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Pseudomyxoma Peritonei

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ABSTRACT *Pseudomyxoma peritonei is a clinical condition that is characterized as a localized or generalized accumulation of abundant gelatinous material within the abdominal and/or pelvic peritoneal cavity which usually shows a protracted clinical course, long-term prognosis is poor and death ultimately occurs as a consequence of intra-abdominal disease progression. We wanted to emphasize the pathological and clinical features of PMP, ideal treatment of the condition, and the outcome.*

The first step to improve the prognosis is to recognize PMP preferably in an early stage. CT imaging should be the choice of radiological assistance in the diagnosis and follow-up. Cytoreductive surgery with intraoperative HIPEC is a treatment strategy with encouraging survival results for selected PMP patients. The pathologic subtype remains the dominant factor for survival. Improvement of survival can be achieved by combination of surgical experience and adequate patient selection. Multi-institutional studies should be recommended. On the other hand, intraperitoneal PDT is potentially an ideal therapy but it needs improvement.

INTRODUCTION:

Pseudomyxoma peritonei (PMP) is a relatively rare clinical entity with an estimated incidence of 1 to 2 million per year (1). It is a poorly understood disease that is known for its production of diffuse intra-abdominal gelatinous collections ('jellybelly') with mucinous implants on peritoneal surfaces and the omentum (3). It is often an intra-operative surprise when gelatinous material is noted while doing laparotomy or diagnostic laparoscopy for ambiguous symptoms.

The term Pseudomyxoma peritonei has been applied broadly and includes a heterogeneous group of pathological lesions from benign to borderline and to frankly malignant lesion. Intestinal mucinous tumours, particularly colorectal cancers and any mucinous neoplasms may present with clinical, radiological and pathological features resembling PMP (2).

CASE REPORT

A 40 yr old male patient presented to our hospital with pain abdomen and mass abdomen for one month. He noticed a small swelling in the umbilical region which was growing gradually. His complaint of pain abdomen was usually after food consumption and it is colicky and sometimes relieved by self induced vomiting. He also had frequent vomitings since 10 days which was non-bilious containing food particles. He had no complaints of fever or jaundice. There was no history of loss of appetite or weight and normal bowel habits.

On examination there were hard nodular masses noted at the umbilicus, both the inguinal regions and suprapubic region. Ultrasound showed mild hepatomegaly and loculated ascites. CECT abdomen showed diffuse hypodensity through out the abdomen with areas of enhancing septa and scalloping noted on the surface of liver and spleen. CT also showed diffuse wall thickening of stomach and pyloric region as well as hypodense leison with enhancing septa noted at umbilicus and bilateral inguinal region. Aspiration and cytology of the abdominal swellings showed homogenous mucin like material with an occasional fibroblast with the possibility of pseudomyxoma.

Colonoscopy was showing normal colonic mucosa with extrinsic compression noted in ascending colon and caecum causing luminal narrowing. Multiple biopsies from caecum and ascending colon showed no evidence of malignancy and granulomata.

Laparotomy and cytoreductive surgery was planned. Intraoperatively over two litres of mucinous gel like substance was emptied (figure 1). There was a hard mass enclosing distal ileum ,caecum and proximal ascending colon with omentum around it making it difficult to identify or isolate any structures in the hard mass. Most of the fibrous disease over the peritoneum along with the greater omentum is removed without injuring the colon and stomach(figure 2). Hard peritoneal mucinous nodules at the umbilicus (figure 3) and at the inguinal regions were removed. Four drains were placed in the abdominal cavity with two drains on either side. Post-operatively intraperitoneal chemotherapy was given.

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DISCUSSION

Pseudomyxoma peritonei has generally been considered benign. However with disease progression over time to massive abdominal distension and obstructive features suggests that it is a borderline malignant disease. PMP is a poorly understood disease with massive amounts of mucinous ascites with peritoneal and omental implants which fills up the entire peritoneal cavity eventually. It was first described by Werth in 1884 as massive amounts of mucinous ascites in association with benign ovarian neoplasm. Werth postulated that it is due to a reaction in response to the jelly-like material released from a ruptured cyst (4).

The natural history in PMP is thought to involve appendiceal adenoma occluding the appendiceal lumen leading to "blow-out" and slow leak of mucus containing epithelial cells from the adenoma(5). These tumour cell surfaces lack adhesion molecules preventing random adherence to peritoneal surfaces and showing its characteristic "redistribution" within the peritoneal cavity (6). Dependent portions of the abdomen and pelvis such as the rectovesical pouch, the right retro-hepatic space and the paracolic gutters, accumulate tumour cells due to gravity. The open lymphatics on the under surface of the right hemidiaphragm and the lymphoid aggregates in the omentum, results in tumour masses classically described as "scalloping" of the liver and as "omental cake"(6). Parts of the gastro-intestinal tract fixed to the retroperitoneum (gastric pylorus, antrum, ileocaecal and rectosigmoid regions) are often involved secondary to the extensive accumulation of tumor cells unlike small bowel which is spared due to peristalsis(7). Ronnett and colleagues, in a retrospective review reported a pathological system wherein they classified low-grade tumors as disseminated peritoneal adenomucinosis (DPAM) and high-grade tumors as peritoneal mucinous carcinomatosis (PMCA), with an intermediate group (IG) demonstrating a mixture of DPAM and PMCA(8). Patients with low grade tumors have maximum survival benefit from aggressive locoregional treatments than those with PMCA(9,10). Earlier, diagnosis was rarely confirmed preoperatively but now with the presence of advanced imaging modalities confirmation is possible preoperatively. Currently high resolution CT scan is considered gold standard investigation for PMP. Computed tomography shows four basic patterns: (1) posterior displacement of intestines with numerous low density masses and calcifications (2) low attenuation mucinous ascites occupying most of the peritoneal cavity or occasionally as localized collections with septae (3) scalloping of intra-abdominal organs due to extrinsic pressure of adjacent peritoneal implants (11,12,13). MRI and PETCT may offer advantages over MDCT in the detection of peritoneal malignancy in select applications when CT alone is inconclusive (14). Due to the indolent nature of the disease most patients usually present in an advanced stage with features of intestinal obstruction or failure to thrive that requires repeated interval debulking for symptomatic relief, with limited expectation of long-term survival and no prospect of cure. Sugarbaker (15,16) introduced and popularized the approach combining Cytoreductive Surgery (aiming for macroscopic complete tumor removal) with HIPEC (Hyperthermic Intraperitoneal Chemotherapy) to address residual microscopic disease. Patients must be medically fit to safely undergo CRS with HIPEC. No clear guidelines exist for patient with disease not amenable to CRS because of tumor extent and distribution, or serious co-morbidity, or age. Even in these patients there is increasing evidence that a major palliative resection i.e, extended right hemicolectomy, greater omentectomy and splenectomy with an ileocolic anastomosis, or total colectomy, showed improved survival and better quality of life (17).

The prognostic value of tumor markers in patients undergoing Cytoreductive Surgery and HIPEC showed a significantly reduced recurrence-free interval for patients who had an elevated baseline CEA prior to surgery and also with at least one elevated marker (among CEA, CA125 and CA19.9) (18).

CONCLUSION

Although PMP is an uncommon condition, all surgeons who operate on abdomen will occasionally_encounter a patient with PMP. The optimal treatment involves a combination of surgery and HIPEC. The treatment strategy is complex, associated with significant morbidity and mortality. The long-term outcomes for CRS with HIPEC in PMP are impressive for patients with low-grade histology amenable to complete cytoreduction. Even when CRS cannot be performed an aggressive palliative resection should be attempted.

Figure: 1

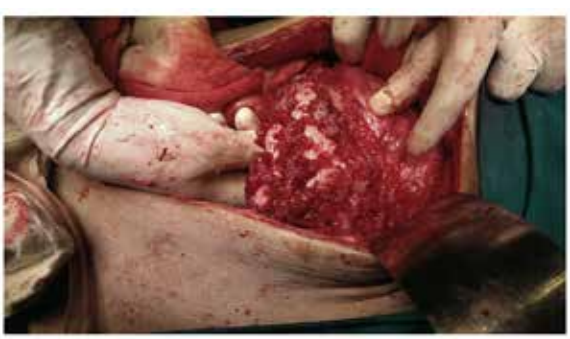

Figure: 2

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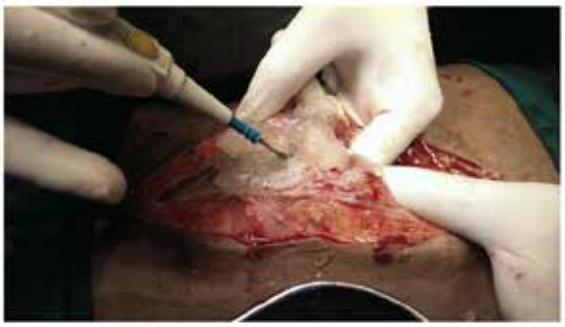

Figure : 3

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