



Surgery

THE CURSE OF THE WHITE PEARL- MUCINOUS APPENDICULAR NEOPLASM PRESENTING AS APPENDICULAR PERFORATION - A CASE REPORT

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KEYWORDS :

INTRODUCTION

Mucinous appendicular neoplasms represent a rare and poorly understood heterogeneous pathology that is present in less than 0.3% of all appendicectomies^[1]. They have a very low incidence in the general population, with age at diagnosis between 50 and 60 years and a male-to-female ratio slightly in favor of the latter^[1]. These types of neoplasms are incidentally discovered during appendicectomies or in routine imaging for other conditions^[1]. From a clinical point of view, patients are often asymptomatic or present with right lower quadrant (RLQ) abdominal pain mimicking appendicitis and in most of the cases, the laboratory findings are nonspecific of the same^[1]. Complications depend on tumor size and histological type and can range from intestinal obstruction and bleeding to spontaneous perforation and peritoneal dissemination causing pseudomyxoma peritonei (PMP)^[1]. In surgical treatment, the open approach is preferred to laparoscopic surgery for large appendicular tumors due to the risk of iatrogenic rupture and mucin spread in the peritoneal cavity that can cause pseudomyxoma peritonei, although minimally invasive approaches such as the robotic-assisted approach and the hand-assisted laparoscopic approach can be considered safe in the resection of appendicular mucocèles in selected patients^[1]. We present our experience in a case of a mucinous appendicular neoplasm as well as diagnostic methods, surgical management and prognosis for this rare pathology and we discuss the main problems encountered in the management of the case.

CASE SUMMARY

A 66 year-old male patient presented with pain in the right lower quadrant of the abdomen since 10 days and fever since 2 days. The patient underwent Computed tomography (CT) imaging using intravenous contrast that revealed an ill defined multilocular peripherally enhancing hypodense collection in RIF and minimal inflammatory changes with segment of appendix within the collection-s/o ? sealed off appendicular perforation with abscess formation.



Figure 1 Computed tomography findings. A: Computed tomography (CT) scan showing collection in RIF with appendix not visualized – sealed off perforated appendix

The patient was taken up for emergency laparotomy during which 30ml of purulent collection was found in the rif region with perforated tip of the appendix and features of early mass formation.

A simple appendicectomy was performed after careful adhesiolysis. Fluid was sent for microbial culture and the resected mass was sent to the pathological department for further study.

The histopathological report of the specimen showed mucinous glands with focal villiform pattern. cells are tall columnar with mild nuclear atypia and abundant mucinous cytoplasm. Lymphoid tissue is atrophic. Extracellular mucin is noted. Base of the appendix is free of tumour. Postoperative staging revealed a stage 0 LAMN. The patient did well after surgery, was discharged after a week and is presently included in our follow-up program, which consists of alternating abdominal ultrasound and CT scans every 6 months.



figure 2 : Intra op: Perforated tip of the appendix

DISCUSSION

Appendicular lesions are a rare entity; in a 10-year Dutch study, the incidence of appendicular lesions was approximately 9 per million persons per year, with one in 113 resected appendices presenting a lesion (benign or malign in nature)[1]. The benign-to-malignant ratio was approximately three to one, although more recent studies present a higher disease incidence, malignant lesion rate and decreased age at diagnosis[1,12,13]. A consensus on classification has been reached by the Peritoneal Surface Oncology Group, grouping mucinous appendiceal neoplasms into adenoma, LAMNs, high-grade appendiceal mucinous neoplasms and mucinous adenocarcinomas [14]. Patients with appendiceal tumors have nonspecific clinical signs or are asymptomatic, and in almost half of cases, they are diagnosed with acute appendicitis; therefore, preoperative imaging plays an important role not only in diagnosis but also in operative strategy[12]. Ultrasound, which is fairly sensitive, is best suited for suspected cases and when complemented by CT, is largely used for the diagnosis, staging and follow-up of appendiceal tumors. However, magnetic resonance imaging (MRI) is more sensitive in identifying peritoneal disease[15]. The standard of treatment in appendicular tumors for many years has been appendicectomy (for tumors under 2 cm) and right hemicolectomy (for tumors larger than 2 cm), although studies have suggested that an intact mucocèle with a normal base of implantation in the cecum is in most cases a rather benign lesion and simple, radical appendicectomy can suffice with no difference in survival between these types of procedures[3,7-11,16,17].

In situations where total removal of the primary tumor cannot be performed by simple or radical appendicectomy, where tumor lymph node involvement is found, or when a non mucinous neoplasm type is present during pathological exam, right hemicolectomy is

recommended as the standard procedure[7-11]. Laparoscopic surgery has gained ground over the open approach in the last two decades, although open surgery can be more reliable in large and giant tumors due to the risk of iatrogenic rupture and mucin spread in the peritoneal cavity[7-11]. Having this in mind, we preferred the open approach in our case in order to perform a complete local resection and then determine whether additional surgery is needed based on the final pathological results of the excised specimen.

We also believe that especially for large lesions the open surgical approach allows a better damage control of the potential rupture and iatrogenic lesions.

CONCLUSION

Mucinous appendicular neoplasms are a rare pathology that present with nonspecific clinical signs and, in many cases, are asymptomatic. Preoperative diagnosis or suspicion is important for the operative strategy and choice of operating technique. Based on histological findings and careful staging, simple or radical appendicectomy can be safely performed for Low grade mucinous appendicular neoplasm if the criteria are met.

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