

A rare presentation of caviar appendix in an Amyand's hernia

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ABSTRACT

Amyand's hernia is a rare condition where the appendix is found within the sac of an inguinal hernia. The occurrence of an appendicular neoplasm within such hernia is even rarer. We highlight the case of a 70-year-old male who presented with an appendicular neoplasm masquerading as a strangulated inguinal hernia. The rarity and heterogenic clinico-pathological presentation of an appendiceal neoplasm renders it potentially lethal in the hands of an unassuming hernia surgeon.

Keywords: Appendicectomy, hernia, mucinous cystadenoma, mucocoele, mesh repair

INTRODUCTION

Mucinous cystadenoma of the appendix is a rare clinical entity accounting for less than 0.3% of resected appendectomy specimens and is commonly referred to as "mucocoele" of appendix.¹ The intraluminal mucin leads to gross cystic dilatation of the appendix followed by obstruction. Although it is largely an incidental intraoperative finding during appendicectomy, it should be considered as a differential diagnosis in case of a palpable right iliac fossa mass. Myxoglobulosis is seen in less than 0.5% of reported cases of mucinous cystadenoma.² It presents as a distended appendix with subcentimetre fish-egg like mucinous spheres occupying its lumen, hence the name caviar appendix. This case high-

lights a rare presentation of myxoglobulosis with underlying appendicular mucinous cystadenoma presenting as an Amyand's hernia.

CASE REPORT

A 70-year-old male presented to the Emergency Department with complaint of painful right inguinal swelling for the past 24 hours. Examination revealed a tender non-pulsatile swelling with increased warmth and redness of the overlying skin. He was admitted and subjected to emergency open hernia repair for the diagnosis of strangulated inguinal hernia. At surgery, an inflamed appendix was found within the hernial sac. Appendicectomy and mesh free hernia repair was performed. Large amount of mucinous pearl like spheres measuring 0.2 cm to 0.4 cm in diameter were noted to extrude from the appendicular lumen following gross examination of the resected

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specimen (Fig. 1). There was no intraperitoneal spillage of the mucinous substance. Histopathological examination revealed an enlarged appendix measuring 5.0 x 2.0 x 1.5 cm with multiple subcentimetre viscid mucus balls. Microscopic examination showed epithelium of crowded columnar cells, hyperchromatic nuclei with large amount of apical mucin noted in areas of papillary configuration. Surgical margins were clear with no evidence of malignancy.

Final diagnosis was reported as myxoglobulosis with underlying mucinous cystadenoma of the appendix. Postoperative recovery was uneventful and patient was discharged home, three days later. Surveillance colonoscopy at one year was unremarkable and the patient has been well since.

DISCUSSION

Claudius Amyand first described Amyand’s hernia in 1735. It refers to the presence of a vermiform appendix within the sac of an inguinal hernia. ⁶ It is rare and accounts for less than 1% of adult inguinal hernias. The incidence of an inflamed appendix within an Amyand’s hernia have been reported to 0.1%. ⁷ Preoperative diagnosis is often not possible and is often made incidentally during emergency surgery for strangulated hernias. Decision to perform appendicectomy and type of hernia repair depends on the intra-



Fig. 1: Intraoperative photograph of a grossly inflamed and distended appendectomy specimen with fish egg like mucin filled spheres extruding from the appendicular lumen.

operative finding, guided by the Losanoff and Basson’s criteria (Table 1). Appendicectomy through the hernial orifice is reserved for Amyand’s hernia types 1 and 2, while types 3 and 4 warrants an exploratory laparotomy due to the risk of peritoneal contamination. Mesh repair of the inguinal hernia is generally avoided but may be considered in the absence of appendiceal inflammation such as in type 1 Amyand’s hernia. The non-inflamed appendix in a type 1 Amyand’s hernia may be reduced manually into the peritoneal cavity avoiding the need for an appendicectomy thus enabling mesh repair of the hernial orifice. However, appendicectomy is recommended in

Table 1: Pathological types of Amyand’s hernia and respective management. ⁸

Types of Hernia	1	2	3	4
Salient Feature	Normal appendix	Acute appendicitis localized in the sac	Acute appendicitis, peritonitis	Acute appendicitis, other abdominal pathology
Surgical Management	Reduction or appendicectomy (depending on age), mesh hernioplasty	Appendicectomy through hernia, endogenous repair	Appendicectomy through laparotomy, endogenous repair	Appendicectomy, diagnostic workup and other procedures as appropriate

the younger age group because of higher lifetime risk in developing appendicitis.⁸

Appendicular mucocoele is classified into neoplastic and non-neoplastic types. The non-neoplastic variant is caused by intraluminal obstruction secondary to increased mucin production, while the neoplastic variant may be triggered by benign or malignant epithelial changes. Mucinous cystadenoma is a benign neoplastic variant of mucocoele, and mucinous cyst adenocarcinoma is the malignant neoplastic variant.^{9, 10} Myxoglobulosis is a rare manifestation of the appendicular mucocoele and was described by Latham in 1897, nearly 55 years following Rokitanski's report of the first case of appendicular mucocoele.^{2, 11} It has a female preponderance with the highest incidence reported in the sixth decade of life.¹² Infective, inflammatory and mucinous nidus formations have been implicated as causative factors for development of the fish egg like spherical structures similar to myxoglobulosis. Despite the various hypotheses, the exact pathophysiology remains a mystery.⁹ Careful and complete resection of the appendicular mucocoele is advocated to prevent the development of pseudomyxoma peritonei. Pseudomyxoma peritonei (PMP) is a dreaded complication of appendicular mucocoele. It occurs within 2 to 15 years following primary surgery and commonly presents as intestinal obstruction. Largely a benign disease, PMP behaves in a malignant manner when left untreated. Long-term survival outcome of PMP is poor with 10 year overall survival reported to be 10% to 30%.¹³ Death is often slow and due to terminal starvation. Combined approach of cytoreductive surgery and perioperative loco-regional chemotherapy has been shown to improve overall survival in patients

with PMP.¹⁴

Myxoglobulosis and mucocoele of the appendix is commonly reported as an incidental intraoperative finding during emergency appendicectomy. Unless, there is evidence of a palpable right iliac fossa mass noted during preoperative examination, radiological assessment is not routinely performed. In case of a palpable mass, a plain abdominal radiograph may be able to demonstrate calcification within the mucinous spheres. Complementary use of ultrasound and contrast enhanced computed tomography (CT) of the abdomen increases the specificity of detection and diagnostic accuracy to almost 90%.¹⁵ CT appearance of mucocoele has been described as round, encapsulated, low-density cystic mass that communicates with the caecum.¹⁶ Varying mass wall thicknesses has not been shown to correspond to any malignancy. However, contrast-enhancing nodules within the appendix is suggestive of mucinous cystadenocarcinoma.¹⁷

Standard appendectomy is often sufficient for appendicular mucocoele with emphasis on meticulous resection, avoidance of intraperitoneal spillage of mucinous material and reducing risk of carcinomatous peritonei. Some authors recommend conversion of laparoscopic appendectomy to open appendectomy for resection of appendicular mucocoele.¹⁷ Right hemicolectomy should be considered in case of perforation, iatrogenic rupture during mobilisation, unhealthy appendicular base, caecal involvement, or suspicion of malignancy.^{9, 18} In case of unfavourable histopathology result following appendectomy, a completion right hemicolectomy is advocated.^{9, 14} Yearly surveillance colonoscopy and moni-

toring with tumour markers has been recommended for appendicular mucocoele due to its association with adenocarcinoma of the colon via the adenoma carcinoma pathway as well as risk of carcinomatosis peritonei.²⁰

When dealing with a culmination of rare pathologies, the best management strategy is to adopt an individualised approach for each clinical entity. Decision between standard appendectomy and right hemicolectomy for appendiceal mucocoele should be made based on intraoperative finding. In this case, the decision to proceed with an appendicectomy followed by mesh-free repair of the Amyand's hernia was based on the Losanoff and Bosson's criteria for Type 2 Amyand's hernia.⁸ The argument for and against the use of mesh during repair of Amyand's hernia remains controversial. In our institution, we avoid mesh fixation when dealing with evidence of local infection, perforation or strangulation. Application of mesh in such circumstance has been associated with a high incidence of mesh infection, surgical site infection, hernia recurrence and fistula formation.^{7, 21}

In conclusion, we report a rare case of a mucinous appendiceal adenocarcinoma presenting as an Amyand's hernia. Amyand's hernia is rare and is often not diagnosed until surgery. Management of the hernia involves judicious intra-operative judgment, taking into consideration the extent of appendiceal inflammation, co-existence of neoplasm and associated intra abdominal pathology. Intraoperative handling of a suspected appendiceal neoplasm is of utmost importance to prevent catastrophic complications that can be otherwise avoided.

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