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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.[1–3] 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.[1–3] 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.[4–11] 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 bilateral inguinal hernia. Apart from an unreductable inguinal hernia, no other sign to suggest strangulation was detected on the right side. Biochemical studies and full blood count revealed no abnormality. An upright plain radiography showed no air-fluid levels. Ultrasonography of the right inguinal region showed intestinal loops with reduced peristaltism inside a hernia sac. In the light of available clinical manifestations and ultrasonographic features, a diagnosis of incarcerated right inguinal hernia was made, and the patient was

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urgently 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 vizualize 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 periappendicular 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 periappendicular 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 Garengeot's hernia) hernia sacs. Albeit rare, it may also protrude into such openings to form obturator hernia, umbilical hernia, spigelian hernia, laparoscopic port site hernia, drain site hernia, incisional hernia, and diaphragmatic hernia.[1]

Amyand's hernia was first described in 1735 by the French surgeon Claudius Amyand in an 11-year-old boy; thereafter, this rare entity was registered in his name, i.e Amyand's hernia.[1,3,8] Although the range of proportion of the Amyand's hernia among all inguinal hernias is so wide, i.e between 0.19% and 1.7%, it is mostly below 1%.[2,3,8] On the other hand, appendicitis inside an Amyand's hernia constitutes 0.07%–0.13% of all appendicitis, nearly around 0.1%. Amyand's hernia is seen in all age groups (3 weeks-92 years) and its prevalence in children is three times of that in adults, probably because of the presence of patent processus vaginalis in the pediatric population.[1,2] Amyand's hernia does have a bimodal distribution, one in the newborn period and the other over 70 years of age, and it is more common in men compared to women.[1,2]

Amyand's hernia is mostly an indirect inguinal hernia, however, it may rarely appear as a direct inguinal hernia. Generally, only appendix vermiformis is found inside the hernia sac in children, in the contrary, cecum or other small bowel loops may be found in adults. Amyand's hernia is almost always seen in the right inguinal region, but left Amyand's hernia may also rarely be encountered. The most common causes of Amyand's hernia on the left are mobile cecum, situs inversus totalis, midgut malrotation, or a long appendix vermiformis extending to the left side of the midline.

Amyand's hernia usually appears with inguinal swelling that may be spontaneously or manually reduced, and most patients remain asymptomatic for years.[1] Depending on the width of the neck of the hernia sac, blood supply to appendix vermiformis inside the sac may be disrupted, which may cause acute appendicitis, perforation, abscess formation, 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.[1–10] 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, appendicitis, perforation, abscess, apparent peritonitis). 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 appendectomy, 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 periappendicular 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.[4–11] 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 appendiceal tumor inside a hernia sac. Neuroendocrine tumors (NETs) are detected in 0.3–2.27% of the appendectomy specimens in patients operated for suspected acute appendicitis, and 1.8–2.3% of the appendectomy specimens in patients operated for incidental appendicitis.[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 invasion, grade II proliferation, suspected or positive surgical borders, and mixed histology (goblet cell carcinoid, adenocarcinoid) are present.[8,12] Simple appendectomy would be adequate in NETs of the appendix 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 periappendicular fat tissue was infiltrated.

Right hemicolectomy in the same session, or after the pathology report was declared, is the most appropriate approach for all adenocarcinoma s of the appendix, irrespective of their location and size. Even the appearance is benign, mucoceles should be treated with surgical 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 mucocele's retention cyst, mucosal hyperplasia, and cystadenoma, right hemicolectomy is necessary for complicated mucoceles comprising the cecum and

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
Reynu	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)
NA	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[a] Closure	Appendectomy	Carcinoid	10	NA	NA
Open	Lichtenstein	Appendectomy	Mucinous Cystadenoma	30	NA	NA
Open	Bassini	Appendectomy	Adenocarcinoma[b]	25	NA	NA
Open	Shouldice	Appendectomy	Perforated App + Villous Adenoma	3	Base	No (12 mo)

M: Male, NA: Not available.
[a] TAPP hernia repair with mesh was performed six weeks after the first operation.
[b] Right hemicolectomy was performed a few days after the pathology report was approved.

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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