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Epithelial Neoplasms of the Cecal Appendix

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Abstract: Cecal appendix carcinoma is characterized as an extremely rare condition and is found in only 1% of appendectomies and accounts for only 0.5% of neoplasms of the intestinal tract. The aim of the study is to discuss Appendix Carcinomatosis by presenting a case of Cecal Appendix Epithelial Neoplasm seen during the Surgery Residence.

Keywords: Appendix Vermiformis, Acute Abdomen, Appendix Neoplasm.

Introduction

Cecal appendix carcinoma is characterized as an extremely rare condition. It is found in only 1% of appendectomies and accounts for only 0.5% of neoplasms of the intestinal tract[1,2]. However, this number is underestimated since its incidence depends directly on the rates of appendectomies performed and the referral of the surgical specimen to the anatomopathological examination.

Regarding the symptomatology, the clinic, when present, is inseparable from the acute appendicitis, causing the surgical intervention to have its genesis in the suspicion of acute inflammatory abdomen. However, the complementary exams also show little specificity for the diagnosis of this neoplasm[1,3,4].

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Consequently, the assertive clinical diagnosis is greatly hampered, and it is postponed intraoperatively in the macroscopic analysis of the local or postoperative alterations at the time of the histological study of the part.

Based on the histological characteristics, the World Health Organization[5] sought to classify appendix neoplasms into nine classes through the "Protocol for examining specimens of patients with carcinoma of the appendix". Despite this initiative, there is still no consensus and homogeneity of nomenclature among the specialists, knowing that they are predominantly represented by epithelial tumors and neuroendocrine tumors[6,7].

As a result of the aforementioned scenarios, surgical services end up performing surgeries too often without any planning or adequacy to the tumor typology, location, degree and evolution, negatively impacting the therapeutics used, as well as the clinical condition and the prognosis of the patients. However, there is still an incipience of publications aimed at the study of this pathology - which contributes even more to the early diagnosis and consequent effectiveness of the treatment[8].

In view of the above, the present study aims to discuss Appendix Carcinomatosis through the presentation of a case of Epithelial Neoplasms of the Cecal Appendix seen during Surgery Residency.

Case Presentation

A 71-year-old male patient, looking healthy, sought emergency care on January 29, 2018 because of severe abdominal pain, nausea and vomiting. On physical examination, pain was reported on decompression of the right iliac fossa, with signs of peritoneal irritation. The complementary exams indicated a slight leukocytosis, with a predominance of the segmented ones. The Abdominal ultrasound revealed signs compatible with the inflammatory disease of the appendix, with consequent rupture of the same.

In view of the aforementioned findings, a laparoscopic appendectomy was performed, which evidenced an important inflammatory process in the topography of the cecum, involving a cecal appendix, which was destroyed. After excision, the surgical specimen was sent for analysis.

In the postoperative period, he developed an abscess in the right iliac fossa, undergoing surgical drainage and laminar drainage on February 2, 2018. The patient left hospital, but returned to the hospital on 03/03/2018, with abdominal distension, inappetence and excretion of fecaloid secretion through the drainage orifice.

At the time, the histopathological report of the surgical specimen removed previously suggested the diagnosis of a neoplasia with little differentiation, invasive, with multinucleated giant cells, with surgical margins compromised by the lesion. In view of the inconclusively of the report, an immunohistochemical study was carried out for definitive and therapeutic conclusion. After analysis, the occurrence of malignant epithelioid neoplasia was observed, with giant cells multinucleated type osteoclasts, but without possibility of determination of the histogenesis of the neoplastic cells, considering the negativity for all the markers tested (S100, EMA, DESMINE, AE1AE3, AML, CD34, INI1 and CD68).

In front of the table, we performed Computed Tomography of the total abdomen, being: moderate volume ascites; thickening of small intestine loops with formation of conglomerates of intestinal loops with right predominance; voluminous expansive lesion in the right iliac fossa, with blurring and multiple mesenteric nodulations without a clear cleavage plane with the distal ileum, with foci of emphysema. No primary neoplastic foci were found.

In view of the findings, a new surgical approach was proposed, which was contraindicated in view of the clinical deterioration due to the spoliation by the pathology. As a result, the patient evolved to death on 03/21/2018, 51 days after the onset of symptoms.

Discussion

According to the specialized literature, carcinoma of the cecal appendix is characterized as a rare condition, which affects individuals in the sixth and seventh decade of life predominantly - this characteristic is presented by the patient in question[1,2,9,10].

Regarding the clinical picture, this pathology presents initially as a form of acute appendicitis, considering the possibility of distension, inflammation and consequent rupture of the cecal appendix. Only in rare or advanced cases, the disease presents with a

tumor in the lower right quadrant of the abdomen[1,3,4,11]. As a result, the assertive diagnosis ends up being impaired, minimizing the chances of successful recovery of the affected patients, as could be evidenced in the case described above.

In this context, despite this non-specificity of the signs and symptoms of cecal appendix carcinoma in relation to acute appendicitis, it is possible to infer that the rupture of this organ is in several cases the first symptom of appendix cancer[12]. This fact corroborates for a more careful conduct in the postoperative period of the patient, thus avoiding complications resulting from the late diagnosis of this pathology, as in the case in question.

Depending on the histological form presented, the cells can produce lymphatic and blood lymphatic metastases to other regions of the intestine, distant organs and local lymph nodes. For staging of the disease, the following nomenclature is usually used[13]:

- Stage I (17%) - mucosal neoplasia limited;

- Stage II (36%) - neoplasia invading all layers, but not exceeding serosa;

- Stage III (17%) - carcinoma invades the cecum, peritoneum, ileocecal ganglia and/or appendicular perforation;

- Stage IV (17%) - occurrence of metastases in other organs

Considering the occurrence of cecal, mesenteric and lymphatic involvement, associated with intestinal perforation with elimination of the fecaloid content of the patient in question, there is Stage III Neoplasia - which presents a reserved prognosis, even with a more invasive intervention. Some authors [5,9,13] still consider the existence of a correlation between biological behavior and tumor size: the larger the size, the greater the probability of malignancy and the consequent aggressiveness.

It is not frivolous to say that if the correct diagnosis was eventually established at the time of the onset of symptoms, unfavorable patient outcomes could be avoided. However, as the literature indicates[2,3,9], early diagnosis is the exception, given the low specificity of the symptoms, as well as the insipidity of surgeons with previous contact in the conduction of similar cases. In spite of this, freezing biopsy should be performed whenever the appendix is atypical, in order to provide greater procedural safety if the histopathological examination reveals a positive result in the future[10,11]. In addition, the aid of Computed Tomography can influence the early diagnosis, since the visualization of calcifications or air bubbles increases the probability that the process is malignant and not infectious or inflammatory[12].

Despite the unusual pattern of appendix neoplasm, another aspect that evokes attention in the case described is the cellular typology presented in the histopathological report: giant epithelioid cells of type osteoclasts - typical cell types typical of bone neoplasms. According to the literature, the three main types of appendix malignant cells are those involved in adenocarcinomas, carcinoids and adenocarcinoids, with a rarity of other cell types (epitheliomas, leiomyosarcomas, fibrosarcomas and liposarcomas) - a feature that enhances the relevance of discussion of the case in question[14].

Regarding the treatment after diagnosis, the right hemicolectomy would be the procedure applicable to all cases, with a significant increase in the survival of the affected patients, compared to appendectomy with cecal/colonic preservation. In cases of confirmed carcinoma after histopathological examination, surgical re-placement to perform right hemicolectomy is recommended, as early as the first month of follow-up. Unfortunately, in the case described, there was not enough time for such, impossibility affected in large part by the delay in the release of anatomopathological reports observed in the geographic region of the study. In addition, adjuvant therapy is not recommended, since neoplasias of this nature are not very sensitive to chemotherapy and radiotherapy - which is why they were not indicated in the case under discussion[8,15,16].

It should be emphasized that all patients diagnosed should be staged, with periodic reassessment and reestablishment in cases in which hemicolectomy is feasible. It is recommended that it be quarterly in the first year, every six months in the following five years and annual after five years[17]. Clinical evaluation should include opaque enema, colonoscopy, and carcinoembryonic antigen (CEA), chest X-ray, and computed tomography to confirm the return of the disease or possible metastases[7,10].

In relation to the prognosis, the greater survival of the patients diagnosed in stages I and II of the disease is well known, with those patients having a 5-year survival of 91 to 100%. Patients in stages III and IV - with neoplastic proliferation outside the appendix, as in the case in question - this expectation falls to 44% of the cases. In addition, 10-year survival in advanced cases of this pathology is generally less than 10%, thus justifying the need for adequate and early management in order to avoid unfavorable outcomes[18].

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