ABSTRACT:
Pseudomyxoma peritonei is a relatively rare and poorly understood condition in which mucus accumulates within the peritoneal cavity. The presence of cells in the mucin, either inflammatory or neoplastic, distinguishes it from simple acellular mucus ascites caused by mucinous spillage. There is widespread seeding of the peritoneal and omental surfaces with a heavy cancerous glaze. This is principally a complication of borderline or malignant neoplasm of the ovary and/or appendix. We report a case with findings of mucinous ascitis and frozen pelvis. Primary lesion could not be identified.

1. INTRODUCTION:
Pseudomyxoma peritonei characteristically arises from ruptured, primary ovarian, and appendiceal adenomas or (Mucinous cyst) adenocarcinomas, but can have an indeterminate site. Despite the abdominal viscera being thickly coated with the mucus-secreting tumour cells, invasion into the substance or extra peritoneal sites does not occur. Instead, the abdominopelvic cavities become filled with tenacious, semisolid, neoplastic mucus, rich in glycoproteins. This often forms large loculated cystic masses. Fox and Langley in 1976 proposed the development of Pseudomyxoma peritonei to be due to a foreign body reaction following spillage of mucus from ruptured cysts into the peritoneal cavity. This has now been disproved, as the presence of mucus-secreting tumour has been shown to be responsible for the development of this condition. In the past there has been much discussion regarding the definition and pathology of Pseudomyxoma peritonei. A recent paper by Sugarbaker et al defined Pseudomyxoma peritonei as an intestinal grade 1 mucinous adenocarcinoma that arises from a primary adenoma.

They describe a redistribution phenomenon in which cancer cells from appendix tumours are found localized at predetermined sites within the abdomen and pelvis, although in some cases the primary tumor may be small and inconspicuous.

2. CASE REPORT:
We report a case aged 50 years with history of abdominal distensions for the last 3 months and difficulty in breathing since 2 months. She has been undergone hysterectomy 5 years back and details of the surgery not available. On examination mass in the left iliac fossa extending to hypogastrium and left lumbar region, surface smooth, free fluid in the abdomen. No organomegaly, on per vaginal examination mass in the left fornix palpated, not fixed to the pelvic wall and no tenderness. Digital rectal examination done no evidence of growth or tenderness. Ultrasound abdomen revealed mixed echogenic mass lesion in left adnexa measuring 2.5x6.5x6.1 cm, left side hydronephrosis and ascitis. No organomegaly. X ray chest shows left pleural effusion. Pleural tapping revealed reactive mesothelial cells along with lymphocytes in the background of RBCs. Malignant cells seen in clusters and singles noticed 200cells/mm³, ascitic fluid analysis revealed predominant tumor cells in clusters.
150 cells/mm³. CA 125 estimation done is 967.50 U/ml (Normal<35 U/ml). CAT abdomen revealed a well defined heterogenous peripherally enhancing lesion with few necrotic areas within the lesion noted in pelvis measuring 9.4x8.9x8.9 cm, fat planed around the lesion with pelvic structures preserved. Left ovary is normal and right ovary shows a small cyst 2.8x2.5 cm. Patient was planned for diagnostic laparoscopy, on examination ascitis moderate, omental caking and nodules all around the parietal peritoneum, and frozen pelvis noticed. Biopsy of the omentum revealed adenocarcinoma with psammoma bodies. Figures(1-4)

3. DISCUSSION:

Pseudomyxoma peritonei is an indolent disease and is most prevalent in women aged between 50 and 70 (1). Until now, it has always been thought that cases of ovarian origin outnumber those of appendiceal origin (2). This theory has been put into question due to a recent report by Ronnett et al (3) which suggests that women actually have synchronous appendiceal and ovarian tumours. Moreover, a large proportion of these ovarian tumours have been shown immunohistochemically to be secondary to the appendiceal tumours. Either way, the fact remains that more women then men appear to suffer from this condition. Clinically, although painless, deterioration of general health begins long before diagnosis. Acute presentation during advanced stages of the disease is common and along with a host of non-specific symptoms, the main complaints are those of abdominal pain and distension. Inflammatory changes associated with peritoneal tumour implants can lead to fistula formation and adhesions (4, 5, 6) which in turn can cause intermittent or chronic partial bowel obstruction. Localized masses are frequently present in Pseudomyxoma peritonei of appendiceal
origin. Surprisingly, signs and symptoms of cancer such as cachexia are rare. Diagnosis is seldom absolute until laparotomy is performed. This is despite the presence of a distended abdomen with non-shifting ascites on physical examination. Laboratory studies are also of little help but fortunately, over the past few years, there have been many reports based on radiological imaging techniques (7), which are proving to be extremely useful in reaching a correct preoperative diagnosis. For example, in later stages of the disease, plain films used when the abdomen is distended with mucus show central displacement of the bowels with obliteration of the psoas muscle border. Occasionally, small calcific lesions can be seen widely disseminated throughout the abdomen. As the disease progresses, plain films become invaluable in following inevitable bowel obstruction and assessing the need for emergency debulking. Furthermore, when used in conjunction with barium studies, the proximal extent of the disease can be assessed and a possible extrinsic tumour causing large bowel obstruction can be ruled out. Conversely, ultrasonography is more useful and generally has similar features to computed tomography images showing abdominal echogenic masses with ascites, multiple septations, and scalloping of the liver.

Computed tomography shows four basic patterns (7-11): posterior displacement of the intestines with numerous low density masses and calcifications; diffuse peritoneal infiltration appearing similar to ascites with septated fluid pockets filling the peritoneal cavity; intrahepatic low density attenuated lesions; and scalloping of intra-abdominal organs due to extrinsic pressure of adjacent peritoneal implants. Scalloping of the liver has been widely described but in 1987, Parikh et al (11) reported the first case of splenic scalloping in Pseudomyxoma peritonei.

Finally, magnetic resonance imaging, which is still being investigated, may prove more helpful than computed tomography especially in assessing the rare visceral invasion by mucinous tumours.

This is based on the limited number of patients reported. One major disadvantage is the poor cost effectiveness compared with computed tomography. In summary, preoperative diagnosis could therefore be made with careful physical examination in conjunction with ultrasound and computed tomography. However, explorative laparotomy still remains the main diagnostic tool of choice. A positive finding is indicated by the presence of liters of yellowish-grey mucoid material involving both the omental and peritoneal surfaces. The pathological features of appendiceal mucinous cystadenocarcinomas closely mimic their ovarian counterparts. Histologically these neoplasms contain solid growths with conspicuous epithelial cell atypia and stratification, loss of gland architecture and necrosis, and are similar to colonic cancer in appearance. The pertinent cytological features of Pseudomyxoma peritonei include a mucinous background with mesothelial cells and histiocytes (13). The well differentiated columnar epithelial cells producing mucin usually display Minimal nuclear features of malignancy.

Even though the origins and nature of the parent neoplasms may be variable, this is not reflected in the cytological features of Pseudomyxoma peritonei (13). Prompt and aggressive treatment, including drainage of the mucus, surgical debulking of the primary and secondary tumour implants, and resection of the omentum should be instituted in all patients (14). Commonly, at
laparotomy a right hemicolectomy is performed. In order to prevent recurrence, resection of both ovaries and the appendix must be carried out in all female patients where the primary site is not found.

4. REFERENCES:


