

A Case of Mucinous Cystadenocarcinoma Ovary Mimicking Pseudomyxoma Peritonei

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Received: Feb. 16, 2018

Accepted: March 18, 2018

ABSTRACT

BACKGROUND: Pseudomyxoma peritonei means false mucinous tumour of the peritoneum characterized by the accumulation of mucinous ascites within the peritoneal cavity. Appendicular disease and and cystadenoma of ovary sometimes coexist in some cases . Diagnosis is seldom absolute until a laparotomy is performed despite the presence of distended abdomen with non shifting ascites. **CLINICAL CASE:** A 70 year old post menopausal woman with complaints of generalized distension of abdomen since 2 years with tense and gross distension of abdomen with no clinical evidence of ascites .CT abdomen and pelvis_pelvi-abdominal complex cystic lesion with septations with no solid components with dehiscence right lateral wall likely ruptured adnexal cyst and ill defined enhancing bilateral hypochondrial nodular lesions ? Peritoneal deposits With a preoperative diagnosis of pseudomyxoma peritonei, patient underwent optimal debulking surgery. Histopathology report revealed poorly differentiated grade 3 mucinous cystadenocarcinoma of ovary with omental deposits. **CONCLUSION:** Pseudomyxoma peritonea is a rare disease characterized by the spread throughout the abdomen of mucinous ascites with peritoneal and omental mucinous implants. Mucinous cystadenocarcinoma of ovary can be sometimes be misdiagnosed for PMP as most of the clinical ,radiological and pathological features overlap.PMP from mucinous cystadenocarcinoma.The differentiation is very important as the mode of adjuvant therapy varies.

CASE REPORT

A 70 year old post menopausal woman came with complaints of generalized distension of abdomen since 2 years .Associated with complaints of vomiting , constipation and weight loss with no relevant family or personal history .On examination patient was hemodynamically stable with a BMI of 24kg/m².Per abdomen examination revealed tense and gross distension of abdomen with everted umbilicus with no clinical evidence of ascites .Per speculum and BME was unremarkable

LABORATORY STUDIES :CA 125 - 10.22U/ml ,CEA - 118.9ng/ml,BETA Hcg - 0.98U/ml,AFP - 2.26 U/ml,LDH- 211 U/ml

Peritoneal fluid analysis revealed thick straw coloured mucinous material with no evidence of malignant cells.

CT abdomen and pelvis showed 20 x 17x 25 cm pelvi-abdominal complex cystic lesion with septations with no solid components with dehiscence right lateral wall likely ruptured adnexal cyst and ill defined enhancing bilateral hypochondrial nodular lesions ? Peritoneal deposits



FIG 1: preoperative image



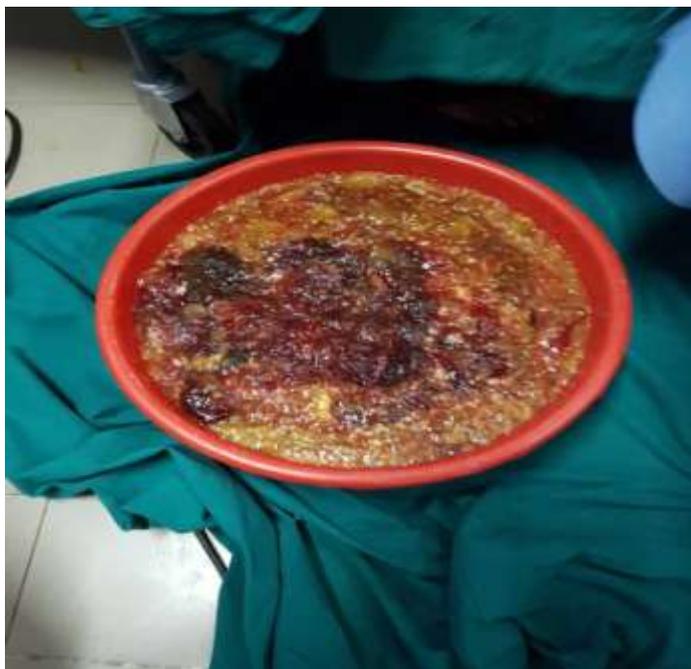
(Fig 2 and 3) : Contrast CT image showing pelvi-abdominal complex cystic lesion with dehiscent lateral abdominal wall

TRETEMENT

With a preoperative diagnosis of pseudomyxoma peritonei, patient underwent optimal debulking surgery.(Total abdominal hysterectomy + Bilateral salpingo oophorectomy + Pelvic Peritonectomy+ debulking of left ovarian tumour + appendectomy + Total Omentectomy + drainage of 30 litres of gelatinous material)



(fig 4). Intra operative image showing removal of gelatinous substance



(fig 5). Gelatinous fluid after removal

Histopathology report revealed poorly differentiated grade 3 mucinous cystadenocarcinoma of ovary with omental deposits. The cyst wall showed papillae and cribriform glands lined by highly atypical cells with frequent mitosis, confluent gland and high grade nuclear features.

Post operative recovery was uneventful and patient was discharged on POD 13 , Patient was advised 6 cycles of adjuvant chemotherapy with carboplatin and paclitaxel .The patient is under strict follow-up & presently doing well untill last F/U a month ago.

DISCUSSION

Pseudomyxoma peritonei (PMP)

The term pseudomyxoma peritonei means false mucinous tumour of the peritoneum with an incidence of 1-2 per million annually . PMP is a clinical term. It is characterized by the accumulation of mucinous ascites within the peritoneal cavity. Most common cause of PMP is appendiceal neoplasm. Most primary appendiceal adenocarcinomas are of the mucinous type, arise from an adenomatous polyp or serrated adenoma or within the appendiceal lumen and consequently the lumen per se becomes occluded. This occlusion finally causes a rupture in the wall of the appendix and therefore mucus containing epithelial cells is spilled within the abdominal cavity.

In vast majority of cases Synchronous ovarian lesions are frequently seen in female patients, which has led to confusion about the true origin of PMP. Most cases of PMP showed positive expression for cytokeratins 18 and cytokeratin 20 whereas the reaction was mostly negative for CK 7. The expression of human alveolar macrophage (HAM) 56 tended to be negative and that of carcinoembryonic antigen (CEA) positive. The abundant expression of the MUC2 and MUC5AC genes were determined in PMP . Appendiceal goblet cells express both MUC2 and MUC5AC, but mesothelial cells and the cells of the ovarian surface express only MUC5AC suggesting PMP is of appendiceal origin and not of ovarian or mesothelial origin.

Different classification schemes of PMP.

Classification	Year	Number of classified groups	Groups
Ronnett	1995	3	DPAM: Disseminated peritoneal adenomucinosis PMCA-I/D: Peritoneal mucinous carcinomatosis with intermediate or discordant features PMCA: Peritoneal mucinous carcinomatosis
Bradley	2006	2	MCP-L: Low grade mucinous carcinoma peritonei MCP-H: High grade mucinous carcinoma peritonei
Pai & Longacre	2005	4	mucinous adenoma M-UMP: mucinous neoplasm of uncertain malignant potential M-LMP: mucinous neoplasm of low malignant potential mucinous carcinoma
WHO	2010	2	Low grade: Low-grade pseudomyxoma High grade: High-grade pseudomyxoma

Clinically PMP manifest as increased abdominal girth, which is caused by the accumulation of gelatinous ascites with “jelly belly” appearance .The chief complaint may be a newly-onset hernia as a consequence of increased intra-abdominal pressure. sometimes cause intestinal obstruction or lesions may also cause pain in the flank due to obstruction of the ureter.

The natural progression of the disease is usually moderately slow, although rapid advancement is occasionally seen .Spreads usually on the peritoneal surfaces, but invasion of the organs is also seen, especially in cases with a high-grade histology. Haematogenous metastases are rare. Maintstay of treatment include complete cytoreductive surgery to be followed by intraoperative HIPEC(hyperthermic intraperitoneal chemotherapy). Cytoreductive Surgery includes the following (described by Sugarbaker)

- Greater omentectomy-splenectomy
- Left upper quadrant peritonectomy, right upper quadrant peritonectomy
- Lesser omentectomy,cholecystectomy with stripping of the omental bursa,
- Pelvic peritonectomy with sleeve resection of the sigmoid colon
- Antrectomy.

The burden of the peritoneal disease is calculated by PCI(peritoneal carcinoma index).The completeness of the surgery is calculated by CC score (completion of cytoreduction score) Optimal cytoreduction is defined when we attain a CC score 0 or 1 after complete surgical resection .Only patients who attain a CC score of 0 and 1 are ideal candidates for post operative HIPEC.

ovarian tumors

There are four major categories of ovarian tumors:

1. Epithelial tumors (65-75%) - serous or mucinous cystadenoma/carcinoma, clear cell carcinoma, Brenner tumor
2. Germ cell tumors (15%) - dysgerminoma, embryonal cell cancer, choriocarcinoma, teratoma
3. Sex-chord-stromal tumors (5-10%) - granulosa cell tumor, thecoma, fibroma
4. Metastatic tumors (10%) - uterine, stomach, colon, breast, lymphoma

Mucinous tumours of ovary are divided into three categories: benign, borderline, and malignant.Malignant mucinous cystadenocarcinomas are rare, and comprises 10% of mucinous ovarian tumors and 5-10% of primary malignant ovarian neoplasms. They are bilateral in 15-30% of cases and have a peak incidence between 40-70 years of age.They can attain a very large size and may be multilocular .Adhesions to adjacent tissues are not present unless there is degenerative changes in the wall.The cysts are lined by tall columnar cells and these secrete a mucous – a glycoprotein rich in neutral polysaccharide.

A rare complication is cyst which ruptures and spills into the peritoneum ,a condition in which epithelial cells of the tumour invades omentum and also spreads over the visceral and parietal peritoneum. From these sites they secrete semisolid mucin into the abdominal cavity and eventually causing distension .The

appearance of the epithelium is remarkably like that of the glands of cervix or of the intestine .The fluid content is thick in consistency yellow-green to brown in colour depending on the presence of blood pigments from previous intracystic hemorrhage.Even the causal tumour is benign and the mucious material is removed , the abdomen tends to refill and is might be due to mucinous metaplasia of the peritoneum.

Appendicular disease and and cystadenoma of ovary sometimes coexist in some cases . Diagnosis is seldom absolute until a laparotomy is performed despite the presence of distended abdomen with non shifting ascites . Mainstay of treatment is cytoreductive surgery along with intra peritoneal or intravenous chemotherapy with cisplatin based regimens.

Recent randomized trials have concluded a definite improvement in DFS & OS with addition of postop HIPEC in all stage III ovarian cancers.

CONCLUSION

Pseudomyxoma peritonea is a rare disease characterized by the spread throughout the abdomen of mucinous ascites with peritoneal and omental mucinous implants. Most often PMP is a clinical diagnosis .In most patients with PMP, the appendix is the site of primary tumour.

Mucinous cystadenocarcinoma of ovary can be sometimes be misdiagnosed for PMP as most of the clinical, radiological and pathological features overlap.

Only immunohistochemistry can at times differentiate PMP from mucinous cystadenocarcinoma.The differentiation is very important as the mode of adjuvant therapy varies. The current recommended therapy for PMP is optimal cytoreductive surgery followed by HIPEC.Adjuvant platinum based doublet chemotherapy is the standard of care for mucinous cystadenocarcinoma of ovary.Appropriately managed PMP has got a good overall prognosis in the form of DFS and overall 10 year survival compared to mucinous cystadenocarcinoma of ovary.

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Leadership is the capacity to translate vision into reality.

~ Warren G. Bennis