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PSEUDOMYXOMA PERITONEII: A CURIOUS ENTITY

General Surgery

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ABSTRACT

Pseudomyxoma paritoneii (PMP), also known as jelly belly, is a rare clinical entity, majority of which arise from mucoid neoplasms of the appendix and is known to affect females more than males. Here we present a case of PMP in a 40yr old female arising from a cystadenoma of the ovary with no gross pathology involving the appendix.

KEYWORDS

Pseudomyxoma peritonei – appendiceal neoplasm – mucinous cystadenomas – intraperitoneal spread - cytoreductive surgeries – chemotherapy

INTRODUCTION

The term pseudomyxoma peritonei was first coined by Werth in 1884. It was thought to arise from a perforated cystadenoma of the appendix. Based on recent studies it has been more widely used to describe peritoneal dissemination of mucus-producing neoplasm mostly originating from the appendix, but also small and large bowel, stomach, pancreas, lung, breast, gallbladder, fallopian tubes, and ovaries. [1][2][3]

PMP has an indolent course and often discovered incidentally with a relatively advanced stage during laparoscopy, laparotomy or imaging studies for other medical concerns. Pseudomyxoma peritonei merits consideration as a 'borderline malignancy' with changing prognosis based on the site of origin. [4] Three categories of PMP were agreed—low grade, high grade, and high grade with signet ring cells. [3]

The incidence of pseudomyxoma peritonei is estimated approximately 1 to 4 out of a million annually. [2] The primary site is identified predominantly as mucinous appendiceal adenocarcinoma. [5] Patients have an average age of 53 years at the time of diagnosis. Females are affected more frequently than males. [6] Up until a decade ago, the annual incidence of PMP was estimated to be approximately 1.7–2 per million person-years in the Netherlands and about 1 per million person-years in Japan [7]. Recent data by Patrick-Brown et al. calculated an incidence of 3.2 per million, with a corresponding estimated prevalence of 22 per million annually, which was obtained from the number of people who had undergone surgery for PMP in Norway and England. [8]

The tumor cells arising from the mucinous epithelium of the appendix continuously produce mucus into the appendiceal lumen and form a mucocele, which eventually ruptures. Free-floating mucinous epithelial tumor cells get deposited in various parts of peritoneal cavity following the intraperitoneal fluid current and gravity. It is termed as the so-called 'redistribution phenomenon.'[9] The greater and the lesser omentum are the major sites of tumor deposits during the early stage of the disease. [5] But the end-stage results in involvement of the entire peritoneal cavity, including extraperitoneal spread mostly to the pleural cavities. [10,11,12] In females, there may be excessive growth on both ovaries. [13]

Case History

A 43 year old, married, hindu female presented with complaints of gradual abdominal distension over the past 2 years. It was associated with mild abdominal discomfort, and difficulty in breathing. There was no history of hematemesis, melena, jaundice, hemoptysis, previous blood transfusion. The patient did not have any known co-morbidities, history of TB, or contact history of TB, use of contraceptives. There is no family history of carcinoma of the biliary tree, carcinoma head of pancreas, carcinoma colon. There is no history of any addictions. The patient had a history of infertility with regular menstrual cycles and attained menopause 2years back.

Fig.1:- Depicts The Grossly Distended Abdomen In A Patient With PMP.

On examination, the patient had a medium built, had mild pallor and bilateral non-pitting pedal edema, no icterus, no clubbing, no lymphadenopathy. Per abdomen examination revealed, protuberant abdomen with flanks full, and centrally placed umbilicus, no engorged veins or any visible swelling. On palpation organomegally could not be appreciated, a dull note on percussion. No audible bruits were heard on auscultation. CVS and Respiratory system examination did not reveal any abnormal findings.

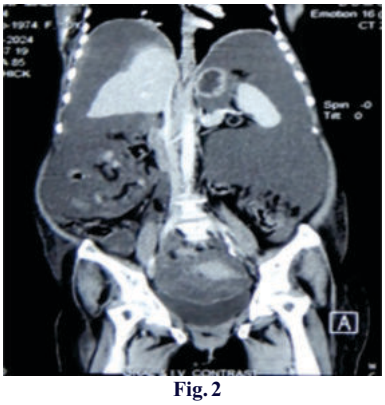
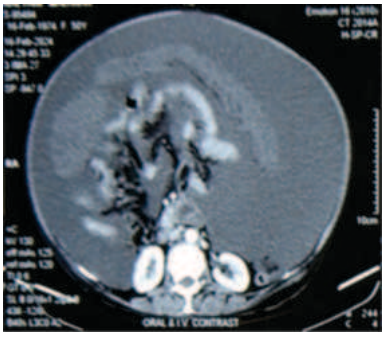


Fig. 2



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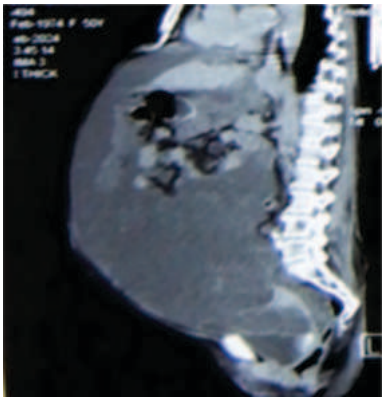

Fig. 4

Fig. 2- Shows The Scalloping Of The Liver Margins, With An Ovarian Sol; Fig. 3 And 4- Shows The Axial And The Lateral Cuts.

CECT whole abdomen revealed scalloping of liver margins with gross ascites and ovarian SOL. Cancer markers like CA-125 was raised, LFT and PT/INR were normal. CECT thorax showed bilateral moderate pleural effusions. Diagnostic tapping was done and revealed a jelly-like substance instead of ascitic fluid. The patient was prepped, and along with a team of gynaecologists, taken up for exploratory laparotomy and TAH-BSO. Per-operatively around 15L of jelly-like material extracted out, a multiloculated mucinous cystadenoma of ovary identified, with no appendiceal pathology and diffuse mesenteric infiltration noted. Post operatively the patient was shifted to CCU, due to unresolving hypotension inspite of giving colloids and multiple blood transfusions. Following that the patient developed respiratory distress on POD-3 was put on mechanical ventilation, then she went into cardiac arrest but could not be resuscitated. Histopathology of the ovarian tumour shows high grade mucinous carcinoma with high cellularity, nuclear atypia and mucin pools.

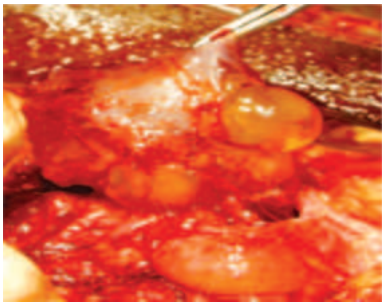

Fig. 5

Fig. 6

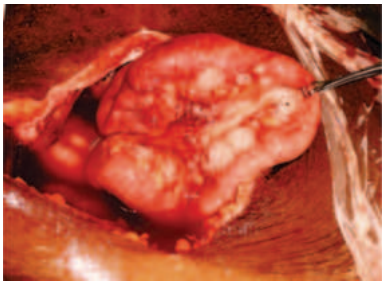

Fig. 7

Fig. 5: Shows The Left Ovarian Multi-loculated Tumour; Fig.6: Shows The 'jelly-like" Material Extracted After Incision, Around 15l Were Extracted; Fig.7: Shows The Mesenteric Infiltration By The Tumour

DISCUSSION

At the initial stages psedomyxoma peritonei presents with non-specific symptoms or is asymptomatic and often misdiagnosed. So along with physical examination, radiological investigations (like CECT, contrast MRI and in some cases PET scan), and biochemical markers (like CA-125 for ovarian origin and CEA and CA19-9 for appendiceal origin) helps in early diagnosing and staging of the disease and further helps outline the treatment options and determine the prognosis.

However histopathological examination gives the definitive diagnosis based on which the treatment protocols can be individualised. It was Ronnett et al. who first divided pseudomyxoma peritonei into two groups: disseminated peritoneal adenomucinosis (DPAM) and peritoneal mucinous carcinomatosis (PMCA). [14] DPAM is characterized by abundant mucus containing scanty mucinous epithelial cells with minimal cytological atypia and mitotic activity, while PMCA is featured by more abundant mucinous epithelial cells with high-grade cytological atypia and mitotic activity. Later on in 2010, the World Health Organization (WHO) further refined the grading system. [15][16]

- Acellular mucin: Mucin within the peritoneal cavity without neoplastic epithelial cells.
- Low-grade mucinous carcinoma peritonei (synonymous with DPAM) presents as mucin pools with low cellularity (less than 10%), bland cytology and non-stratified cuboidal epithelium. Tumor cells are arranged in strips or gland-like structures. Infiltrative growth is not present.
- High-grade mucinous carcinoma peritonei (synonymous with PMCA): Mucin pools with high cellularity, moderate/severe cytological atypia, numerous mitoses, and cribriform growth pattern. Destructive infiltrative invasion of underlying organs is often present.
- High-grade mucinous carcinoma peritonei with signet ring cells: Any lesion with a component of signet ring cells, classified separately because of their worse prognosis.[17][18]

Treatment protocols at present aim at complete resection in terms of cytoreductive surgery (CRS) combined with hyperthermic intra-peritoneal chemotherapy (HIPEC). Previously adopted measures like periodic surgical debulking have been discouraged due to inevitable disease recurrence and repeated surgeries. In a study Viara et al showed that if preoperative chemotherapy was performed it had a negative prognostic factor with statistically significant impact both on overall survival (OS) and disease free survival (DFS). [19]

Another study where 104 patients with PMP were treated with combined CRS and HIPEC, it showed that favorable outcome after comprehensive treatment can be expected in patients with DPAM, not treated with preoperative systemic chemotherapy and amenable to adequate cytoreduction. [20]

Another point of debate that has evolved in the recent times, is regarding the origin of PMP whether it has a predominant ovarian or gastro-intestinal (appendiceal) origin. In a study by Brigitte et al, it was stated that of the 68 cases of women having PMP 30 of them had tumors involving ovaries. After taking into consideration certain gross and microscopic features it was concluded that 28 of them had appendiceal tumors as the primary and the remaining two were either consistent with intestinal or ovarian origin.[22]

The treatment guidelines however does not differentiate on the origin

of the tumor and consensus reveals a combined approach of CRS with HIPEC to have shown greater overall survival.

CONCLUSION

Though the patient did not survive, we present this case to highlight its rarity, and to emphasize that early diagnosis can be achieved if the index of suspicion is kept high and that significant survival is achievable by combination therapy, particularly in lower grade tumors.

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