Case Report

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Radiology Section

Pseudomyxoma Peritonei: Radiological Aspect

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ABSTRACT

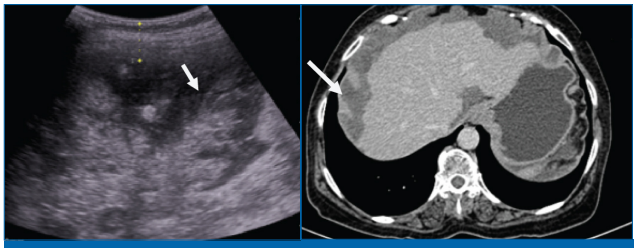
Pseudomyxoma peritonei is one of the complications of mucin secreting tumours arising from appendix or ovary. Malignancy of biliary tree, urachus and colon may also have similar presentation. It can also be a post operative presentation in aforementioned malignancy. Excess of mucin may impair function of viscera. Early diagnosis, management and postoperative outcome is major prospective of radiological imaging. We hereby present a case of 61 years old female who presented with the complaint of pain abdomen for last 2 months which was not relieved by taking medicines. Computed Tomography (CT) scan was done which suggested it as umbilical metastatic deposit.

Keywords: Cystadenocarcinoma, Mucinous, Ovary

CASE REPORT

A 61-year-old female patient came with complain of pain abdomen (predominantly in lower abdomen) since last 2 months. Pain was dull aching, non radiating in nature and did not relieved by taking medicine. No history of fever and bladder or bowel complains. Patient underwent exploratory laparotomy 2 years back, followed by excision of ovarian mass with right abdominal panhysterectomy. On histopathology, it was diagnosed as mucinous cystadenocarcinoma of bilateral ovary (R>L) with omentum and myometrial infiltration. Later patient received 6 chemotherapy cycles.

Patient was referred to the Department of Radiology for ultrasound and CT-scan. Ultrasound showed a large mixed echogenic mass in right flank extending to right subhepatic region with ill defined outline with scattered cystic component and intervening solid component. Colon and small bowel loops were seen adherent to it. Evidence of gross ascites with echogenic content scalloping on surface of liver and spleen [Table/Fig-1].



[Table/Fig-1]: USG showing hypoechoic collection (arrow) in right paracolic gutter with heteroechoic content.
[Table/Fig-2]: Axial post contrast CECT shows hypodense lesions (arrows) in perihepatic region causing scalloping on the surface of liver.

CT-scan was done later on ill defined cystic lesion in right subphrenic, sub hepatic, perisplenic, right paracolic gutter and pelvis with multiple enhancing soft tissue component. Lesion was displacing bowel loops centrally and scalloping surface of liver and spleen. Multiple enhancing soft tissue density lesions in peritoneum and omental surface. A well defined enhancing soft tissue density lesion measuring 1.5 cm x 1.3 cm in umbilical region suggestive of umbilical deposit (metastatic deposits) [Table/Fig-2-5].

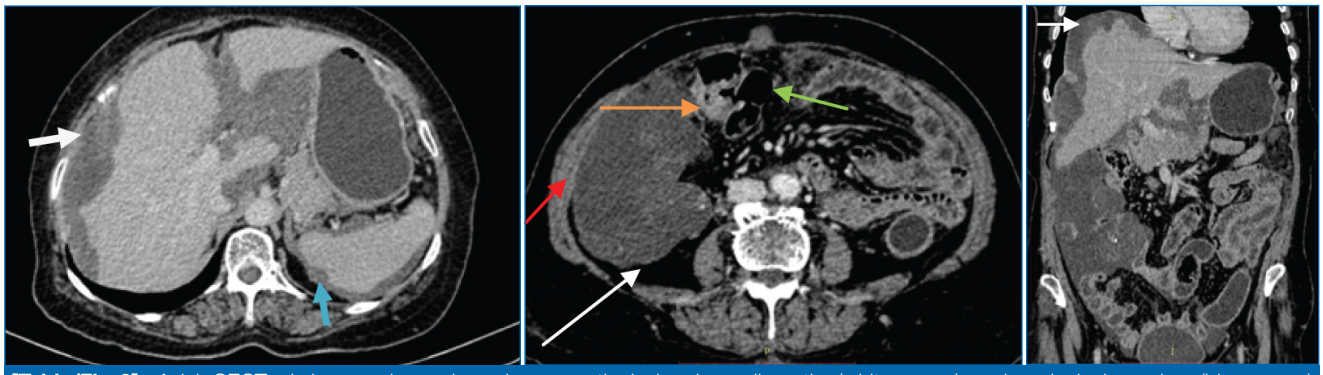
FNAC was performed, fluid aspiration was done and the findings were consistent with pseudomyxoma peritoeni.

The patient was then referred to the Surgery Department for further management, where she was lost of further appointments.

DISCUSSION

The term pseudomyxoma peritonei was coined by Werth in 1884 [1], which means a false mucinous tumour of the peritoneum. Pseudomyxoma peritonei or a false mucinous tumour of peritoneum is term to describe pathologically any benign or malignant tumour as a result of intraperitoneal accumulation of mucin [2] characterised by mucinous ascites along with peritoneal and omental deposits.

Incidence rate of pseudomyxoma peritonei is 1 case per million population per year [3]. and common in women than men. Mean age of diagnosis is 49 years (range, 23-83 years) [4]. Common presentation is progressive abdominal pain, increased abdominal girth and weight loss. Patient may also present pain in lower abdomen on right side mimicking appendicitis [5].



[Table/Fig-3]: Axial CECT abdomen shows hypodense cystic lesion in perihepatic (white arrow) and perisplenic region (blue arrow) causing scalloping on surface of liver and spleen. **[Table/Fig-4]:** Axial CECT abdomen shows enhancing omental nodule (orange arrow) with peritoneal thickening (red arrow). Hypodense cystic lesion with enhancing soft tissue component in right paracolic gutter displacing bowel loops medially (white arrow). Enhancing soft tissue component noted in umbilicus (green arrow) suggestive of umbilical deposit. **[Table/Fig-5]:** Coronal CECT abdomen shows hypodense cystic lesion (arrow) scalloping on the surface of liver.

In rare cases, ureteric obstruction and oedema in lower limbs may be seen secondary to venous obstruction. Recurrent disease may be seen on bowel surfaces presenting as intestinal obstruction or obstructive jaundice secondary to fibrosis and intestinal adhesions, that can be fatal [6,7].

Ronnet BM et al., [8] analysed the 109 patients with pseudomyxoma peritonei and based on clinicopathological features divided it into two categories i.e., Disseminated Peritoneal Adenomucinosis (DPAM) and Peritoneal Mucinous Carcinomatosis (PMC). Disseminated Peritoneal Adenomucinosis (DPAM) was characterised by abundant extracellular mucin with scanty proliferative mucinous epithelium, showing little cytological atypia or mitotic activity. Peritoneal Mucinous Carcinomatosis (PMC), shows abundant mucinous epithelium with cytological features of carcinoma.

Pseudomyxoma peritonei commonly arise from neoplasm of appendiceal or ovarian origin. Appendiceal lesions seen with pseudomyxoma peritonei include mucosal hyperplasia, benign cystadenoma and cystadenocarcinoma. Primary ovarian cystadenocarcinoma or mucinous borderline tumours were earlier thought to be the site of origin. Site of origin is controversial as both of organs are involved in the majority of female patients [9]. Several studies showed that ovaries are involved secondary to appendix origin [10,11] which is supported as 25-50% of the patients are male and ovarian lesions are usually bilateral or mainly on right side with tumour implants were found to be on the surface rather than within ovarian tissue in 75% of patients. Morphological and immunophenotypical differences between synchronous lesions of ovary and appendix also support two site origin of tumour [12].

Apart from appendix and the ovary, rare sites of origin of pseudomyxoma peritonei includes colon, stomach, gall bladder, pancreas, urachus, urinary bladder, breast, fallopian tube, uterine corpus and lung [13,14].

On imaging, radiography in pseudomyxoma peritonei patients is similar to massive ascites shows increased opacity throughout the abdomen, with poorly defined margin of abdominal viscera and obliterated psoas margins, obscured inferior hepatic margin and liver tip displaced medially (Hellmer's sign), centrally displaced ascending and descending colon and the properitoneal fat stripe displaced laterally when large amounts of mucin are in the paracolic gutters' opacity on either side of the bladder. Calcification is also reported within pseudomyxoma peritonei after chemotherapy, they may also be present before chemotherapy [15].

On sonography, pseudomyxoma peritonei may present as echogenic ascetic fluid with non mobile echoes and echogenic septations. Echogenic masses secondary to omentum and parietal peritoneum involvement may also be seen. Bowel loops are displaced centrally surrounded by echogenic ascetic fluid [16]. Scalloping of hepatic and splenic margins may also be present.

CT characterised by hypodense multiple complex cystic masses in the peritoneum, scalloping of the liver and occasionally splenic margins, omental thickening/omental caking and peritoneal implants with septated pseudo ascites compressing the visceral organs. Soft tissue attenuation area also be present suggesting solid tumour elements, fibrosis, or compression of the mesentery [16]. Scalloping of the visceral representing the indentations on visceral capsule margins by extrinsic pressure of the mucinous implants.

Differential diagnois of pseudomycoma peritonei includes peritoneal carcinomatosis, peritoneal sarcomatosis and peritonitis. MRI may be more useful than CT-scan especially in assessment of visceral invasion by mucinous tumours. Major disadvantage of MRI is the poor cost effectiveness in comparison to the CT-scan.

CONCLUSION

Pseudomyxoma peritonei commonly arise secondary to ovarian/appendix malignancy. These patients may require

repeated removal of mucinous material. Knowledge of this rare condition is an important for early diagnosis and proper management.

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