

TITLE: PSEUDOMYXOMA PERITONEI – A RARE CASE REPORT



INTRODUCTION

Pseudomyxoma peritonei (PMP) is a rare condition with an estimated incidence of one to two cases per million annually. It is characterized by gelatinous abdomen often referred to as "jelly belly." It commonly arises from mucinous tumors of the appendix or ovary & clinical manifestations include abdominal distension, pain, and bowel obstruction, though many cases are discovered incidentally during surgery or imaging for other conditions. Histopathology and tumor markers such as CA 19-9 and CEA assist in diagnosis.

Management typically involves complete cytoreductive surgery (CRS) combined with hyperthermic intraperitoneal chemotherapy (HIPEC) to address residual microscopic disease. Despite advancements, long-term survival remains limited, with five- and ten-year survival rates reported between 50% and 30%.

OBJECTIVES: To present a rare occurrence of pmp and to discuss the individualized diagnostic and management strategies.

CASE REPORT: A 33 years old , multipara came with complaint of mass per vagina since 1 year

On examination, abdomen was distended extending upto 32 weeks gravid uterus size, doughy in consistency and a vague mass felt in the lower abdomen, more in the left iliac fossa and suprapubic region.

On per vaginal examination, 3rd degree uterovaginal prolapse was present with grade 1 cystocele.

ULTRASONOGRAPHY REPORT:

- 1.? Mucinous cystic neoplasm originating from left ovary
2. Cystic collection of below mentioned fluid
Gross fluid with internal septations and low level echoes is noted in peritoneal cavity extending into perihepatic and perihilar spaces forming cystic bubbly appearance. It also causes diffuse scalloping of visualized liver surfaces and shows echogenic deposition over visualized liver surface. From above findings possibility to be considered

1. Pseudomyxoma peritonei
2. Peritoneal carcinomatosis

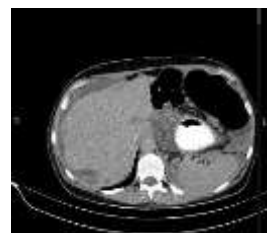
CT SCAN REPORT:

- * Multi cystic lesions in left side of pelvis, left ovary not separately visualised ? mucinous left ovarian lesions.

- * Gross amount of dense fluid in peritoneal cavity with few calcifications and causing scalloping of hepatic and splenic contours - likely pseudomyxoma peritonei.

- Elongated cystic lesion in region of appendix ? appendiceal mucocoele ?? loculated collection.

- * Omental thickening with multiple soft tissue deposits.



OPERATIVE FINDING:

On staging laparotomy, massive mucinous ascites of about 5 litres present. Left ovarian mucinous cyst of 10-12 cms present but already ruptured. Dense adhesions present between the uterus and bladder anteriorly and posteriorly with the bowel. Mucocoele of the appendix present.

Frozen section was reported as low grade / borderline mucinous cystadenoma of ovary. Fluid cytology negative for malignancy.

Adhesiolysis was done after removing as much mucin deposits as possible and a complete cytoreductive surgery with total abdominal hysterectomy with bilateral salpingo-oophorectomy with infracolic omentectomy with appendectomy was done.

FINAL HISTOPATHOLOGY REPORT:

Borderline left ovarian mucinous tumor with mucocoele of the appendix. IHC recommended to confirm primary appendiceal neoplasm.



DISCUSSION :

Pseudomyxoma peritonei (PMP) poses significant diagnostic and management challenges, as its slow progression often leads to advanced-stage presentation characterized by extensive mucinous deposits across the peritoneal surfaces. In our patient also, the diagnosis was incidental and the patient did not have any symptoms related to PMP.

Normally there are no adhesions in a case of PMP due to the Redistribution phenomenon. But in our case, there were dense adhesions which were released with great difficulty.

IHC is confirmatory to diagnose whether the primary is in the ovary or the appendix. Though advised in our case, the patient refused.

Patient was referred to a higher centre for further management with HIPEC.

CONCLUSION : The lack of a universal approach due to its rarity and incidental diagnosis most of the times underscores the complexity of managing PMP and highlights the necessity for multidisciplinary involvement to tailor effective therapeutic interventions for affected patients.

REFERENCE:

- 1) Garg P. Pseudomyxoma peritonei leading to "jelly belly" abdomen: a case report and review of the literature. Journal of Medical Case Reports. 2024 Jun 28;18(1).
- 2) Tsoukalas N, Tsapakidis K, Tolia M, Kiakou M, Galanopoulos M, Aravantinou-Fatorou E, et al. Pseudomyxoma Peritonei: A Challenging Clinical Diagnosis. Case Report and Review of the Literature. Cancer Diagnosis & Prognosis. 4(2):198–203.