Case Report

Appendiceal Carcinoid Tumor within Amyand's Hernia: A Case Report and Review of the Literature

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ABSTRACT

Introduction: Amyand's hernia is defined as the herniation of the whole or a part of appendix vermiformis into an inguinal hernia sac. Amyand's hernia is very rare, constituting about 1% of all inguinal hernia patients and 0.1% of all acute appendicitis patients. In one-quarter of Amyand's hernia patients, inflammatory changes are detected in appendix vermiformis. Detecting an appendiceal tumor in an Amyand's hernia sac is extremely rare, with only eight cases having been reported so far in the English literature.

Case presentation: A 64-year-old man with an appendiceal carcinoid tumor inside an incarcerated Amyand's hernia sac. As the tumor was localized to the radix of the appendix, resection was carried out with the help of a linear-cutter stapler in a way to contain a part of the cecal wall. As the pathology examination revealed a carcinoid tumor infiltrating the periappendicular fatty tissue, right hemicolectomy was performed in a second session, in compliance with the oncological principles. No metastasis was present at the 24th month. Case reports published in the English literature regarding the Amyand's hernia with an appendiceal tumor were also analyzed in this study.

Conclusion: Amyand's hernia containing an appendiceal tumor is an extremely rare clinical entity. Clinical manifestations mostly mimic the incarcerated hernia and thus accurate diagnosis is made incidentally during hernia surgery. The basic oncological principles apply for the approach to tumors inside a hernia sac.

1. Introduction

Whole or part of appendix vermiformis being inside an inguinal hernia sac is termed as Amyand's hernia. Amyand's hernia is very rare, constituting about 1% of all hernia cases and 0.1% of all acute appendicitis cases.1 Amyand's hernia's clinical signs and symptoms typically resemble those of incarcerated inguinal hernia and thus most patients are operated with a preliminary diagnosis of incarcerated or strangulated inguinal hernia.1-2 Hence, a preoperative diagnosis can be made with an ultrasonography (US) or contrast-enhanced computerized tomography (CT) in a very small proportion of patients.1-3 Amyand's hernia may remain asymptomatic and complication-free for years. However, the appendix vermiformis may be strangled by hernia sacs with a narrow neck, which may result in signs and symptoms of appendicitis, perforation, or peritonitis.1-2 The treatment of an Amyand's hernia typically varies whether the appendix vermiformis in a hernia sac is complicated or not. Detection of an appendiceal tumor inside an Amyand's hernia is an extremely rare situation, and only a few cases have been reported.6-11 In this study, we aimed to report a 64-year-old man who was found to have a carcinoid tumor inside an incarcerated Amyand's hernia sac.

2. Case report

A 64-year-old man presented to our emergency department with severe pain in the right inguinal region. He states that the swelling had begun two years ago, and it had spontaneously disappeared and returned from time to time but had intensified for the last two days. Apart from congestive heart failure, he had no history of chronic prostatism or chronic obstructive pulmonary disease. An upright physical examination revealed a carcinoid tumor infiltrating the periappendicular fatty tissue, right hemicolectomy was performed in a second session, in compliance with the oncological principles. No metastasis was present at the 24th month. Case reports published in the English literature regarding the Amyand's hernia with an appendiceal tumor were also analyzed in this study.
urgentley taken to the operation theatre. After a right inguinal incision was made, spermatic cord and its contents were returned with a part of Penrose drain. The indirect hernia sac adhered to the cord that contained intestinal loops was isolated. After opening the sac, an edematous appendix vermiformis and a short ileal segment were noticed. As there was no sign of ischemia, the ileal segment was returned to the abdominal cavity. However, since the appendix vermiformis was edematous and a firm mass was palpated in its root, a resection was performed using an 80 mm linear cutting stapler to visualize a part of cecum wall neighboring the appendix. Then, the hernia sac was reduced a graft-free hernia repair was performed. The patient was discharged on the 5th postoperative day uneventfully. The pathology report revealed a grade I NET infiltrating the peripendiccular fatty tissue [CgA: diffuse (+ + +), CD56: diffuse (+ + +), PanCK: focal (+), Ki-67: < %1, Mucicarmine: negative]. Since the tumor was close to the cecum wall and infiltrated the peripendiccular fatty tissue, right hemicolectomy with ileocolic anastomosis was performed. The histopathological examination of the colon specimen revealed no tumor tissue. The patient was followed by medical oncology and general surgery departments, and remained free of tumor metastasis during the 24-months of follow-up.

3. Discussion

Appendectomy is the most common emergency surgical procedure performed worldwide. Depending on the position of its free end, the appendix vermiformis may have a retrocecal, pelvic, preileal, postileal, subcecal, subileal, or subcecal location. Despite being rare, appendix vermiformis may protrude into natural or iatrogenic openings due to several predisposing risk factors. Appendix vermiformis most commonly protrude into the inguinal (Amyand’s hernia) and femoral (De Basson). If the appendix vermiformis inside the hernia sac (Type I) is ruptured, which may cause acute appendicitis, perforation, abscess for- mation, or peritonitis. 1–3 Majority of patients with Amyand’s hernia are urgently operated for a preliminary diagnosis of incarcerated or strangulated hernia, and Amyand’s hernia is an incidental finding during surgery. 1–10 Therefore, the role of radiological tools is very limited.

Cases without an urgent condition may be diagnosed with US or CT. 1–3 In the presented case, despite similar anatomical descriptions in the official sonographic examination report, the preliminary diagnosis of the consulting radiologist was incarcerated inguinal hernia on the right.

The treatment option depends on the status of the appendix vermiformis (normal, appendixitis, perforation, abscess, apparent peritoni- tis). The most widely accepted algorithm is proposed by Losanoff and Basson. If the appendix vermiformis inside the hernia sac (Type I) is normal, hernia content should be reduced without performing appendect- omy, followed by hernia repair with a suitable mesh. 1–4 In contrast to that view, some authors advocate the application of appendectomy even if appendix vermiformis appear to be normal, given the risk of acute appendicitis. 1–4 If acute appendicitis is present inside the hernia sac (Type II), appendectomy must be performed, and the defect should be repaired using one of the primary repair techniques without using a mesh. In contrast to that view, some authors have performed hernia repair without using a mesh, and no infectious complications were observed. 1–4 Acute appendicitis inside the hernia sac and signs of peritoneal irritation (Type III) necessitate open appendectomy and primary hernia repair without using a mesh. 1–4 Another approach is performing appendectomy, primary closure of the internal ring during laparotomy, and repairing hernia with a mesh a few weeks later, after the inflammation subsides. For patients with acute appendicitis inside the hernia sac plus another intraabdominal condition (Type IV), the same approach as for Type III is applied. Additionally, adjunctive treatment options are applied depending on intraabdominal pathology.

The most commonly encountered pathological features are normal appendix vermiformis, acute appendicitis, perforated appendicitis, and peripendiccular abscess. However, a very limited number of cases of an appendiceal tumoral lesion inside the Amyand’s hernia sac has been reported in the literature. We encountered 8 case reports while searching the English literature from the PubMed and Google Scholar databases. 6–11 Two cases had carcinoid tumor; two cases had goblet cell carcinoid tumor; two had mucinous cystadenoma; and one case had adenocarcinoma, and one had villous adenoma. The patient diagnosed with adenocarcinoma later underwent right hemicolectomy while the rest of the patients underwent no additional operation considering tumor’s properties (Table 1).

Both the above-mentioned classification and oncological principles should be followed during the management of patients with appendi- ceal tumor inside a hernia sac. Neuroendocrine tumors (NETs) are de- tected in 0.3–2.27% of the appendectomy specimens in patients oper- ated for suspected acute appendicitis, and 1.8–2.3% of the appendectomy specimens in patients operated for incidental appendici- tis. 12 Right hemicolectomy is the most appropriate approach for NETs with mesoappendix invasion, or with a diameter equal to or greater than 20 mm. Among tumors with a diameter ranging between 10 to 19 mm, right hemicolectomy is the most appropriate approach if one or more of the parameters such as mesoappendix invasion, vascular in- vasion, grade II proliferation, suspected or positive surgical borders, and mixed histology (goblet cell carcinoid, adenocarcinoid) are pre- sent. 6,12 Simple appendectomy would be adequate in NETs of the ap- pendix with no mesoappendix invasion and a diameter of 10–19 mm, and NETs with a diameter of less than 10 mm. 12 The present case was operated with right hemicolectomy since the tumor was in close proximity to the cecum wall, and peripendiccular fat tissue was in- filtrated.

Right hemicolectomy in the same session, or after the pathology report was declared, is the most appropriate approach for all adenocarci- noma s of the appendix, irrespective of their location and size. Even the appearance is benign, mucoceles should be treated with sur- gical resection with clean borders, since some of them have been shown to have a potential for malignancy. Hence, while simple appendectomy is adequate for some histological subgroups, such as mucocoele’s reten- tion cyst, mucosal hyperplasia, and cystadenoma, right hemicolectomy is necessary for complicated mucoceles comprising the cecum and...
terminal ileum, and for mucinous cystadenocarcinomas.

In conclusion, Amyand's hernia, and appendiceal tumor inside the hernia sac are two rare clinical entities. The majority of Amyand's hernia mimic signs and symptoms of incarcerated inguinal hernia, and, thus, diagnosis is made incidentally during surgery. The basic oncological principles should be followed in the management of tumors inside a hernia sac.

**Author contribution statement**

TP and BS: performed the surgical procedure; SA: wrote the paper, analyzed and interpreted the data.

**Conflict of interest statement**

The authors declare no conflict of interest.

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| Table 1 Summary of the clinicopathological characteristics of patients with Amyand's hernia and an appendiceal tumor in the English literature. |
|---|---|---|---|---|---|---|---|---|
| References | Year | Age | Sex | Preliminary Diagnosis | Hernia Side | Hernia Type | Hernia Status | Intraoperative Findings |
| Christodoulidis 2017 | 52 | M | Inguinal Hernia | Right | Indirect | Incarcerated | Acute Appendicitis |
| Yahya 2017 | NA | NA | NA | NA | NA | NA | Acute Appendicitis |
| Elbanna 2015 | 81 | M | Amyand's Hernia | Right | Indirect | Incarcerated | Acute Appendicitis |
| Reymu 2015 | 70 | M | Inguinal Hernia | Right | NA | Strangulated | Acute Appendicitis |
| Nahmias 2013 | 50 | M | Amyand's Hernia | Right | Indirect | Strangulated | Acute Appendicitis |
| Shabeeb 2010 | 62 | M | Inguinal Hernia | Right | Indirect | Incarcerated | Acute Appendicitis |
| Wu 2010 | 62 | M | Inguinal Hernia | Right | NA | Incarcerated | Acute Appendicitis |
| Salemis 2006 | 61 | M | Inguinal Hernia | Right | Indirect | Strangulated | Acute Appendicitis (Perforated) |

| Surgery | Hernia Repair | Approach to Appendix | Histopathological Evaluation | Tm Size (mm) | Tm Location | Recurrence |
|---|---|---|---|---|---|---|
| Open | Bassini | Appendectomy | Goblet Cell Carcinoid | 22 | Tip | No (12mo) |
| Open | NA | NA | Goblet Cell Carcinoid | NA | NA | NA |
| Open | Bassini | Appendectomy | Carcinoid | 15 | Tip | NA |
| Open | Primary | Appendectomy | Mucinous Cystadenoma | NA | NA | NA |
| Laparoscopic | Internal Ring Closure | Appendectomy | Carcinoid | 10 | NA | NA |
| Open | Lichtenstein | Appendectomy | Mucinous Cystadenoma | 30 | NA | NA |
| Open | Bassini | Appendectomy | Adenocarcinoma | 25 | NA | NA |
| Open | Shouldice | Appendectomy | Perforated App + Villous Adenoma | 3 | Base | No (12 mo) |

M: Male, NA: Not available.

* TAPP hernia repair with mesh was performed six weeks after the first operation.

† Right hemicolectomy was performed a few days after the pathology report was approved.

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