CASE REPORT

Mucocoele of the appendix: A case report

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Abstract

Mucinous cystadenoma is a rare tumour of the appendix. It accounts for only 0.4% of the gastrointestinal tract malignancies and is reported rarely in the literature. Therefore, surgical management is not yet established. Here we report the case of a 65-year-old female who presented with a dragging sensation and a feeling of a mass in the right iliac fossa. Her computed tomography (CT) suggested formation of an abscess in the parietal peritoneum. She was scheduled for laparotomy and upon exploration, a mass was found arising from the tip of the retroperitoneal appendix. The whole of the appendix was studded with mucoid material. Right hemicolectomy was performed and histopathology of the appendix showed mucinous cystadenoma with no evidence of malignant changes. The recovery was uneventful and the patient was discharged on the fourth post-operative day. The unusual presentation of retroperitoneal pseudomyxoma without any intraperitoneal pathology prompted us to report this case.

Keywords: Appendiceal tumours, Mucinous cystadenoma, Pseudomyxoma peritonei.

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Introduction

Appendicular tumour is a rare entity, as it is an incidental finding on histopathology in patients who are operated for appendicitis and the histopathology of the specimen shows the tumour. Very few among them have malignant potential, while most are of benign pathology. A report was published by the National Cancer Institute, USA, in which Surveillance, Epidemiology and End Results (SEER) database identified that appendiceal neoplasms account for approximately 0.4% of total gastrointestinal tumours.[1]

Besides rarity, a wide range of histopathology is involved, mainly mucinous epithelial neoplasms (also called adenomas), cystadenoma, and benign neoplastic mucocoele. Radiologists and surgeons are quite familiar with this condition as appendectomy is a commonly performed surgery. However, appropriate management is still crucial.[2]

Removal of the whole appendix is a vital step in the management of mucocoele of the appendix as it can spread if it is crushed or ruptured accidentally, resulting in a peritoneal spread and the condition is known as pseudomyxoma peritonei in which epithelial cells are deposited in the peritoneal cavity. The frequency of primary malignant tumour of the appendix is even less and are reported in < 2% patients.[3]

The size of a mucocoele is also important as a cyst <2cm in diameter is a simple retention cyst; however, a cyst >2cm is likely to be a cystadenoma or cystadenocarcinoma. Hyperplastic epithelium is also seen in size greater than 2cm.[4]

Case Presentation

A 74-year-old female, resident of Rawalpindi, Pakistan, presented at the Shifa International Hospital on May 23, 2018, with complaint of lower abdominal pain for the last one month. The pain was sudden in onset and was colicky in nature. She complained of intermittent dragging sensation that lasted for two to three hours; there was no aggravating factor and pain was relieved after some time on its own. There was no relation with food, and no vomiting or significant weight loss was noted. On abdominal examination, a soft, non-tender mass was felt in the right hemiabdomen with well- defined margins of 7x10 cm in size. Percussion over mass was dull, otherwise the abdominal examination was unremarkable. The patient's written informed consent as well as approval from the ethical committee was taken for publishing the case report.

Her laboratory investigations were within normal limits. However, her Ca-125 level was 125 units/mL (normal values are 0-35 units/mL). Her CEA levels were 11.9 ng/ mL (normal values are 3 (ng/mL), while Alpha-fetoprotein (AFP) was normal. Her CT scan showed a well-defined rounded mass in the right lower abdomen measuring 9.5x11.5x10.2cm in size, showing marginal calcification, few intralesional calcific foci were also seen. The rest of the lesion contained mainly fluid and very few areas were soft tissue. Extensive degenerative changes in the bones

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were seen while lung fields were clear.

The patient was scheduled for laparotomy by a gynaecologist's team. Spinal anaesthesia was opted for, as she was at high risk for general anaesthesia. Pfannenstiel incision was given. Both the ovaries and uterus were found to be normal and so a general surgeon was called in. A large mass was found at the distal end of the ileum, due to which right hemicolectomy was performed.

The patient was comfortable postoperatively and was discharged on the fourth postoperative day. She was followed up on the 10th post-operative day and then after one month. The patient remained symptom free.

Discussion

The epithelial lining of the vermiform appendix is mucous-secreting columnar cells. The pathological changes can result in 1) retention cyst, 2) villous hyperplasia, 3) cystadenoma, and 4) cystadenocarcinoma. During these pathological changes, the mucocele of the appendix is formed, resulting in abnormal dilatation of the appendix due to excessive mucinous production by epithelial cells. These tumours were classified by the WHO (World Health Organisation) and AJCC (American joint commission on cancer) in 2010,[5] the lesions that are limited to the appendix are referred to as adenoma (mucinous), irrespective of the cytological appearance and there is no relation to obtained surgical margins. Once the tumour is beyond muscularis mucosae or there is mural perforation, only then it is labelled as cystadenocarcinoma.[6]

Mucinous cystadenoma has an atypical presentation besides normal appendicitis. The patient may present with an abdominal mass, rectal bleeding, haematuria, ureteric obstruction or even atypical features of intussusceptions can be encountered.[7]

The treatment of this tumour is controversial; surgical resection of the appendix is the preferred treatment, though the extent of the surgery is controversial. Removal of the caecum is suggested in some cases. Laparoscopic appendectomy has shown a faster recovery rate than open appendectomy.[8]

The option is appendectomy if there is retention cyst, hyperplasia or even cystadenoma. If the cystadenoma resides at the base of the appendix, resection of caecum is indicated. The option of right hemicolectomy is reserved for cystadenocarcinoma. However, no survival benefit is noted.[9] Indications of right hemicolectomy are: 1) need to clear primary tumour or complete cytoreduction, 2) lymph nodes are involved, 3) non-mucinous cyst is observed histologically.[10]

The gross enlargement of the appendix or if the lumen is studded with mucous raises clinical suspicion. The clinical incidence of mucocele is 0.2-0.3% in resected appendices in Europe and the United State.10 No local data is available and few cases have been reported in Pakistan.[11] It is difficult to differentiate whether the disease is benign or malignant before surgery; however, it is important to avoid spillage of content as it may lead to pseudomyxoma peritonei.[12]

Three pathological types are seen. Hyperplasia, cystadenoma and cystadenocarcinoma. The surgical treatment remains controversial as at times it has been observed that undue surgery has been performed. The fear of dissemination causes surgeons to overdo. Laparoscopic approach is also a surgical option for mucocele appendix.[13]

Under consideration of pseudomyxoma peritonei, we prefer open approach as the prognosis is bad with this pathology. Five-year survival is 53-75% and 10-year survival is 10-32%. There is no confirmation whether it is benign cystadenoma or cystadenocarcinoma. Some surgeons still prefer an open approach rather than laparosopy.[14]

Volcano sign on colonoscopy is diagnostic of mucocele of the appendix. Nodularity in the cyst and enlarged lymph node may direct malignant factors.[12]

Conclusion

The presented case was diagnosed before surgery as appendiceal mucinous cystadenoma and treated by open resection. Recognising volcano sign and the absence of malignant factors preoperatively, plus intraoperative pathological diagnosis allowed us to avoid operating excessively for benign appendiceal mucocele.

Disclaimer: None to Declare

Conflict of Interest: None to Declare

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