AN INTERESTING CASE OF ASCITES

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Abstract:
Pseudomyxoma peritonei (PMP) is a rare condition characterized by the presence of mucin producing tumors in the abdominal cavity. PMP is listed among the rare or orphan diseases acknowledged by the National Association of rare diseases. It is generally accepted that appendix cancer is the most common source of the tumors that cause PMP. We present this case report of a man who presented with a rare disease of PMP and with the likely origin of PMP from appendix which has been removed four years back for details not known.

Keyword: pseudomyxoma peritonei, ascites, mucin jelly, hyperthermic intraperitoneal chemotherapy, sugar baker.
A 45 year old male presented with painless progressive abdominal distension for one year. Patient had loss of appetite and significant weight loss. Ascites was not associated with pedal edema and oliguria. No history of jaundice/bowel symptoms. Patient took alcohol 30-40 g once in a month for 4 years. No H/O DM/HT/IHD. H/O Appendectomy done four years back. On Examination patient had generalized muscle wasting. No evidence of pallor, icterus or pedal edema. On examination abdomen was swollen, flanks were full and umbilicus everted. There was firm, non tender, mobile non tender masses were seen involving whole abdomen. Ascites was evident. Complete blood count, sugar, renal function test and liver function test were normal. HBsAg and Anti HCV were negative. Diagnostic aspiration of ascetic fluid revealed thick gelatinous material. Ascitic fluid analysis showed protein of 5.3 gm/dl and albumin 3 mg /dl. Upper endoscopy and colonoscopy were normal.

USG abdomen showed normal echotexture of liver with scalloping, ascites and multiple cystic lesion throughout abdomen. CECT abdomen shows scalloping of liver with omental and peritoneal deposits. Ascitic fluid cytology and umbilical nodule FNAC were negative for malignancy.

Diagnostic laparoscopy findings:
Multiple mucinous deposits all over the peritoneum, omentum, liver surface, small and large bowel and spleen

Omentum grossly thickened
Hemorrhagic ascites – 4 litres

Primary could not be made out
Hence the Diagnosis of Pseudomyxoma peritonei was made. Procedure done was omentectomy and debulking of the mass. Excised omentum was sent for biopsy. Patient was started on systemic chemotherapy.

BIOPSY FINDINGS:
Section shows a multiloculated lesion lined locally by columnar epithelial cells enclosing pools of extravasated mucin. Focal areas show clusters of floating cells having moderate amount of cytoplasm and round to oval nuclei with occasional cells having vacuolated cytoplasm and peripherally placed nuclei

IMPRESSION: Low grade Mucinous neoplasm

Females are 2-3 times more common than males. With the history of appendicectomy four years back likely reason is appendiceal etiology. Pseudomyxoma developing 35 years after appendicectomy have been reported. Here We present this case report for its rarity.
DISCUSSION:

Pseudomyxoma peritonei (PMP) is a poorly understood disease. It refers to a progressive disease process within the peritoneum which originates from the appendix or ovaries and is characterized by the production of copious amounts of mucinous fluid resulting in a "jelly belly". PMP is a rare indolent disease (1), which is most prevalent in women aged between 50 and 70 years. Clinically although painless, clinical deterioration of health begins long before diagnosis. The main complaints are abdominal pain and distension besides non-specific complaints. Inflammatory changes associated with peritoneal tumor implants can lead to fistula formation and adhesions which can cause intermittent or chronic partial obstruction.(1)

The characteristic PMP dissemination within the peritoneal cavity is defined by Sugarbaker (2) as a complete redistribution phenomenon indicating a complete and sequential invasion of the peritoneal cavity with large tumor volume localization at predetermined anatomical sites and minimal invasion at other sites.

There is continued debate regarding the origin of PMP. Most commonly associated with mucinous tumors of the appendix or ovary. (3) Can occur rarely with mucinous tumors of the bile duct, stomach, pancreas etc.

Diagnosis of PMP is often difficult. Ultrasonography detects immobile ascites. Gelatinous masses can mark the hepatic and splenic margins (scalloping) by extrinsic pressure. (4) A CT exam is the best to diagnose and stage PMP. Sometimes progressive punctuate calcifications may be seen with scalloping. There is currently no accepted standard treatment for PMP. Surgical debulking and appendectomy is widely regarded as the mainstay treatment of PMP. Sugarbaker from the Washington Cancer Institute describes an aggressive surgical intervention to improve prognosis. He combined maximal surgical debulking with maximal regional chemotherapy. Peritonectomy is the proposed surgery which includes complete removal of the tumor. (5) Removal of all tumor tissues from the parietal and visceral peritoneum is done. Large tumor nodules must be resected and all visible tumors removed. Small cancer deposits on the visceral peritoneum are also individually electroevaporated

Hyperthermic intraperitoneal chemotherapy (6) is only required to eradicate microscopic residual disease for its complete success. The combined use of hyperthermia and intraperitoneal chemotherapy enhances the cytotoxicity of chemotherapeutic agents and increases tissue penetration by chemotherapy in cancerous tissue as compared to normal tissue. The intraperitoneal temperature is maintained at 42.5 degree celcius. Different chemotherapeutic agents are used depending on the tumor histological characteristics. Patients with PMP may benefit from the Sugarbaker procedure and have an estimated 5-year and 10-year survival of approximately 50% and 18%, respectively.

We are presenting this case as PMP as itself is a rare disease. Indian case reports are less but present (7). PMP is usually seen in females. In our case, patient was a male with history of appendectomy four years back. Case reports have been published with history appendectomy as long as 35 years back.(8)
REFERENCES:

1 Werth R. Pseudomyxoma peritonei. Arch Gynaecol 1884; 24:100-118


8 Pseudomyxoma peritonei occurring after 35 years of appendicectomy -World Journal of Surgical Oncology 2004, 2:19