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A rare case report on pseudomyxoma peritonei

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Abstract
Pseudomyxoma peritonei (PMP) is a rare, low-grade neoplastic condition characterized by mucinous ascites and peritoneal implantations. It often originates from an appendiceal mucinous neoplasm, though other sources may contribute. Due to its infrequent occurrence and diverse clinical presentations, PMP poses diagnostic and therapeutic challenges.

Keywords: Pseudomyxoma peritonei, abdominal gelatinous collections, mucinous cystic tumours, mucinous secreting adenocarcinoma

Introduction
Pseudomyxoma Peritonei (PMP), nicknamed "Jelly Belly", is a very rare disease with an incidence of one to two in million, yearly [1]. It is characterized by presence of diffuse intra-abdominal gelatinous collections (jelly belly) with implants of mucinous material on peritoneal surfaces and the omentum. Many cases are diagnosed accidentally when investigating or operating for other reasons. PMP is generally considered as benign; however it should be considered as borderline malignancy due to its progression over time, massive abdominal distension and nutritional compromise leads the long term survival poor in most patients [2].

Case report
A 59-year-old male, operated case of lap converted to open appendectomy, presented to Surgery OPD with complaints of abdominal distension, abdominal pain in Right Iliac fossa and anorexia with 5-6 kg of weight loss since one month.

- On examination a 5 cm x 4 cm hard, irregular, non-tender, non-mobile mass was present in the Right lliac fossa.
- USG abdomen was suggestive of appendicular lump with mild ascites with multiple internal echos.
- His CA 19.9 was >400, rest other routine blood investigations were within normal range.
- On suspicion of malignancy CT scan was done which showed hypodense mass like wall thickening involving lateral wall and base of the caecum with multiple loculated fluid collections throughout the peritoneal cavity, with involvement of omentum, mesentery, liver, spleen and pancreatic head suggestive of pseudomyxoma peritonei.
- Patient was posted for surgery for Right Hemi-colectomy and Evacuation of mucinous material but greater omentectomy and debulking was done due to in-operability.
- Post-operative period was uneventful and he was advised chemotherapy at higher centre.
- Histopathology of omentum was suggestive of mucinous secreting adenocarcinoma.

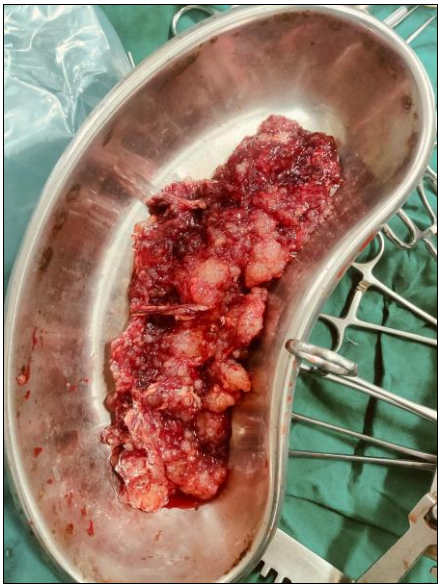

Fig 1: Intra-operative photographs of Gelatinous Material

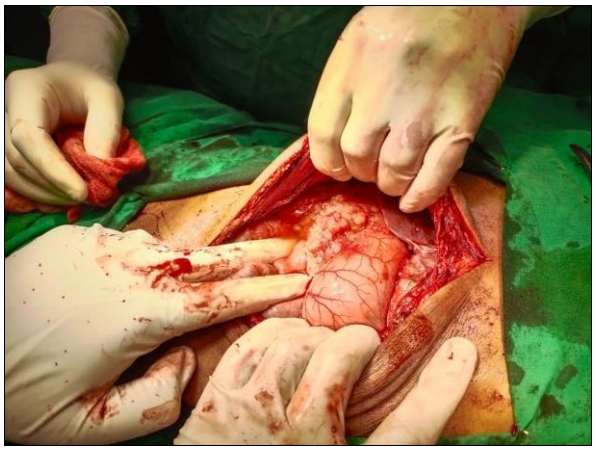

Fig 2: Intra-operative photographs of Gelatinous Material

Discussion

PMP is rare condition occurs more frequently in women, which is associated with mucinous cystic tumours of Ovary and Appendix [3]. The majority of patients are diagnosed during, or after, a laparotomy or laparoscopy, for suspected appendicitis, peritonitis or gynaecological cancer. Imaging modality for the diagnosis and staging of PMP is CT scan. The treatment modalities are appendectomy with Right hemi-colectomy/Pan-hystrectomy with omentectomy are done. Intra peritoneal instillation of chemotherapeutic agents like 5FU, Mytomicin C, Oxaliplatin and mucolytic agents like Dextran, Urokinase after surgery are beneficial [4].

Conclusion

Pseudomyxoma peritonei known for its rarity and non-specific abdominal presentation. Despite of the current standards of treatment, PMP frequently recurs, with limited treatment options. Appropriate timed imaging studies and multidisciplinary expertise are crucial in the diagnosis and management. Considering the substantial role of mucin in the pathogenesis of PMP, development of strategies for targeting mucin and its biology seems to be of particular significance [5].

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Conflict of Interest

Not available.

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