THE SURPRISE BEHIND AN EMERGENCY APPENDECTOMY

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CASE REPORT

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Abstract

Malignant tumors of the appendix can be classified into mucinous epithelial neoplasms, neuroendocrine tumors (typical carcinoids), goblet cells or composite carcinoids, lymphomas, adenocarcinomas and lymphoid or mesenchymal sarcomas. Histologically, more than half of appendicular tumors are of neuroendocrine origin, while the rest are adenocarcinomas (mucinous, signet ring or non-mucin-secreting). These tumors are rarely suspected before surgery, the final diagnosis being intraoperative and established by histopathological examination. Appendiceal mucinous neoplasms represent an extremely small percentage of all appendectomy specimens. We present the case of a 44-year-old patient who presented an altered general condition, severe pain in the right iliac fossa associated with episodes of nausea, vomiting and fever. The result of the contrast computed tomography of the abdomen and pelvis identified acute appendicitis with right parietocolic appendicular plastron. Emergency surgery was decided and under general anesthesia, laparoscopic appendectomy with drainage of the peritoneal cavity was performed. The postoperative evolution was favorable and the patient was discharged on the 7th postoperative day. The histopathological examination of the appendectomy specimen established the diagnosis of infiltrative mucinous adenocarcinoma at the level of the adventitia. After multidisciplinary team meeting, given the histopathological findings, right ileohemicolectomy with ileo-transverse anastomosis was decided and performed. The patient is under oncological surveillance with periodic assessment, being currently disease free. Appendicular mucinous neoplasms are a rare disease for which diagnostic and therapeutic management is a challenge. Appendiceal mucocele remains a rare cause fo acute surgical abdomen.

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Introduction

Malignant tumors of the appendix can be classified into mucinous epithelial neoplasms, neuroendocrine tumors (typical carcinoids), goblet cells or composite carcinoids, lymphomas, adenocarcinomas and lymphoid or mesenchymal sarcomas. Histologically, more than half of appendicular tumors are of neuroendocrine origin, while the rest are adenocarcinomas (mucinous, signet ring or non-mucin-secreting). These tumors are rarely suspected before surgery, the final diagnosis being intraoperative and established by histopathological examination [1].

Appendiceal mucinous neoplasms have a low incidence among cases requiring an appendectomy, but they are the second most common cause of appendiceal tumors after carcinoid tumors [2].

The theory of appendicular mucocele appeared for the first time in the medical literature in 1876 described by Feren, although its first description was given by Rokitansky in 1842 [3].

Appendiceal mucinous neoplasms are classified into low-grade or high-grade. They have an unpredictable evolution, with a variable recurrence rate. Patients with low-grade lesions have a low recurrence rate, whereas those with positive post-appendicectomy margins and appendicular perforations have an increased risk of developing malignant pseudomyxoma peritonei (PMP) [4-6].

As for the treatment, there are still controversies, ranging from a simple appendicectomy when the tumor is located only in the appendix to right ileohemicolectomy in more advanced stages, associating it with chemotherapy treatment [2].

Case presentation

We present the case of a 44-year-old male patient who came to the Emergency Unit of "Saint John" Emergency Clinical Hospital with an altered general condition, severe pain in the right iliac fossa associated with episodes of nausea, vomiting, fever and chills, symptomatology that had started 12 hours before presentation.

On admission, the clinical examination of the patient revealed a spontaneously painful abdomen, exacerbated on palpation of the right iliac fossa that associated signs of peritoneal irritation with accelerated intestinal transit.

Biological constants indicated:

- microcytic hypochromic anemia with hemoglobin=10.3g/dL
- hematocrit=30.50%
- leukocytosis (WBC=19.65x1000/uL) with neutrophilia (14.14x1000/uL)

The result of the contrast computed tomography of the abdomen and pelvis identified acute appendicitis with right parietocolic appendicular plastron (Figure 1,2).

Emergency surgery was decided and under general anesthesia, laparoscopic appendectomy with drainage of the peritoneal cavity was performed.

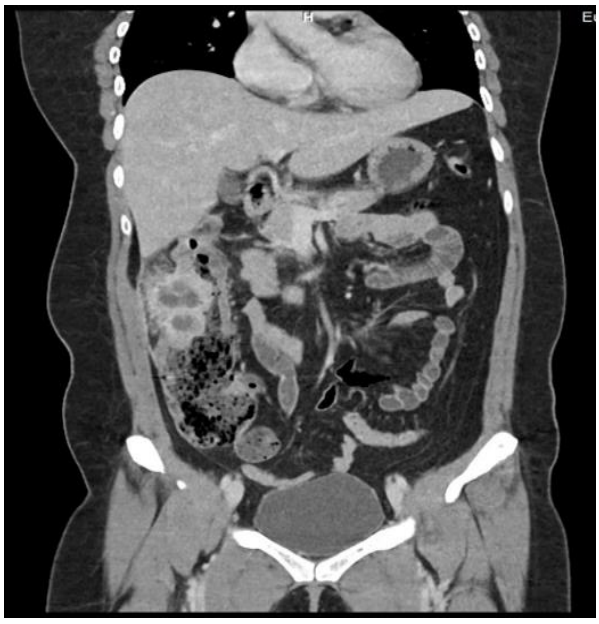

Figure 1 - Abdominal CT, frontal view, appendicitis with right parietocolic appendicular plastron

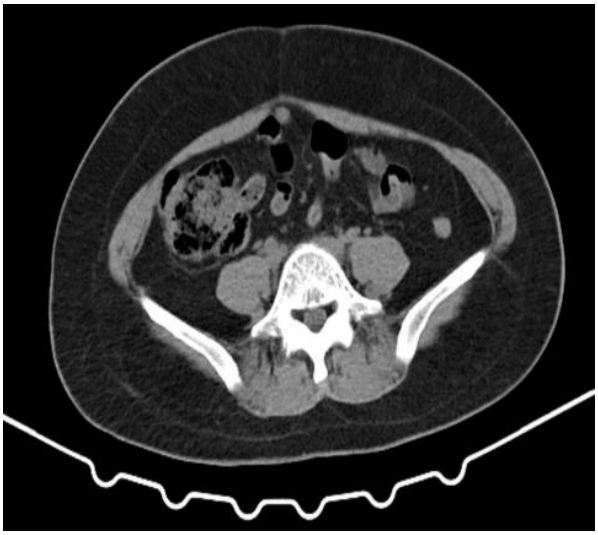

Figure 2 – Abdominal CT, transversal view, appendicitis with right parietocolic appendicular plastron

The postoperative course was uneventful. He was discharged on the 7th postoperative day with a good general status, no abdominal complaints, resumed intestinal transit and food tolerance.

The histopathological examination of the appendectomy specimen established the diagnosis of high-grade (G3) infiltrative mucinous adenocarcinoma at the level of the adventitia (pT3) (Figure 3).

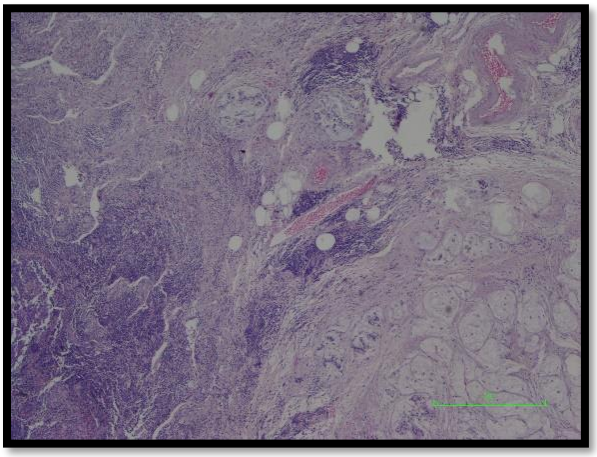

Figure 3 - High-grade (G3) infiltrative mucinous adenocarcinoma at the level of the adventitia

After multidisciplinary team meeting, given the histopathological findings, right ileohemicolectomy with ileo-transverse anastomosis was decided and performed. The patient is under oncological surveillance with periodic assessment, being currently disease free.

Discussions

Mucinous neoplasms are becoming more common, with a higher proportion of cases being found in young people. Appendicular mucocele is discovered by chance in more than half of the cases during imaging, endoscopic examinations, or surgical interventions for other reasons [7].

The symptomatology is non-specific, with acute abdominal pain in the right iliac fossa being the most common; a tumor mass can also be palpable at times. These can be associated with other unusual symptoms and signs, such as intestinal fistulas, intestinal occlusion, and gastrointestinal hemorrhage [8,9].

The first paraclinical investigation used is abdominal ultrasound, which usually shows cystic formations with variable echogenicity depending on the mucin composition. The "onion skin sign" is a specific sign for appendicular mucocele, which is manifested by numerous echogenic layers along a dilated appendix (greater than 15 mm on ultrasonography, supporting the diagnosis of appendiceal mucocele). This sign is not always present [10].

Computed tomography (CT) is another important diagnostic modality in the diagnosis of appendicular mucocele. In the right lower quadrant of the abdomen, a hypoechoic, well-encapsulated mass with smooth regular ridges can be seen. Mural calcifications, a specific sign of mucocele in most cases, can be highlighted in approximately half of the cases [11].

Colonoscopy is an effective method for determining the presence of a mucocele.

Colonoscopy shows a smooth formation with normal mucosa overlying the appendicular orifice. When air is blown into this formation, it retains its firmness and does not flatten. It has the appearance of a volcano, hence the "sign of the volcano" [8].

The most serious complication is pseudomyxoma peritonei (PMP), which is a rare pathology caused by appendicular perforation after an appendicular mucinous neoplasm and is characterized by mucin dissemination in the peritoneum with peritoneal ascites. In order to avoid this complication, there must always be a high degree of suspicion regarding atypical appendicular pathology [2,6].

For patients with uncomplicated low-grade lesions with localized disease, a simple appendectomy is an option. In the case of mucinous adenocarcinoma with appendicular perforation, positive resection margins and positive lymph nodes, right hemicolectomy is required and cytoreductive surgery (CRS) combined with hyperthermic intraperitoneal chemotherapy (HIPEC) and postoperative chemotherapy is recommended [12-14].

Treatment after appendectomy is satisfactory in patients with low-grade neoplastic lesions, with a 10-year survival rate of up to 90%. Patients with high-grade neoplastic lesions also show good results after surgical resection, but the risk of progression to PMP persists, with a lower survival rate.

Despite the good immediate results of mucinous neoplasm operations, remote monitoring is recommended, even if the lesions turn out to be benign because there have been cases of recurrence and progression to PMP, as well as cases of metachronous colonic neoplasms [12, 14].

Conclusions

Appendiceal mucinous neoplasms are a rare disease with difficult diagnostic and therapeutic management. Appendiceal mucocele is a relatively uncommon cause of acute surgical abdomen.

In an emergency, ultrasound and CT are the investigations of choice for diagnosing appendiceal mucocele, but histopathological examination establishes the definitive diagnosis.

Mucinous ascites is an advanced stage with a poor prognosis. Because surgical management is dependent on the stage of the injury, accurate diagnosis is critical.

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