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Case Report

# Primary peritoneal serous carcinoma – A rare case report

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ABSTRACT

Primary Peritoneal Serous Carcinoma (PPSC) was first described by Swerdlow in 1959. As the epithelial layer of the ovary and peritoneum derives from common embryonic origin, coelomic epithelium, PPSC is often misdiagnosed as serous carcinoma of ovary. Estimated incidence of PPSC in the United States is 6.78 cases/ 1,000,000 individuals. A 67 years female presented with ascitis. Ultrasonography revealed normal uterus with bilateral adnexae. No pathology was mentioned elsewhere. Ascitic fluid revealed features positive for malignancy. On gross examination, omentum was nodular and firm with no gross pathology in bilateral adnexae. Microscopically, omentum revealed features suggestive of High grade serous carcinoma with tumour deposits in cortical stroma of bilateral adnexae (tumour nests <5 mm2). Hence a diagnosis of High grade serous carcinoma, primary peritoneal was made. Clinico-radiological and histopathological correlation is essential in the diagnosis of PPSC. Diagnosis of PPSC is based on Gynecology Oncology Group criteria (1993). Recognition of this entity becomes important as PPSC is aggressive in nature, debulking surgery and chemotherapy is the only available treatment option.



## 1. Introduction

Primary Peritoneal Serous Carcinoma (PPSC) was first described by Swerdlow in 1959.[1] Since the epithelial layer of the ovary and the peritoneum shares a common embryonic origin, deriving from coelomic epithelium, PPSC tends to be misdiagnosed as serous carcinoma of ovary. PPSC and Epithelial ovarian carcinoma were considered as a single entity in the past, but molecular and epidemiological findings have shown that PPSC is distinct from epithelial ovarian carcinoma.[2] Estimated incidence of PPSC in the United States is 6.78 cases/ 1,000,000 individuals.[1,3] The common presenting symptoms of PPSC are abdominal pain, abdominal distension and abdominal discomfort.[1] Clinico-radiological and histopathological correlation is essential in the diagnosis. Diagnosis of PPSC is based on the Gynecology Oncology Group criteria (1993).[1] Recognition of this entity becomes essential for accurate evaluation and management of the patient.

## 2. Case Presentation

A 67 years female presented with abdominal distension with ascitis. We received specimen of bilateral salpingo-oopherectomy and omentectomy with a clinical suspicion of carcinoma ovary. We also received ascitic fluid for malignant cytology.

On gross examination, external surface of omentum was nodular. Cut section was firm, homogenously white in colour. Bilateral adnexae were normal and revealed no gross pathology (Figure 2 a,b,c).

Microscopically, omentum revealed tumour cells arranged in papillae and solid nests, exhibiting pleomorphic vesicular nuclei, prominent nucleoli with few bizzare cells

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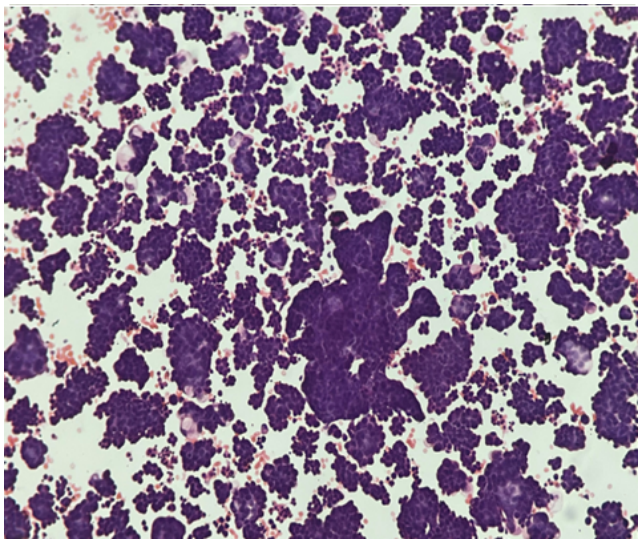

**Figure 1:** Ascitic fluid reveals pleomorphic tumour cellsarranged in 3 dimensional clusters, features positive for malignancy.

and increased mitotic activity, features suggestive of High grade serous carcinoma (Figure 3 a,b). These tumour deposits were also seen in cortical stroma of bilateral adnexae (tumour nests <5 $mm^2$). Rest of the bilateral adnexae was unremarkable. Hence a diagnosis of High grade serous carcinoma, Primary peritoneal was made (Figure 4 a,b). Immunohistochemistry workup was advised, but the patient was lost to follow up.

## 3. Discussion

Primary Peritoneal Serous Carcinoma (PPSC) is a rare epithelial malignancy, first described by Swerdlow in 1959.[1]

The incidence of PPSC is considerably lower than that of epithelial ovarian cancer, 6.78 cases/ million versus 120.5 cases/ million, respectively. According to Teng Hou et al. PPSC predominantly affects postmenopausal women.[4] According to the literature, CT/PET scan plays a role in the diagnosis.[5]

The diagnosis of PPSC mainly relies upon clinico-radiological and histopathological correlation. Diagnosis of PPSC is made based on Gynecology Oncology Group criteria (1993) as follows: i) Both ovaries must be normal in size or enlarged due to benign process; ii) Involvement in extraovarian sites is greater than the involvement of either ovarian surface; iii) Absence of a deep-seated invasive ovarian carcinoma or tumour measuring <5×5 $mm^2$in ovarian cortical stroma; and iv) Histopathological and cytological characteristics of the tumor similar to epithelial ovarian cancer.[1,6] PPSC is a part of the hereditary breast-ovarian cancer (HBOC) syndrome since the frequency of BRCA mutations in peritoneal and ovarian cancer is similar.[3] Diagnostic tests,

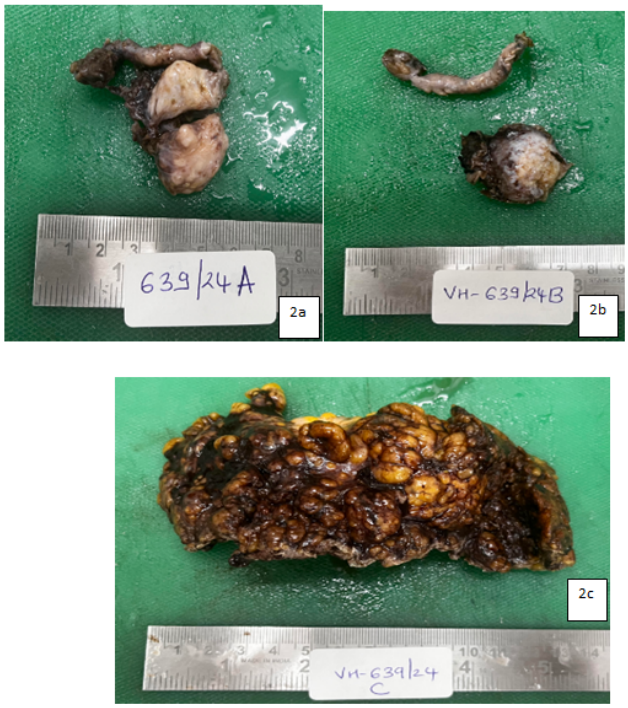


**Figure 2: a)** & **b)** :Gross picture of bilateral adnexae. **c)**: Gross picture showing multiple nodules overomentum.

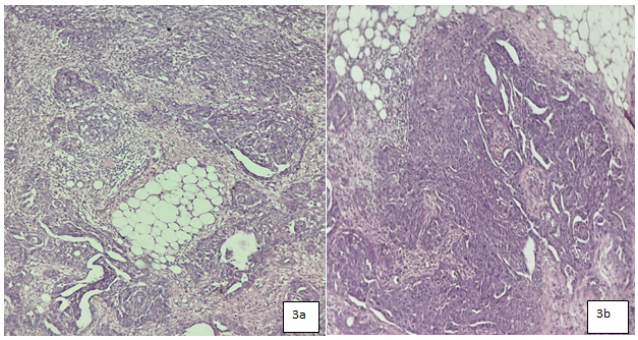


**Figure 3: a)** & **b)** : Microscopically, Omentum reveals tumour cells arranged in papillae and solid nests, suggestive of high grade serous carcinoma.

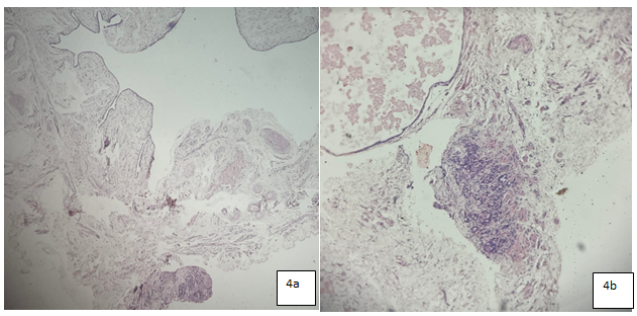


**Figure 4: a)** & **b)** : Tumour deposits<5$mm^2$ in cortical stroma of bilateral adnexae.

such as CT/ 18 Fluorodeoxyglucose positron emission tomography (18 FDG PET), is useful in detecting any abnormal mass or irregular omental thickening, aiding in the diagnosis of PPSC. However, histopathological examination and immunohistochemical staining is the gold standard in diagnosing PPSC. Immunohistochemical studies also aids in differentiating PPSC from its close differentials, which include primary epithelial ovarian cancer (PEOC), peritoneal malignant mesothelioma (PMM) and metastasis.[7] Cytoreductive surgery and platinum based chemotherapy is the only available treatment option for PPSC.[8,9]

## 4. Conclusion

Primary Peritoneal Serous carcinoma (PPSC) is a rare epithelial malignancy. Diagnosis mainly relies upon clinical, radiological and histopathological findings. Diagnosis of PPSC is based on Gynecology oncology Group criteria. Recognition of this entity is important as it's a rare and aggressive malignancy,cytoreductive surgery and platinum based chemotherapy is the only available treatment.

## 5. Source of Funding

None.

## 6. Conflict of Interest

None.

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