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TITTLE: Low Grade Appendiceal Mucinous Neoplasm- A Case Report

Nasir Khan, Sibgha Aimon

Shifa International Hospital, Islamabad, Department of General Surgery.

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INTRODUCTION:

Mucinous neoplasm of appendix accounts for 0.2-1.4% of appendectomies done. [7] These tumors are frequent in female and present in 5th to 6th decade of life. [4,6] Usual presentation is with pain abdomen, and it can rupture leading to pseudomyxoma peritonei. [1,2] Diagnosis is generally made with CT scan with typical features of cystic mass with wall calcification. [4,5] Disease limited to appendix can be treated by simple appendectomy [1], but advance disease with peritoneal spread requires cytoreductive surgery. [7]

We present a case of a 42-year-old female with Low grade mucinous neoplasm.

CASE PRESENTATION:

A 42-year-old female presented in surgical clinic with pain right lower abdomen for one month. Pain was sudden in onset, initially low intensity and intermittent, then it became constant, moderate intensity localized to right iliac fossa. Pain was associated with occasional nausea and vomiting. She did not reported fever, loss of appetite, weight loss or change in bowel habits. There was no past surgical history. On examinations her vitals were normal and abdomen was non distended, but tender in right iliac fossa. Her initial workup showed normal White blood count, and hemoglobin was 10/1g/dl.

Ultrasound abdomen performed which showed Cystic mass in right iliac fossa. CT scan abdomen was ordered to further evaluate the mass.

Ct scan showed A tubular, elongated, blind ending fluid filled structure of size 10x4.3x4.2cm with curvilinear, mural calcification in right iliac fossa at the base of cecum, reaching superiorly up-to subhepatic region with minimal surrounding fat stranding and sub centimeter lymph nodes. (Figure 1, 2)

On these findings' working diagnosis of Appendiceal Mucinous Neoplasm was made. Patient was counselled in detail and informed consent taken for Laparoscopic Appendectomy, with possibility of conversion to Open Appendectomy.

Figure 1

Arrow points toward cystic swelling with mural calcification

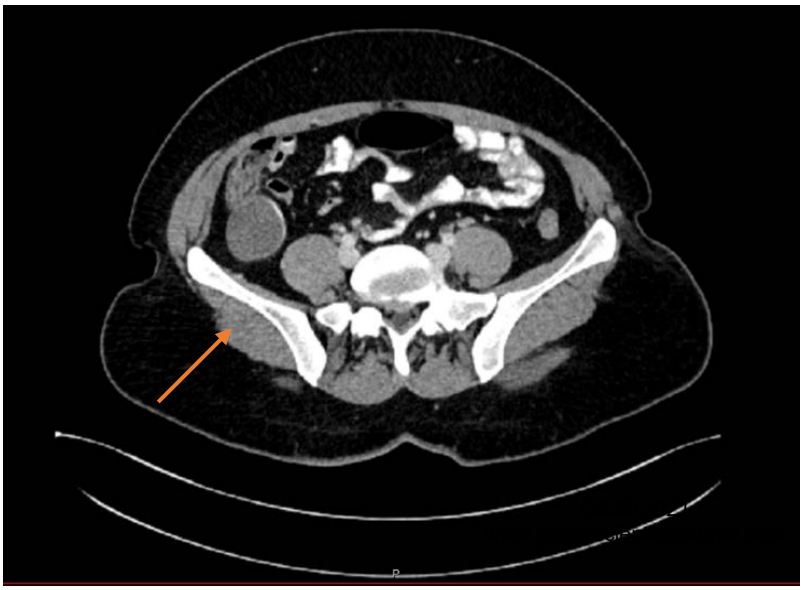

Figure 2

Arrow points toward cystic swelling in appendix

Patient admitted to surgical floor 1 night prior to surgery. On the day of surgery, standard Laparoscopic appendectomy protocols were followed. After Induction of anaesthesia, scrubbing done, and Pneumoperitoneum created via Veress needle. Standard 3 ports inserted for Laparoscopic appendectomy. Per operatively we found large tubular appendix with normal base and no perforation. Figure 3. Laparoscopic appendectomy was performed, and appendix retrieved in an endobag. Appendectomy specimen (Figure 4) was sent for histopathology. Appendicular stump was further biopsied to determine margin status.

Figure 3

Per-operative finding of dilated appendix without any perforation.

Figure 4

Appendectomy Specimen

Patient remained well postoperatively, Diet started the same day, she was mobilized and was discharged the next day.

Her final histopathology showed Low Grade Appendiceal Mucinous Neoplasm (LAMN) with tumor size of 10x4cm. Resection Margins were Uninvolved. Acellular Mucin was

Invading Subserosa but did not extend to serosal surface, so tumor was staged pT3. Patient was advised follow-up at 6 months with CT scan abdomen.

Discussion:

Appendiceal Mucinous Neoplasm are a rare entity and are found in 0.2-1.4% of appendectomies performed for acute appendicitis. It has female predominance with a male to female ratio of 1:4. [3] They are mostly diagnosed in patients at 50-60yrs of age [1,3,4]. Complicated neoplasms have a higher risk of having occult mucinous neoplasm. [1] The Peritoneal Surface Oncology Group International (PSOGI) in 2020 divided Mucinous Neoplasm into serrated polyp, Low grade Appendiceal Mucinous neoplasm (LAMN), High grade Appendiceal Mucinous Neoplasm (HAMN) and Mucinous Adenocarcinoma. Most common presentation is pain right lower quadrant. [7] Occasionally it can present with intestinal obstruction, intussusception, and genitourinary symptoms due to involvement of right ureter. [1]

CT scan abdomen with contrast is diagnostic modality of choice, with findings of cystic mass with mural calcifications, [6]. Rupture of Neoplastic lesions can lead to Pseudomyxoma Peritonei (PMP) due to dissemination of tumor cells [3].

Low grade Appendiceal Mucinous Neoplasm (LAMN) describes the tumor that is not infiltrating and limited to muscularis propria. They have acellular mucin pool [1,7]. Histologically, LAMN has atypical glandular cells with Pushing Invasion. [2]

Treatment depends upon extent of the disease. For local disease involving appendix only, Appendectomy is sufficient. Retrieving appendectomy specimen in endobag is advised as to prevent inadvertent rupture leading to dissemination. [1] . For Pseudomyxoma peritonei, treatment method suggestive is cytoreductive surgery with Heated Intraperitoneal Chemotherapy (HIPEC). [7]

Prognosis of disease limited to appendix, such as in our case, is good with 5-year survival up-to 100%, and of malignant form varies from 30-80% [5,7]

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