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The Great Imposter: A Rare Case of a Pseudomyxoma Peritonei Mirroring Severe Appendicitis

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Abstract

PMP (Pseudomyxoma Peritonei) is an uncommon disease occurring most commonly via pierced Appendiceal epithelial tumour followed by ovaries. The best therapy contains a mix of CRS (Cytoreductive surgery) along with intense HIPEC (Intraperitoneal chemotherapy). There is an argument related to the diagnostic category on its predictive significance. Tomography computer imaging is the best pre-operative method. High tumour indicator is associated with poorer diagnosis and bigger reappearance rates. A 33-year-old female was admitted to the emergency department of Sree Balaji Medical College and Hospital with severe pain over the right iliac fossa with four episodes of vomiting and one episode of fever. On Examination, he had Tenderness and Rebound Tenderness over the Right Iliac Fossa. CECT showed features suggestive of Acute Appendicitis and the patient was taken up for Surgery where a biopsy showed Pseudomyxoma Peritonei. The patient further underwent HIPEC and was followed up for 1 year with no recurrence of the disease.

Keywords: *Appendiceal Mucinous Tumour, Cytoreductive Surgery, Intense Intraperitoneal Chemotherapy, Jelly Belly, Marginal Malignancy and HIPEC, Peritoneal Cancer Index PCI, Pseudomyxoma Peritonei, PMP, Peritoneal Malignancy.*

Introduction

PMP is thought to be harmless, but its actions indicate that it should, in the best of scenarios, be regarded as a marginal tumour, with the syndrome proceeding to a huge abdominal distension and dietary settlement in many instances. The survival in the long term in many patients stays bad, with a report of 5-year and 10-year survival chances of 50% and 10% to 30%, respectively. In recent times there has been a worldwide note in the management of the PMP, especially in the macroscopic elimination of tumours by complicated surgical methods along with intense HIPEC [1].

PMP is a rare medical thing with just one or two in a million in a year appearance [2]. It is categorised by dispersing intra-abdominal jellylike groups, also known as jelly-belly, and with Mucinous inserts on the peritoneal areas and in the omentum [3].

Quite a few cases exhibit without a warning at a laparotomy or laparoscopy and are guessed to be at a cross-section of imaging in the examination or showing a different diagnostic being. Therefore, those who do the operation in the abdominal cavity will come across a random occurrence and will encounter diagnostic ambiguity because of the uncommon aspect of the PMP and due to the absence of verification or agreement on the management.

PMP (Pseudomyxoma Peritonei) is an uncommon disease with a bleak diagnosis if it isn't addressed correctly. It is advised that a first step to better the diagnosis of the patients is to acknowledge this medical condition, if possible, in the initial phase. Expertise in pathogenesis and general diagnostic equipment

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is important here. Radiologist help should be part of the diagnostic process, and CT imaging should be used [2].

After CRS with HIPEC, the chances of survival in 5 years vary from 62.5% to 100% for small evaluations and 0%-65% for higher evaluated diseases. In a treatment, illness and death can be from 12% to 67.6% and 0% to 9%, respectively. The best treatment for PMP is surgery and HIPEC, which, even in the best of scenarios, is a decent 'marginal' peritoneal malignancy [2].

In 1884, Werth created the name PMP about a mucinous tumour in the ovary [4].

Frankel in 1901 described an occurrence related to an appendix cyst. In actuality, the clinical-pathology thing, PMP pattern or jelly belly, signifies a disease. This can be from Mucinous ascites related to an Appendix Cystadenoma, which is the real PMP, an honest Mucinous Adenocarcinoma. In addition, intestinal mucinous tumours, especially colorectal cancers, or other mucinous neoplasms may have medical, radiological, and pathological characteristics similar to PMP [5].

Case Report

A 33-year-old female was admitted to emergency with acute pain on the right iliac fossa with vomiting and about four episodes of non-bilious vomiting. In addition, she had an incidence of fever. She had no other issues in the past.

Upon inspection, tachycardia was observed. As per her abdominal check-up, flabbiness was seen and guarding on the right iliac fossa with tenderness and rebound - tenderness. The rectal and vaginal examinations were normal.

Her blood work showed the total WBC count was higher, ~14,000 × (10)9 / L.

In preoperative, Alvarado's score was measured and was 8/10.

Abdomen CT was indicative of a long and inflamed appendix with a measure of 16.8 mm in diameter with peri-appendiceal fat cut off and flanking the collection. The right ovary was not visible, and, in its place, a 9.0 cm x 4.4 cm x 6.1 cm cystic lesion was visible in the right adnexa, more likely right hydrosalpinx/less likely right para-ovarian cyst (Figure 1; Figure 2). An initial diagnosis of acute appendicitis/mucocele of the appendix was made. A diagnostic laparoscopy was planned. Trocar was non-negotiable due to peritoneal nodularity, thus converted to lower midline laparotomy.

Intra-operative findings: PCI (Peritoneal Cancer Index) score of 14 was calculated intraoperatively (Figure 3).

A 6 cm×5 cm gelatinous matter-filled cavity was detected on the right Iliac fossa at the ileocecal junction, likely to be appendiceal neoplasm (Figure 4). Bilateral ovaries palpable separately, right cystic. Debulking of the sample was completed and sent to pathology (Figure 5).

Dense omental adhesion is seen. Right iliac fossa mass was detected with verification of extra mucinous/gelatinous matter placed on the intra-abdominal cavity, which included the bowel, omentum, and parietal peritoneum, with some traces of it on the liver (Figure 6; Figure 7).

Postoperatively, the patient was started on symptomatic treatment with antipyretics and analgesics, and the patient improved symptoms.

Histopathology showed Low-Grade Mucinous Neoplasm involving muscles and fat (peritoneal fat)/Mucinous Carcinoma Peritonei, Grade 1 (Pseudomyxoma Peritonei, Grade 1).

Immunohistochemistry was completed and returned positive for CK20 and CEA (suggestive of appendiceal origin). The patient was moved to an oncology centre where she underwent HIPEC clinically and was followed up for 1 year, and no recurrence of the disease was observed.

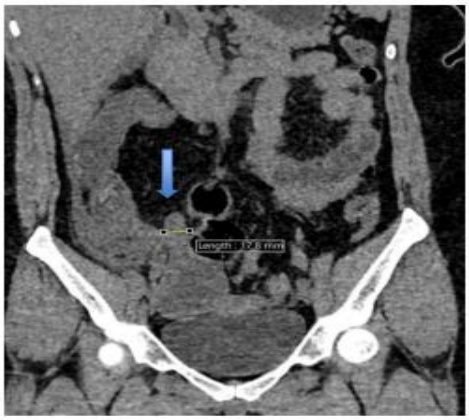

Figure 1. CT Abdomen Coronal Plane Showing Dilated Appendix (Blue arrow)

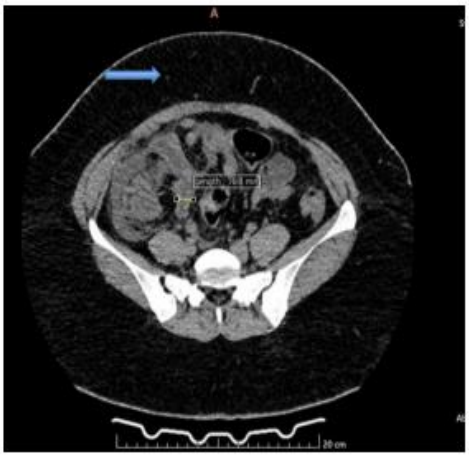

Figure 2. CT Abdomen Transverse Plane Showing Dilated Appendix (Blue Arrow)

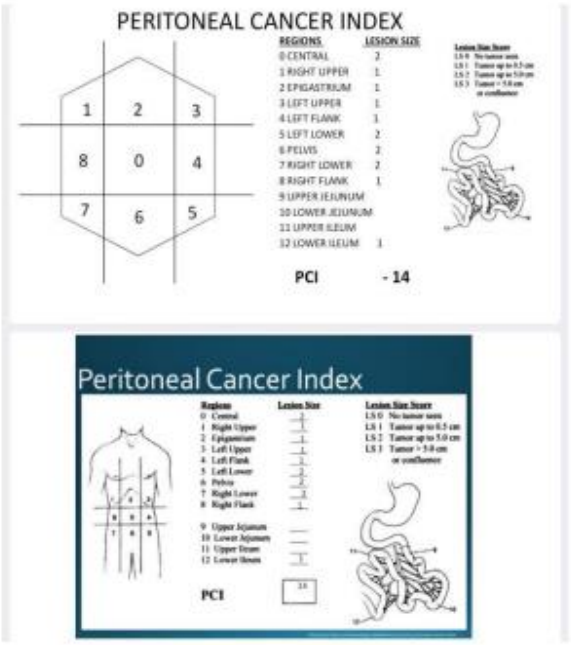

Figure 3. Showing Peritoneal Cancer Index (PCI)

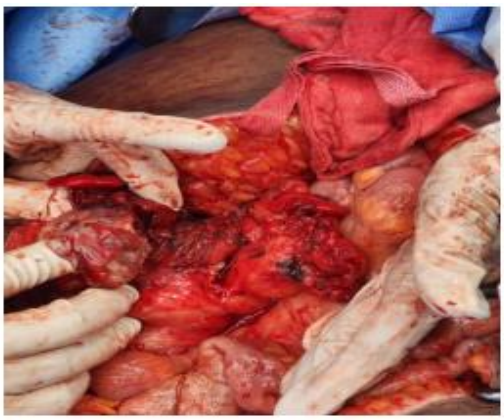

Figure 4. Showing Exhibits Intra-Operative Images of 6cm×5cm Gelatinous Matter Filled Cavity (Black arrow)

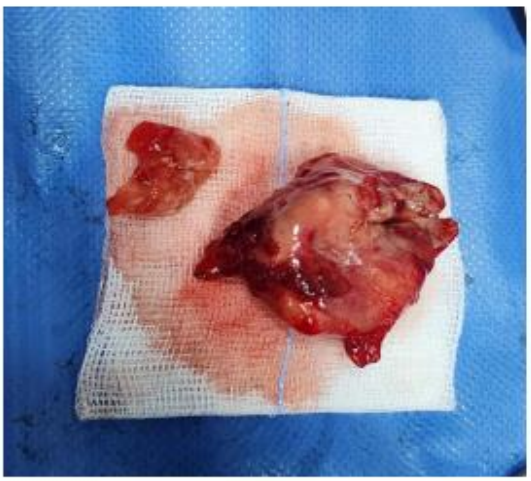

Figure 5. Exhibits Gelatinous Matter Filled Cavity and Appendiceal Neoplasm

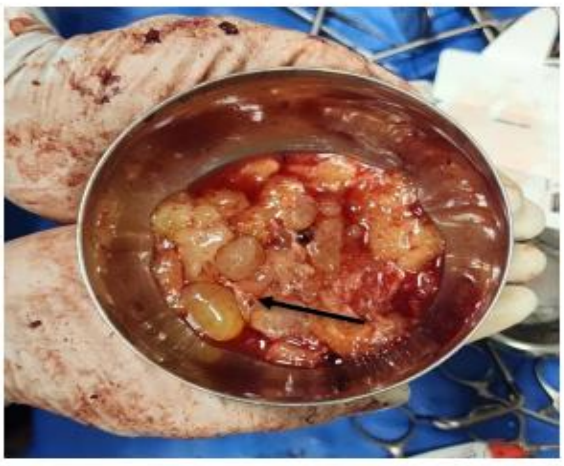

Figure 6. Showing Gelatinous Material Deposits

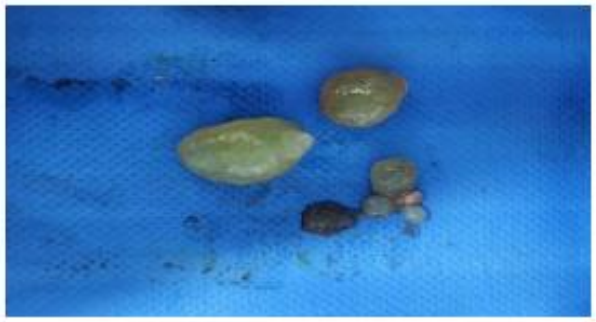

Figure 7. Exhibits Gelatinous Collections 'Jelly Belly' with Mucinous Deposits

Discussion

PMP (Pseudomyxoma Peritonei) is an uncommon illness known for its chances of death if not attended to appropriately. It is advised that a first step to better the diagnosis of the patients is to acknowledge this medical condition, if possible, in the initial phase. Expertise in pathogenesis and general diagnostic equipment is important here. The cure plan for PMP should follow complete cytoreduction and avoidance of reappearance or progress [6].

The merged method of treatment, containing cytoreductive surgery along with hyperthermic intraperitoneal chemotherapy, appears quite effective here. This method is presently being used in most centres in the world with encouraging outcomes and appears to gain support as the customary care method [7].

PMP is a rare disease categorised by Mucinous ascites, typically initiating through a fractured low-grade Mucinous neoplasm of the appendix [8, 9]. PMP narration is from the "restructuring phenomenon," where the mucinous tumour cells collect at distinctive areas with the comparative freeing of the motile small bowel to a smaller degree to additional areas of the abdominal expanse [10].

Peritoneal tumours gather because of the pull and at the areas where there is absorption of peritoneal fluid, which is in the omentum and the area below the diaphragm, mainly to the right. The best cure is completed macroscopic tumour excision, described as cytoreductive surgery (CRS) mixed with HIPEC. The complete operation for CRS plus HIPEC for widespread PMP takes about ten hours and comprises two-sided parietal and diaphragmatic peritonectomies, right-side hemicolectomy, radical greater omentectomy with splenectomy, cholecystectomy, liver capsulectomy, a pelvic peritoneectomy, having or not having rectosigmoid resection, and two-sided salpingo-oophorectomy with hysterectomy in female patients [10].

Conclusion

In conclusion, PMP is a rare and intractable entity. Special attention should be paid to its preoperative assessment, including early diagnosis, pathologic classification, and peritoneal cancer index. Currently, PMP is treated with complete cytoreduction combined with heated intraoperative intraperitoneal chemotherapy as a widely accepted and even standard curative treatment.

Conflict of Interest

There is no conflict of interest.

Acknowledgement

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