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High-Grade Mucinous Neoplasms Of The Apendix- A Case Report

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Abstract:
Introduction: *Mucinous neoplasms of the appendix are rare and present symptoms similar to acute appendicitis, which makes diagnosis difficult. The accumulation of mucin inside the organ can lead to its perforation resulting in peritoneal pseudomyxoma (PMP), a condition that is to be treated.*
Case report: *A 36-year-old man with no previous medical history suddenly experienced mild abdominal pain that spread throughout his abdomen. A full abdominal ultrasound (USG) was carried out, revealing an enlarged appendix with the walls also showing signs of calcification. Laboratory tests showed alterations and a computed tomography (CT) scan revealed a 10 cm-long perforated appendix. An emergency appendectomy was performed via laparotomy. The anatomopathological examination revealed a mucinous neoplasm with a high degree of differentiation, known as HAMN.*
Conclusion: *High-grade mucinous neoplasia of the appendix (HMN) is a rare disease that is difficult to diagnose early due to the similarity of symptoms to acute appendicitis. The use of methods such as USG, CT, and tumor markers contributes to a detailed treatment of the patient before a surgical approach.*
Keywords: *Appendectomy;Appendicitis;Mucinous Neoplasms*

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I. Introduction

Mucinous neoplasms of the appendix are characterized by the accumulation of mucin in the organ, although rare conditions, they have different histological types, and a certain similarity with acute appendicitis in terms of symptoms, making the diagnosis challenging. This situation has been the subject of study in scientific literature. (1, 2, 3)

Mucinous lesions of the appendix are classified as non-neoplastic and neoplastic. (2, 3) Neoplastic lesions are subdivided into Serrated Polyps, Low-Grade Mucinous Neoplasms (LAMN), High-Grade Mucinous Neoplasms (HAMN), and Mucinous Adenocarcinoma. The HAMN present lesions with expansive characteristics with enlarged, hyperchromatic, and pleomorphic nuclei; abundant atypical mitotic activity, and abundant mucin production. However, as it is a recently created pathological category,(4) the scientific data about the behavior of HAMNs is scarce. Also, it is important to remark that this is a highly controversial area, including that the World Health Organization classifies most noninvasive epithelial appendiceal lesions as LAMNSs(5).

Although mucinous neoplasms of the appendix are rare, these tumors have a low incidence ranging from 0.2% to 0.7% of total appendectomies, and have a clinical presentation that resembles non-neoplastic conditions (6). This disease causes an accumulation of mucin within the organ, which may mean falsely acute appendicitis, and, If it perforates and spreads to the peritoneal cavity, it can lead to a condition called pseudomyxoma peritonei (PMP) (7). Diagnosis is challenging and requires an abdominal CT scan or ultrasound, which presents intra- and post-operative risks.

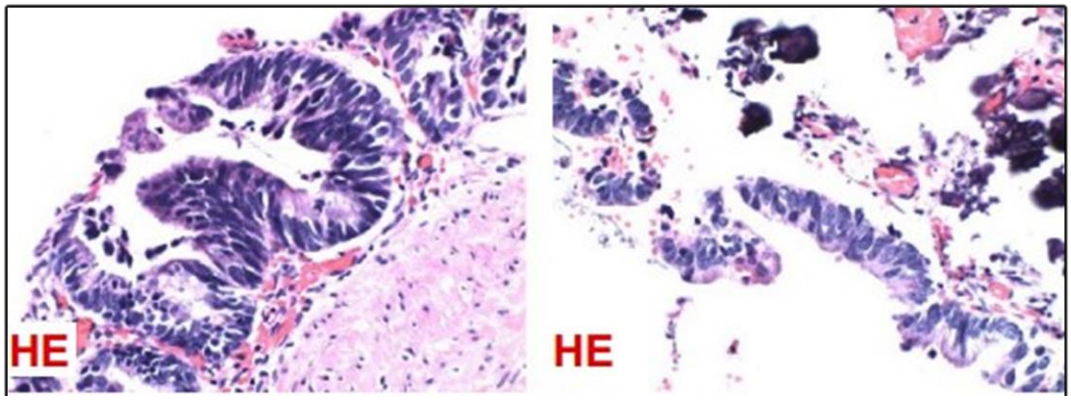
This case report describes a patient who presented with symptoms suggestive of appendicitis and underwent a surgical procedure. Histopathological analysis revealed high-grade mucinous neoplasms (Figure 1).

II. Case Presentation

A 36-year-old man named TC, who has no medical history or medication use reported, suddenly experienced mild abdominal pain that spread throughout his abdomen. He did not have a fever, loss of appetite, or any other symptoms. However, two days later, the pain became more intense and focused on the right iliac fossa. As the pain persisted, he consulted with a family ultrasonographer. The following day, a complete

abdominal ultrasound was conducted, which revealed a dilated appendix measuring 4 cm in diameter- **FIGURE 1**.

The walls of the appendix also showed signs of calcification. Initially, the doctor suspected Merkel's diverticulum or some other pathology of the colon because the symptoms were not typical of acute appendicitis. However, due to these findings, the professional, who was knowledgeable about the symptoms of acute appendicitis, ordered routine laboratory tests with inflammatory markers, Polymerase Chain Reaction(PCR), and tumor markers, Carcinoembryonic Antigen(CEA), as well as a computed tomography scan (CT scan), which was conducted at a private clinic. The laboratory tests showed that the patient had a PCR level of 48 mg/dL (reference value: 1,0-5,0mg/dL), leukocyte count of 9100 mm^3 (reference value: 4.000-10.000mm^3), and CEA level of 113 ng/mL (reference value: <5,5ng/mL). The CT scan conducted on the following day revealed a perforated appendix that was 10 cm long with mucin leaking into the abdominal cavity. As a result, the patient was referred for emergency appendectomy surgery at the regional hospital on the same evening. An uneventful laparotomy appendectomy was performed. Drainage and irrigation were performed to remove leaked mucin and prevent cancer cell growth after a ruptured appendix. The appendix, lymph nodes, and mucin samples were sent for anatomopathological examination- **FIGURE 2**.



The test showed that there was a mucinous neoplasm present with a high degree of differentiation, which is called HAMN. However, the margins were free of cancer cells and no dysplastic cells were found in the lymph nodes. The mucin present was acellular. The oncologist recommended follow-up MRI or CT scans every six months to check for any signs of mucinous ascites. Six months after the procedure, the patient is healthy. CEA and PCR values have returned to reference values(CEA:1,2ng/dL and PCR:2,0mg/mL) and the MRI showed no remission of the neoplastic cells in the peritoneal cavity.

III. Discussion

High-grade mucinous neoplasia of the appendix (HAM) is a rare disease characterized by obstructive dilatation caused by the accumulation of mucoid material in the appendix lumen (8, 9, 10, 11).

It is widely accepted among researchers (1, 7,12,13) that the most significant complication of mucinous neoplasm of the appendix is the perforation of the organ, similar to what occurred with the patient. This leads to the spread of mucin into the peritoneal cavity, resulting in peritoneal pseudomyxoma (PMP). Unfortunately, treating PMP is a complex process with unsatisfactory long-term outcomes. The patient's follow-up as well as the disease's long-term development, we'll report opportunely.

It's worth mentioning that there are many pieces of medical literature available, such as KUMAR(14), that only briefly mention the topic of mucinous neoplasms of the appendix due to the rarity of the condition, meeting common sense among clinicians and researchers (8, 10, 15).

Typically, these articles generalize the clinical and laboratory discrimination of such neoplasms, as was reported by Michael J. (4), KUMAR (14), Nelya (16), and Carpenter SG. (17), Rangarajan (19), Ruiz-Tovar (20), and Demetrashvilli (21). To summarize, the clinical presentation of this condition is not specific and resembles that of conventional acute appendicitis. Patients may not exhibit symptoms, but if they do, they may experience pain in the lower right abdomen, vomiting, nausea, a palpable mass in the abdomen, weight loss, and gastrointestinal bleeding (4, 16), information congruent with the findings in the clinical case reported. It is imperative to remember that as 50% of cases are discovered by chance during appendectomy or subsequent pathology, this cannot be considered a wrong finding, as defended by Overman (7), which was readily corroborated by Gronroos (22) and Wang (23). In fact, in the beginning, the very difficult differential diagnosis constituted an additional barrier to patient care.

The way it is described may lead one to believe that a doctor's role in distinguishing between different cases is minimal. However, this case study shows that professionals with the correct knowledge of the disease's propaedeutics can suspect some percentage of mucinous neoplasms of the appendix in advance. There are indications, supported by studies, that allow this differentiation (10, 12, 16, 24).

For instance, the absence of leukocytosis makes the occurrence of acute appendicitis unlikely, as mentioned by Gronroos (16), except if the disease is in its early stages.

The presence of calcifications in the wall, as seen in Figure 2 and visualized by USG, is an important marker of chronicity and a relevant factor indicating neoplasms. The sensitivity and specificity of this marker are 83% and 92% respectively, as mentioned by Wang (17). Computed Tomography (CT) is considered the most accurate diagnostic method. It can be used to discover specific signs such as an appendix lumen larger than 1.3 cm, cystic dilatation, and calcification of the wall. These signs indicate the presence of mucocele (4,6).

It is possible to measure tumor markers such as CEA before the diagnosis of mucinous neoplasia of the appendix if there is suspicion and time. CEA is a strong indicator in such cases. However, these markers are typically measured only after the diagnosis has been made. This information has been explained by McFarlane (10), and corroborated by others (13, 14, 18).

Thus, although high-grade mucinous neoplasia of the appendix (HMN) is a rare disease, appendectomy is the most common abdominal surgical emergency in the world, with a lifetime risk of 8.6 percent in males and 6.9 percent in females (25). So, even if it represents a low percentage of appendectomies, 0.2%-0.7% (6), the absolute number of people affected is significant. In addition, studies carried out by Marmor (26) indicate an increase in cases of mucinous neoplasia of the appendix.

Furthermore, it is known that the prior diagnosis of a mucinous neoplasm of the appendix is decisive in guiding the surgeon's conduct and avoiding intra-operative and post-operative complications, especially PMP, the treatment of which is a complex process with unsatisfactory long-term results. And is much costly, because one of the best alternatives to combat PMP is HIPEC(Hyperthermic Intraperitoneal Chemotherapy) (27, 28).

IV. Conclusion

It is important to recognize the actions of the medical professional in charge of the treatment for TC, despite being unable to prevent the appendix perforation due to the rapidly deteriorating condition of the patient. The professional was able to suspect beforehand that the condition was a neoplasm of the appendix, rather than a common acute appendicitis, which highlights their expertise in this area. Identifying this condition in advance can improve the patient's prognosis significantly.

It is important to highlight that the initial research, diagnosis, and recommended treatment in this case are supported by a significant body of scientific literature on the topic. Additionally, it's worth noting the significance of this article in fostering discussions and presenting new research and analysis on clinical cases.

High-grade mucinous neoplasia of the appendix (HMN) is a rare disease that presents similar symptoms to acute appendicitis. It is often diagnosed during surgery or on histopathological analysis. Early diagnosis can be challenging as the symptoms are similar to acute appendicitis. However, methods such as USG, CT, and tumor markers can help in a detailed analysis of the patient before the surgical approach. If there are signs of neoplasia,

the surgical approach should be done with caution, paying attention to surgical margins and the dissemination of mucin into the peritoneal cavity. This can prevent PMP, a condition where mucin spreads to the peritoneal cavity.

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