

Perforated Appendix Containing Goblet Cell Carcinoid Tumor inside an Amyand Hernia Managed Laparoscopically

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Background	An Amyand hernia is an inguinal hernia containing the vermiform appendix. A goblet cell carcinoid (GCC) inside an Amyand hernia presenting with acute appendicitis has only been reported three times in the literature, but to the author's knowledge, presentation with a perforated appendix in this setting has yet to be described.
Summary	We present an extremely rare case of a 79-year-old male with an Amyand hernia presenting with a perforated appendix containing a GCC. A 79-year-old male with a heavy smoking history and a longstanding and uncomplicated history of bilateral inguinal hernias presented complaining of a two-day history of increasing bilateral painful groin lumps and generalised peritonitis. As generalised peritonitis was atypical for incarcerated hernia, a CT abdomen was performed and demonstrated acute peritonitis surrounding the appendix that was contained within the right hernia orifice. The patient had a laparoscopic appendectomy that confirmed Amyand hernia with acute appendicitis. Histology revealed a GCC, and the patient went on to receive a completion right hemicolectomy.
Conclusion	A presentation of perforated appendicitis with generalised peritonitis inside an Amyand Hernia and managed laparoscopically is a rare occurrence, made rarer still by the presence of a GCC. It is also a challenging preoperative diagnosis; presentations of incarcerated inguinal hernia with atypical features should warrant consideration of this differential diagnosis.
Keywords	Amyand hernia, appendicitis, goblet cell carcinoma

DISCLOSURE STATEMENT:

The authors whose names are listed immediately above certify that they have NO affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation in speakers' bureaus; membership, employment, consultancies, stock ownership, or other equity interest; and expert testimony or patent-licensing arrangements), or non-financial interest (such as personal or professional relationships, affiliations, knowledge or beliefs) in the subject matter or materials discussed in this manuscript.

PATIENT CONSENT:

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Case Description

A 79-year-old male with a heavy smoking history and a longstanding and uncomplicated history of bilateral inguinal hernias presented complaining of a two-day history of increasing bilateral painful groin lumps right more so than left. The pain had been worsening over the last few months, but rapidly increased leading up to presentation. He was unable to reduce the lumps himself. He had no associated bowel changes or nausea and vomiting.

On examination the patient was stoic although in obvious discomfort on moving. He was afebrile and haemodynamically stable. Examination of the groin revealed bilateral masses, hard and tender to palpation and bilaterally irreducible. Examination of his abdomen demonstrated generalised peritonitis.

Investigations showed a white cell count of $10 \times 10^9/L$ with a mild neutrophilia ($8.16 \times 10^9/L$). Given the degree of abdominal discomfort, a CT abdomen was performed that revealed large bilateral direct inguinal hernias, with the right containing a distended appendix, while both hernial orifices contained collections that extended up to liver.

The diagnosis of perforated appendicitis within an Amyand hernia was made preoperatively and the patient was booked for emergent operative management and commenced on intravenous antibiotics (IVABx).

The patient had a laparoscopic appendicectomy without inguinal hernia repair. Intraoperatively, there was purulent peritonitis throughout the entire peritoneal cavity. There were also large bilateral direct inguinal hernia defects, both containing purulent fluid, with the right containing an inflamed appendix adhered to the hernial sac. Both hernias were able to be reduced once the patient was under a general anaesthetic.

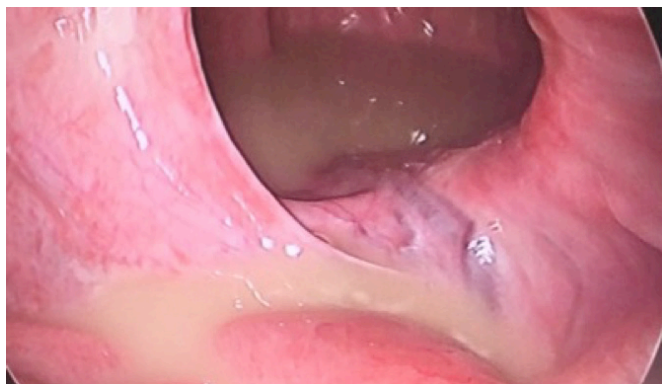


Figure 1. Laparoscopic view of abdomen, collection within the left hernial sac



Figure 2. Laparoscopic view of abdomen, inflamed appendix adhered to right hernial orifice

Postoperatively, the patient made a rapid recovery and was discharged on day 5 with a course of antibiotics. His histopathology revealed a well-differentiated GCC invading the muscularis propria to the serosal surface with involved margins. The patient went on to have a completion right hemi-colectomy and was referred to the oncologists for consideration of adjuvant therapy postoperatively.

Discussion

Although Rene Jacques Croissant de Garengot was the first to describe a caecal appendix within an inguinal hernial sac in 1731, it was the French surgeon to King George II of England, Claudius Amyand, who performed the first appendicectomy in this setting, and who subsequently gained eponymous rights to this hernia type.^{1,2,3} We now estimate that one percent of inguinal hernias contain the vermiform appendix, while 0.1 percent have an inflamed appendix, and just 0.01 percent contained a perforated appendix.^{4,5} The chances of acute appendicitis alone during one lifetime is eight percent.⁶ Traditionally, complicated Amyand hernias have been managed with open surgery, and most reports in the literature describe this approach; however, with the advent of laparoscopic surgery, this has become an increasingly utilised strategy.⁷ Appendiceal tumors are found in roughly one percent appendicectomies and are typically carcinoid tumors or adenocarcinomas. The GCC is a rare and aggressive tumor of the appendix that accounts for less than five percent of appendiceal tumors.⁸

Consistent in the literature with most cases of Amyand hernia are the challenges of diagnosis. Indeed, in a review article examining 50 cases of Amyand hernia, only one was diagnosed preoperatively.⁶ Often, Amyand hernia mimics the presentation of a strangulated inguinal hernia (i.e., a painful irreducible groin lump), although there are not often obstructive symptoms (e.g., abdominal disten-

tion, vomiting). The case presented here would agree with this assessment. The patient presented very much like a non-obstructed, incarcerated inguinal hernia, with only the generalised peritonitis and the bilateral irreducibility and tenderness being clues towards additional pathology necessitating CT scanning.

An often postulated mechanism of appendicitis within an Amyand hernia is based around strangulation of the appendix around the hernial neck causing distention, ischemia inflammation, and potentially perforation.^{1,3,8} In our case, there could also be potential for intraluminal obstruction from the GCC contributing to the distention, inflammation and perforation.

Due to the complexity surrounding the management of dual pathologies (hernia and appendix), a scoring system by Losanoff and Basson has been developed to guide management. Outlined in the accompanying table (Table 1), it essentially details the most appropriate type of hernia repair given the degree of inflammation.^{6,8} The original classification system calls for an open approach to management.^{6,8} However, in our case, the use of laparoscopic techniques allowed for good visualisation as well as adequate peritoneal lavage while avoiding the need for laparotomy.

Type of Amyand hernia	Key features	Appropriate surgical management
Type 1	Non-inflamed, normal appendix	Reduction/ Appendectomy + repair with mesh
Type 2	Acute appendicitis, contained inflammation within hernial sac	Appendectomy through open inguinal hernia approach with non-mesh repair
Type 3	Acute appendicitis with peritonitis extending beyond the hernial sac	Appendectomy through laparotomy, non-mesh hernia repair
Type 4	Acute appendicitis in association with other intra-abdominal pathology	Appendectomy and proceed as appropriate

Table 1. Losanoff and Basson's classification of Amyand hernia

An additional element to our case was the presence of a GCC within the appendix. GCCs are rare tumors of the appendix that often present with acute appendicitis, but can also present with vague abdominal pain/mass, rectal bleeding or intussusception.⁸ They are very aggressive, between 15 and 30 percent metastasis compared to two to five percent of typical appendiceal carcinoids.⁸ They often require further surgical management such as a right hemicolectomy with additional adjuvant therapy, and thus, a multidisciplinary approach is favored in this instance.⁸

Conclusion

A presentation of perforated appendicitis with generalised peritonitis inside an Amyand hernia and managed laparoscopically is a rare occurrence, made rarer still by the presence of a GCC. While in this case, the diagnosis was suspected prior to arriving in theater, difficulty in preoperative diagnosis is well-documented, and any suspicion of complication (as was the case here) should warrant consideration of a diagnostic laparoscopy. In this case, the use of CT imaging was very helpful with diagnosis, and the laparoscopic approach was effective in achieving appendectomy and source control for intra-abdominal sepsis.

Lesson Learned

A perforated appendix within an Amyand hernia is an exceptionally difficult preoperative diagnosis and often mimics an incarcerated inguinal hernia. Presentations of incarcerated inguinal hernia with atypical features should warrant consideration of this differential diagnosis. Traditionally, Amyand hernias have been managed with an open technique, but here, we demonstrate the advantages of a laparoscopic approach.

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