Abstract: Mucinous Adenocarcinoma arising from Appendix with associated Pseudomyxoma peritonei is a rare clinical entity. Here we report a case of 49-year-old male patient, who presented to the OP with an abdominal mass of size 20 cm by 15 cm. USG, CT, MRI showed suspected bowel mass or retroperitoneal mass. CT guided biopsy and laparoscopic biopsy from the mass turned to be inconclusive. Due to the diagnostic dilemma, explorative laparotomy and debulking surgery of the mucinous omental caking were done. After HPE, the final diagnosis is Appendicular Mucinous Adenocarcinoma with Pseudomyxoma peritonei. This case is being presented for its rarity.

Keyword: Pseudomyxoma peritonei, Appendicular mucinous adenocarcinoma

CASE REPORT

49-year-old male patient came with the presenting complaints of abdominal mass & abdominal distention for 6 months. Patient had complaints of dragging abdominal pain, early satiety, loss of weight, loss of appetite. No history of vomiting & alteration in bowel movements. No significant past history & family history.

USG Abdomen:
Complex hyperechoic & cystic like mass. Possibility of bowel mass/Retroperitoneal mass.

CT/MRI Abdomen:
Right Retroperitoneal mass lesion with cystic component & specks of calcification. Adhesions with matted bowel loops. Moderate ascites, peritoneal nodules, peritoneal thickening. Possibility of Retroperitoneal malignancy/Bowel mass/Sarcoma.

MRI Images

CLINICAL EXAMINATION:
DIAGNOSTIC DILEMMA:
CT guided biopsy & Laparoscopic biopsy proved to be inconclusive. Inspite of all investigation modalities the diagnosis still remained elusive.Hence the patient was subjected to Explorative Laparotomy.

PEROPERATIVE FINDINGS:
Gelatinous ascitis ,peritoneal mucinous implants with omental caking were detected. Appendix could not be visualised & was replaced by a tumour mass. Debulking of tumor done.Due to poor general conditions of the patient further exploration could not be done.Specimen sent for HPE.

Histopathological Examination Report:
Mucinous Adenocarcinoma arising from Appendix with Secondary Omental & Peritoneal deposits.

FINAL DIAGNOSIS: Appendicular Mucinous Adenocarcinoma with Pseudomyxoma peritonei

Post operatively Medical Oncologist opinion was obtained.Patient was advised 6 cycles of Cisplatin+Leucovarin+5 Fluourouracil chemotherapy regimen.

Patient had completed first cycle of Chemotherapy.

FOLLOW UP
Adequate health education regarding Mucinous adenocarcinoma of Appendix,its prognosis and appropriate counselling was given to the patient.He is now on regular follow up for further cycles of chemotherapy.

REVIEW OF LITERATURE:
Gelatinous material in the peritoneal cavity was described first in 1884 and termed pseudomyxoma peritonei because the material was not composed of true mucin. Its origin was believed to be rupture of a pseudomucinous ovarian cyst. In 1901, Frankelg first reported pseudomyxoma peritonei secondary to a ruptured cyst of the appendix.Pseudomyxoma peritonei is commonly due to mucinous cystadenocarcinoma of Appendix, Ovarian, Pancreatic cancer. The incidence of pseudomyxoma peritonei is approximately 2 in 10,000 laparotomies. Three of four patients are women. However, when the ovary as primary site is excluded, the gender ratio is nearly 1:1. Histologically, pseudomyxoma peritonei is characterized by a benign appearance, with simple columnar epithelium containing small uniform nuclei and mucin containing Vacuoles.The term pseudomyxoma peritonei should be reserved for those patients with tumors that originate in a mucin producing adenocarcinoma, with epithelial cells found in the peritoneal mucinous implants.

Neoplasias of the appendix with or without association with pseudomyxoma peritonei can be classified as:
1) Simple mucoceles
2) Mucosal hyperplasia and hyperplastic polyps
3) Serrulate adenoma (mixture of hyperplastic and adenomatous polyp)
4) Mucinous cystadenoma
5) Mucinous neoplasia of uncertain malignant potential
6) Mucinous neoplasia of low potential malignancy
7) Invasive adenocarcinoma

Invasive adenocarcinoma (mucinous, intestinal or signet ring) of the appendix is rare and less frequent than mucinous adenomas.
Pseudomyxoma peritonei has been reported to metastasize rarely to the lung.Currently, it is believed that these sporadic instances of extraabdominal spread are secondary to transdiaphragmatic seeding rather than true hematogenous spread.Whether pseudomyxoma peritonei is theoretically benign or malignant has been questioned on the basis of its inability to metastasize and invade tissue. However, it ultimately causes death as a result of extensive disease, bowel obstruction and fistulisation. Preoperative diagnosis of pseudomyxoma peritonei is possible, although it often is mistaken for carcinomatosis, widespread lymphoma, pyogenic peritonitis, and loculated ascites.Paracentesis
occasionally has been successful. The gelatinous nature of the mucin, however, often makes attempts at aspiration futile. Laparoscopy guided biopsy is rarely successful. Much attention recently has been focused on preoperative sonographic and radiographic diagnosis of pseudomyxoma peritonei. Sonographic findings of an echogenic mantle with multiple intraperitoneal multicellular cysts and ascitic septations are specific for pseudomyxoma peritonei.

**Ultrasound features:**
Nonmobile echogenic ascites with multiple semisolid masses. Scallopings of the hepatic and splenic margins

**CT features:**
Mucinous ascites with the density of fat. Omental thickenings, Multiseptated lesions, Scallopings of organs, Curvilinear calcifications.

**Histology remains the golden standard for the diagnosis of Pseudomyxoma peritonei & Mucinous cyst adenocarcinoma.**

Differential diagnoses of pseudomyxoma peritonei & Mucinous cystadenocarcinoma.

Calcifications.

Multiseptated lesions, Scalloping of organs, Curvilinear

Mucinous ascites with the density of fat, Omental thickenings, Multiseptated lesions, Scallopings of organs, Curvilinear calcifications.

**CONCLUSION**

Although preoperative sonography & radiological investigations aid in the diagnosis, Exploratory Laparotomy & Histology still remains the golden standard for the final diagnosis of Pseudomyxoma peritonei & Mucinous cyst adenocarcinoma. The primary treatment of pseudomyxoma peritonei is surgical debulking. The therapy consists of exploratory laparotomy and a cytoreductive procedure. Because of the diffuse nature of the disease, it is impossible to attain wide margins. The primary tumor mass should be identified and resected. As the decum and ascending colon often are surrounded diffusely by the tumor, a right hemicolectomy may be required. Omentectomy is done for two reasons. First, the omentum usually is involved diffusely in the tumor process, and its removal is part of the cytoreductive procedure. Second, these patients often require multiple cytoreductive procedures and omentectomy facilitates subsequent procedures. All gelatinous implants should be removed in an attempt to render the patient disease free. Tumor in the pancreaticoduodenal omentum requires removal of the lesser omentum, and at times the vagus nerve and left gastric artery. When the primary lesion is a mucinous adenocarcinoma of the appendix, macroscopically expressed as a mucocele rupture, with dissemination into peritoneal cavity, the appropriate treatment is right hemicolectomy and lymphadenectomy drainage. Moreover, when the lesion is restricted to the appendix, which can increase the cure. A large percentage of patients have recurrences after their initial operation. When feasible, these patients should undergo repeat laparotomies and aggressive debulking procedures if symptomatic. Repeat laparotomies also are indicated as needed to relieve intestinal obstruction from recurrent or persistent disease. The prolonged survival of these patients supports this aggressive approach, despite the increased morbidity that accompanies these repeat laparotomies. Because of the high recurrence rate after the initial operation, many adjuvant chemotherapeutic regimens have been used. For patients with pseudomyxoma peritonei originating in the colon and appendix, 5-fluorouracil has been the traditional drug of choice. The results of chemotherapy have not been encouraging overall. No improvement in survival has been documented. Because of the unpredictable nature of the disease and the observation that patients can remain disease-free without undergoing adjuvant chemotherapy after a complete workup, the patient should undergo exploratory laparotomy, resection of the primary tumor, and an aggressive cytoreductive procedure. Abdominal CT is an excellent technique to follow these patients for recurrence.

**SUGARBAKER PROTOCOL**

The procedure described by Sugarbaker consists of procedures of peritonectomy.

The peritonectomy procedures include:

1. Greater omentectomy and splenectomy
2. Left subdiaphragmatic peritonectomy
3. Right subdiaphragmatic peritonectomy
4. Lesser omentectomy and cholecystectomy with stripping of the omental bursa
5. Complete pelvic peritonectomy
6. Partial or complete gastrectomy

Occasionally has been successful. The gelatinous nature of the mucin, however, often makes attempts at aspiration futile. Laparoscopy guided biopsy is rarely successful. Much attention recently has been focused on preoperative sonographic and radiographic diagnosis of pseudomyxoma peritonei. Sonographic findings of an echogenic mantle with multiple intraperitoneal multicellular cysts and ascitic septations are specific for pseudomyxoma peritonei.

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