ADENOCARCINOMA OF APPENDIX MIMICKING ACUTE APPENDICITIS: A CASE REPORT AND LITERATURE REVIEW

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ABSTRACT

Adenocarcinoma of the appendix is a rare neoplasm, often diagnosed in patients of the older age group, who present with a clinical picture of acute appendicitis. This paper presents a case of adenocarcinoma of the appendix in a case initially treated as acute appendicitis.

KEYWORDS: Adenocarcinoma of appendix, Acute appendicitis, Appendicectomy, Right hemicolectomy, Krukenberg tumour.

INTRODUCTION

Primary cancer of the appendix is diagnosed in 0.9 – 1.4% of appendicectomy specimens.[1] The majority of primary malignant appendiceal tumours are carcinoid (up to 90%), followed by mucinous adenocarcinoma.[2,3] In general, primary appendiceal cancer accounts for only 0.4% of all gastrointestinal cancers.[4,5] The most common clinical picture involves features of acute appendicitis, however an abdominal mass or anaemia may also feature. Appendiceal cancer is histologically classified into carcinoids, goblet cell adenocarcinoma (GCA), colonic and mucinous adenocarcinoma.[4,6,7] Right hemicolectomy may be necessary to ensure proper staging and management. This paper presents a case of adenocarcinoma of the appendix in a female patient aged 83 years.

CASE REPORT

An 83-year old female patient presented to the Accident & Emergency Department with a 2-day history of right iliac fossa pain and reduced appetite. She was known to have congestive cardiac failure, atrial fibrillation (on warfarin) and was classed as ASA 3. Blood results showed white cell count 8x10^3/L and C-Reactive Protein 36mg/L. CT abdomen revealed a fluid-filled dilated (11mm) appendix, surrounded by inflammatory changes [Fig.1&2].

The patient underwent laparoscopic appendicectomy. Intraoperatively, the appendix was found to be inflamed. However, approximately 1 inch distal to the base, there was a tense, pale, grey nodule, possibly cystic in nature. After resection, the lump was measured to be 26mm distal to the base and 16 mm in length [Fig.3]. Histology was consistent with a mucinous, moderately-differentiated adenocarcinoma. The tumour invaded the muscularis propria into the subserosa, but not into the serosal surface. There was no evidence of lymphovascular invasion [Fig.4]. The tip and base of the appendix were free of tumour involvement, however showed severe transmural acute inflammation with associated serositis. Immunostaining for synaptophysin was negative, but chromogranin staining showed a few cells were positive, with Ki67 positivity in more than 90% of cells. The tumour was graded as T3NxMx. The postoperative histology was discussed with the patient, and further treatment with right hemicolectomy and surveillance was offered. The patient expressed a firm desire to have no further surgery or interventions.

Several months later, a follow-up CT scan showed a malignant-looking lesion in the descending colon [Fig.5&6], with signs of peritoneal deposits [Fig.7]. There was also a complex adnexal mass, suggestive of the presence of a Krukenberg tumour or the possibility of a synchronous ovarian malignancy. The patient was referred to the oncological team for palliative care.

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Fig 1: CT abdomen - coronal view: The appendix is dilated to 11 mm, fluid-filled, and surrounded by minor inflammatory changes.

Fig 2. CT abdomen - axial view: The appendix is dilated to 11 mm, fluid-filled, and surrounded by minor inflammatory changes.

Fig. 3: A cystic mass seen distal to the base of the appendix during laparoscopic appendectomy, initially diagnosed as a mucocele.

Fig. 4: Appendix showing extensive infiltration of the wall by a mucinous adenocarcinoma.

Fig. 5: CT Abdomen - axial view: Malignant looking stricture within the descending colon.

Fig. 6: CT Abdomen - Coronal view: Malignant looking stricture within the descending colon.
DISCUSSION

Adenocarcinoma of the vermiform appendix is a rare neoplasm, usually diagnosed incidentally and most often affecting adults.\[^{8}\]\ It was first described by Berger in 1882\[^{4}\]; the patients often present with the clinical picture of acute appendicitis, particularly with right iliac fossa abdominal pain.\[^{9}\]\ Other rare presentations also reported include an abdominal mass\[^{10}\], right thigh mass\[^{5,11}\], caecal intussusception\[^{12}\], peritonitis, anaemia\[^{13}\], hydromephrosis\[^{14}\], or Krukenberg tumour of the ovary.\[^{15}\]\ In general, the incidence of appendiceal neoplasm varies from 0.01 to 0.2 per 100,000 persons per year.\[^{7}\]\ The majority of primary appendiceal cancers occur in the age group of 55–65 years, with the exception of the malignant carcinoid, which has been reported in patients with a mean age of 38 years.\[^{16}\]\ Patients of an older age group and presenting with symptoms of acute appendicitis should be investigated for the possibility of an underlying neoplasm\[^{8}\]; a CT scan is suggested in such scenarios. The tumour may be diagnosed either as an isolated diagnosis or associated with another condition, such as ulcerative colitis\[^{17}\], ovarian malignancy\[^{18}\] or De Garengot hernia (femoral hernia containing the appendix).\[^{19}\]\ It is also possible for adenocarcinoma of the appendix to arise in a post appendicectomy stump.\[^{20}\]\ The reported complications from appendiceal adenocarcinoma include torsion, disease dissemination, hydromephrosis\[^{14}\], intussusception, and bowel perforation.\[^{3}\]\

The most common primary appendiceal malignant tumour is carcinoid (up to 90\%)\[^{2,3}\]; followed by mucinous adenocarcinoma.\[^{2,3}\] This mucinous variety most often presents following perforation of the primary tumour, with a spread of mucin and tumour cells throughout the peritoneal cavity. This results in a clinical syndrome of abdominal distention, secondary to mucinous ascites; it is known as pseudomyxoma peritonei. The condition is usually diagnosed at operation for appendicitis, peritonitis, possible ovarian malignancy, or on diagnostic imaging.\[^{21}\]\ Several other conditions of the appendix must also be differentiated from primary mucinous appendiceal adenocarcinomas, including acute appendicitis, mucocele\[^{22}\]; carcinoid, caecal carcinoma, lymphoma\[^{5}\] or rarer tumours such as ganglioneuroma.\[^{23}\]\ The colonic type is found in less than 0.1\% of cases, where the cystic sub-variety is characterised by its tendency for perforation and dissemination.

The surgical management of appendiceal carcinoma is controversial; is appendicectomy enough or should it be extended to right hemicolecctomy? Appendicectomy alone is justified when the tumour is limited to the mucosa and does not extend beyond the muscularis mucosae and has no vascular infiltration. The only exception for this is in goblet cell adenocarcinoma, as 20 - 40\% of these cases show regional lymph node metastasis. Therefore, for appropriate management, right hemicolecctomy and staging investigations are highly recommended for adequate management regardless of primary tumour size.\[^{21,24}\]\ For all cases of goblet cell adenocarcinoma, locally advanced adenocarcinoma or carcinoids, postoperative staging is suggested, followed by right hemicolecctomy and systemic chemotherapy as required.\[^{23}\]\ Positive results have also been reported by Sugarbaker, following development of a definitive cytoreductive surgery using hyperthermic intraoperative intraperitoneal chemotherapy.\[^{25}\]\

After surgery and adjuvant treatment, patients should be followed up with clinical assessment and endoscopy imaging in the same way as for colonic cancer.

CONCLUSION

Patients of an older age group presenting with symptoms of acute appendicitis should be investigated for the possibility of an underlying malignancy; a CT scan is suggested in such scenarios. Detection of a vermiform appendiceal adenocarcinoma is essential, as it ensures the appropriate management of a right hemicolecctomy instead of appendicectomy alone.
REFERENCES