small bowel. The patient evolved well, being discharged, and returned to postoperative follow-up without signs of relapse of the disease.

Conclusion: The relative inaccessibility of the small bowel and the malignant neoplasms' non-specific symptoms make an early diagnosis difficult. Discovery of the disease, often only at an advanced stage, results in complications and less effective therapy. The laparoscopic approach might be advantageous and effective in the treatment of advanced small bowel cancer with invasion of adjacent structures.

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Fístula enterovesical causada por adenocarcinoma ileal primário

R E S U M O

Palavras-chave:

Adenocarcinoma mucinoso
Fístula do sistema digestório
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Introdução: As neoplasias do intestino delgado são entidades raras com apenas 3,2% dos tumores gastrointestinais localizando-se neste segmento. O adenocarcinoma é o segundo tipo histológico mais comum entre as neoplasias do intestino delgado. Os sintomas da doença são inespecíficos, sendo a dor abdominal o mais comum, dificultando o diagnóstico precoce. Dentre as complicações, as fístulas são de ocorrência ainda mais rara.

Relato de caso: Mulher de 86 anos, hipertensa, diabética e ex-tabagista. Quadro de dor em abdome inferior, hematúria, fecalúria, febre eventual e perda ponderal. Ao exame físico pele e mucosas hipocoradas. Ressonância Magnética de abdome evidenciou espessamento parietal em segmento do íleo distal com fístula para a parede vesical anterior. Ausência de metástases. Submetida à biopsia endoscópica transuretral da lesão vesical, cujo estudo anatomopatológico evidenciou adenocarcinoma mucinoso invasivo. Realizada enterectomia com enteroanastomose primária, associada à cistectomia parcial por videolaparoscopia. Estudo anatomopatológico das peças cirúrgicas concluiu adenocarcinoma mucinoso com células em anel de sinete localizado em intestino delgado e bexiga. A Imuno-histoquímica identificou achados compatíveis com adenocarcinoma mucinoso de origem em intestino delgado. A paciente evoluiu bem no pós-operatório, recebendo alta e retornando para seguimento pós-operatório sem sinais de recidiva da doença.

Conclusão: A relativa inacessibilidade do intestino delgado e os sintomas inespecíficos das neoplasias malignas deste sítio dificulta o diagnóstico precoce. O reconhecimento da doença, muitas vezes somente em estágio avançado, resulta em complicações e sequelas com terapêutica menos eficaz. A abordagem laparoscópica pode ser vantajosa no tratamento do câncer avançado do intestino delgado com invasão de estruturas adjacentes.

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Introduction

Small bowel neoplasms are rare entities, representing about 3.2% of gastrointestinal tumors.¹ It is estimated 10,190 new cases of small bowel tumors and 1390 deaths resulting from this neoplasm in the United States for the year 2017.¹ It affects more frequently people between the 6th and 8th decade of life. Due to the low incidence of the disease, the diagnostic suspicion is often relegated to the background, and its diagnosis is almost always carried out during the investigation of other gastrointestinal pathologies or non-specific symptoms.²

Regarding the small bowel neoplasms histological types, an American study³ showed that in the period between 1973 and 2005 the carcinoid tumor exceeded adenocarcinoma. In 2005, the carcinoid tumor appeared in 44.3% of the cases, followed by adenocarcinoma (32.6%), lymphomas (approximately 15%) and stromal tumors (approximately 7%). The general incidence of small bowel tumors has practically duplicated in the last 40 years, with the development of radiographic and endoscopic diagnostic techniques⁴ as one of the possible reasons for this increase, especially the increased incidence rates of carcinoid tumors when compared to other histological types. However, when analyzing the frequencies per small bowel segment, adenocarcinoma remains the most frequent duodenal tumor while carcinoid tumors are the most common in the ileus.³

Adenocarcinoma is most often found in the duodenum (55–82% of cases), followed by jejunum (11–25%) and ileum (7–17%).⁴ Some hypotheses try to explain this different incidence among small bowel segments blaming the exposure time of the mucosa to carcinogens, dilution of carcinogens by enteric secretions, characteristics of intraluminal microbiology, presence of microsomal enzymes in enterocytes that would have a protective function to carcinogenesis, among other factors. However, there is still no clear explanation to justify this differentiated pattern of segmental neoplastic small bowel involvement adenocarcinoma.⁴ In patients with concomitant Crohn's disease and adenocarcinoma, there was a greater occurrence in the distal ileum.⁵

Among the risk factors for the appearance of primary small bowel adenocarcinoma, the main one is the presence of previous adenoma,⁶ although the presence of clinical conditions such as hereditary non-polytopic colorectal cancer, Peutz-Jeghers syndrome, Crohn's disease, familial adenomatous polyposis, Cowden's syndrome, celiac disease and juvenile polyposis^{2,5} also constitute as important risk factors. Excessive alcohol consumption might also be related to increased risk of developing this neoplasm.⁵

The symptomatology of primary small bowel adenocarcinoma is nonspecific, with abdominal pain being the most common symptom, followed by nausea, vomiting, fatigue, anemia, upper or lower gastrointestinal bleeding, jaundice and weight loss.⁷

The small bowel tumors diagnosis can be a real challenge because, in most part of the time, the disease is asymptomatic or behaves with nonspecific symptoms. Nowadays, with the advent of new diagnostic technologies, the detection of both the primary tumor and the secondary metastases has been optimized through the use of complementary tests such as computed tomography, magnetic resonance imaging, digestive endoscopy, endoscopic capsule, ultrasonography, electronic enteroscopy, barium contrast studies and double balloon enteroscopy. However, there is still no standardized diagnostic approach due to the rare occurrence of these neoplasms. It is relevant to emphasize the therapeutic role of endoscopic exams.

Despite of these different diagnostic techniques, the diagnosis is often made only intraoperatively, mainly in acute abdomen context,^{8,9} such as in subocclusive/occlusive conditions, intestinal perforation¹⁰ or digestive hemorrhage.

Laboratory tests generally reveal nonspecific results suggestive of intestinal bleeding leading to iron deficiency and consequent anemia. Carcinoembryonic Antigen (CEA) and Carbohydrate Antigen 19-9 (CA 19-9) serum levels may be elevated in 30–40% of patients with small bowel adenocarcinoma, but with limited sensitivity and specificity in diagnostic, but important for the monitoring of the disease.^{2,11}

Intestinal obstruction (more frequent in tumors of the distal ileum), digestive hemorrhage and intestinal perforation¹⁰ are among the mainly complications reported. Fistulas due to neoplasms of the small bowel are rare entities, with few reports in the literature, often related to the concomitant presence of Crohn's disease.¹²⁻¹⁶

The objective of this report is to present the clinical case of a female patient with primary ileal mucinous adenocarcinoma complicated with enterovesical fistula.

Case report

A 86-year old woman came to our unit with a 1-year history of lower abdominal pain, associated with hematuria, dysuria, fecaluria and sporadic fever, losing in the period a total amount of 12 kg. She was feeling a lower abdominal "heavy" sensation and a burning pain around the vaginal and anal areas. The patient had a history of diabetes and hypertension. Also referred herself as a former smoker. On physical examination, the patient presented mild paleness and pale mucous membranes.

Nuclear magnetic resonance showed parietal thickening in the distal ileum segment with a fistula to the anterior bladder wall. The differential diagnosis of tumoral lesions and inflammatory bowel disease was made. Colonoscopy showed no findings. Further investigations with cystoscopy revealed a vegetative lesion located on the left and superior wall of the bladder, measuring approximately 10 cm, although computed Tomography of thoracic and upper abdomen showed no evidence of metastatic foci.

The results of a transurethral bladder biopsy were compatible with a invasive mucinous adenocarcinoma, reaching submucosa, without vascular, lymphatic or perineural involvement. Based on these findings, the patient was subsequently submitted to a ileus loop segmental enterectomy

with primary end-to-end enteroanastomosis, associated with a wide margin partial cystectomy and primary rafia. Both procedures were performed by videolaparoscopy.

The biopsy samples obtained from the surgery were sent to a new study, which diagnosed mucinous adenocarcinoma with signet ring cells located in the small bowel and bladder, and a invasion of adjacent structures, without vascular, lymphatic and perineural involvement, but with free margins, pT4pNx staging. Immunohistochemical exams identified KS20.8, DAK-CDX2, 1D2C3, MIB1 (60%) clones, which is compatible with mucinous adenocarcinoma with signet ring cells of intestinal origin.

The patient evolved well, being discharged on the 7th postoperative day (POD). She was followed up in the Onofre Lopes University Hospital's coloproctology and urology departments for 6 months, without signs of relapse of the disease.

Discussion

The small bowel adenocarcinoma imposes a real diagnostic challenge on the physician, due to the lack of early stage symptoms, often requiring endoscopic or radiological analysis, which limits its adequate diagnostic and therapeutic approach to centers with better structure and qualified professionals. This harsh reality ends up delaying the early diagnosis of the tumor and pushes worse prognostic perspective on patients.

Despite the insufficient level of evidence in the treatment of small bowel tumors in consequence to the scarcity of randomized studies¹⁷ and the rarity of the disease, the cornerstone of ileal adenocarcinoma therapy nowadays is its surgical resection with adjuvant chemotherapy. Although few studies have evaluated the role of chemotherapy in the treatment of this neoplasm, adjuvant chemotherapy might compensate for poor prognostic factors, without, however, a statistically significant benefit. There is a need for high-level evidence studies to better clarify the role of adjuvant chemotherapy in the context of small bowel adenocarcinoma.¹⁸

For jejunal and ileal tumors, curative surgery (R0) consists of a wide resection with free cancer margins, associated with regional lymphadenectomy, similar to colonic adenocarcinomas.⁶

Video-laparoscopy and its application in cancer surgery has been gaining greater acceptance among surgeons and consolidating as an effective and safe approach for the surgical treatment of malignant tumors of the small intestine. Among the advantages of laparoscopy are: lower intraoperative bleeding rate, postoperative morbidity and paralytic ileus complication, less need for analgesics, lower immunological and metabolic stress, shorter hospital stay, early return to daily activities and a better postoperative aesthetic aspect.⁶ In our case, we proceeded with the extensive videolaparoscopic enterectomy of the ileal tumor with posterior partial cystectomy, with free broad cancer margins. The patient had a good evolution in the postoperative period, being discharged from hospital in the 7th POD.

As with most upper gastrointestinal neoplasms, the prognosis of small bowel adenocarcinomas is usually bad, mainly due to diagnostic delay, with a median survival of 9–11

months⁷ in metastatic disease. The presence of compromised surgical margins, vascular or perineural invasion, and lymph node involvement are predictors of poor prognosis,¹⁹ which was not evidenced in the patient in this case.

This report becomes relevant to the literature because of the low incidence of small bowel neoplasms complicated with enterovesical fistula, with special emphasis on the uniqueness of the case when reporting the presence of adenocarcinoma as the histological type in the distal ileum

In addition, the case also illustrates the feasibility of a satisfactory outcome in the immediate and late postoperative period through laparoscopic surgical tumor resection in small-bowel adenocarcinomas with invasion of adjacent organs. High-level evidence is needed to define the true role of video-laparoscopy in resecting malignant small-bowel tumors.

Conflicts of interest

The authors declare no conflicts of interest.

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