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Case Report

Low Grade Primary Peritoneal Serous Carcinoma: A Case Report

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ABSTRACT:

Background: Primary peritoneal serous carcinoma (PPSC) is a rare malignancy, predominantly occurring as high-grade tumors. This case report discusses a rare presentation of low-grade PPSC, emphasizing its unique clinical, radiological, and pathological characteristics.

Case Presentation: A 64-year-old female presented with a year-long history of intermittent abdominal pain, distension, anemia, and melena. Physical examination revealed an indurated mass in the supra-pubic region. Imaging studies showed an infiltrative mass in the infraumbilical region. Serum CA-125 was elevated. Diagnosis was established post-operatively after laparoscopic examination revealed peritoneal nodules. Histopathological and immunohistochemical studies confirmed features of low-grade serous carcinoma. Despite initial treatment response, the patient's condition deteriorated, leading to eventual mortality.

Discussion: LGSC of the peritoneum is extremely rare. Their presentation with non-specific abdominal symptoms poses diagnostic challenges. Differential diagnosis should include other causes of peritoneal carcinomatosis and metastatic serous carcinomas. While treatment aligns with that of epithelial ovarian cancer, early diagnosis remains a challenge due to non-specific presentation.

Conclusion: Clinicians should consider PPSC, even its low-grade variant, in differential diagnosis for postmenopausal women with unexplained abdominal symptoms, especially when imaging studies are inconclusive.

Keywords: Primary peritoneal serous carcinoma, Low-grade serous carcinoma, Peritoneum, CA-125, Differential diagnosis, Postmenopausal women.

INTRODUCTION:

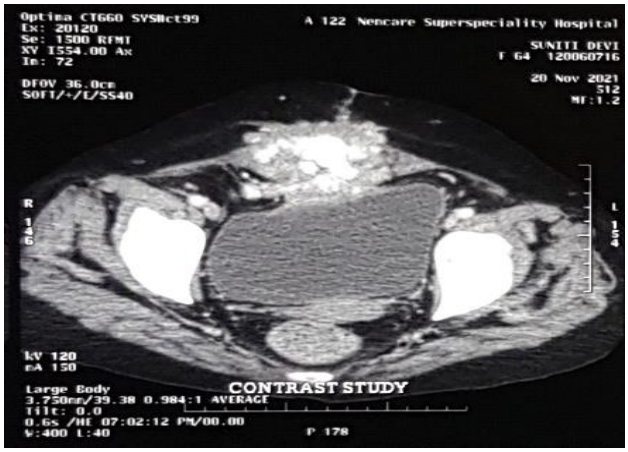
Primary peritoneal serous carcinoma (PPSC) is a rare malignancy that originates in the peritoneal lining of the abdomen and pelvis. Remarkably, it arises in the absence of overt disease in the ovaries or fallopian tubes. It's believed to stem from embryonic remnants of müllerian cells within the peritoneum[1,2]. First identified by Swerdlow in 1959[3], PPSC closely resembles papillary serous ovarian carcinoma in its clinical and pathological manifestation. Moreover, serous carcinomas of the peritoneum, ovary, and fallopian tube are further delineated into high-grade serous carcinoma (HGSC) and low-grade serous carcinoma (LGSC) based on their molecular pathogenesis and therapeutic response[4]. Notably, the literature predominantly reports high-grade cases of serous peritoneal carcinomas[5]. This report presents a case of low-grade primary peritoneal serous carcinoma, underscoring its clinical, radiological, and pathological characteristics to differentiate it from other causes of peritoneal carcinomatosis.

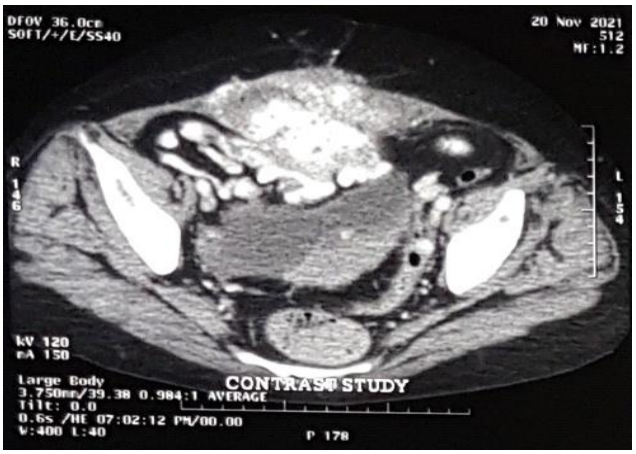
Case Presentation:

A 64-year-old female, of average build, reported a year-long history of intermittent abdominal pain, distension, anemia, and melena. Accompanied by colicky pain and passage of black stool, she had managed long-standing diabetes with insulin, hypertension with medication, and showed symptoms of hypothyroidism and chronic kidney disease. Despite undergoing conservative treatment for recurrent subacute intestinal obstruction and anemia for a year, her symptoms would periodically alleviate only to return. The patient had been admitted to the emergency room twice with acute abdominal pain and features suggesting subacute intestinal obstruction. A colonoscopy returned inconclusive results. A CT scan, however, indicated lower abdominal fibrous adhesions and potential ulcerations & omentum involvement. Her medical history revealed a cesarean section 36 years prior, a laparoscopic infraumbilical ventral hernia repair a decade ago, and an emergency laparotomy for incarcerated infraumbilical omentocele with peritonism 18 months back. Post-surgery, she

experienced a discharging sinus from the incision for about three months. Physical examination revealed a puckered infraumbilical scar, and a palpable indurated mass in the supra-pubic region, measuring 10x6 cm, which was hard, fixed, and non-expansile upon coughing.

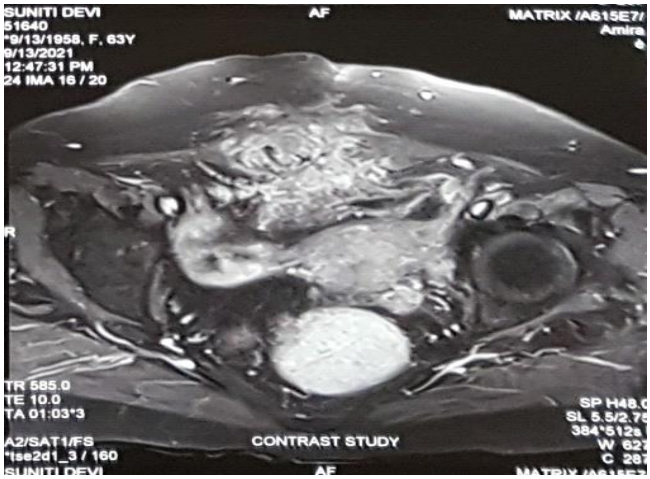
Repeat CECT abdomen revealed infiltrative mass with internal calcification in infra umbilical region of anterior abdominal wall which was adherent to distal small bowel loops and anterior urinary bladder wall with non-dilated small bowel. Bilateral ovaries and uterus are normal.





Infiltrative mass with internal calcification in infra umbilical region of anterior abdominal wall, adherent to distal small bowel loops and anterior urinary bladder wall.

MRI of whole abdomen reveals large inflammatory mass in the lower anterior abdominal wall with marked adhesion with pelvic small bowel loops causing sub-acute small bowel obstruction.no abnormality seen in bilateral ovaries and uterus.



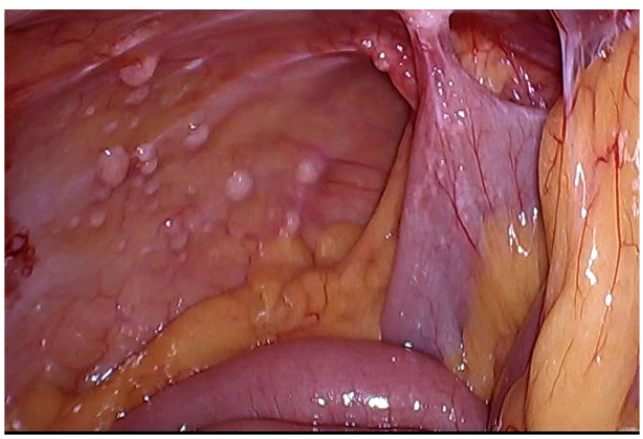
Inflammatory mass in the lower anterior abdominal wall with marked adhesion with pelvic small bowel loops, uterus & ovaries not involved.

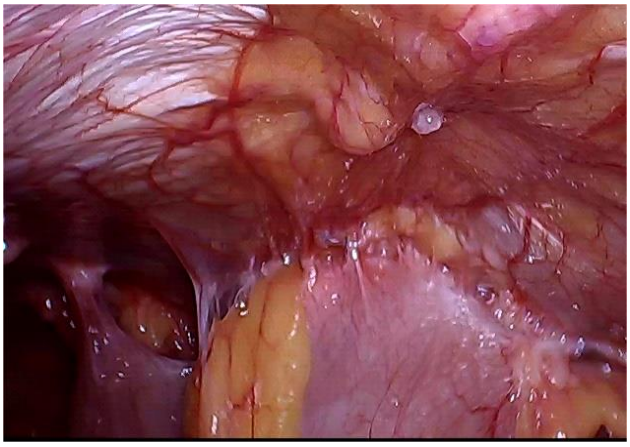
CA-125 was raised (217 U/ml).

Image guided trucut biopsy from lower anterior abdominal wall lesion revealed foreign body granulomas with fibrosis. On the basis of history and supporting investigations pre op diagnosis of SAIO with inflammatory mass lesion adhered to lower anterior abdominal was made and decision for diagnostic laparoscopy and proceed was taken.

On Diagnostic Laparoscopy Following Findings Were Noted:

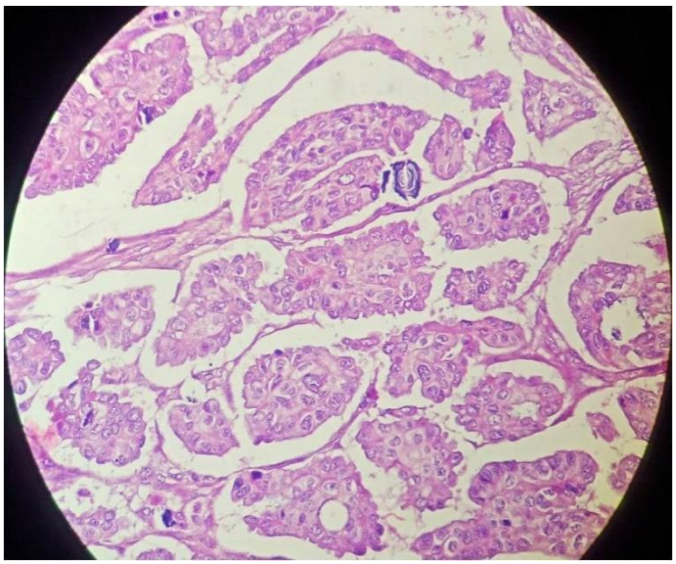
- Minimal ascites/intra-abdominal collection
- Few peritoneal nodules in the lower abdominal wall near the bowel adhesions. Tissue from the nodules and adjacent area was taken for histopathological examination and immunohistochemistry
- Conglomerated mass comprising of omentum, loops of small bowel and part of sigmoid colon densely adhered to infraumbilical part of anterior abdominal wall in the previously operated ventral hernia site (mesh adhesion).
- Liver, stomach, large intestine and rest part of small bowel are normal.





Laparoscopic adhesiolysis was done and the crowded small gut loops were separated from abdominal wall with sharp and blunt dissection. Previous mesh was removed with great difficulty due to dense adhesions. During the process of adhesiolysis we encountered enterotomy due to in tight adhesion intra bowel adhesion. So the procedure was converted to open technique. The conglomerated mass was excised leaving and resection anastomosis was done. Haemostasis maintained and abdomen was closed.

Microscopic examination of serosal nodules with densely adherant areas revealed features of low grades serous carcinoma composed of invasive nests of malignant cells having mild cytologic atypia with multiple foci of psammomatous calcification. No necrosis, mitosis identified. Mucosal areas shoed focal areas of mucosal necrosis and haemorrhage. Immunohistochemistry was confirmative as the tumour cells expressed Pan CK, CK 5/6, HBME-1, CK -7, PAX-8,WT 1, P16, ER and focal P53. Tumour cells were negative for Calretinin, CK20, CEA, CD 56, Synaptophysin, chromogranin.



Patient recovered well post operatively and oral feeding started on 4th post op day . pt was discharged on 7th post op day. Medical oncologist took over the treatment after two weeks.

She was treated with oral etoposide 50mg and cyclophosphamide 50 mg daily for two weeks with one week off. Total six cycles were given. She showed initial good response but started deteriorating after sixth cycle. She developed ascites not responding to treatment and ultimately succumbed to the disease.

DISCUSSION:

PPSC is a rare tumor that diffusely involves peritoneum having similar histopathological features of primary ovarian serous carcinoma. Incidence of PPSC is about one-tenth of tuboovarian serous carcinomas. Most PPSCs are HGSCs which share same frequency of germline BRCA mutations as their tuboovarian counterparts [5,6] . LGSCs of peritoneum are extremely rare which occur de novo or after diagnosis of serous borderline tumour. The MAPK pathway is involved in pathogenesis of LGSCs [6]. Histologically LGSCs are characterised by mild nuclear atypia, mitotic index less than 12 mitoses/ 10 high power fields and lack necrosis. Whereas HGSCs have widespread nuclear pleomorphism, high mitosis, aberrant p53 immunophenotype (diffuse or null positivity). Our case had both histological and immunophenotypical features of LGSC.

Moreover PPSC need to be distinguished from other causes of peritoneal carcinomatosis notably peritoneal malignant mesothelioma (PMM) and metastatic serous carcinomas from ovary/ fallopian tube. In 1993, the diagnostic criteria of PPSC described by the Gynecology Oncology Group include ovaries must be either absent or normal size in size; extra ovarian involvement is must be greater than the surface involvement of either ovary; absence of a deep seated invasive ovarian carcinoma or invasive disease in ovarian cortical stroma with tumour that measure less than 5x5 mm^2 ; and histological and cytological features of ovarian epithelial carcinoma [7]. In our case bilateral ovaries were atrophic. Malignant mesothelioma was ruled out due to tumour being calretinin negative and estrogen receptor positive. Metastasis from uterus could be ruled out due positivity of tumour cells for WT1.

Most cases of LGSC of peritoneum present with non-specific symptoms of abdominal pain and distension which was also seen in our case. In our situation the patient presented to emergency department with features of sub acute intestinal obstruction and diagnosis was made post operatively. Therefore a possibility of serous carcinomas of peritoneum must be considered in differential diagnosis of chronic abdominal pain and distension. Serum tumour marker CA 125 is found to be raised in serous tumours which was elevated in this case.

Clinically, women diagnosed with PPSC are treated using the same surgical and chemotherapeutic

approach as epithelial ovarian cancer because of the similarities in biological behavior. The management of PPSC consists of combining optimal surgical debulking with an intravenous Taxol and Platin doublet chemotherapy of six cycles may offer the patient the most effective treatment.

CONCLUSION:

Although rare, PPSC should always be in the differential diagnosis in postmenopausal women presenting with unexplained abdominal symptoms, especially in the context of a non-conclusive imaging study. It's essential for clinicians to be aware of this entity to ensure timely diagnosis and appropriate management. The combination of clinical presentation, imaging, and histopathology is paramount in arriving at the correct diagnosis. However, early identification remains elusive due to its non-specific presentation, making prompt treatment difficult.

Ethical approval:

Institutional and international research regulations were observed during the time of interacting with the patient and the preparation of the manuscript.

Consent:

A written informed consent was obtained from the patient for the case details to be published, and it has been kept by the corresponding author.

Conflicts of interest

The authors declare that there are no conflicts of interest regarding the publication of this article.

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