Case Report

A Rare case report on Pseudomyxoma peritonei

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Abstract

Pseudomyxoma peritonei (PMP), nicknamed "Jelly Belly", is a very rare disease characterized by presence of jelly like fluid called mucin in the abdominal cavity. The natural history of PMP revolves around the "redistribution phenomenon", whereby mucinous tumor cells accumulate at specific sites with relative sparing of the motile small bowel and to a lesser extent other parts of the gastrointestinal tract. This paper reports a case of an elderly woman, who was diagnosed with pseudomyxoma peritonei, who underwent surgery for the same and intraoperatively she was suspected to have primary arising from the ovary, postoperatively the histopathology report said that the primary was identified to be arising from the appendix. This also shines light on how cytoreductive and debulking surgery play a major role in prolonging the life span in such patients.

Key words

Pseudomyxoma peritonei, Redistribution phenomenon, Cytoreductive surgery, Debulking surgery, HIPEC (hyperthermic intraperitoneal chemotherapy), Appendicular carcinoma.

Introduction

Pseudomyxoma peritonei (PMP), its incidence is approximately 2 per million annually. However, experience from high volume centres suggests that the actual incidence may be higher at 3–4 operable cases per million per year [1]. PMP appears to be more common in women, who often present with rapidly enlarging ovarian masses, secondary to transcoelomic spread [2, 3]. Nonetheless, due to its indolent nature, it is usually discovered at an advanced stage and severely impacts quality of life [4]. First case was reported by a gynecologist, Dr R Werth, in 1884. Carl Rokitansky was the first to describe

an appendiceal mucocele in 1842 [5]. Earlier the cause was unknown and the discussion regarding the etiology emerged very recently, in the first quarter of the 20th century. The disease has a wide spectrum, from slow growing benign lesions to rapidly progressive infiltrative disease [6]. It is increasingly noted that more patients are currently being diagnosed on imaging for unrelated pathology or as an unexpected finding at laparoscopy/laparotomy for an acute abdomen [3, 7]. In 1 to 20% of the patients it is coincidently detected radiologically or at laparotomy or during hernia repair when mucus is found in the hernia sac.The classical distribution of PMP deposits has been termed the "redistribution phenomenon". This phenomenon results from movement of free-floating epithelial cells with little, or no, adhesional properties. Cells move with the peritoneal fluid and by gravity. Proliferation of tumor deposits in predetermined sites of the peritoneal cavity is therefore dictated by gravity and concentration at the sites of peritoneal fluid absorption [8]. It is not a primary peritoneal disease. Its a disseminated disease from a primary lesion of one of the intraperitoneal organs, which being Appendix, Ovary, Colon, Stomach, Pancreas, and Urachus. From these organs the mucin break out and spread into the peritoneal cavity. The interval between the discovery of the primary tumour and the clinical diagnosis of PMP vary significantly (2 years to 20 years) making it extremely difficult to diagnose in early stages. The CT scan of the chest, abdomen and pelvis with intra-venous and oral contrast is the imaging modality of choice for PMP [9, 10]. The complications associated with this condition are dreadful. As only 50% present with symptoms, majority lies in the undetected zone, which can lead to serious complications like intestinal obstruction, severe ascites, menstruation problems and infertility. So long term survival can be achieved by surgery alone. In cases where complete tumour removal is not feasible, maximum tumour debulking can still result in long term survival. PMP is challenging, complex but nevertheless the recommended treatment for PMP is a combined strategy of complete macroscopic tumor removal (entitled complete cytoreductive surgery, CCRS), combined with HIPEC [11-13].

Case report

An elderly hypertensive lady presented to our OPD in February 2020, with distension and pain in abdomen for a period of 8 months. She also gave history of swelling of both lower limbs, vomiting, fever, cough, loss of appetite, weight and breathlessness. On examination abdomen was grossly distended, with dilated veins over it, everted umbilicus (**Figure - 1**) with presence of generalised mild tenderness. On radiological examination, USG Abdomen and pelvis revealed multiple septations and loculations with echogenic content within peritoneal cavity, with omental thickening with scalloping of liver and spleen with thick collection overlying the surface, suggestive of PMP. On further investigation, On CECT Abdomen and pelvis, there was gross collection noted in peritoneal cavity with proteinaceous contents, displacing bowel loops due to gross ascites, features suggestive of peritoneal carcinomatosis (**Figure - 2a, 2b**). CEA levels were 1800.

Figure - 1: Dilated veins and umbilicus everted and pushed downwards.

Patient underwent Surgery on 12th of February 2020, exploratory laparotomy (debulking procedure) under general anesthesia, in which 12 litres of yellowish jelly was removed (**Figure - 3**). Parietal peritoneum involvement was seen and was accompanied by excision of the mass

(**Figure - 4**), appendectomy (**Figure - 5**), type A hysterectomy (**Figure - 6**), omentectomy (**Figure - 7**), parietal peritonectomy (**Figure - 8**). Intraoperatively the primary appeared to be arising from the ovary as the gross appearance of appendix was normal. Post histopathological assessment the primary organ involved was seen to be the appendix (well differentiated, grade 1, low grade appendiceal mucinous adenocarcinoma). IHC markers came strongly positive for CK-20 and CDX-2, and negative for CK-7, PAX-8 and TTF-1. Post-operative period was uneventful and she was advised chemotherapy and follow up, however patient did not undergo chemotherapy and patient expired in the first week of April i.e.; during the nationwide COVID 19 lockdown.

Figure - 2a: CECT abdomen and pelvis (axial view) showing gross collection in the peritoneal cavity with proteinaceous contents, displacing the bowel loops anteriorly.

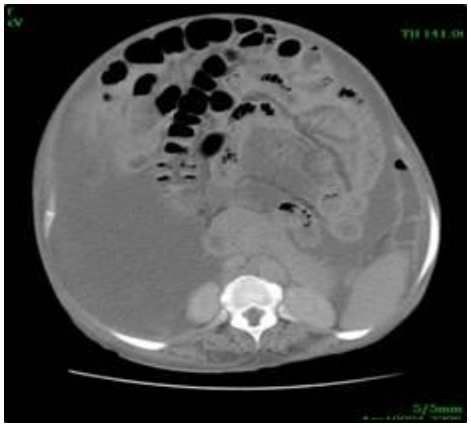

Figure - 2b: CECT abdomen and pelvis (lateral view).

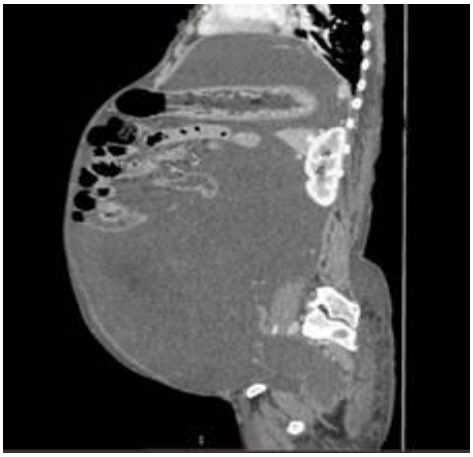

Figure - 3: Approx. 12 litres mucinous material removed.

Figure - 4: Mass cut after clamping**.**

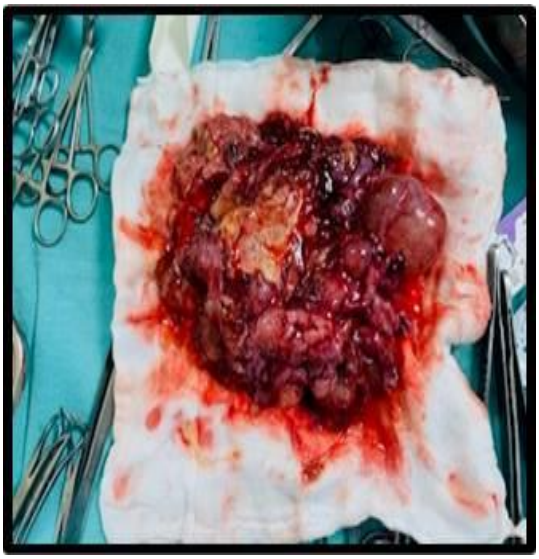

Figure - 5: Appendectomy.

Figure - 6: Type A hysterectomy.

Figure - 7: Omentectomy.

Figure - 8: Parietal peritonectomy.

Discussion

PMP is primarily described as a peritoneal disease looking like a myxoma but it's neither a real myxoma nor a primary peritoneal disease. It's a disseminated disease from a primary lesion of one of the intraperitoneal organs. It has two theories: 1) Theory of secretion (intraperitoneal extravasation of mucus secretion). 2) Theory of collection (intraperitoneal collection of gelatinous material and mucinous tumour cells). Gravity accumulates deposits in the pelvis. 50-80% patients will have symptoms reflecting the location of the primary or the metastatic tumor. 20-30% Female patients will be detected with ovarian mass and will be on evaluation for low abdominal pain or menstruation problem or infertility. 1-20% patients are coincidentally detected on USG or CT scan or at laparotomy for any other reason when mucus is found in the abdomen, less than 1% patients present with suspected bladder tumour or right femoral neuropathy. In most female patients it is commonly misunderstood that the primary arises from the ovary, likewise understood in our case, post-surgery after histopathological examination we found the primary was arising from the appendix, and the post op immunohistochemistry markers suggested the same. Initially described by Sugarbaker, et al. [8] in 1995, cytoreductive surgery consists of six resections that aim to decrease tumor burden from peritoneal surfaces. The resections include: greater omentectomy-splenectomy, left upper quadrant peritonectomy, right upper quadrant peritonectomy, lesser omentectomy-cholecystectomy, pelvic peritonectomy with resection of the sigmoid colon, and antrectomy [8]. The treatments are cytoreductive surgery with peritonectomy to ensure removal of all visible evidence of the disease from the abdomen and pelvis. As the mucinous tumor is so widely distributed throughout the abdomen and pelvis, the entire surgery takes 10-12 hours. Then, to prevent reimplantation of malignant cells, the abdomen is washed with a warm chemotherapy solution. This is usually referred to as hyperthermic intraperitoneal chemotherapy or HIPEC. The hyperthermic intraperitoneal drugs usually used include mitomycin C and oxaliplatin. The chemotherapy is heated to 42 degrees in the abdominal cavity to accentuate the penetration of drugs into the mucinous tumor and to increase

the local cytotoxicity. Suitability for surgery is based on a peritoneal cancer index (PCI), with a PCI ≥ 20 representing unresectable disease [14]. The importance of this grading system is that tumors larger than 2.5 cm, if not resected during CRS, will not be eliminated by HIPEC [15]. Resections are deemed complete if surgeons are able to achieve CC0 or CC1. Serum tumor markers are helpful in predicting aggressiveness of disease. In patients who are secretors, elevated tumor markers help in follow-up and early identification of recurrence [16]. The three tumor markers commonly used in PMP are the carcino-embryonic antigen (CEA), carbohydrate antigen 125 (CA 125) and carbohydrate antigen 19.9 (CA 19.9) [17]. There has been a recent pilot study reporting that CEA levels reduce, or normalize, at 7 days after complete CRS [18].

Conclusion

Pseudomyxoma peritonei known for its rarity and non-specific abdominal presentation, clinician's awareness of this disease entity is helpful in early diagnosis and prompt action. The treatment modality is focused on cytoreductive surgery with hyperthermic intraperitoneal chemotherapy (HIPEC). Our case also highlights the need for a multidisciplinary approach in achieving a good palliation and long term survival.

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