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A 70 years Woman Presented With Pseudomexoma Peritoni Secondary to Primary Ovarian Mucinous Cystadenocarcinoma: A Case Report

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Abstract

Pseudomyxoma peritonei (PMP) is a rare and life-threatening condition of the parietal peritoneum that produces abundant mucin or gelatinous ascites. The tumors cause fibrosis of tissues and impaired digestion or organ function, and if left untreated, the tumors and mucin they produce will fill up the abdominal cavity. This will result in compression of organs and will destroy the function of the colon, small intestine, stomach, and other organs. Prognosis with treatment in many cases is optimistic, but the disease is lethal if untreated, with death occurring via cachexia, bowel obstruction, or other types of complications. Symptoms can be non-specific and ignorable for several years such that most cases are diagnosed during explorative surgery.

A 70 yrs old woman admitted in Popular Medical College Hospital with anorexia, discomfort with abdominal distention and constipation. She was diagnosed clinically,with the help of CT scan and tumor marker as multi septated large ovarian tumor with incisional hernia.She had past history of ovarian cystectomy 2 years back. After laparotomy huge amount of mucin was found with in the abdominal cavity, probably from ovarian origin and then we had done total abdominal hysterectomy with bilateral salpingo-ophorectomy, appendectomy, removal of all mucin from abdominal cavity with hernia repair. Hystopathologically it was detected as low grade mucinous cyst adenocarcinoma and she completed 6 cycles chemotherapy. Her oneyear follow up period was completely uneventful. We report an interesting and rare case of Pseudomyxoma peritonei who had undergone successful surgery and post oparetive chemotherapy. She is under regular follow up yearly. She is now symptom and disease free. This case is presented with review of Literature here.

Conclusion: *It is a rare disease. Reporting the case may help the physician to understand the outcome of pseudomyxoma peritonei.*

Keywords: *Pseudomyxoma peritonei,Mucinous cystadenocarcinoma.*

Introduction

Ovarian cysts occur commonly in women of childbearing ages [1].They are usually benign and therefore tend to be asymptomatic in the majority of the population. The differential diagnoses of benign ovarian cysts include serous cyst, dermoid cysts, brenner cysts, and mucinous cysts. Of all the ovarian tumors, mucinous cystadenomas account for 15%-20%. The overall incidence of pseudomyxoma peritonei was previously estimated at 0.5 to 1 case per 100,000 people per year [2]. Pseudomyxoma peritonei is a rare type of neoplasm. It is characterized by the presence of mucinous tumors on the peritoneal surface, with excessive mucin production [3]. Generally, the primary tumor originates in the mucinous epithelium of the appendix [3,4]. In the latter

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stages of the disease, it completely involves the peritoneum, leading to symptoms [3,4]. It can also originate from the ovary, pancreas, bile ducts, colon, and gallbladder.

Recent research in Europe indicates that the previous estimation of 1-2 persons per million.[5] May be underestimating the actual rate by approximately half, with the real incidence being approximately 3.2 persons per million, and the prevalence being 22 persons per million. It is slightly more common in women than men (male:female ratio of approximately 1:1.3[6], although the actual ratio is difficult to identify due to potential misdiagnoses and possibly inclusion bias in reported studies. The median age at presentation is typically about 50 years with a range of 20-25 years, but PMP may strike persons of any age.[6] [7]

Pseudomyxoma peritonei arise from the ovarian surface epithelium and have smooth inner and outer thin walls. Specifically, benign mucinous cystadenomas comprise 80% forming the majority of ovarian mucinous tumors; 10% of these tumors are found to be malignant and the remaining 10% are borderline [8]. A striking feature for these benign mucinous cystadenomas is that they could become massive in size ranging from 5 to 28 cm with larger size increasing the risk of malignancy [9]. Pseudomyxoma peritonei deposits are consistently seen within the greater omentum, lesser omentum, and beneath the right hemi-diaphragm. However, the tumor is absent on the peritoneal surfaces of the intestine and mesentery due to peristaltic activity [10].

The purpose of our case report was to evaluate outcome and long-term survival of our patient after cytoreductive surgery (CRS) and chemotherapy as it is rare case.

Case Report

Mrs. Halima, widow 70 years postmenopausal woman who presented with known case of ovarian tumour with incisional hernia and with the complaints of lower abdominal pain for 5 months along with abdominal distention and constipation. She was mother of 2 children and referred to OBGYN department of PMCH for further management. The patient's symptoms were neither associated with lower back pain, swelling of her feet, nor with excessive straining, dysuria, urinary frequency, and urgency. She had no complaint about nausea, vomiting, dizziness, diarrhea or any liver disease. On her past surgical history she had ovarian cystectomy by general surgeon 2 years back. But she could not supply any previous histopathology report of that operation. She had no family history of bowel, breast, ovarian, and endometrial cancer. Her physical examination revealed abdominal distension with a large mass of 30 weeks pregnancy size with irregular surface, margin not well defined, firm inconsistency, non-mobile and was fixed to underlying structure but not fixed to overlying skin. On percussion there is dullness over the mass but no shifting dullness and fluid thrill as well as no focal tenderness or rebound tenderness or guarding of the abdomen upon examination. On per vaginal examination Cervix flashed and vaginal atrophy was noted. On bimanual examination- uterus bulky for her age, all fornixes and pouch of douglus were full, ill-defined mass was felt. On per rectal examination - rectal mucosa was free. Her CA 125 was78.70u/ml and CA 19.9 was with in normal level. Trans abdominal ultrasound scan (USS) revealed a multi septet cystic lesion in the lower abdomen mainly on right side, measuring about more than 128x110 mm size. Fluid collection is seen in the peritoneal spaces suspected pseudomyxoma paritonei. Uterus bulky within homogenous myometrium.

Figure1: *Transabdominal ultrasonography showing a multi septate cystic lesion in the lower abdomen-pseudomyxoma paritonei. Diffuse loculated cystic structure is seen in lower abdomen mainly on right side. Measureing about more than 128x110 mm size. Fluid collection is seen in the peritoneal spaces of the lower abdomen & intestinal loops float within it.*

We had done CT-scan whole abdomen for farther confirmation.

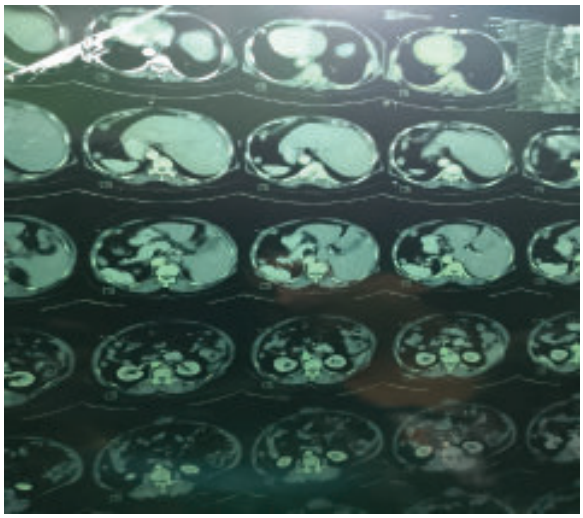

Figure 2: *Abdominal CT in axial (A) and coronal (B) sections showing large, well-defined, unilocular cystic mass measuring 14x12 cm. Uterus bulky, moderate ascites, within the ascetic fluid a curved hair line septa like area is seen giving cystic appearance at the pelvis (131x97) ovarian cyst. Large pad of soft tissue mass (Metastasis / soft tissue sarcoma) over the linea alba with in the subcutaneous fatty tissue.*

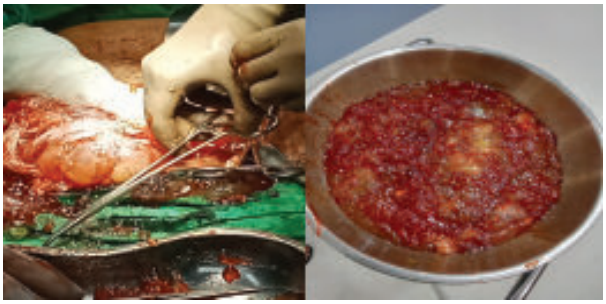

Figure 3: *Jelly like structure come out.*

Operative findings: Abdomen was opened with vertical incision. After opening the peritoneal cavity there was huge amount of mucinous material which was sucked out. Peritoneal toileting was done with normal saline, then left ovarian cyst about 14x12 cm with intact capsule was found .Uterus was bulky and right ovary was atrophied and exploration of whole abdomen including cecum, ascending colon and under surface of liver was done. After successive clamping, cutting & ligating total abdominal hysterectomy with bilateral salphingo-ophrectomy and appendicectomy was done.

Histopathology Report: low grade mucinous cystadenocarcinoma and adenomyosis with endometrial polyp, chronic cervicitis. Appendix was normal. Histological staging was $T_4N_0M_0$.

Post oparetive period: Her postoperative period was uneventful. She got 6 cycle chemotherapy.

Discussion

Ovarian neoplasm has different subtype's these include surface epithelium, germ cell, or sex cord-stromal tissue [6]. The majority of the diagnosed ovarian neoplasms are benign and 95% of all ovarian malignancies are epithelialorigin. Of all the ovarian epithelial neoplasm, 80% of ovarian mucinous cystadenomas are benign and 5%-10% present are bilateral [7].

It is slightly more common in women than men (male:female ratio of approximately 1:1.3,[6], although the actual ratio is difficult to identify due to potential misdiagnoses and possibly inclusion bias in reported studies. The median age at presentation is typically about 50 years with a range of 20-25 years, but PMP may strike persons of any age.[6] [7]. In my patient she is 70 years female.

The size and laterality play a major role in the determination of tumor origin whether primary or metastatic. In contrast with metastatic tumors, the primary tumors tend to be larger and unilateral [4]. The large size of the ovarian cyst is highly suggestive of mucinous cystadenoma. Likewise, in this case, the patient had large left ovarian cyst about 14x12 cm. So this tumour maybe primary origin.

The initial diagnosis of large adnexal mass is obtained by the pelvic ultrasound (US). Inparticular, the transabdominal US and the transvaginal doppler US become essential tools for evaluating ovarian masses. CT scan and MRI can further helpful in visualizing the specificity of ovarian cysts. Cancer antigen CA125, CA-199 are an important tumor marker that helps to differentiate between benign and malignant ovarian masses [11].

In my patient, I have done USG and the finding was showing a multi septate cystic lesion in the lower abdomen- pseudomyxoma paritonei. Diffuse loculated cystic structure is seen in lower abdomen mainly on right side, measureing about more than 128x110 mm size. Fluid collection is seen in the peritoneal spaces of the lower abdomen & intestinal loops float within it. Abdominal CT in axial (A) and coronal (B) sections showing large, well-defined, unilocular cystic mass measuring 14x12 cm. Uterus bulky, moderate ascites, with in the ascetic fluid a curved hair line septa like area is seen giving cystic appearance at the pelvis.

Large pad of soft tissue mass (Metastasis / soft tissue sarcoma) over the linea alba with in the subcutaneous fatty tissue. Her CA 125 was78.70u/ml and CA 19.9 was with in normal level.

Management of ovarian masses depends on a combination of factors, such as age, medicalhistory, symptoms, size of the cyst, and menopausal state of the patient. The mucinous cystrupture can lead to mucinous deposits filling in the entire peritoneum as the complication commonly called pseudomyxoma peritonei [7]. The best treatment after menopause is total abdominal hysterectomy with bilateral salphingo-ophrectomy .In my case, total abdominal hysterectomy with bilateral salphingo-ophrectomy and appendicectomy was done and exploration of whole abdomen including cecum, ascending colon and under surface of liver was done. However, performing surgical interventions for large masses, as in ourgeriatric patientis often associated with fatal consequences, such as sepsis, pulmonary embolism, and cardiac failure [7].Therefore, appropriate monitoring and follow-up are highly recommended in the postoperative period. In my patient, her postoperative period was uneventful.

Treatment is variable, both due to its rarity and to its frequently slow-growing nature. Treatment ranges from watchful waiting to debulking and hyperthermic intraperitoneal chemotherapy (HIPEC, also called intraperitoneal hyperthermic chemotherapy, IPHC) with cytoreductive surgery. [12].

But In our patient oncologist suggested 6(six) cycles systemic chemotherapy depending upon the histopathology report. After completion of chemotherapy within one year follow up patient was completely symptom free with normal general and per abdominal examination findings.

Conclusion

Pseudomyxoma peritonei (PMP) is a rare disease, proper diagnosis and treatment can prevent fatality. Careful evaluation of clinical symptoms with USG, CT Scan, CA-125 marker are helpful for diagnosis Surgical debulking is the standard treatment for pseudomyxoma paritonei, with intraperitoneal chemotherapy or systemic chemotherapy can be choice after histopathological grading. Some recurrence cases of pseudomyxoma paritonei, palliative care aims to improve the quality of life by alleviating symptom of Cancer.

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