

ADENOCARCINOMA OF THE APPENDIX - A CASE REPORT AND REVIEW OF THE LITERATURE**James Lucocq, Natalia Torres De Araujo Albuquerque and Darren J. Porter***

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ABSTRACT

Neoplasms of the vermiform appendix are rare and may be encountered at any elective or emergency operation. It is estimated that 50% of appendicular neoplasms present as appendicitis and are diagnosed on pathologic examination of the surgical specimen. The incidence is 0.4% to 1% of gastrointestinal malignant neoplasms. Carcinoid tumors are the most common primary tumor identified in the appendix. Adenocarcinoma of the appendix is very rare and occurs at a frequency of 0.08% to 0.1% of all appendectomies. In this study, we report a case of appendiceal adenocarcinoma diagnosed histologically following a laparoscopic appendectomy and review the literature on this rare pathology.

KEYWORDS: Appendectomy, Right hemicolectomy, Appendiceal adenocarcinoma, Carcinoid tumours, Pseudomyxoma peritonei.

INTRODUCTION

Adenocarcinoma of the vermiform appendix is a rare disease that was first described by Berger in 1882.^[1] It occurs in 0.1% to 0.2% of all appendectomies with an incidence of 0.2 per 100000 per year. This condition is most common in the 6th to 7th decades of life and has a slight male predominance.^[2]

The aetiology of appendiceal adenocarcinoma is unclear but there is an association with neoplasia at other locations in the colon. 75% of cases of appendiceal adenocarcinoma present with clinical symptoms but in 25% it is an incidental finding. The most common symptoms include acute pain in the right lower quadrant with fever and anorexia.^[3] If an unexpected appendiceal mass is encountered during laparoscopy an appendectomy should be performed with frozen-section analysis of the mass.^[4] If an adenocarcinoma is present, the patient should be discussed at the colorectal multi-disciplinary team (MDT) meeting where appropriate pre-operative staging and management will be discussed.^[5]

In this case report we present a case of an adenocarcinoma of the appendix diagnosed by histology following a laparoscopic appendectomy, and review the current literature as to the optimal investigation and management of these rare cases.

CASE REPORT

A 51-year-old male with no significant past medical or surgical history was admitted with a three-day history of severe lower abdominal pain, fever, nausea and anorexia. On admission observations were normal and the patient was afebrile. This patient appeared systemically well; however on examination, he had localised tenderness in the right iliac fossa, most pronounced over McBurney's point with rebound guarding. Admission bloods were normal with the exception of an elevated C-reactive protein of 150mg/L. This patient was diagnosed with acute appendicitis and intravenous antibiotics were commenced. A Computerised Tomography (CT) scan of the abdomen and pelvis was requested given the patient's age. This demonstrated a distended appendix measuring 11mm in diameter with surrounding fat stranding and prominent regional lymph nodes, in keeping with acute appendicitis (Figure 1)



Figure 1: Axial CT scan demonstrating a distended appendix with surrounding fat stranding and prominent regional lymph nodes, suggestive of acute appendicitis.

This patient underwent an emergency laparoscopic appendicectomy. The appendix had perforated and was adherent to an erythematous segment of distal ileum. Despite some difficulty in dissection the appendicectomy was performed laparoscopically, the appendix was sent to histopathology for analysis, and a thorough intraperitoneal lavage was performed. This patient made an unremarkable recovery and was discharged home on day three post-operatively.

Histopathological analysis demonstrated an adenocarcinoma involving the base of the appendix (Figure 2). This patient was discussed at the colorectal MDT meeting and thereafter underwent a colonoscopy

and a staging CT thorax. The staging CT scan did not show any evidence of metastatic disease. The colonoscopy demonstrated a 3mm polyp at the ano-rectal margin and a polypectomy was performed. The remainder of the colonoscopy was normal (Figure 3). The patient was re-discussed at the colorectal MDT meeting following full staging and subsequently underwent a laparoscopic right hemicolectomy with extracorporeal side-to-side stapled anastomosis. The patient had an uncomplicated post-operative recovery and was discharged home on day five post-operatively.

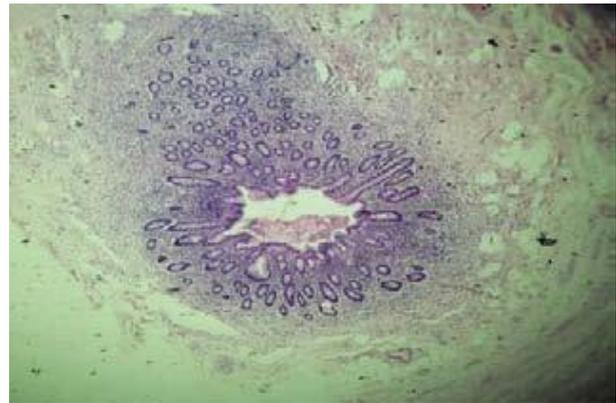


Figure 2: Histopathological examination of the specimen demonstrating an adenocarcinoma of the appendix. Immunohistochemistry shows negativity for chromogranin A.

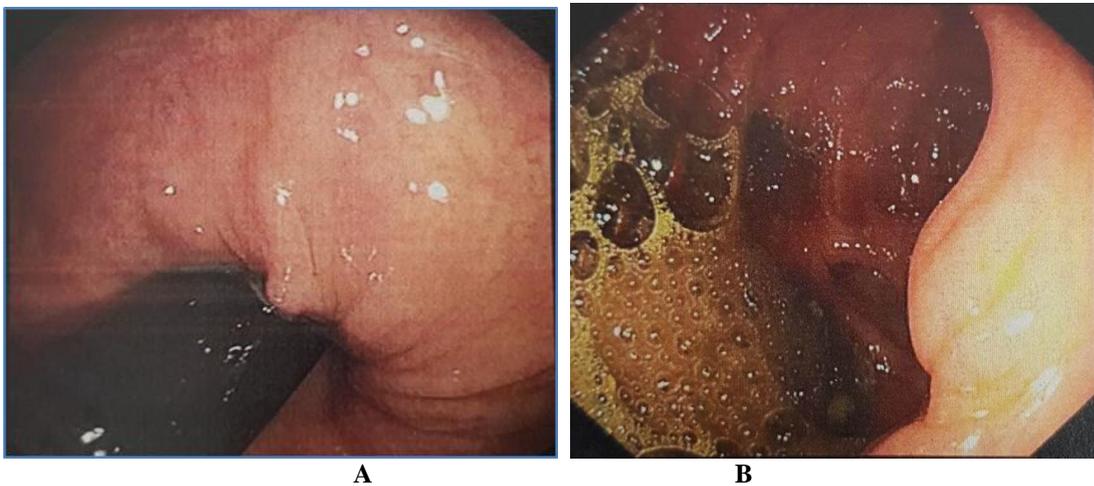


Figure 3: Colonoscopy demonstrating a small polyp at the ano-rectal junction (A) and a normal caecum with ileo-caecal valve (B).

DISCUSSION

Primary adenocarcinoma of the vermiform appendix is a rare disease that was first described by Berger in 1882.^[1] It occurs in 0.1 - 0.2 % of appendicectomies with an incidence of 0.2 per 100000 per year. This condition is most common in the 6th to 7th decades of life and has a slight male predominance.^[2]

The aetiology of appendiceal adenocarcinoma is unclear but there is an association with neoplasia at other locations in the colon. The majority of appendiceal tumours are carcinoids, whilst the remaining 10-20% are mucinous cyst-adenocarcinomas, adenocarcinomas, lymphosarcomas, paragangliomas, or granular-cell tumours.^[2]

Seventy five percent of cases of appendiceal adenocarcinoma present with clinical symptoms but in 25% it is an incidental finding. The most common symptoms include acute pain in the right lower quadrant with fever and anorexia.^[3] If an unexpected appendiceal mass is encountered during laparoscopy an appendectomy should be performed with frozen-section analysis of the mass.^[4] Most masses are benign mucocèles and require no further management. If however lymphoma or carcinoid is identified, chemotherapy or a right hemicolectomy may be required. If an adenocarcinoma is present, the patient should be discussed at the colorectal multi-disciplinary team (MDT) meeting where appropriate pre-operative staging and management will be discussed.^[5]

Pre-operative detection of appendiceal adenocarcinoma is invariably difficult since the clinical presentation is usually non-specific, unless they are large tumors involving the stump of the appendix.^[6] Adenocarcinomas of the appendix show characteristic morphological variations, some resemble a colorectal carcinoma, some arise from a carcinoid, some appear similar to mesenteric cysts, and some are mucinous tumours that may appear well differentiated and indistinguishable from an adenoma. The diagnosis of primary appendiceal carcinoma usually depends on the histopathological analysis following appendectomy or other explorative surgical procedures.^[7] Primary adenocarcinomas of the appendix are often perforated and pseudomyxoma is assumed to be a perforated state.^[6]

The optimal surgical treatment of primary appendiceal carcinoma is controversial. Simple appendectomy appears to be sufficient for early, small (<2cm) non-invasive carcinomas of the appendix, however many carcinomas are diagnosed on the basis of the pathology from an appendectomy and are advanced, large (>2cm) and invasive, in such cases a right hemicolectomy is the treatment of choice.^[8] For mucinous-type adenocarcinoma with pseudomyxoma peritonei, a right colectomy is the preferred procedure.^[9]

Adjuvant chemotherapy in adenocarcinoma of appendix is not yet established in clinical practice; therefore it is recommended for patients with positive lymph node affection or locally advanced tumors to be treated in accordance with the recommendation for colon cancer.^[10]

Mucinous-type appendiceal adenocarcinoma with pseudomyxoma is a separate disease entity. The diffusely scattered tumour cells in the peritoneal cavity usually stay in the peritoneal surface and grow slowly. Definitive cytoreductive surgery with hyperthermic intraoperative intraperitoneal chemotherapy has shown promising results.^[11]

The five year survival rate for all patients was 54.4% and was associated with the stage of disease, the grading and the histological subtype.^[12]

CONCLUSION

Primary adenocarcinoma of the appendix is a rare tumour. Clinical presentation is usually similar to acute appendicitis. Pre-operative diagnosis can be difficult. Treatment is with right hemicolectomy, even if this is a secondary procedure following an initial appendectomy. Surveillance for synchronous or metachronous tumors, especially in the gastrointestinal tract, is warranted.

Grant

None.

CONFLICTS OF INTEREST

We the authors of this case report have no conflicts of interest to declare.

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