

Carcinoid tumor of the appendix with clinical picture of acute appendicitis, urgently operated on by laparoscopic appendectomy. Presentation of clinical case

Tumor carcinoide de apéndice con cuadro clínico de apendicitis aguda, intervenido de urgencia por apendicetomía por laparoscopia. Presentación de caso clínico

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Abstract

Cecal appendix tumors are extremely rare (less than 0.5%), with carcinoid tumors being described as the most common type of appendix tumor. A case of a 32-year-old male patient is reported, with no significant pathological history, with acute abdomen data. Laparoscopic appendectomy was decided, presenting findings of an appendicular inflammatory process, resulting in a neuroendocrine tumor histopathology report. Despite the fact that appendix neoplasias are rare, appendicitis is a common presentation, hence the importance of conducting histopathology studies on every piece of cecal appendix obtained.

Keywords: Acute abdomen. Acute appendicitis. Carcinoid tumor. Laparoscopy.

Resumen

Los tumores de apéndice cecal son extremadamente raros (menos del 0.5%), y los tumores carcinoides se describen como el tipo de tumor apendicular más frecuente. Se reporta el caso de un paciente de 32 años, sin antecedentes patológicos de importancia, con datos de abdomen agudo. Se decidió apendicetomía laparoscópica, presentando hallazgos de proceso inflamatorio apendicular, con reporte de histopatología de tumor neuroendocrino. Pese a que las neoplasias de apéndice son raras, la apendicitis es una presentación común, y de ahí la importancia de realizar estudios histopatológicos de toda pieza de apéndice cecal obtenida.

Palabras clave: Abdomen agudo. Apendicitis aguda. Tumor carcinoide. Laparoscopia.

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Introduction

Cecal appendix tumors are extremely rare and less than 0.5% of the appendectomy specimens correspond to appendix cancer. This type of tumors is rarely diagnosed before surgery, and oftentimes they are found intraoperatively or by pathologic analysis¹. Statistically, most studies mention that carcinoid tumors were the most frequently found appendiceal tumor, while 10-20% correspond to adenocarcinoma, adenocarcinoid or lymphosarcoma, among others².

Carcinoid tumor of the appendix was described for the first time in 1882. Its incidence ranges from 0.08% to 0.1% of all appendectomies and from 4% to 6% of malignant appendiceal neoplasms. Epidemiologically, the mean age of onset is around 50 years of age, with male predominance³. Appendicitis is a common occurrence in men and women, but peritoneal dissemination of the appendiceal mucinous neoplasia is a significant initial manifestation⁴. However, right hemicolectomy is the treatment of choice in most cases. The prognosis of appendiceal neoplasia is determined by the degree of extension in the appendiceal wall, the presence or absence of metastatic nodes, and the presence of distant metastases, especially hepatic ones⁵.

Clinical case

32-year-old male patient, without significant pathological history, with ailment onset 32 hours before admission, with abdominal cramp-like pain in the umbilical and periumbilical region, with a 10 out of 10 pain scale. He visits a medical office, he is diagnosed with urinary tract infection, and is prescribed a medical treatment based on sulfonamide and non-steroidal anti-inflammatory agents. However, his progress is not favorable, which is the reason why he visits the medical unit emergency department at the Regional Hospital of Tlalnepantla.

His vital signs on admission are the following: heart rate 72 b.p.m., respiratory rate 16 r.p.m., temperature 37.5 °C, and blood pressure 103/70 mmHg.

Awake, conscious, oriented, with discrete dehydration of mucous membranes, adequate coloration of integuments, normocephalic, neck without alterations, precordium with rhythmic heart sounds of good intensity without alterations, well-ventilated lung fields, without aggregate phenomena, flat abdomen, without peristalsis, soft and depressible, painful on palpation in the right iliac fossa, with voluntary muscle resistance, positive McBurney's point, positive psoas, positive



Figure 1. Standing abdominal radiograph showing multiple air-fluid levels and bilateral psoas effacement, as well as preperitoneal fat.

obturator and Rovsing sign, genitalia according to age and sex, eutrophic extremities, osteotendinous reflexes present, immediate capillary filling and distal pulse present.

Complete blood count: leukocytes 17.74; neutrophils 82.6%; hemoglobin 17.7 g/dl; hematocrit 53.4; platelets 287,000; glucose 106.7 mg/dl; prothrombin time 13.4 s; INR 1.24; thromboplastin time 29.5. General urine analysis: pH 7; sediment with leukocytes 4-6 per field; few epithelial cells and scarce bacteria.

Standing abdominal radiograph shows multiple air-fluid levels and bilateral psoas effacement, as well as preperitoneal fat (Fig. 1).

His hospitalization in the emergency department in charge of general surgery is decided, and his management with analgesic, third generation cephalosporin, and intravenous solutions is initiated. He is prepared for surgery and goes to the operating room.

Surgery performed: laparoscopic appendectomy. Hasson technique approach through a 1 cm umbilical incision; then, a 12 mm umbilical trocar is placed and capnoperitoneum is initiated up to 12 mmHg. A 30° lens is introduced and a diagnostic laparoscopy is performed. Then, 5 mm trocars are placed in the suprapubic region and left flank.

Findings: lax omental adhesions to the wall. Purulent free fluid of approximately 20 ml. Vermiform appendix of 10 cm, hyperemic, edematous throughout, with fibrinopurulent plaques. Appendectomy is performed, and the stump is managed with Halsted technique. Without complications, minimal bleeding (Figs. 2 and 3).

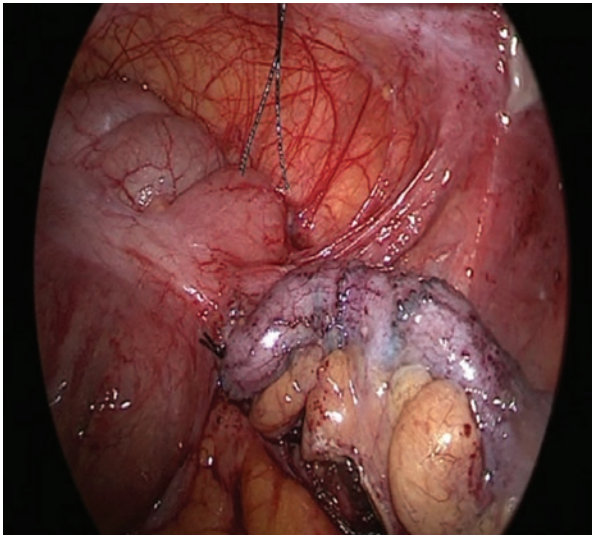


Figure 2. Dissection and ligation of the appendix base.

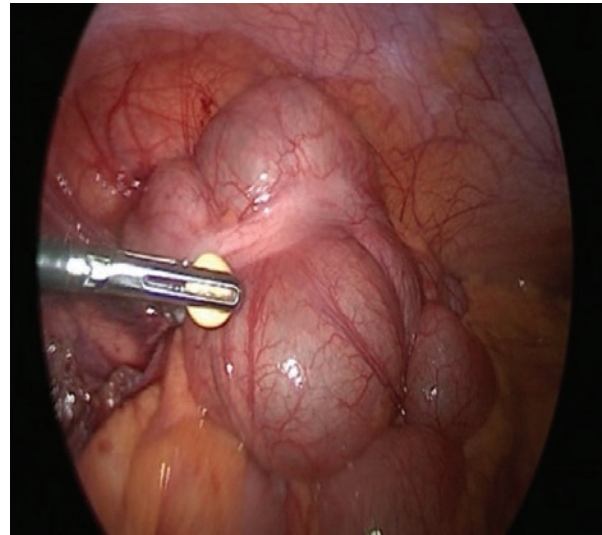


Figure 4. Neuroendocrine tumor (appendiceal carcinoid tumor).

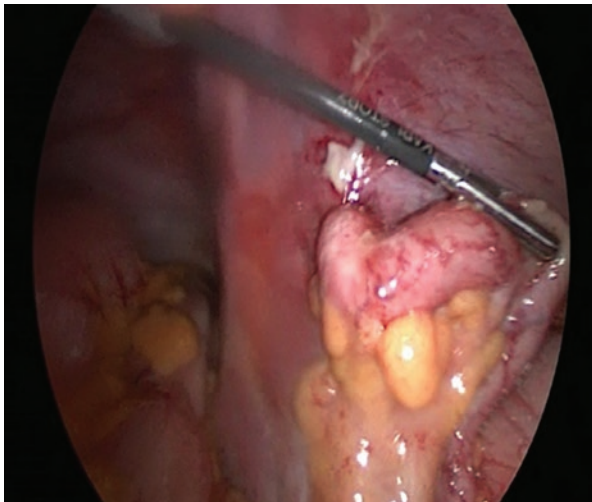


Figure 3. Laparoscopic appendectomy with Halstead technique stump management.

After the surgical event without complications, the patient is asymptomatic, tolerating oral route at 12 hours, channeling gases and evacuating at 18 hours. His discharge with outpatient follow-up is decided.

The pathologic specimen is sent to ordinary analysis, receiving an urgent result with the following tissue description: intestinal mucous membrane with preserved intestinal epithelium, lamina propria with hyperplasia, lymphoid tissue with active lymphoid follicles, congestive serosa. The middle third shows preserved colonic epithelium and crypts with mucus production, thickened serosa with leukostasis and neutrophilic aggregates and

congestive mesoappendix. Tip sections show neoplasia arranged in nests of organoid appearance, medium-sized cells, with scanty and moderate cytoplasm, and “salt and pepper” nuclei. The tumor extension is of up to 5 mm in its major axis. It runs through the submucosa and the muscular layers individually, which in serial sections does not exceed the outer muscular layer, serosa or mesoappendix. Neuroendocrine tumor (appendiceal carcinoid tumor) (Fig. 4).

Discussion

Carcinoid tumor is the most frequent malignant histological type in the appendiceal pathology, with a prevalence of about 0.32% among digestive tumors. Since only a minimal percentage of the population undergoes appendectomy, it can be assumed that most patients with appendiceal carcinoid tumors do not show any clinical manifestations and do not require treatment, without that affecting their life expectancy, in spite of the abdominal tumor⁵.

Approximately 10% are placed in the cecal appendix distal third, where they have low possibilities of causing obstruction. If located at the cecal appendix base, obstruction may occur, causing appendicitis. Therefore, most patients remain asymptomatic. Symptoms are more probable in patients with large tumors and in rare cases with metastases beyond the regional lymph nodes⁵.

The clinical behavior and progress depend, mainly, on the tumor size. Tumors smaller than 2 cm (found

in approximately 95% of patients) have low probability of metastases⁵.

For 1-2 cm appendiceal carcinoid tumors, or even in higher grade tumors, formal resection of the right colon does not seem to improve survival rate. However, follow-up of these patients is suggested if poor prognosis factors, such as nodal metastasis, lymphovascular invasion, mesoappendiceal invasion or mixed pathology, are identified⁶.

Those tumors larger than 2 cm produce distant metastases in up to a third part of patients in the moment of diagnosis or during surgical intervention, which mainly affect the area nodes. These tumors can also produce invasion of the mesoappendix. In these cases, it becomes necessary to perform a hemicolectomy, resecting the cecal appendix lymph nodes and any residual disease that could remain in the cecal appendix base or in the mesoappendix. Liver metastases are rarely produced, but right hemicolectomy must be completed as tumor treatment in those cases presenting them⁶.

Although the postoperative surveillance strategies and frequency vary, Murray et al.⁷ showed that there was no tumoral recurrence or disease specific mortality in the follow-up period, and conclude that post-resection surveillance may not be necessary for ≤ 1 cm tumors with R0 resection.

In most cases, like in ours, tumors are effectively treated with appendectomy, and no recurrence and no disease-specific deaths have been described in patients with a median follow-up of 5 years and even 25 years⁷.

Conclusions

Cecal appendix tumors are extremely rare and less than 0.5% of appendectomy specimens are diagnosed as appendiceal cancer. Even though appendiceal neoplasms are rare, appendicitis is common in men and women, hence the importance of conducting histopathological analysis of every cecal appendiceal specimen obtained. Right hemicolectomy is the treatment of choice in most cases.

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Conflicts of interest

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Ethical disclosures

Protection of people and animals. The authors declare that no experiments have been made on human beings or animals for this research.

Data confidentiality. The authors declare that they have followed the protocols of their work center regarding the publication of patient data.

Right to privacy and informed consent. The authors have received an informed consent from the patients and/or subjects referred to in this article. This document is in possession of the corresponding author.

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