Research Paper

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Pseudomyxoma Peritonei: A Case Report

MEDICAL SCIENCE

KEYWORDS : Pseudomyxoma peritonei, mucinous carcinomatosis, hyperthermic chemotherapy, gelatinous material, irregular masses

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ABSTRACT

Pseudomyxoma peritonei (PMP) is an uncommon condition characterized by abundant extracellular mucinous material in the peritoneal cavity and tumoural implants on the peritoneal and epiploic surfaces. Aim of this case report is to create awareness among the clinicians regarding this uncommon disease presented with ascites and irregular masses in the abdomen. PMP is a rare disease, may be benign or malignant, presented with ascites and irregular masses in the abdomen which is infrequently encountered in our clinical practice.

Introduction:

Pseudomyxoma peritonei (PMP) was first described by Rokitansky in 1842 . It is an uncommon

condition characterized by abundant extracellular mucinous material in the peritoneal cavity and tumour implants on the peritoneal and the epiploic surfaces. Incidence is one per million per year3 and encountered in 2 of 10,000 laparotomies with gelatinous masses called "Jelly belly. It is three to four times more in women than in men . The median age of presentation is 54 years. The origin of pseudomyxoma peritonei is very controversial. Tumour often arises from goblet cells of large bowel or appendix. Over expression of the gene MUC-2 responsible for mucin secretion is noted in PMP.

Case Report:

68 year, female P4L4 presented with complaints of abdominal distension , pain and discomfort since 2 months, difficulty in walking since 2 months, swelling of both lower limb since 8 days. Patient was a known case of Diabetes Mellitus with HTN since 13years on regular treatment K/C/O IHD – Angioplasty done 12 years back CABG done in8 years back.Patient had history of umbilical hernia repaired 10-12 years back. History of abdominal tubal ligation 35 years back. Patient also gave a history of decreased Appetite since last few months. Bladder and bowel movements were normal. Patient was vitally stable when she presented to the hospital. Per abdomen examination showed gross distension, umbilical hernia and ascites. Clinical diagnosis of Ovarian Malignancy with Ascites with umbilical hernia with CHD with H/O CABG/DM/HTN was made. All routine investigations were sent which were normal . Tumour markers CA 125- 31.11u/ml CEA- 17.35 ng/ml .USG– s/o umbilical hernia with Right ovarian cyst with ascitis .USG guided paracentesis was done which was suggestive of mesothelial cells in gelatinous fluid (scanty**). CT abdomen** showed Large complex cystic collection in mid abdomen s/o mesentric/ovarian cyst 20x20x16 cms, Appendicular calcification [mucocele], Gross ascites , Umbilical hernia(herniation of bowel loop) .

Fitness from physician posted for surgery--- Explorative Laprotomy with omentectomy with appendectomy with right ovarian cyst removal done.

Histopathology report suggestive of:**Ovary**-Pseudomucinous cystadenoma (benign) **Appendix**- mucinous neoplasm limited to mucosa **Omentum** – mucinous deposit devoid of epithelial cell **Abdominal wall** localised pseudomucinoma .

Patients stay in the hospital was uneventful and was discharged on postoperative day 15.Followed up 6 monthly with CA 125,CEA (chorionic embryonic antigen)and USG. CA125- 5u/ml CEA - 2.46ng/ml USG: Normal ,No ascites ,Grade 1 medical renal disease.

Discussion :

The clinical presentation of pseudomyxoma peritonei is caused by overwhelming increase in MUC-2 secreting cells and accumulation of excessive mucin secreted by these cells in the peritoneal cavity with no place of drainage12. The accumulated mucinous material gives the appearance of gelatinous masses. So the condition is called "Jelly Belly". The mucin sets on the peritoneal surfaces, gives compression on the vascular structures, hepatic hilum, gastrointestinal tract and other abdominal structures. There is distortion and impaired function of the visceral organs in the abdomen.The origin of PMP is very controversial. This is an unusual low grade malignancy often arising from the goblet cells of large bowel or appendix. Commonly it arises from mucinous tumour of the appendix and occasionally from ovary, colon, rectum, stomach, gall bladder, bile duct, small intestine, urinary bladder, lung, breast, pancreas and fallopian tube. The benign cases are called disseminated peritoneal adenomucinosis and when malignant features are present the term peritoneal mucinous carcinomatosis is used. This low grade malignancy may spread through the peritoneal cavity. Visceral invasion is rare and metastatic spread by haematogenons or lymphatic route is not seen. Metastasis to lungs by transdiaphragmatic seeding has been reported. Debulking surgery combined with intraperitoneal hyperthermic chemotherapy is the standard treatment. Systemic chemotherapy is given occasionally. Ronnett et al observed 75% and 68% of 5 years and 10 years survival rate respectively for cases of disseminated peritoneal adenomucinosis, 14% and 3% for cases of peritoneal mucinous carcinomatosis respectively.

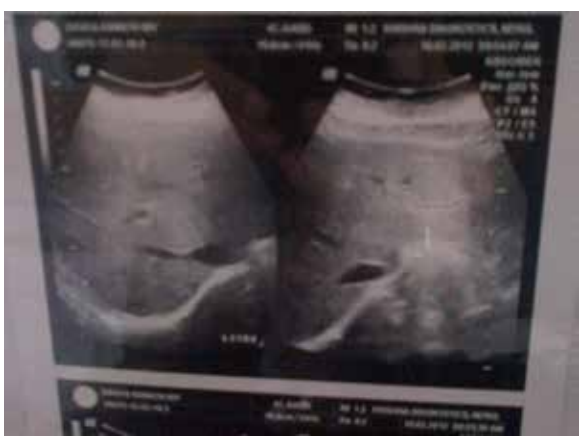
Conclusion:

Pseudomyxoma peritonei is a rare disease, may be benign or malignant. The reported patient presented with features of ascites, which is common in our country. After investigation it was diagnosed as a case of pseudomyxoma peritonei, which is infrequently found in our clinical practice.

Figure 1. RIGHT OVARIAN MUCINOUS-CYSTEDENOMA

Figure 2. CYSTEDENOMA WITH MUCINOUS COLLECTION

Figure 3. CT SHOWING RIGHT OVARIAN CYST



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