CASE REPORT

Primary Mucinous adenocarcinoma of the appendix- An update on the diagnosis, pathological features of appendiceal mucinous adenocarcinoma

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Abstract:

Primary Mucinous Adenocarcinoma is a rare tumour. Out of 43 appendix specimens received in our laboratory, there was one case of mucinous adenocarcinoma (2.32%). We report a case of primary mucinous adenocarcinoma in a 40-year male patient, admitted for appendicectomy. The patient presented with an appendicular mass, four months previously. The initial treatment was conservative, with the mass considerably reducing in size. The patient was requested to come for a follow up, but disappeared following reduction in the size of the appendicular lump and improvement of his symptoms. He contacted consultant again four months later, as the pain had re-appeared. On examination there was a persistent residual lump in the right iliac fossa. Appendicectomy was done and histology confirmed mucinous adenocarcinoma. Right hemicolectomy was done as a second stage procedure. All surgical specimen, including normal looking appendix be submitted to histological examination as not to miss a sinister diagnosis, requiring a follow up and even second surgical intervention,

Received
Date: 7th November, 2019
Accepted
Date: 6th March, 2020

Keywords : Appendicitis, mucinous adenocarcinoma, appendicular lump

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Introduction:

The primary neoplasm of the appendix are present in less than 2% of surgical appendicectomy specimen.[1] The neoplasm includes epithelial tumours, mesenchymal tumours and lymphoma. Mucinous neoplasm of the appendix are a complex, diverse group of epithelial neoplasm, causing cystic dilatation of the appendix due to accumulation of gelatinous material morphologically referred to as mucocoele.[2] The tumour was first described by Rokitansky in 1842.

Cystic dilatation of the appendix results from luminal obstruction either by non-neoplastic or neoplastic conditions. Simple cyst are retention cysts that can result from an obstructing faecolith. An appendicular mucinous neoplasm refers to a tumour associated with a neoplastic adenomatous growth- adenoma or adenocarcinoma. Various classification schemes by Pai and Longacre, Misdraji et al., Carr Wand Sobin and the 2010 WHO classification have been proposed. These are similar in that they all define benign neoplastic adenoma as confined to the mucosa,[3] without mucin or cells penetrating the muscularis mucosa or evidence of perforation. Invasive adenocarcinoma is a frankly invasive neoplastic lesion with cellular invasion beyond the muscularis mucosa.

We present here a case of invasive mucinous adenocarcinoma, presenting acutely as appendicular mass, resolving to some extent with the relief of symptoms. Later pain recurs, residual lump is felt and appendicectomy done.

Case report:

History of a patient who presented with mucinous adenocarcinoma of the appendix. A 40-year male patients was admitted for interval appendicectomy. About 4-months back he was treated for an appendicular mass. The appendicular mass reduced in size on conservative treatment over several weeks. As the lump and pain lessened, patient failed to keep the follow up appointment, The pain recurred and patient

contacted consultant again. On examination of the abdomen, a residual small lump persisted in the right iliac fossa. The patient was admitted for appendicectomy. The appendix labelled as mucocoele of the appendix was sent for the histological examination.

On bisecting the specimen, the fundus of the appendix was found to be ruptured and mucinous material had perforated the serosa and had collected outside the fundus. The appendix specimen was distorted fibrosed and lumen was twisted instead of a linear one. There was evidence of haemmorhage around the ruptured fundus. The mucinous material had formed a wall around itself and was intact.

The ultrasound beautifully shows the lumen of the appendix with the tip of the appendix giving way and surrounded by fluid – mucus.

Ultrasound report: 4.5 cms tubular blind ending structure seen in the right iliac fossa with its tip into well-defined complex cystic area measuring 5.1X 2.5 cm. The appearance suggests acute appendicitis with the tip of the inflamed appendix in the cavity of previous residual abscess.

The complex cystic area surrounding the tip of the appendix was mucus which had accumulated after the serosa had given way.

Discussion:

Mucinous Adenocarcinoma of the appendix is an exceedingly rare tumour.[4] Out of 700-cases of appendix specimen, between the years 2002-2008, only one case of mucinous adenocarcinoma was detected. Presentation of the conditions is with acute appendicitis or appendicular lump.[5] Mucinous adenocarcinoma and colonic adenocarcinoma are the most common types that are found in appendix. 40% of them are mucin secreting mucinous adenocarcinomas. Classifying the type of adenocarcinomas is important because colonic adenocarcinoma has a bad prognosis as compared to mucinous adenocarcinoma.[6] Appendiceal mucinous neoplasms with signet ring cell features (in which signet ring-shaped cells are seen floating within the mucin nodules) or poorly differentiated histology are believed to be more aggressive and have a worse prognosis.

In our case the patient presented with an appendicular lump, which reduced in size over a period of 4 months, but did not completely subsided. The diagnosis is not suspected either pre-operatively or even at the time of surgery. The diagnosis is established on histo-pathological examination. Pre-operative investigations particularly ultrasound examinations are helpful but not diagnostic. Ultrasound of this case confirmed the acute appendicitis surrounded by non-echoic mass thought to be appendicular abscess. The surrounding non echoic lesion was presence of mucus outside the appendix. The mean age of presentation is 50 years and male to female ratio is 4:1 with predominance of male.[7] The presence of mucin outside the appendix can lead to pseudomyxoma peritonei with dissemination of mucinous tumour deposit all over the peritoneum. In our case the mucinous collection outside the appendix was walled off without any spread.

The pseudomyxoma peritonei ideally should be labelled when there is widespread of peritoneal involvement with gelatinous sticky ascites.[8]

Treatment of invasive: mucinous adenocarcinoma appendix is right hemicolectomy.[9] In case of pseudomyxoma peritonei, the treatment will consist of debulking surgical procedures with perhaps chemotherapy.[10,11] The case is reported here because of its rarity and interesting ultrasound and pathological findings.

Conflict of interest: None

Funding source: None

Role and contribution of authors:

Dr S Khurshid, collected the data, references and did the initial writeup

Dr A Memon, collected the data, references and helped in introduction writing.

Dr Tasir Mumtaz Ahmed, critically review the article and made useful changes

Dr Zakiuddin G Oonwala, critically review the articel and made final changes.

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