Unusual Textbook case of PSEUDOMYXOMA PERITONEI of the ovary and its management

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A 46 yr old, perimenopausal lady was admitted from the OPD with complaints of abdominal distension and swelling 2-3 months and pain abdomen off & on specially after meals. Her periods were irregular for last 2 years and her LMP was 10 months back.

She was thinly built, poorly nourished.

There was a large lump filling the abdominal cavity extending till the base of sternum, firm , non tender, restricted mobility .

On per vaginal examination the lump felt to be arising from the pelvis till above the umbilicus, uterus and bilateral adnexa not felt separately from the mass.

CECT (Whole Abdomen) showed a large Left ovarian cyst 20 by 19 cm with solid and cystic areas, uterus and right adnexa normal

Serum CA125 was Normal

A provisional diagnosis of Ovarian Tumor was made.

Patient was managed by a team of Gynecologist, Cardiologist, General Surgeon, and Anesthetist.

FINDINGS

A large solid cystic mass seen filling the whole abdomen with yellowish jelly like material in the cavity, on exploration the cyst was found to have ruptured with spill into the cavity, uterus and right ovary with tube appeared normal. The appendix was enlarged 5x6 cm with rupture and similar jelly like material oozing from that end. Peritoneal cavity appeared
normal and so was the rest of the gastrointestinal tract. Liver and Spleen appeared normal.

Ovarian cystectomy with abdominal hysterectomy with right salpingo oophorectomy and appendicectomy done. Good Peritoneal lavage done.

**HISTOPATHOLOGY REPORT**

Mucinous cystadenoma-left ovary with pseudomyxoma peritonei and mucinous neoplasm of the appendix

She was advised follow up and referred to oncologist for further treatment.

**DISCUSSION**

The incidence of *pseudomyxoma peritonei* is 2 in 10000 laparotomies, with a female to male ratio of 3 to 1.

Pseudomyxoma peritonei (PMP) first described by Werth is an uncommon and poorly understood disease characterized by abundant extracellular mucin in the peritoneum. The "myxomatous" appearance is attributed to the associated fibroblastic and vascular proliferation that is probably incited by the mucin. This results in multifocal peritoneal, serosal and ommental implants admixed with copious amounts of mucin accumulation within the abdomen and pelvis resulting in the belly full of jelly – "the jelly belly".

PMP is a broad descriptive term embracing a wide spectrum of biological behavior of neoplasms from the benign to the borderline to the frankly malignant lesion. Other terminologies to reflect this spectrum of biological behavior ranges from disseminated peritoneal adenomucinosis (DPAM) – the benign variant to peritoneal mucinous carcinomatosis (PMCA) – the malignant variant. A definitive diagnosis of PMP requires the presence of

a) mucinous neoplastic cells/epithelium, and

b) mucinous ascites – diffuse intraabdominal mucin.

Some authors also require the presence of diffuse mucinous implants for this diagnosis.

It is more commonly seen in women who usually present with increasing abdominal girth and this tends to be related to underlying ovarian lesions which are usually mucinous tumors that can be associated with a teratoma. Though uncommon in men, these cases are virtually all associated with a lesion in the appendix. Other possible primary sites include colorectum, gallbladder, pancreas, urachus, urinary bladder, breast and lung, but these are uncommon. PMP can occur years (range from 5–35 years) later after the initial presentation of an appendiceal event and, therefore, accurate diagnosis prior to surgery is often delayed and inaccurate. The disease may be localized in the right lower quadrant initially and then become
more generalized with mucinous peritoneal, serosal and omental implants. 10% of patients die of PMP within 5.5 years of their initial presentation while others experience recurrent and/or residual disease. Advanced abdominal disease leading to intestinal obstruction accounts for majority of the patients' morbidity and mortality.

**TREATMENT**

Ranges from **watchful waiting** to **debulking** and **cytoreductive surgery**

Chemotherapy (typically the agent Mitomycin C) may be infused directly into the abdominal cavity to kill remaining microscopic cancerous cells. The heated chemotherapy (HIPEC) is perfused throughout the abdominal cavity for an hour or two as the last step in the surgery, or ports are installed to allow circulation and/or drainage of the chemicals for one to five days after surgery, it is typically given in multiple cycles for several months after surgery.

Systemic chemotherapy may be administered as additional or adjuvant treatment. Due to the increased availability of new chemotherapies developed for colon and colorectal cancer patients, some patients have experienced stability in tumor growth with systemic chemotherapy. Systemic chemotherapy is reserved for patients with advanced disease, recurrent disease, or disease that has spread to the lymph nodes or distant sites.

This disease may recur following surgery and chemotherapy. Periodic post operative CT scans and tumor marker laboratory tests are used to monitor the disease for any tumor regrowth.

Additionally recent (2003) publications linking the MUC2 enzyme over expression to the cell reproduction has launched research efforts into additional drug treatments.