TUMOR MUCINOSO DE APÊNDICE CURSANDO COM PSEUDOMIXOMA PERITONEAL: UM RELATO DE CASO

Appendix mucinous tumor cursing with peritoneal pseudomixoma: a case report

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RESUMO

As neoplasias de apêndice cecal tem rara incidência dentro do espectro de neoplasias intestinais, sendo um importante diagnóstico diferencial aos pacientes de 5ª a 6ª década de vida, principalmente, que apresentem quadro clínico sugestivo de apendicite. Os achados radiológicos de massa em região periapendicular, implantes metastáticos e ascite podem sugerir o diagnóstico, mas este só é confirmado após abordagem cirúrgica e realização de biópsia. Este relato de caso traz um caso de tumor de apêndice mucinoso de baixo grau com pseudomixoma peritoneal. Tem sua importância devido a sua baixa incidência e, portanto, merece destaque ao retratar os principais aspectos encontrados no ato cirúrgico que serão fundamentais para o diagnóstico macroscópico intraoperatório, escolha da técnica de recessão e tratamento.

Palavras-Chave: Apêndice Cecal; Pseudomixoma Peritoneal; Tumor Mucinoso.

ABSTRACT

As neoplasias de apêndice cecal tem rara incidência dentro do espectro de neoplasias intestinais, sendo um importante diagnóstico diferencial aos pacientes de 5ª a 6ª década de vida, principalmente, que apresentem quadro clínico sugestivo de apendicite. Os achados radiológicos de massa em região periapendicular, implantes metastáticos e ascite podem sugerir o diagnóstico, mas este só é confirmado após abordagem cirúrgica e realização de biópsia. Este relato de caso traz um caso de tumor de apêndice mucinoso de baixo grau com pseudomixoma peritoneal. Tem sua importância devido a sua baixa incidência e, portanto, merece destaque ao retratar os principais aspectos encontrados no ato cirúrgico que serão fundamentais para o diagnóstico macroscópico intraoperatório, escolha da técnica de recessão e tratamento.

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RELATO DE CASO

INTRODUCTION

The cecal or vermiform appendix consists of a tubular structure of approximately 2-20 cm, which originates in the posteromedial portion of the caecum, with possible immune function[1]. The group of vermiform appendage tumors is heterogeneous and of variable prognosis, with low incidence in relation to intestinal neoplasms, being most often an anatomopathological finding in patients undergoing surgery for acute appendicitis[2].

Primary malignancies of malignant character are rare and their histological classification is controversial and can be divided into epithelial tumors and neuroendocrine tumors. Tumors of epithelial origin may be benign or malignant (adenomas and adenocarcinomas, respectively), and about 0.1% of appendix removal surgeries performed have adenocarcinoma[3]. Among epithelial neoplasms, low grade mucinous and mucinous adenomas are the most frequent. Carcinoids, on the other hand, are tumors derived from neuroendocrine cells widely found in the appendix, the most common site in the gastrointestinal region[4].

CASE REPORT

R.N.C., male patient, 47 years old, single, hearing impaired, treated at the Emergency Room of the Palmas General Public Hospital (HGPP) with diffuse abdominal pain, beginning 4 months ago with worsening 4 days ago, associated with nausea and vomiting. On physical examination, the patient had a flat abdomen, with normotympanic airborne noises, deep painful palpation in the epigastric region, right hypochondrium, and right iliac fossa, with no signs of peritonitis.

In the laboratory tests the blood count was within normal range and the other tests, such as liver injury and function, pancreatic enzymes, renal function and electrolytes were also within the reference values. Among the altered results, there was an increase in C-reactive protein (18.69 mg/L) and serum glycemia (141.0 mg/dL). No tumor markers were requested.

Total abdomen ultrasound showed signs of mild deposition liver disease of probable fatty etiology, vesicular lithiasis, bilateral calculus and/or renal calcifications, moderate ascites and cystic formations adjacent to the bladder. Upper abdominal computed tomography showed calculous cholecystopathy, as well as a moderate amount of free intra-abdominal fluid. Pelvic computed tomography showed cystic-shaped oval formations with calcified walls adjacent to the caecum measuring 2.7 cm to 4.0 cm.

During surgery, there was mucinous ascites, 5 cm appendage tumor (with ulceration) and diffuse tumor implants throughout the intestine and peritoneum. Due to the macroscopic findings in the surgery, suggestive of appendix neoplasia, a right hemicolectomy was performed followed by ileotransversoanastomosis. Histopathology concluded low-grade mucinous neoplasia, reaching subserosa and mesoappendix, with 9 intraperitoneal metastatic isolates, staging as T3, N0, M1, G3 (IVB) 10. No angiolymphatic or perineural invasion was identified.

DISCUSSION

Low-grade mucinous neoplasms have a higher incidence in the population aged 50 to 70 years, with women being slightly more affected than men. It is clinically similar to appendicitis, which is its main differential diagnosis, found in 50 to 70% of cases[1,3,4]. When low grade mucinous neoplasms spread in the peritoneal cavity and excessive mucus accumulation occurs in the intraperitoneal region, we have a peritoneal pseudomyxoma[4]. In these cases, there is mucinous ascites and tumor redistribution, which may involve peritoneum, omentum, and bowel loops. Symptoms such as diffuse abdominal pain, palpable mass, gastrointestinal or genitourinary obstruction, intestinal bleeding, and distension secondary to peritoneal pseudomyxoma may also occur less frequently. Appendicular torsion is a rare manifestation and presents symptoms similar to those of appendicitis. In some cases, they are asymptomatic, and the diagnosis is made through findings during surgeries with appendectomy[5].

Features on mucinous adenocarcinoma imaging exams include thickening of the appendicular wall or irregular nodule and periapendicular soft tissue deposits. The appendix is usually distended thanks to the presence of mucin, whereas in some cases it is also possible to visualize intraperitoneal metastasis. Features of appendix pseudomyxoma include mucinous ascites, peritoneal implants, and gastrointestinal, genitourinary, or ovarian metastases. There is also the presence of free or intra-abdominal liquid collections[6,7,8,9].

For surgical planning, it is important to perform tumor staging (Tables 1 and 2). Peritoneum-confined disease, including peritoneal pseudomyxoma with disease M1a, is considered a regional metastatic site[10].

Table 1: TNM staging of mucinous adenocarcinoma of the appendix, adapted from *Kelly*[10].

	Definition
T stage (T)	
TX	Primary tumor cannot be assessed
T0	No evidence of primary tumor
Tis	Carcinoma *in situ*
T1	Invades submucosa
T2	Invades muscularis propria
T3	Invades through muscularis propria until subserosa or mesoappendix
T4	Invades visceral peritoneum, including right lower quadrant parietal peritoneum and / or directly invasion of other organs or structures
T4a	Invades visceral peritoneum including right lower quadrant parietal peritoneum tumor
T4b	Directly invades other organs or structure
Nodes (N)	
NX	Nodes cannot be assessed

N0	No regional lymph node involvement
N1	Metastasis to 1–3 regional nodes
N2	Metastasis to ≥ 4 regional nodes
Metastasis (M)	
M0	No distant metastasis
M1	Distant metastasis
M1a	Intraperitoneal metastasis in the right lower quadrant including intraperitoneal pseudomyxoma
M1b	Extraperitoneal distant metastasis
Histological grade (G)	
GX	Cannot be determined
G1	Well-differentiated
G2	Moderately differentiated
G3	Poorly differentiated
G4	Undifferentiated

Table 2: Staging of Appendix Mucinous Neoplasia, adapted from *Kelly*[10].

Stage	
0	Tis, N0, M0
I	T1/2, N0, M0
IIA	T3, N0, M0
IIB	T4a, N0, M0
IIC	T4b, N0, M0
IIIA	T1/2, N1, M0
IIIB	T3/4, N1, M0
IIIC	Any T, N2, M0
IVA	Any T, N0, M1a, G1
IVB	Any T, N0, M1a, G2/3 Any T, N1/2, M1a, any G
IVC	Any T, any N, M1b, any G

The optimal treatment for epithelial mucinoid neoplasms depends on the histological characteristics of the primary tumor and peritoneal disease, as well as the TNM stage10. The treatment for appendage-confined cancers is appendectomy. Cases of low-grade mucinous neoplasia with high grade lymph node involvement are treated with appendectomy, right hemicolectomy and lymph node dissection.

Patients with peritoneal dissemination are submitted to laparoscopy and based on the extent and histological grade of the peritoneal tumor, cytoreductive surgery (CRS) and hyperthermic intraperitoneal chemotherapy (HIPEC)[10,11]. Extraperitoneal metastatic disease to the liver, retroperitoneal lymph nodes or other sites is considered by many surgeons to be a contraindication to curative-intentional surgery[10]. Patients ineligible for complete CRS should undergo systemic chemotherapy with regimens used for metastatic colorectal cancer[10].

In patients with mucinous neoplasms and peritoneal dissemination of the disease, a combined approach consisting of laparoscopy, CSR and HIPEC is considered the standard of treatment[10,12,13]. The aim of CRS is the complete resection of all visible tumors, defined as removal of all tumor deposits larger than 2.5 mm[14].

CONCLUSION

The diagnosis and management of appendix tumor goes through important stages: recognition of its clinical manifestations, adequate request and interpretation of complementary exams and, finally, therapeutic planning.

The mucinous tumor of the appendix has a very variable and nonspecific clinic, and confusion with the clinical picture of an acute appendicitis, such as mild colic associated with anorexia, nausea and vomiting with usual duration of 4 to 6 hours, is common with pain initially in the periumbilical or epigastric region, with irradiation to the right lower quadrant as inflammation spreads[1].

Thus, it is important that it be thought of as a diagnostic hypothesis when there are signs and/or symptoms related to the more chronic gastrointestinal tract, as presented here, since it is a pathology with evident prognostic benefit when it has a precocious diagnosis.

As resources for diagnostic investigation, scientific evidence indicates benefit in the research of biochemical markers, such as CEA, CA 19-9 and CA-125. However, in the case presented, this research was not performed due to unavailability of markers during the present case.

In the choice of imaging methods, ultrasound has high sensitivity and specificity, making it useful for appendix evaluation and may also show secondary signs of malignancy, such as appendicular mucocele, regional adenopathy, ascites and abdominal mass[15,16]. The latter two were described in the examination of the case reported here, and are therefore useful in constructing clinical reasoning during case management.

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