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Pathology

PRIMARY PERITONEAL SEROUS CARCINOMA- A RARE PRESENTATION & ITS DIAGNOSTIC CHALLENGES

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ABSTRACT Primary peritoneal serous carcinoma is a rare primary malignancy of peritoneum with wide clinical and histologic concordance with epithelial ovarian malignancy. Proper radiologic evaluation with laparotomy and extensive histologic study with a good degree of clinical suspicion can help in arriving at a definitive diagnosis.

KEYWORDS : PPSC, High Grade Serous Carcinoma, ER, Ki-67.

INTRODUCTION:

Primary Peritoneal Serous Carcinoma (PPSC) is a very rare primary malignancy of peritoneum, with an age-adjusted incidence rate of 6.78 per million[1]. It diffusely involves the peritoneum & is clinically & histopathologically indistinguishable from primary serous ovarian carcinomas[2]. This can be attributed to the fact that epithelial layer of the ovary & the peritoneum shares a common embryonic heritage, deriving from coelomic epithelium early in life[3].

Here we present a case of this rare entity, in a 60 years old female patient.

CLINICAL PRESENTATION:

A 60 years old female patient presented in medicine OPD, with complaints of abdominal distention, nausea, constipation, loss of appetite, bloating & significant weight loss over a period of 3 months. Patient did not have any significant past medical & surgical history. She had menopause 10 years back.

She was advised complete blood count, that showed mild anemia, whereas Liver Function Test (LFT), Serum Urea & Creatinine showed values within normal ranges. On Ultra-sonography of whole abdomen, there was moderate amount of ascites, patchy hyper echoic nodular thickening of omentum, surfaces of bladder & pouch of Douglas. Bilateral ovaries were of normal size of 3 cm diameter each. All other findings were within normal limits. She was later advised for Serum Ca-125 levels, that showed values within normal range.

She was later prepared for laparoscopic biopsy of omental, other peritoneal nodules, bilateral ovaries and fallopian tubes & tissues were sent for histopathological examination in Department of Pathology.

HISTOPATHOLOGICAL STUDY:

Sections from all omental & peritoneal nodules showed similar histologic properties of the neoplastic pathology. There were tumor cells arranged in sheets, vague papillary (Figure 1) & transitional pattern (Figure 2). The cells are highly pleomorphic with moderate cytoplasm, focal clear cell change, marked nuclear atypia, prominent nucleoli, atypical & brisk mitosis. There were also areas of necrosis. The histologic picture corroborated with High Grade Serous Carcinoma of Ovary (Figure 3). However, histological sections from bilateral ovaries and fallopian tubes (Figure 4) were within normal limits, which was in concordance with the imaging study.

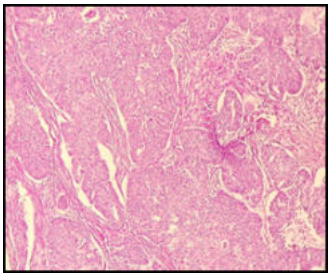

Figure 1: Papillary architecture in peritoneal nodule

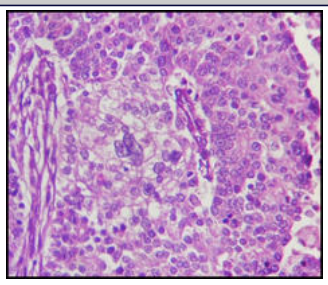

Figure 2: Transitional pattern of arrangement with focal clear cell change

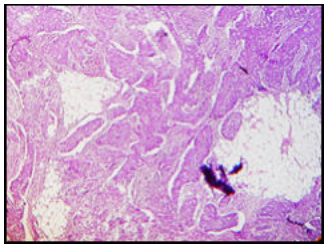

Figure 3: Omentum showing involvement with PPSC

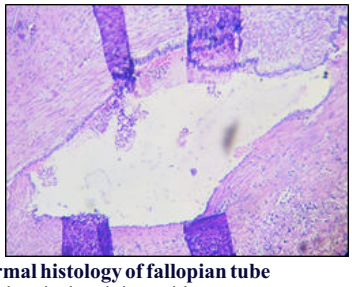

Figure 4: Normal histology of fallopian tube

Immunohistochemical staining with estrogen receptor (ER) showed diffuse positivity (Figure 5). Ki-67 staining pattern showed the high proliferative nature of the tumor (65% positivity) (Figure 6).

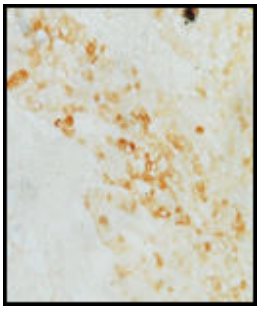

Figure 5: ER positivity in PPSC

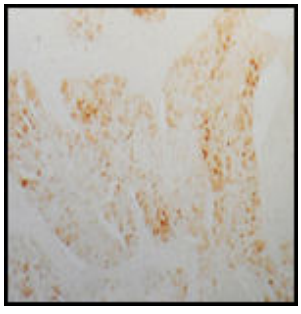

Figure 6: High Ki-67 positivity (65%)

From the above histopathological picture, in corroboration with imaging & immunohistochemical study, a diagnosis of **Primary Peritoneal Serous Carcinoma (PPSC)** was offered.

DISCUSSION:

Primary peritoneal serous carcinoma is classically defined as malignant serous epithelial neoplasm arising in peritoneum without any gross and microscopic involvement of bilateral adnexae. It is a fairly unusual tumor of secondary müllerian system with marked similarity in clinical presentation, histology & progression with epithelial ovarian neoplasm, as was evident from this case[4]. A multifocal origin of PPSC was mentioned by Muto *et al*, as was also seen here with disseminated nodules throughout the peritoneal cavity[5]. There are ongoing studies to demonstrate association of such cases with *BRCA* mutation[6].

Most cases of PPSC were reported in elderly postmenopausal women, with a median age range of 57-66 years[7] which as at par with the age of this case. Very occasional cases have been demonstrated in male subjects[8]. With better imaging resolutions and understanding of this entity, there has been a substantial increase in frequency of this neoplastic process and conclusive laparotomy outcomes.

Some of the morphological mimickers include malignant mesothelioma of peritoneum, metastatic peritoneal carcinomatosis, peritoneal psammocarcinoma & advanced stage ovarian carcinomas with peritoneal and omental spread[7]. Diagnostic laparotomy with extensive histopathological & immunohistochemical assessment can solve the diagnostic dilemma often encountered in these cases[9].

In our scenario, the clinical picture with presenting age was that of an ovarian pathology, which was discordant with imaging and tumor marker profile. The most difficult conundrum was to identify the origin of primary neoplasm. The histopathological diagnosis was that of High Grade Serous Carcinoma. In absence of primary focus at other site, as delineated by imaging studies and grossly normal ovaries seen in laparoscopy, imaging and in histology, PPSC was the definitive diagnosis that was delivered.

CONCLUSION:

Clinical presentation of PPSC may be that of malignant epithelial neoplasm. A sole histologic picture may be that of High Grade Serous Carcinoma. Thus, PPSC should always be considered a possibility in the backdrop of our mind in cases with grossly normal bilateral adnexae, but disseminated peritoneal carcinomatosis and malignant tumor ascites.

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