A RARE CASE OF LOCALLY ADVANCED MUCINOUS ADENOCARCINOMA OF THE APPENDIX CONFINED IN THE INGUINAL CANAL: CLINICAL PRESENTATION AND OPERATIVE MANAGEMENT

Michalis Tsimaras*, Vasiliki Ziozia*, Stefanos Koffas*, Napoleon Xirokostas*, Dimitrios Filippou**, Panagiotis Skandalakis** and Vasileios Komporozos△

*4th Surgical Department, Evaggelismos General Hospital, Athens, Greece Department of Anatomy and Surgical Anatomy, Medical School, National and Kapodestrian University of Athens, Greece.; **Department of Anatomy and Surgical Anatomy, Medical School, National and Kapodestrian University of Athens, Greece.; △4th Surgical Department, Evaggelismos General Hospital, Athens, Greece.

ABSTRACT Background: An Amyand’s hernia is a right inguinal hernia that contains the appendix in the hernia sac. Mucinous adenocarcinoma of the vermiform appendix is a rare neoplasm commonly discovered through the histology report of a presumed appendicitis specimen or as the cause of pseudomyxoma peritonei. It is very rare for the two entities to co-exist and present as an incarcerated right inguinal hernia.

Case Summary: We report a rare case of a mucinous appendiceal adenocarcinoma that ruptured towards the confined anatomical space of the inguinal canal. Initial presentation of the patient was with small bowel obstruction, and clinical examination revealed a co-existing right inguinal hernia. Exploratory laparotomy findings revealed an appendiceal mucinous adenocarcinoma is invading the adjacent bowel loops and identified an encapsulated collection of mucinous material extended into the inguinal canal and lateral abdominal wall. He underwent a right hemicolectomy with excision of the adjacent small bowel loops and the affected abdominal wall.

Conclusion: As the patient presented with a long-standing history of inconsistent symptoms, he underwent thorough preoperative investigations which revealed the true extent of the underlying disease and led to appropriate surgical intervention. The fact that a tumour extended towards the confined space of the inguinal canal prevented the development of pseudomyxoma peritonei. It remains to be examined if such cases, where the mucinous material is contained bare a favourable prognosis.

KEYWORDS mucinous, appendiceal carcinoma, Amyand’s hernia, pseudomyxoma peritonei

Introduction

An Amyand’s hernia is a right inguinal hernia that contains the appendix in the hernia sac. Mucinous adenocarcinoma of the vermiform appendix is a rare neoplasm commonly discovered through the histology report of a presumed appendicitis specimen or as the cause of pseudomyxoma peritonei. It is very rare for the two entities to co-exist and present as an incarcerated right inguinal hernia.

We aim to present a case of a rare clinical presentation of mucinous adenocarcinoma of the vermiform appendix; initially presenting with symptoms consistent with an indirect inguinal hernia causing small bowel obstruction. We elaborate on clinical
presentation and appropriate preoperative investigations until optimal surgical treatment.

**Case Report**

A 53-year-old patient presented to our Surgical Accident and Emergency Department with a 48-hour history of diffuse abdominal pain and vomiting. His past medical history was unremarkable. He reported similar episodes of colicky abdominal pain and distension with associated constipation over the past two months that had resolved spontaneously.

Bowel sounds were increased consistently with an obstructive picture. Clinical examination revealed a palpable mass in the right inguinal region. There was no cough impulse. It should be noted that, according to the patient, that “swelling” had been present for the past six months, but as there was no associated pain, he did not investigate it further. CT scan of the patient’s abdomen depicted the distended loops of small bowel and a soft tissue mass at the terminal ileum, which appeared to be the cause of the obstruction. It also noted a 3.7cm encapsulated fluid collection at the lateral abdominal wall at the same level as the ileocaecal valve. No other abnormalities were detected.

Imaging findings were further investigated with colonoscopy; a caecal mass was visualised occupying approximately one-third of the bowel lumen. Biopsy of the above lesion revealed villus appearance of the surface epithelium with underlying oedema, recent haemorrhage and dysplastic features. It raised the suspicion that the above histologic features could be indicative of malignancy in the adjacent area from where the biopsy was taken. Therefore, we proceeded to exploratory laparotomy.

Caecum with adjacent small bowel loops formed a mass that was adherent to the internal inguinal ring. That was the point where the CT described fluid collection was starting, expanding towards the abdominal wall. The appendix was not identified. Frozen section sent intraoperatively from the above region showed pools of mucin with glandular formations suspicious of malignancy. The patient underwent a right hemicolectomy with excision of the adjacent small bowel loops and the affected abdominal wall. All macroscopically evident mucinous material was meticulously removed from the abdominal cavity. Final histology was reported as high grade appendiceal mucinous adenocarcinoma with the extended invasion of the mesentery and focal invasion of the adjacent abdominal wall.

He recovered well and was discharged on the eighth postoperative day. He was referred to our Oncology department for further treatment. The decision was for him to receive Folfox adjuvant chemotherapy for six months. He is now fifteen months post-surgery and remains asymptomatic and clinically well. His follow up CT scan depicts a 2x3 cm mass at the inguinal region for which our MDT advised on active surveillance.

**Discussion**

An inguinal hernia is a common surgical disease and repair of an inguinal hernia is one of the most common operations performed by general surgeons. Presence of the appendix vermiformis in the sac of an inguinal hernia is referred to as Amyand’s hernia. This variation is encountered in less than 1% of all adult inguinal hernia cases and 0.2% of appendicitis cases. The incidence of a normal appendix in the sac of an inguinal hernia is approximately 0.13%, and it is even more uncommon to encounter an inflamed appendix in an inguinal hernia sac.

The management of the appendix when present in an Amyand’s hernia is debatable. Losanoff and Basson developed a classification system to be used as a treatment guideline for Amyand’s hernias. They specify four types of Amyand hernias. In type I, where there is no evidence of inflammation, a mesh repair is recommended. In type II Amyand’s hernia, there is inflammation of the appendix confined within the hernia sac, and, thus, the proposed operation is appendectomy and endogenous repair without mesh placement. Type III has defined as the presence of co-existing peritonitis, and, therefore, laparotomy,
appendicectomy and a primary hernia repair are required. In the last category, type IV, there is evidence of underlying abdominal pathology. In this case, apart from appendicectomy and hernia repair, an appropriate diagnostic and treatment sequence of the underlying pathology is warranted. Currently, the most popular approach to inguinal hernia repair is a Lichtenstein mesh repair with the trend favoring laparoscopic mesh repair. Routinely, the hernia sac is neither explored nor ligated; after detachment of the hernia sac from the neighbouring structures, the sac is reduced intact in the peritoneal cavity. In some cases, however, a macroscopic abnormality is suspected in the hernia sac or its contents, and the sac should be explored. It has been suggested that if a gross abnormality is seen, microscopic examination of the abnormality should be performed so as not to miss occult metastatic cancer.

There have been other cases of rare intra-abdominal conditions presenting as incarcerated inguinal hernias; especially in elderly patients. Lymphomas, retroperitoneal sarcomas and other pathological entities as lymph nodes, psoas abscesses, femoral aneurysm and pseudoaneurysm, dermoid and epidermoid cysts, all have been described as inguinal masses.

The accidental discovery of a neoplasm in the inguinal hernia sac also remains a rare during elective repair. Malignant masses in inguinal hernias appear in less than 0.5% of excised sacs. The majority of intrasaccular neoplasms are colonic, and the most commonly reported site is the sigmoid colon, usually in a left inguinal hernia. Primary tumour of the inguinal canal, such as Sertoli-Leydig cell tumour, and metastasis of peritoneal tumours to the inguinal hernia sac are occasionally reported. These metastatic tumours typically originate from the colon, ovary, appendix, prostate and the gallbladder.

Coexistence of an Amyand’s hernia and neoplasia is quite rare. According to a report published by the National Cancer Institute, appendiceal neoplasms constitute a rare entity of gastrointestinal neoplasms, accounting for approximately 0.4% of gastrointestinal tumours.

The majority of them are histological findings of appendicectomies performed for presumed acute appendicitis. In the literature, the reported incidence ranges from 0.9% to 5% and carcinoid of the appendix is the predominant histologic type (66-80%). However, a recent review by SEER(Surveillance, Epidemiology and End Results) for the period from 1973 to 2007 identifies the mucinous adenocarcinoma as the most common type with a percentage of 37%. Most common clinical presentation of appendiceal adenocarcinomas is that of acute appendicitis, ascites, and intra-abdominal mass or non-specific abdominal pain. In less than 20% of cases, the tumour is an incidental finding.

Adenocarcinomas of the appendix have three histological subtypes: mucinous adenocarcinoma, with abundant mucin production, colonic type adenocarcinoma, with the similar clinical course and signet ring cell adenocarcinoma, which bears a worse prognosis.

As far as the colonic type is concerned, the primary form of spread is a local invasion to adjacent structures. Controversy exists regarding the lymphatic spread of the disease, with the majority of the reports stating that the colonic type has a greater tendency for lymphatic spread. Mucinous adenocarcinoma, however, tends to diffusely sprawl in the peritoneal cavity through mucin pools, resulting in peritoneal mucinous carcinomatosis. In the case presented above, the mucinous material was sooner confined in the right inguinal fossa. During disease progression, small bowel obstruction developed due to tumour invasion in the adjacent bowel loops and an encapsulated collection of mucinous material extended into the inguinal canal and lateral abdominal wall. It was feasible for all macroscopic mucinous material to be removed from the abdominal cavity.

To our knowledge, there has been a reported case where, during primary hernia repair, a mucinous tumour of unknown origin was incidentally discovered. Similarly, the mucinous material was locally removed; the patient proceeded to adjuvant treatment and remained free of disease recurrence during follow up. Pseudomyxoma peritonei (PMP) is a rare, progressive disease of unknown origin. The incidence is estimated at 1/2-10000 per year. The primary tumour site is usually discovered in the appendix or- in case of women- in ovaries, appearing as tumours of low malignancy. Making an accurate diagnosis causes difficulties—symptoms tend to be misleading, suggesting more frequent pathologicals of the abdominal cavity. It is also not rare that the patient is for a long time asymptomatic.

Although optimum treatment is debatable, most expert opinion favours extensive surgical debulking with or without adjuvant therapy. Today, the treatment of peritoneal surface malignancies (PSM) by cytoreductive surgery (CRS) combined with heated intraperitoneal chemotherapy (HIPEC) is the preferred treatment option and has shown promising oncological outcomes.

The presence of PSM in the groin may be the result of a pre-existing inguinal hernia or, if it originates from the ovary, the result of regional lymphatic spread. Tumour deposits in the inguinal canal may be identified on preoperative imaging as well as at the time of inguinal surgery and should be considered in surgical planning. In all cases, complete excision is necessary. However, it has been recently suggested that perfusion of the inguinal canal during HIPEC should be considered, as part of the adjuvant treatment.

Conclusion

The presence of an underlying malignancy in the inguinal canal, however rare, should be taken into account when the symptoms are not typical of a hernia, and the patient reports similar longstanding symptoms. A high index of suspicion will lead to an adequate surgical resection through a midline rather than a groin incision. It remains to be examined if such cases, where the inguinal canal contains the mucinous material and lateral abdominal wall bare a favourable prognosis.

Competing Interests

None

Funding

None

References
