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CASE REPORT

Appendicular adenocarcinoma: palpable mass in the right iliac fossa

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ABSTRACT
Primary neoplasia of the appendix is a rare entity that is difficult to diagnose preoperatively. The adenocarcinoma represents only 10% and occurs mainly in patients between 62 and 65 years of age. We present the case of a 73-year-old female patient with pain in the right iliac fossa of 5 months' duration associated with a palpable mass. Computed tomography showed irregular circumferential parietal thickening of the cecum and the tumor markers were elevated. A laparoscopic right colectomy was performed with oncological criteria due to a presumptive diagnosis of appendiceal neoplasia. Histopathology reported adenocarcinoma of the appendix, intestinal type, with cecal and ileal invasion and 2/14 positive adenopathies, T4N1Mx. The initial clinical course was good. The patient is currently undergoing adjuvant treatment.

Keywords: *Appendicular Adenocarcinoma; Palpable Mass in Right Iliac Fossa; Right Colectomy*

INTRODUCTION

Primary neoplasia of the appendix is a rare entity that is difficult to diagnose preoperatively. It corresponds to less than 1% of all appendectomies.[1] The most common histology is neuroendocrine neoplasia, which accounts for approximately 50% of all malignant tumors of the appendix, while adenocarcinoma only represents around 10%.[2] Mucinous adenocarcinoma is the most common type, while intestinal or colonic variants are the least frequent. The mean age of presentation of non-carcinoid appendiceal neoplasms ranges between 59 and 63 years. Colonic-type adenocarcinoma of the appendix occurs at a mean age of 62 to 65 years, with a slight predominance in males. These tumors are usually a secondary finding after appendectomy for acute appendicitis..

CLINICAL CASE

A 73-year-old woman consulted for abdominal pain predominantly in the right iliac fossa of 5 months' duration, without concomitant symptoms. On physical examination, she presented a 6 x 6 cm palpable mass in the right iliac fossa that was frankly painful and partially mobile. She has not had significant weight loss in the last 3 months.

Colonoscopy showed a bulge covered by normal mucosa in topography of the ileocecal valve, with a "cushion sign". It was interpreted as a submucosal lesion of the ileocecal valve (Fig. 1).

Computed tomography of the abdomen and pelvis showed circumferential and irregular parietal thickening of the cecum. It was associated with subtle changes in the density of the adjacent fatty planes, engorgement of the regional blood vessels, and two enlarged lymph nodes with a hypodense center attributable to necrosis in the ileocolic territory, the largest measuring 13 mm (Figs. 2 and 3).

Carcinoembryonic antigen: 7.83 ng/ml (NV up to 5.0). Ca 19-9: 80 U/ml (NV up to 37).

With a high presumption of a neoplastic process, it was decided to perform an exploratory laparoscopy in which a hard mass originating in the cecum was observed in the right iliac fossa, adhered to the parietal plane, without being able to identify the cecal appendix. Laparoscopic right colectomy was performed with oncological criteria (Fig. 4).

Histopathology revealed an exophytic mass at the base of the appendix occupying 50% of the lumen (Fig. 5). The diagnosis was semi-differentiated adenocarcinoma of the appendix, colonic type, with colonic and ileal wall infiltration and involvement of 2 out of 14 lymph nodes found: pT4N1Mx.

DISCUSSION

Malignant neoplasms of the appendix are rare and until 1985 the world literature reports only 300 cases of non-carcinoid carcinomas (adenocarcinomas).[4] Appendicular carcinomas are reported with an age-adjusted incidence of 0.12 cases per 1,000,000 per year.[3] In 1350 appendectomies in our series during the 2010-2020 period, only 1 case of appendiceal adenocarcinoma occurred. They represent only 0.4% of all gastrointestinal tumors.

The authors declare no conflicts of interest

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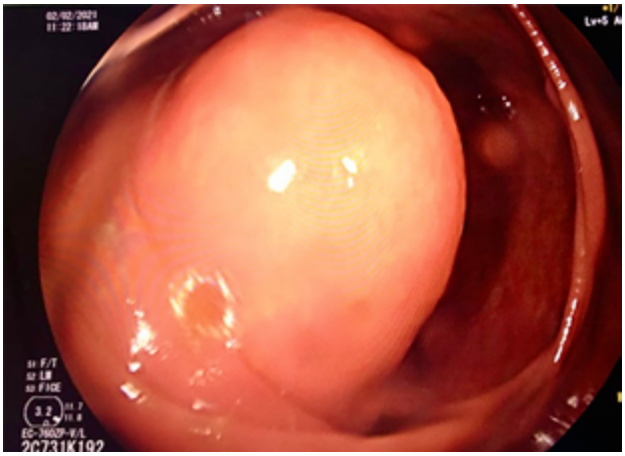

Figure 1: Endoscopic image showing a bulge covered with normal mucosa ("cushion" sign), which is interpreted as a submucosal lesion of the ileocecal valve.

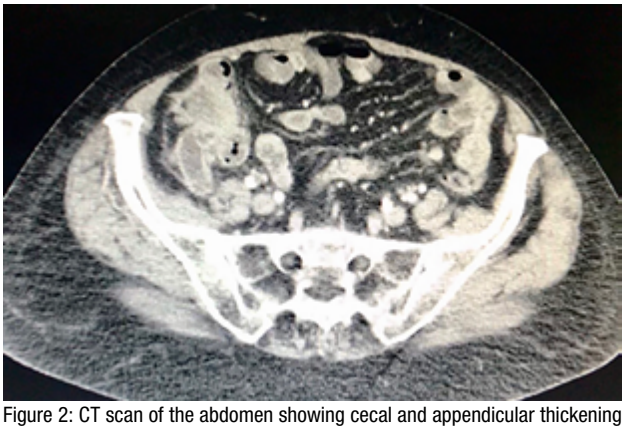

Figure 2: CT scan of the abdomen showing cecal and appendicular thickening with local lymph node involvement.

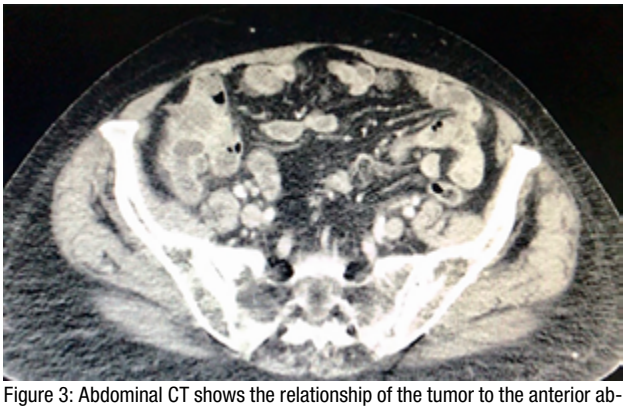

Figure 3: Abdominal CT shows the relationship of the tumor to the anterior abdominal wall.

Most present with pain in the right iliac fossa simulating acute appendicitis and are diagnosed incidentally in the histological evaluation of the surgical specimen (50% of cases). Other forms of presentation are palpable mass (13%), ascites, nonspecific gastrointestinal or genitourinary discomfort (5%).[3] In a study by Nitecki et al. at the Mayo Clinic,[4] none of the 94 patients could be diagnosed preoperatively. By the time the patient becomes symptomatic, the disease process is usually advanced.

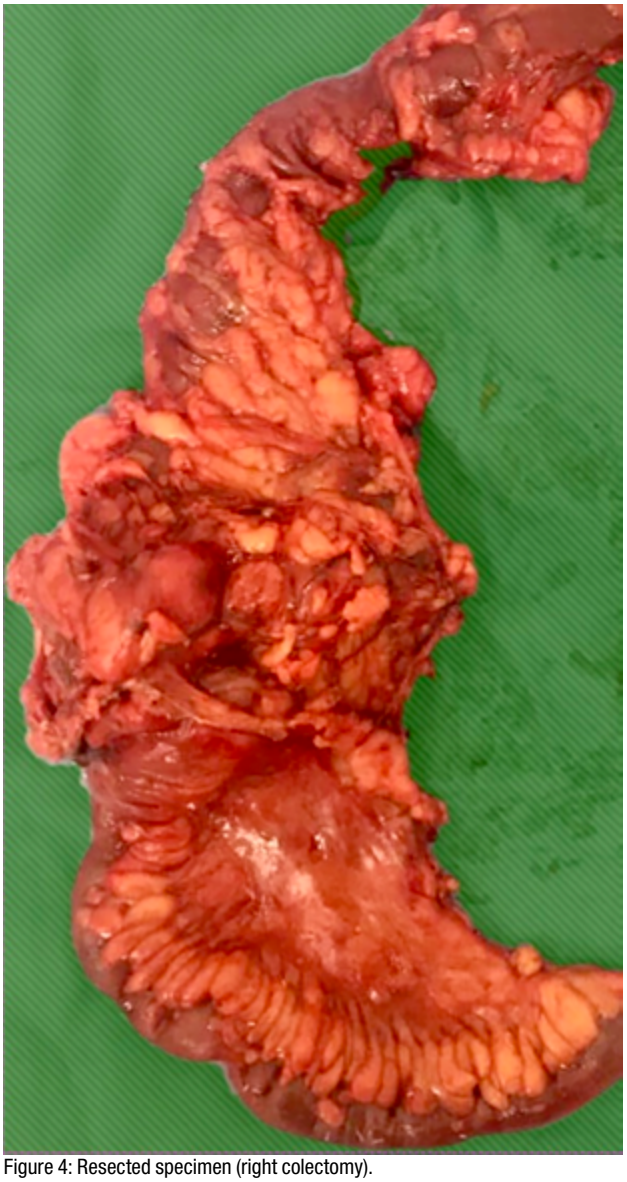

Figure 4: Resected specimen (right colectomy).

The differential diagnosis of primary appendix cancer includes: adenocarcinoma (colonic or mucinous), neuroendocrine carcinoma, and mixed tumors. Adenocarcinoma differs from cystadenocarcinoma in that it invades the colon wall and lacks mucin. In addition, it is mostly poorly differentiated, so it has a worse prognosis.[5]

The survival of appendiceal carcinomas is related to the type and size of the tumor. In this sense, the survival of non-carcinoid tumors is significantly worse than that of carcinoid tumors.[3,5,6]

Lymphatic metastases typically occur in 30% of cases and warrant adjuvant systemic chemotherapy. The frequency of distant metastases is not exactly known given the rarity of the disease, although it is estimated at 23-37%. The ovaries are the most frequent site affected and more exceptionally the liver and lung. In those patients

with T1 tumors with favorable characteristics (well-differentiated and without angiolymphatic invasion), appendectomy may be considered. On the other hand, unfavorable T1 tumors (angiolymphatic invasion, high grade and/or positive margins) or those with higher staging (by CT of the chest, abdomen and pelvis) should be treated by right colectomy.[7] Tumor invasion is the most important determinant of treatment for adenocarcinoma of the appendix. Although it is debated whether a simple appendectomy or a right colectomy should be performed, the latter procedure prevails.[6] Thus, in simple appendectomies, subsequent colonic resection should be considered after histopathological confirmation, while in the presence of a presumptive neoplastic process primary right hemicolectomy, with removal of 12 or more lymph nodes, is recommended for accurate staging.[5-7]

CONCLUSION

A rare case of colonic-type appendiceal adenocarcinoma is presented. On this occasion, the diagnosis was not made after the appendectomy, but was rather a preoperative suspicion because it presented as a palpable mass with imaging and humoral evidence. This allowed resection with oncological criteria by videolaparoscopy and subsequent adjuvant treatment. This sequence makes the case even less frequent.

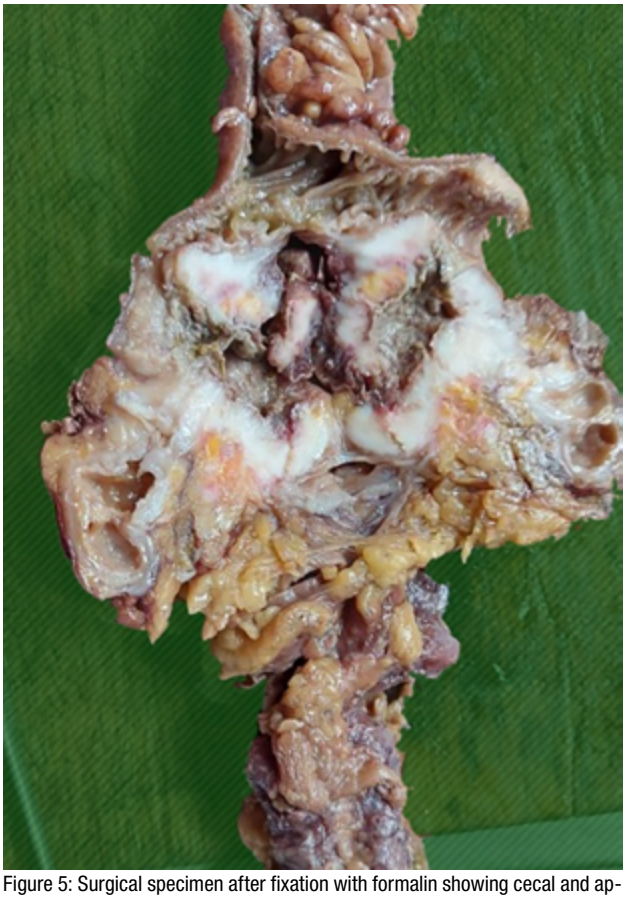

Figure 5: Surgical specimen after fixation with formalin showing cecal and appendicular parietal infiltration.

REFERENCES

1. Ozakyol AH, Sariçam T, Kabukçuoğlu S, Cağa T, Erenoğlu E. Primary appendiceal adenocarcinoma. Am J Clin Oncol 1999; 22:458-59.
2. Butte B. Tumores del apéndice cecal. Análisis anatomoclínico y evaluación de la sobrevida alejada. Rev Chil Cir 2007; 59:217-22.
3. McGory ML, Maggard MA., Kang H, O'Connell JB, Ko CY. Malignancies of the appendix: Beyond case series reports. Dis Colon Rectum 2005; 48, 2264–71.
4. Nitecki SS, Wolff BG, Schlinkert R, Sarr MG. The natural history of surgically treated primary adenocarcinoma of the appendix. Ann Surg1994; 219:51-7.
5. Benedix F, Reimer A, Gastinger I, Mroczkowski P, Lippert H, Kube R. Primary appendiceal carcinoma--epidemiology, surgery and survival: results of a German multi-center study. Eur J Surg Oncol 2010; 36:763-71.
6. Deans GT, Spence RAJ. Neoplastic lesions of the appendix. Br J Surg 1995; 82: 299–306.
7. Kelly KJ. Management of appendix cancer. Clin Colon Rectal Surg 2015;28:247–55.