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A Case Report: Pseudomyxoma Peritonei due to Ruptured Appendicular Mucocele

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Abstract: ***This article details a unique case of Pseudomyxoma Peritonei PMP, a rare condition characterized by mucinous ascites and peritoneal implants, originating from a ruptured appendicular mucocele in a 36 - year - old female with no significant past medical history. Initially presenting with abdominal pain and distension, investigations led to surgical intervention, revealing extensive intra - abdominal mucinous material. The histopathology confirmed high - grade appendicular mucinous neoplasm with omental involvement. The discussion emphasizes the importance of specialized, multidisciplinary care for PMP patients, highlighting the prognostic value of tumor markers like CEA, CA 19.9, and CA 125 in appendiceal and ovarian PMP, respectively. The article advocates for the combined treatment approach of cytoreductive surgery CRS with hyperthermic intraperitoneal chemotherapy HIPEC as the gold standard for PMP management, offering significant survival benefits. It also outlines the necessity of long - term surveillance due to the potential for late recurrences and considers systemic chemotherapy for specific high - grade cases.***

Keywords: Pseudomyxoma Peritonei, Appendicular Mucocele, Cytoreductive Surgery, Hyperthermic Intraperitoneal Chemotherapy, Tumor Markers

1. Introduction

Pseudomyxoma peritonei (PMP) is a rare clinical entity; characterized by diffuse intra - abdominal gelatinous ascites with mucinous implants on peritoneal surfaces. WERTH in 1884 coined the term PMP, describing it in association with a mucinous tumor of ovary. The annual incidence is 1 - 2 cases per million. Here I present a case of PMP, admitted in our hospital with a different aetiology.

2. Case Report

A 36 Years old female attended our emergency with a history of dull aching pain at lower abdomen for last 4 months and abdominal distension since 2 weeks. There was no significant past medical and surgical history. There was no history of comorbidities. Her menstrual cycle was regular, History of 3 normal deliveries with the last child birth 12 years ago.

On general examination, only pallor was present. Rest was essentially normal.

On local examination there was voluntary guarding at right lower abdomen with an elongated diffuse mass felt which was tender, nonmobile, with ill - defined margins.

Investigations revealed: CECT (W/A) - elongated hypodense lesion in Right iliac fossa suspicious of Appendicular Mucocele & mesenteric fat stranding in lower abdomen and Pelvis. Mild ascites present.

Routine Blood investigations were within normal limits.

Abdominal exploration was planned after optimisation. On Laparoscopy there was gelatinous material with white patches all over the peritoneal cavity, omentum and bowel loops. Conversion to open Laparotomy was decided. Appendectomy and omentectomy was performed. The Jelly like material was scooped off as much as possible.

3. Conclusion

It was a case of Pseudomyxoma Peritonei due to ruptured appendicular mucocele. on HPE report it turned out to be - High Grade appendicular mucinous neoplasm with omental involvement.

4. Discussion

Patients with PMP should be referred to a specialist centre with multidisciplinary expertise in the assessment and management of patients with peritoneal malignancy.

Tumor markers are found to have prognostic value and are useful for follow up of patients after treatment. CEA and CA 19.9 are found to be high in PMP with an appendiceal origin. CA 125 is also found high but with ovarian involvement.

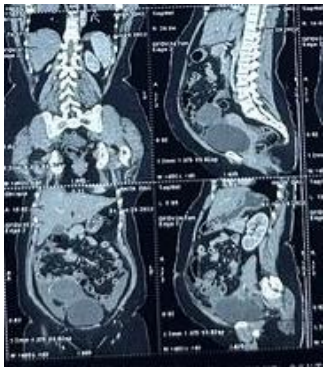
The accepted treatment is CRS combined with HIPEC (Sugarbaker). This approach combines multiple peritonectomy procedures with multivisceral resections as required to achieve a complete surgical clearance of the tumour (complete cytoreduction), which is augmented by HIPEC (typically mitomycin C or oxaliplatin) to eradicate presumed residual microscopic disease.

The combined operation may require total abdominopelvic peritonectomy, greater and lesser, omentectomy, bilateral salpingo - oophorectomy, hysterectomy, cholecystectomy, splenectomy, partial gastrectomy, colectomy and anterior resection of the rectum.

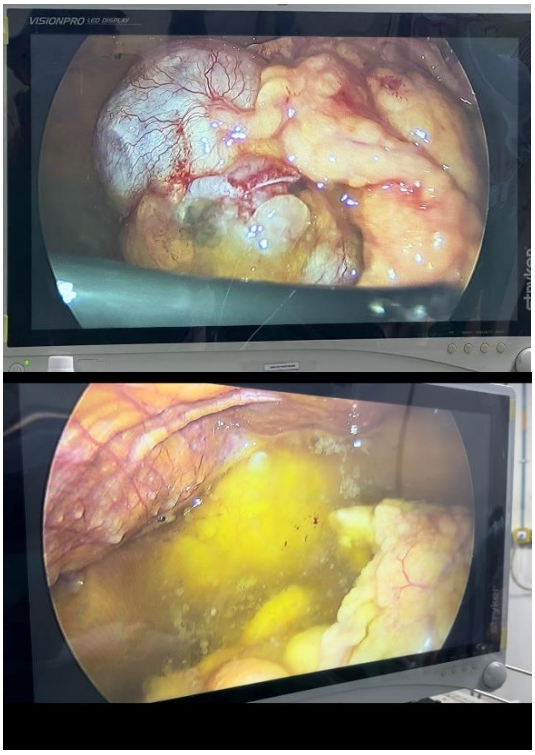
Following a complete cytoreduction 5 - and 10 - year survival rates of 87% and 70%, respectively, can be achieved

Follow - up typically comprises at least annual clinical evaluation, monitoring of tumour markers and CT scan. Surveillance should be continued for at least 10 years as late recurrence is documented.

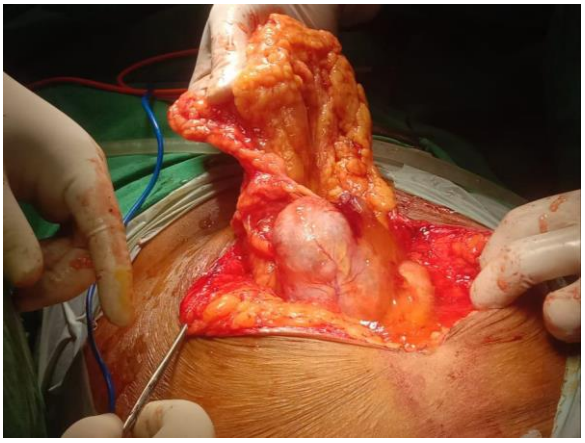
Systemic chemotherapy may be considered as first - line treatment in patients with high - grade or invasive unresectable disease or in the adjuvant setting following CRS/HIPEC, in patients with high - grade tumour. It is not typically considered in patients with low - grade PMP.



Picture 1: CECT Whole Abdomen showing elongated hypodense lesion seen in right iliac fossa region suspicious of appendicular mucocele with mild ascites



Picture 2 & 3: Laparoscopic picture showing appendicular mucocele and gelatinous material in peritoneal cavity with white deposits over peritoneum.



Picture 3: Intra Op open laparotomy picture showing the appendicular mucocele with gelatinous material all around and omental deposits

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