CASE REPORT

Amyand's hernia containing an appendiceal goblet cell carcinoid tumour

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SUMMARY

Amyand's hernia is a rare occurrence where the appendix is trapped within an inguinal hernia. Appendicitis within the hernia is even rarer. However, the presence of an appendiceal neoplasm in an inguinal hernia is almost unheard of with only two cases reported in the literature. We present an extremely rare case of an inflamed appendix within an Amyand's hernia, which was found to be a goblet cell type carcinoid tumour requiring further oncological resection and treatment.

inguinal region towards the scrotal sac, and attempts of reduction in the emergency department were unsuccessful.

INVESTIGATIONS

Initial investigations revealed a white cell count of $13.7 \times 10^9/L$ which predominantly consisted of neutrophils ($10.5 \times 10^9/L$). At this point, the diagnosis of a Richter's type hernia was made given the lack of obstructive bowel symptoms. Given that the patient was age-appropriate, a CT scan with oral

BACKGROUND

In 1735, Claudius Amyand performed the first ever recorded appendicectomy in an 11-year-old boy presenting with a right inguinal hernia. Intraoperatively, he found the appendix containing a stone-encrusted pin sitting within the inguinal hernia sac.¹ Since then numerous cases of the eponymously named Amyand's hernia have been described in the literature. Appendiceal neoplasms are also very rare, often present as appendicitis and found incidentally in 1% of appendicectomy specimens.² We present an extremely rare case of an inflamed adenocarcinoid appendiceal tumour within an Amyand's hernia which has only been reported twice in the literature.

CASE PRESENTATION

A 67-year-old man was referred to the acute general surgical unit with a painful right-sided groin lump resembling an incarcerated hernia despite normal bowel motions and absence of nausea or vomiting. He described a sudden appearance of the right groin lump 3 days prior to presentation while lifting heavy objects. Attempts to reducing the lump by himself and his local general practitioner were unsuccessful and subsequent worsening of his pain with onset of erythema prompted his attendance to the emergency department.

He is otherwise in very good health with a medical history of type II diabetes mellitus and hypertension, both of which are well controlled by oral medications. Besides having had a total knee replacement, he has not had any previous surgical procedures.

On examination, he appeared well although in obvious discomfort and promptly indicates the right groin being the source of his pain. His vital signs were normal with a soft, non-tender abdomen and active bowel sounds on auscultation. The tender lump appeared to arise from the right

Figure 1 Abdominal CT axial views showing an inflamed appendix within a right inguinal (Amyand's) hernia.

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Figure 2 Abdominal CT coronal view showing an inflamed appendix exiting the internal ring into a right inguinal sac.

Figure 3 A swollen hernia sac containing intraperitoneal structures during open right inguinal hernia repair.

contrast was ordered which showed an indirect incarcerated inguinal hernia containing an inflamed appendix without evidence of perforation or collection (figures 1 and 2).

DIFFERENTIAL DIAGNOSIS

- Richter's hernia
- Incarcerated hernia containing omentum or small bowel
- Femoral hernia

TREATMENT

As his vital signs remained stable and his pain manageable with oral analgesia, he was admitted as an inpatient overnight on intravenous antibiotics. He proceeded to the operating theatre in the morning where an oblique groin incision was made so as to access the inguinal hernia. Intraoperative findings were consistent with an incarcerated right inguinal hernia containing an inflamed appendix without evidence of perforation (figures 3 and 4). An appendicectomy was performed through the groin incision and a modified Bassini's repair was performed without

Figure 4 An inflamed appendix identified on entering the hernia sac. No perforation or pus was identified.

mesh. The patient had an uneventful recovery from surgery and was discharged on day four.

OUTCOME AND FOLLOW-UP

Histopathological examination of the specimen revealed a goblet cell carcinoid (GCC) tumour measuring 10 mm at the proximal stump extending deeply into the muscularis propria involving the proximal resection margin. Perineural invasion was detected with a Ki-67 rate of 60%. Changes consistent with acute appendicitis were also seen. A postoperative staging chest, abdomen and pelvis CT demonstrated a soft tissue nodule at the caecum, presumably the stump of the appendix, with positive uptake on a subsequent fluorodeoxyglucose-positron emission tomography (FDG-PET) scan. There was no other evidence of distant disease. A colonoscopy performed was essentially normal with an incidental finding of a hyperplastic rectal polyp.

The patient was referred to a local specialised cancer centre where he subsequently went on to have an open right hemicolectomy, cytoreduction and hyperthermic intraperitoneal chemotherapy (HIPEC) 5 months after his initial presentation. Histopathology examination of the resected caecal nodule was confirmed to contain a 4 mm focus of residual GCC with clear surgical margins and no other diseases were identified. He continues to visit his colorectal surgeon and medical oncologist, and was scheduled to have a repeat CT and a repeat colonoscopy 12 months after his surgery. Of note, he did not proceed to having adjuvant chemotherapy as there is currently not enough evidence to suggest this may be necessary given surgical clearance of disease.

DISCUSSION

An inguinal hernia may contain intraperitoneal structures such as omentum, bowel and a Meckel's diverticulum (Littre's hernia). Less commonly herniation of extraperitoneal structures such as the ureter and bladder can occur. The finding of an appendix in an inguinal hernia is rare and was first described in 1735 by Claudius Amyand, Sergeant Surgeon to King George II of England, who successfully performed an appendicectomy in an 11-year-old boy who had a perforated appendix.¹

The reported incidence of Amyand's hernia is varied ranging from 0.19% to 1.7% of hernias, while the incidence of appendicitis occurring within an inguinal hernia is estimated at 0.07–0.13% of inflamed appendices.^{3 4} The relationship between

hernial incarceration and inflammation of the appendix is not clear; however, Weber *et al*⁵ have suggested that inflammatory swelling may lead to progressive incarceration, causing impaired blood supply, ischaemia and bacterial overgrowth within the faeces-containing appendix.

Management of an Amyand's hernia is dependent on the condition of the appendix as assessed intraoperatively (table 1). In the case of a normal appendix, a simple mesh (Lichtenstein's) repair can be performed, with or without an appendectomy based on age and surgeon's preference. In our case, a type 2 Amyand's hernia based on Losanoff and Basson's classification, acute appendicitis warrants an appendectomy via a hernia repair incision followed by primary hernia repair with no mesh. A Bassini's repair is favoured where the defect is closed by suturing the conjoint tendon to the reflected part of the inguinal ligament.⁶ Milanchi *et al*⁷ go one step further and recommend a laparoscopic appendectomy followed by an open hernia repair without prosthetic mesh if appendicitis is diagnosed preoperatively by CT imaging.

The evidence for non-mesh repair in the setting of potential contamination is not conclusive. A systematic review by Hentati *et al* looking at repair techniques in strangulated inguinal hernias, with and without small-bowel resections, reported a lower rate of wound infection as well as hernia recurrence in mesh repairs compared to non-mesh repairs.⁸ Despite this, in the case of Amyand's hernias, most authors recommend appendectomy and hernia repair without prosthesis implantation due to contamination of the operative field inherent with surgery to the appendix.

The other unique facet to our case is the histopathological finding of an appendiceal neoplasm. Carcinoid tumours of the appendix occur in one in 300 of appendectomies.⁹ GCC tumours, a rare neoplasm of the appendix, have been described as a separate entity from adenocarcinoma and carcinoid tumours. It was first recognised in 1974 and has been described under a number of different names such as mucinous carcinoid, intermediate type carcinoid and crypt cell carcinoma.¹⁰ GCCs account for <5% of all primary appendiceal tumours¹¹ with equal sex predilection and higher incidence among Caucasians.⁹ While diagnosis is made on histological grounds, most patients present with acute appendicitis as tumour cells proliferate sparsely leading to fibrous thickening, contraction and obstruction of the appendiceal lumen. Other less common presentations include intussusception, per rectal bleeding, intermittent lower abdominal pain, secondary genitourinary complication and presence of a palpable mass, often as Krukenberg tumours in women.¹² A GCC tumour is identified histologically by the presence of individual glands separated by smooth muscle, with lining cells containing intracytoplasmic mucin.¹³

GCCs originate from a pluripotent cell that differentiates into mucinous and neuroendocrine cells and have an unpredictable natural history.¹⁴ Although previously considered a variant of carcinoid tumours, current literature suggest that GCCs are a distinct tumour more appropriately termed 'crypt cell carcinoma' because it reflects its more aggressive clinical behaviour and have greater similarity to adenocarcinoma rather than carcinoid.¹⁵ GCC tumours metastasise in 15–30% of cases compared with 2–5% in appendiceal carcinoids,¹⁶ commonly to the ovaries but can also occur in the pelvis, abdominal cavity, rib, vertebra and lymph nodes through lymphatic, transcoelomic and intraperitoneal invasion.¹⁷

Management of appendiceal GCC neoplasms is essentially surgical regardless of the site of tumour within the appendix.¹⁸ Even though GCC has been shown to be more aggressive than carcinoid tumours, prognosis is generally good and is thought to be between carcinoid and well-differentiated appendiceal adenocarcinoma.¹⁹ Peritoneal spread from GCC is as invasive as peritoneal disease from colorectal adenocarcinoma origin, hence such cases warrant complete surgical removal and should be considered for intraperitoneal chemotherapy.²⁰ Goede *et al* have described acceptable indications in an attempt to identify patients with appendiceal carcinoid tumours who are suitable candidates for right hemicolectomy (box 1).¹¹ In our case, the diagnosis of GCC with positive margins and residual disease on staging CT and PET scans post appendectomy is a strong indication for a right hemicolectomy, cytoreduction and intraperitoneal chemotherapy.

In terms of adjuvant therapy, there is limited evidence to support chemotherapy in GCC tumours after complete surgical resection. Adjuvant chemotherapy has shown a survival benefit only in a subgroup of patients with ovarian Krukenberg tumours secondary to appendiceal GCC.²¹

Presentation of a GCC tumour within an Amyand's hernia is certainly rare, and has only been described once in the literature. Wu *et al*²² described a 62-year-old man who underwent an emergency open repair for an acutely tender, clinically incarcerated right inguinal hernia. An incidental finding of an inflamed appendix warranted an appendectomy and a primary hernia repair with no mesh. Histopathology examination showed an adenocarcinoid tumour of goblet cell type measuring 25 mm in length and 10 mm in diameter and as clearly indicated a right hemicolectomy was performed.

Table 1 Losanoff and Basson's classification of Amyand's hernia⁶

Type of hernia	Salient features	Surgical management
Type 1	Normal appendix	Reduction or appendectomy (depending on age), mesh hernioplasty
Type 2	Acute appendicitis, localised in sac	Appendectomy through hernia, endogenous repair
Type 3	Acute appendicitis, peritonitis	Appendectomy through laparotomy, endogenous repair
Type 4	Acute appendicitis, other abdominal pathology	Appendectomy, diagnostic workup and other procedures as appropriate

Box 1 Goede *et al*'s indications for right hemicolectomy in patients with appendiceal carcinoid tumour¹¹

Acceptable indications for right hemicolectomy

- Lesions larger than 2 cm in diameter
- Histological evidence of mesoappendiceal invasion
- Tumours at the base of the appendix with positive margins or involvement of the caecum
- High-grade malignant carcinoids, including those with high mitotic index and Ki-67 levels
- Goblet cell adenocarcinoid

Relative indications for right hemicolectomy

- Isolated finding of angioinvasion
- Tumours at the base of the appendix, 1–2 cm in diameter, with clear margins
- Mucin-producing tumours

A case report by Elbanna *et al* describes an 81-year-old man who presented with incarcerated right inguinal hernia containing distal ileum, caecum and appendix on CT imaging. Intraoperatively, only an inflamed appendix was found with no bowel ischaemia, hence an appendicectomy and a modified Bassini's hernia repair were performed. Histopathological examination revealed a 15 mm well-differentiated, low-grade carcinoid tumour of the appendix tip with invasion down to subserosa. In the absence of invasion into peritoneum or mesoappendix, lymphovascular invasion and involvement of resection margins, further resection or treatment was not warranted.²³

The incidental finding of an appendiceal carcinoid tumour in a patient with an Amyand's hernia is extremely rare, with only few cases described in the literature. To be highly suspicious of this occurrence in every inguinal hernia repair is unreasonable; however, practising good surgical principles such as the prudent use of prosthetic mesh is recommended. Ultimately, the diagnosis of an appendiceal neoplasm is made after histopathological examination. Hence, it is appropriate to manage such a scenario as two separate conditions, first intraoperatively and then post-operatively, preferably in a multidisciplinary approach.

Learning points

- ▶ Amyand's hernia is a rare presentation of an inguinal hernia, in which the appendix is incarcerated within the hernia sac.
- ▶ Surgery is diagnostic as well as therapeutic, and the choice of operative technique depends on the condition of the appendix. Use of prosthetic mesh should be avoided in an inflamed appendix where there is likely contamination.
- ▶ Appendiceal tumours are rare, usually present as appendicitis, and are diagnosed on histopathological examination.
- ▶ The two conditions pose diagnostic and management challenges and in the event of having both concurrently, it is appropriate to manage them separately.

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