

## Case Report

# A rare case of jelly belly (pseudomyxomaperitonei)

Uday Kiran Mokati\*, Kalandi Barik, L. Hari Prasada Rao, P. Vijaya, P. Avinash

Department of General Surgery, KIMS and RF, Amalapuram, East Godavari District, Andhra Pradesh, India

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**\*Correspondence:**

Dr. Uday Kiran M.,

E-mail: [drudaykiran.m@gmail.com](mailto:drudaykiran.m@gmail.com)

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### ABSTRACT

Pseudo myxomaperitonei (PMP) is a rare case with progressive dissemination of mucinous tumours and mucinous ascites in the abdomen and pelvis with an estimated incidence of 1 to 2 out of a million. Pseudomyxomaperitonei is a neoplastic disease originating from a primary mucinous tumor of appendix, mucinous cystadenoma of ovary with a distinctive pattern of the peritoneal spread. Computed tomography and histopathology are the most reliable diagnostic modalities. We present a case of pseudomyxomaperitonei in a 55 year old male presented with pain abdomen and abdominal distension with ascites. Ultrasonography localized collection noted in right iliac fossa and left iliac fossa and ascites present. Computed tomography showed localized collection noted in right iliac fossa and left iliac fossa which is arising from appendix and ascites present. Exploratory laparotomy peritoneal cavity filled with mucinous substance along with ruptured cyst wall in right iliac fossa was found. The mucinous substance drained and the ruptured cyst was resected with appendix at base. In conclusion, pseudomyxomaperitonei is a rare presentation. Despite the current standard of treatment modalities as extensive surgical resection combined with chemotherapy, PMP frequently recurs with treatment options being limited at recurrence and with severe impact on quality of life.

**Keywords:** Appendix, PMP

### INTRODUCTION

With an estimated incidence of 1-2 out of a million, pseudomyxomaperitonei is also known as adenomucinosis or gelatinous ascites listed as a rare NIH office of rare diseases research (ORDR) and national organization of rare disorders (NORD).<sup>1</sup> As an indolent neoplasm with unspecific manifestations, PMP tends to be misdiagnosed, or discovered at advanced stages. Despite a multidisciplinary approach composed of extensive surgical procedure and chemotherapy, PMP frequently recurs and increasingly decreases the quality of life.

### CASE REPORT

A 55 years male presented to emergency department with pain abdomen for 1 month with increased intensity of

pain and distension of abdomen since 5 days. On admission patient was stable with pulse rate: 70 beats/min and BP 140/90mm of Hg with tenderness over the right iliac fossa and left iliac fossa with no guarding and rigidity. No history of similar attack in the past. No abnormality detected in X-supine and erect abdomen.

Ultrasonography showed organized collection in right and left iliac fossa and ascites. Computed tomography shows Localized collection noted in right iliac fossa and left iliac fossa which is arising from appendix and ascites present. On exploratory laparotomy- peritoneal cavity was filled with thick gelatinous mucinous substance (1.5 to 2 Liters) with ruptured cyst arising from vermiform appendix at base and ileocecal junction. Gelatinous peritoneal substance was drained and whole of the ruptured cyst along with the appendix was resected at base of appendix and followed by thorough washing of peritoneal cavity with 2 liters of normal saline. Whole of

the peritoneal cavity and gastrointestinal tract from duodenojejunal flexure to rectum inspected specimen sent for histopathology examination.



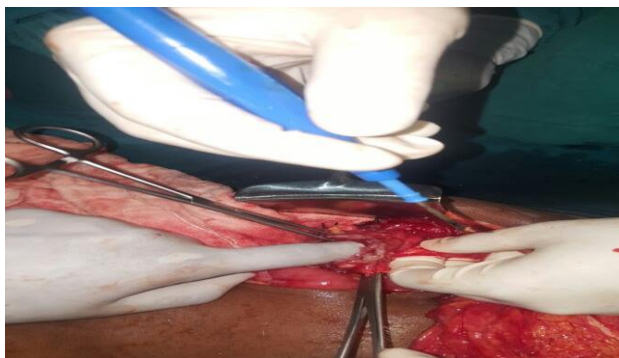
**Figure 1: Gelatinous substance in the abdomen.**



**Figure 2: Specimen of appendix and cyst wall.**

Histopathology report showed low grade mucinous neoplasm of the appendix with high risk of recurrence and pseudomyxomaperitonei changes.

Post-operative period was uneventful and patient was discharged on 10<sup>th</sup> post-operative day, patient sent for chemotherapy and follow-up done for every 3<sup>rd</sup> month.



**Figure 3: Dissecting the appendix with cyst wall.**



**Figure 4: IC junction with appendix and cyst wall.**

## DISCUSSION

Pseudomyxomaperitonei is characterized by dissemination of mucinous tumour cells on peritoneal surfaces and progressive accumulation of mucinous ascites throughout the peritoneal cavity resulting in the so called 'jelly belly'. An ovarian tumor (or) an appendicealmucocoele (or) a pre-existing intraperitoneal mucinous neoplasm has been implicated as primary cause of PMP.<sup>2,3</sup> As follows emerging evidence supports the appendiceal origin rather than ovarian origin of the disease.

Ronnet et al suggested a classification of multifocal peritoneal tumours in to three groups.<sup>4</sup>

- Group I: Disseminated peritoneal adenomucinosis (DPAM).
- Group II: Peritoneal mucinous carcinomatosis (PMCA).
- Group III: Peritoneal mucinous carcinomatosis with intermediate or discordant features.

It usually presents with lower abdominal pain, abdominal distension, ascites with pelvic pressure and gynaecological complaints in females due to the ovarian deposits of mucinous tumours.

Ultrasonography shows echogenic peritoneal masses or ascites with echogenic particles. However, conclusions cannot be drawn from ultrasonography alone since the mucinous ascitis resembles free peritoneal fluid.<sup>5</sup>

Computed tomography shows higher densities of collections (mucinous ascites) compared to non-mucinous collections, which is characteristic pattern of mucinous accumulation and the extent of disease for preoperative planning and prognostic purposes.<sup>5-7</sup>

MRI shows location of mucocoele and its morphologic criteria identically to computed tomography. T<sub>1</sub> and T<sub>2</sub> weighted MRI are more sensitive in distinguishing between mucin and fluid ascites.<sup>8-10</sup>

- T1: shows typically low signal,
- T2: shows typically high signal,
- T1C + (Gd): may show enhancement.

Treatment consists of repetitive surgical debulking for recurrence in combination with hyperthermic and intraperitoneal chemotherapy.

## CONCLUSION

Pseudomyxomaperitonei is a rare disease. Even with extensive resection combined with chemotherapy it recurs and limits the quality of life. Frequent follow ups of the patient are necessary.

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