

## CASE SERIES OF PSEUDOMYXOMA PERITONEI (PMP) IN A COLORECTAL CENTER HTJS

 Author: **Jaganiswaran B**<sup>1</sup>, Farizan A<sup>1</sup>, Ash-Shafhawi A<sup>1</sup>, Vishnu M<sup>2</sup>, Rosli I<sup>2</sup>

Affiliation: Colorectal Surgery Unit, Department of Surgery, Hospital Tuanku Ja'afar, Seremban, Negeri Sembilan, Malaysia



### INTRODUCTION

Pseudomyxoma peritonei is a rare type of cancer that affects the peritoneal cavity. It translates as "false mucinous tumour of the peritoneum". The term 'false tumour' is used as this cancer does not grow into solid tumours. Instead, it spreads by continuously producing mucin, a jelly-like substance that gradually accumulates in the peritoneal cavity.[1]

Classically, PMP tumours are not primary, but secondary to ruptured mucinous tumours of other organs, especially the appendix. Occasionally, PMP arises from adenocarcinomas of other sites within the gastrointestinal tract. Typically, this disorder is characterized by an abundant accumulation of mucinous ascites developing from mucin secretion by a primary tumour. The primary tumour ruptures and tumour cells then spread to implant throughout the peritoneal cavity, which results in the typical "jelly belly" appearance.

PMP does not spread through blood or lymphatic system like other cancers, but it can spread through the abdominal cavity with implant of the abdominopelvic organs surfaces. This makes it difficult to treat.[2]

### CASE PRESENTATION

We present our experience in managing four PMP cases originating from low grade mucinous neoplasm of appendix (LAMN) treated with cytoreductive surgery and hyperthermic intraperitoneal chemotherapy (HIPEC) at our center from February till October 2023. Case 1 and Case 3 presented with progressive abdominal distention for years and had undergone laparotomies and drainage of mucin at the referral hospitals. Case 2 was diagnosed with LAMN following TAHBSO for initially thought to be mucinous ovarian carcinoma and Case 4 presented with acute right sided abdominal pain at our center and CT abdomen showed primary tumour of the appendix with features of PMP. The clinical and operative data of all patients as well as post operative outcomes are as summarized in Table 1 and 2. Following pre-operative optimization, all patients underwent CCR-O Cytoreduction which includes total peritonectomy and multi-visceral resection followed by hyperthermic intraperitoneal chemotherapy with Mitomycin C (15mg/ m<sup>2</sup> BSA) over 60 minutes with temperature of 41.5°C. All patients were discharged well and are currently under close surveillance.

PATIENT	AGE/ GENDER	PCI SCORE	CCR	OPERATI NG TIME (min)	BLOOD LOSS (mls)
Case 1	64/M ECOG 1	21	CCR-0	627	2500
Case 2	46/F ECOG 0	14	CCR-0	817	3400
Case 3	51/F ECOG 2/ASA 2	30	CCR-0	567	3500
Case 4	55/F ECOG 2/ASA 2	31	CCR-0	668	3500

PATIENT	ICU STAY	TOTAL STAY	30-DAY MORTALI TY	MORBIDIT Y (CTCAE)	COMPLIC ATION TYPE
Case 1	5	15	0	1	Ileus
Case 2	5	19	0	2	Haematuria
Case 3	3	16	0	1	Ileus
Case 4	2	14	0	3	Urosepsis Ileus

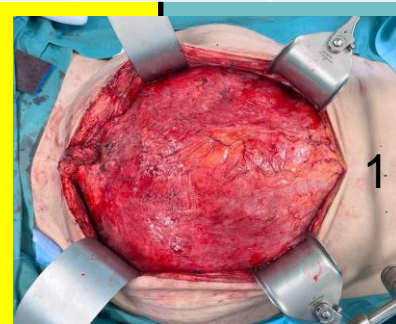


FIGURE 1 & 2 ARE FROM CASE 2  
 FIGURE 1 SHOWS PMP (PRE-PERITONEAL DISSECTION)  
 FIGURE 2 SHOWS INTRAPERITONEAL MUCINS

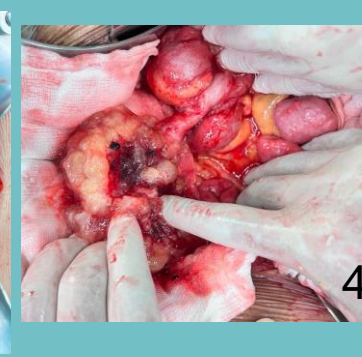


FIGURE 3 & 4 ARE FROM CASE 4  
 PMP - PCI SCORE 31 PRIMARY TUMOUR FROM LAMN

### DISCUSSION

PMP from low grade mucinous neoplasm of the appendix (LAMN) typically presents with progressive gross abdominal distention (wet type of PMP) which at the later stage cause debilitating pain, respiratory distress as well as malnutrition.

Surgical treatment with CRS-HIPEC for PMP is the gold standard treatment, with established center reporting 10-year survival rate of more than 90%. The surgical technicality of cytoreduction for high volume PMP is challenging and highly morbid as it requires complete peritonectomy with multi-visceral resection including total colectomy, splenectomy, cholecystectomy, greater and lesser omentectomy, TAHBSO in female, occasionally distal gastrectomy and patient will have multiple anastomoses created. Such surgery must be performed by team of surgeons trained in peritoneal surface surgery to limit intra-operative complications and ensure good intra-operative surgical judgement.

Mortality may result from massive intraoperative bleeding with shock and organ dysfunction or post-operative complications from anastomotic leaks, bladder and pancreatic fistulations, intra-abdominal collections/abscesses. HIPEC with Mitomycin C is associated with myelosuppression and septic complications.[3,4,5] Established high volume CRS-HIPEC centres reported mortality of 3% and severe post-operative morbidity (CTCAE 3-5) of 30%.

Patient with PMP undergoing CRS-HIPEC requires detailed pre-operative planning and pre-rehabilitation to ensure optimal physical and physiological fitness before this highly morbid intervention. Intra-operative period often lasting 10-15 hours requires good collaboration between surgeons and anaesthetists with use of invasive cardiac index monitoring for goal directed therapy to ensure patient's homeostasis is within normal limits. Post-operative period requires ICU support, intensive rehabilitation, and nutritional support by multidisciplinary teams.

### CONCLUSION

Selected patients with high volume PMP from low grade mucinous neoplasm of the appendix (LAMN) is best treated with CRS-HIPEC with acceptable post-operative complications by centers with trained surgeons and multidisciplinary support. The low-grade nature of LAMN confers long oncological survival following CRS-HIPEC.

### REFERENCES

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