Mucinous Cystadenoma of the Appendix—An Unusual Cause of Intestinal Obstruction

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Abstract

***Introduction**: Report of a rare presentation of mucinous cystadenoma of the vermiform appendix. **Clinical Picture**: A lady who presented with intestinal obstruction and peritonitis was found to have gangrenous small bowel caused by strangulation by a tumour of the appendix. **Treatment**: Right hemicolectomy. **Outcome**: The histology of the appendicular tumour was mucinous cystadenoma. The patient recovered well. **Conclusion**: Mucinous cystadenoma of the appendix can present in many ways and it is important to recognise the pathology at operation.*

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Key words: Cystadenocarcinoma, Mucocele, Pseudomyxoma peritonei

Introduction

Mucinous cystadenoma of the vermiform appendix is an uncommon entity. It is a benign lesion but may disseminate and lead to the development of pseudomyxoma peritonei. The condition is usually discovered during surgery, as the presentation is varied and non-specific. We present a case of mucinous cystadenoma of the appendix who presented with small bowel obstruction.

Case Report

A 65-year-old Malay lady presented with complaint of a sudden onset of colicky central abdominal pain associated with vomiting and constipation for 4 days. There was no loss of appetite or weight. She has no family history of colorectal malignancy.

On examination, she was afebrile and her vital signs were normal. She had mild pallor and was not jaundiced. Her abdomen was soft and not distended. Her lower abdomen was tender and guarded and a mass was felt in the right iliac fossa. Her bowel sounds were sluggish. Digital rectal examination revealed a rectum packed with brown stools.

Blood tests revealed a normal white cell count and a haemoglobin level of 11.4 g/dL. Her liver function test was normal except for a raised aspartate transaminase and lactate dehydrogenase levels. Abdominal X-ray showed dilated loops of small bowel with faecal shadows present in the rectum.

The patient underwent an emergency exploratory laparotomy where the pedunculous appendix was found wrapped around an infarcted loop of terminal ileum (Fig. 1). At the end of the appendix was a tumour measuring 8 x 5 cm.

Fig. 1. Mucinous cystadenoma of the appendix with infarcted terminal ileum.

The liver, ovaries and omentum were unremarkable. A limited right hemicolectomy with an end-to-end ileocolic anastomosis was performed.

Pathological examination showed a dilated appendix which, when cut, revealed a thin-walled cyst filled with

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translucent gelatinous substance. On microscopic examination, the appendix was found to contain an amorphous myxoid substance with scattered foci of calcification, cholesterol clefts and chronic inflammatory cells. There was no evidence of malignancy in the appendix, ileum and colon. No mucin deposits were noted in the peritoneal cavity.

After surgery, the patient had an uneventful recovery and was discharged on the seventh day. The patient remained asymptomatic 1 year after the surgery.

Discussion

Mucocele of the appendix is an uncommon lesion. It has a dilated lumen which is filled with mucin to form a cystic mass. The reported incidence has been quoted to be 0.2% to 0.3% of all appendicectomy specimens.[1] The median age of presentation is 55 years but this condition can occur from the 2nd to the 9th decade. The female to male ratio is about 2:1.

Mucoceles are divided into four groups depending on the characteristics of their lining epithelium. They are classified into retention cysts, mucoceles with mucosal hyperplasia, mucinous cystadenomas and mucinous cystadenocarcinomas.[1,2] Of these groups, mucinous cystadenomas are the commonest, accounting for 63% to 84% of all mucoceles.[2,3] Mucinous cystadenomas and cystadenocarcinomas are said to be linked to ovarian and colonic neoplasms in some reports.[1,4]

Mucinous cystadenoma is often asymptomatic but it can present as pain in the right lower quadrant or as a palpable mass on physical examination. Other presentations include gastrointestinal bleed,[2] obstruction of the right ureter leading to hydronephrosis or intestinal obstruction as a result of volvulus or appendiceal intussusception.[5]

Preoperative diagnosis is difficult due to the lack of or non- specificity of its symptoms. The tumour marker, carcinoembryonic antigen (CEA), has been reported to be raised in mucinous cystadenocarcinomas[6] but in practice, the usefulness of this test is questionable. Most of the mucinous cystadenomas detected prior to surgery are done so via radiological and endoscopic means. On barium enema, there is partial or no filling of the appendix with contrast. A submucosal or extrinsic mass may be shown to indent or displace the caecum. Ultrasonography may show a cystic lesion with variable echogenicity with or without calcifications. Computed tomographic scan may show a mass with or without calcifications or septation. Attenuation of the contents of the mass may range from near-water density to soft tissue density pending on the amount of mucin.[7] Colonoscopy may show a submucosal or extrinsic mass in the caecum or an intussusceptum. The appendiceal orifice may be obliterated and not found, or it may be seen in the centre of a mound surrounded by normal-appearing mucosa—the so-called "volcano sign".[3]

At the time of operation, the diagnosis will usually become obvious. It is important to handle these tumours with care and avoid rupture. Mucinous cystadenoma is not discernable from mucinous cystadenocarcinoma just by gross examination and rupturing a mucinous cystadenocarcinoma will lead to the dissemination of the mucin and the mucin-secreting malignant cells into the peritoneal cavity, causing pseudomyxoma peritonei. A search for co-existing ovarian or colonic tumours should also be performed.

An appendicectomy is adequate in dealing with an uncomplicated mucinous cystadenoma but care must be taken not to rupture the tumour. When the tumour extends to the caecal wall and frozen section study is not available, a right hemicolectomy is preferred.[1] In cases where the tumour has ruptured, in addition to resecting the tumour, aggressive removal of all gross spillage including bilateral oophorectomy and omentectomy should be performed.[8] Perioperative intraperitoneal chemotherapy has also been advocated to improve survival.[9]

The prognosis of mucinous cystadenomas is good with a 5-year survival rate of 91% to 100% but it decreases to 25% in mucinous cystadenocarcinomas.[1]

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