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Case Report

Primary Peritoneal Serous Carcinoma Often Clinically Misdiagnosed As Primary Serous Ovarian Carcinoma

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Abstract
Primary peritoneal serous carcinoma (PPSC) is a rare entity that diffusely involves the pelvic peritoneum seen predominantly in elderly postmenopausal women. The worldwide incidence is unknown because of the lack of large multicentre studies. PPSC are difficult to diagnose preoperatively as radiological investigations fail to interpret peritoneal infiltration. The histopathological criteria laid down by Gynaecology Oncology Group GOG helps to distinguish between primary peritoneal carcinoma and peritoneal metastasis from primary ovarian carcinoma.
Keywords: *Primary peritoneal serous carcinoma, Ovary, peritoneal deposits.*

Introduction

Primary peritoneal carcinoma is a rare malignancy arising from peritoneal epithelium and it was firstly described by Swerdlow in 1959 as "mesothelioma of pelvic peritoneum"[1]. Primary peritoneal serous papillary carcinoma (PPSPC) is the most common histologic type among primary peritoneal malignancies. The worldwide incidence is unknown because of the lack of large multicentre studies[2,3]. It is very difficult to distinguish primary peritoneal serous carcinoma from primary serous ovarian carcinoma as the patient presents with same clinical presentation in both PPSC and PSOC. The diagnostic criteria of this entity to differentiate it from primary serous carcinoma of ovary has been defined by the Gynaecology Oncology Group includes: - (a) Ovaries must be normal in size or enlarged as a result of benign process. (b) Extraovarian involvement must be greater than the surface involvement of either ovary (c) ovarian involvement must be absent, confined to the ovarian surface epithelium without stromal invasion, or involve the cortical stroma with a maximal tumour dimension of less than 5x5 mm. (d)The tumor in the extra-ovarian site should be serosal both histologically and cytologically[4].

Case Report

A 64-year-old female menopausal for 2 years presented with abdominal distension for 1 month. On per abdomen examination abdomen was uniformly distended with superficial veins visible and fluid thrill present. Ascitic fluid analysis revealed low SAAG ascites and possibility of malignancy and tuberculosis was kept. CECT Abdomen revealed gross ascites with omental thickening with abdominal and pelvic lymphadenopathy and uterus and bilateral adnexa were normal**.** [FIG-1A,B,C] Ascitic fluid cytology showed metastatic adenocarcinoma and CBNAAT was negative. CA-125 level was

markedly raised (17162.2 U/ml) and CEA was normal. Mammography and colonoscopy studies were normal. Considering the above findings a clinical diagnosis of carcinoma ovary was kept and platinum based chemotherapy was started and CA-125 levels were monitored routinely. After receiving 6cycles of chemotherapy patient underwent exploratory laparotomy with total abdominal hysterectomy with bilateral salpingo-opherectomy with infracolic omentectomy. On opening the abdomen there was evidence of adhesion of gut with uterus, fallopian tubes and ovaries. Uterus was post-menopausal in size and bilateral adnexa were normal and there were metastatic deposits in gut and omentum. We received a specimen of uterus with cervix measuring 6x4x2.5 cm with attached bilateral adnexa. Right and left fallopian tubes measured 4.5 cm and 6 cm in length respectively and right and left ovaries measured 2.5x1x0.7 cm and 1x0.5x0.5 cm respectively. Endometrium was atrophic, myometrium, bilateral fallopian tubes and ovaries were grossly within normal limits.[FIG-2] We also received two globular fibrofatty soft tissue pieces measuring 5x4x2 cm and 16x8x2 cm respectively. On cut section of both showed multiple grey white grey brown firm areas were seen along with yellowish areas. On microscopic examination endometrium showed cystic atrophy, myometrium, cervix and bilateral fallopian tubes were within normal histological limits. Right side ovary showed psammoma bodies involving the ovarian surface and no epithelial component was seen while the left ovary showed psammoma bodies and occasional malignant glands on the surface measuring <5mm in diameter with adherent omental tissue.[FIG-3,A,B] Sections from the omentum revealed sheets, papillae of tumor epithelial cells showing pleomorphism, high N/C ratio, prominent nucleoli, increased mitotic activity, moderate to abundant vacuolated cytoplasm, abundant psammoma bodies, fibrosis and moderate chronic inflammatory infiltrate [.FIG4.A,B] On immunohistochemistry tumor cells were positive for CK7,CA125 and negative for CK20 and vimentin. Diagnosis of primary peritoneal serous carcinoma post NACT was given.

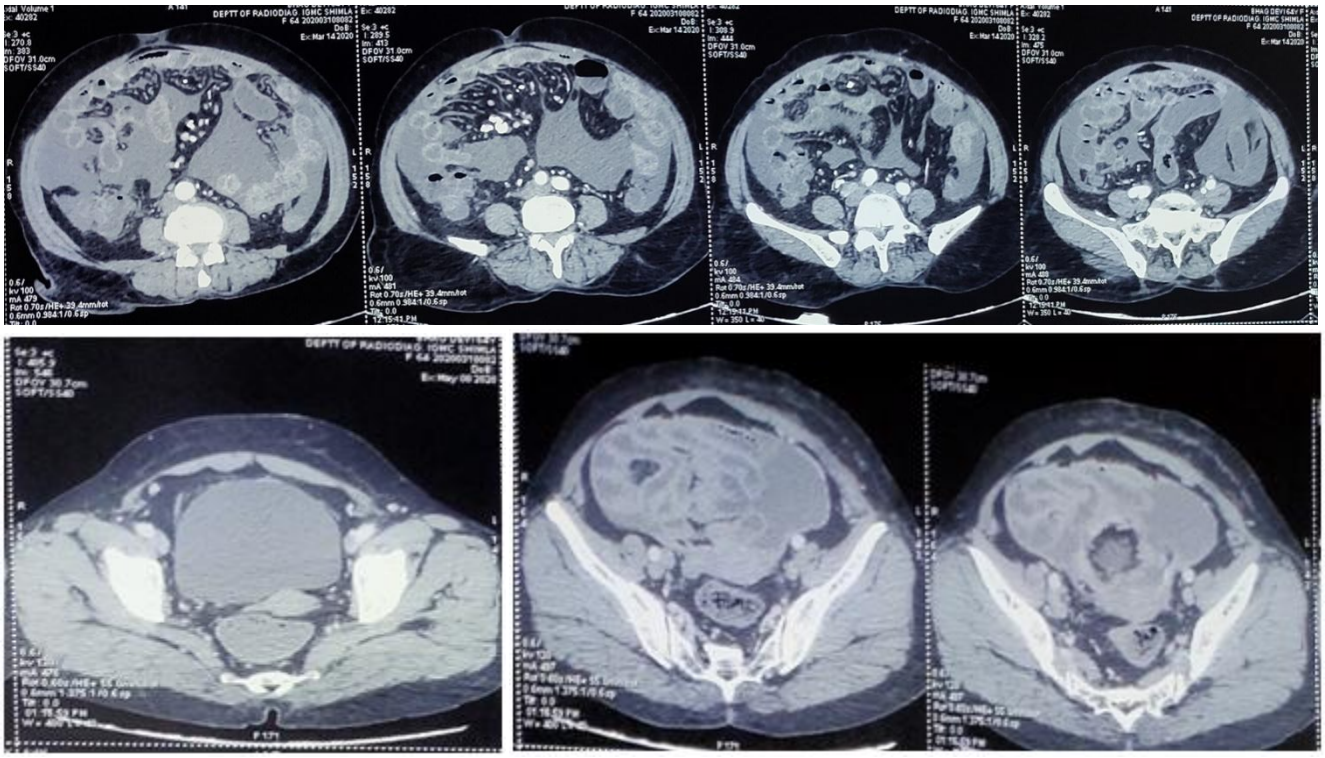

Fig 1 Ct abdomen and pelvis showing gross ascitis and omental thickening in the abdomen and post menopausal uterus with no enhancements and grossly normal bilateral adnexa.

Fig 2 Gross picture of the total hysterectomy specimen with solid, firm grey white areas in the omentum and grossly normal uterus and bilateral adnexa.

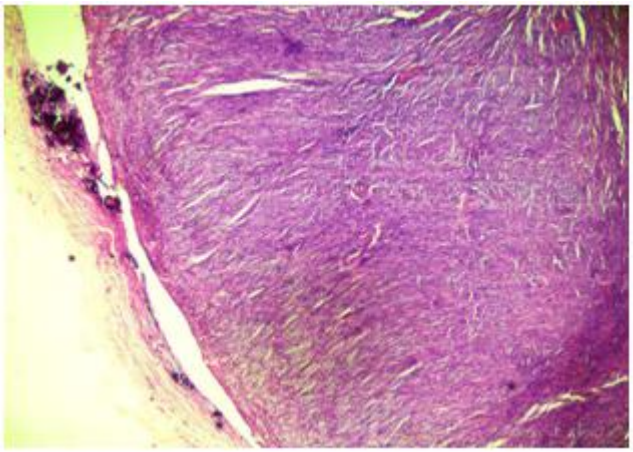

Fig-3A Photomicrograph of Right side ovary showing psammoma bodies involving the ovarian surface and no epithelial component H&E X 40

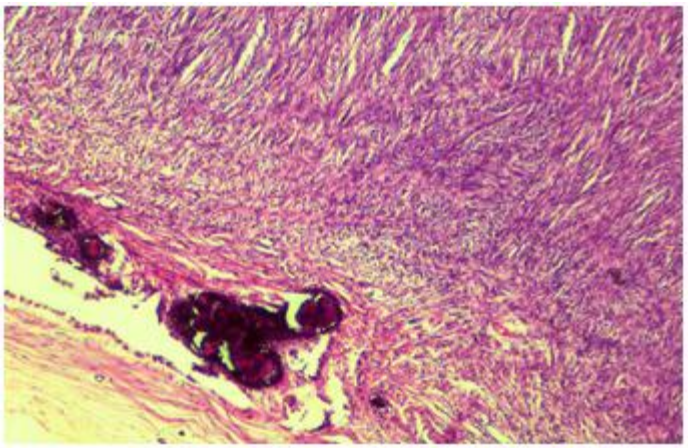

Fig-3B Photomicrograph of left side ovary showing psammoma bodies involving the ovarian surface and no epithelial component H&E 40X

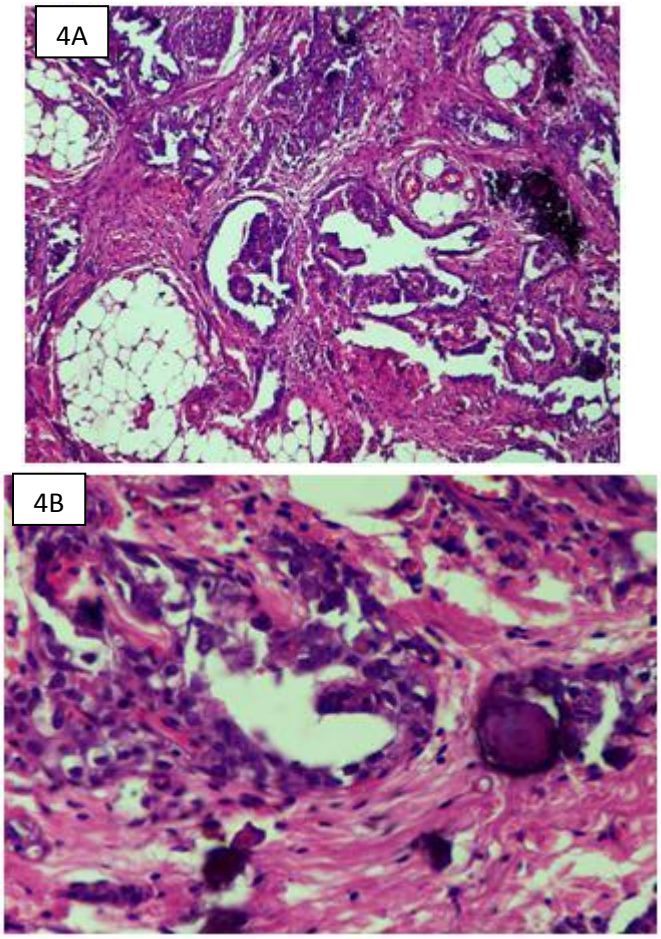

Fig-4 Photomicrograph of omentum shows sheets, papillae of tumor epithelial cells showing pleomorphism, high N/C ratio, prominent nucleoli, increased mitotic activity, moderate to abundant vacuolated cytoplasm, abundant psammoma bodies H& E 100X [FIG 4-A &B].

Discussion

The invasion of the serous membrane lining the abdominal cavity, viscera, and coelom by malignant cells is called peritoneal surface malignancy or peritoneal cancer (PC). It is divided into primary and secondary types. The de novo origin of cancer in the mesothelium of the abdomen causes primary mesothelioma. In contrast, the dissemination of tumor cells in the peritoneal cavity from other sites results in secondary peritoneal cancer[6].

Primary peritoneal serous carcinoma (PPSC) also known as serous surface papillary carcinoma, primary peritoneal carcinoma, extra-ovarian serous carcinoma, that diffusely involves the peritoneum, indistinguishable from primary serous

ovarian carcinoma. It is a rare primary malignancy of abdominal cavit[5]. The age specific peak incidence of peritoneal cancer is 75-79 years, similar to that ovarian cancer but older compared to fallopian tube cancer (70-74 years)[3].

Primary peritoneal serous carcinoma (PPSC) affects predominantly elderly and postmenopausal women. Although early stages of the disease may be asymptomatic, most patients in advanced stages complain of abdominal distension, abdominal lump, diffuse nonspecific abdominal pain, vomiting, weight gain and dyspnoea secondary to massive ascites. Elevated Ca-125 levels and scan findings consistent with ascites, omental involvement, and parietal peritoneal nodules without ovarian pathology may indicate PPSC. Given that PPSC shares the same clinical presentation with primary ovarian serous carcinoma and the two entities are indistinguishable immunohistochemically, the Gynaecology Oncology Group has set specific criteria in order to set the diagnosis of PPSC. Apart from ovarian serous carcinoma, PPSC is crucial to be distinguished from other primary peritoneal cancer subtypes, such as malignant mesothelioma, pseudomyxoma peritonei and clear cell carcinoma of peritoneum, as well as from secondary peritoneal carcinomatosis or inflammatory peritoneal diseases such as actinomycosis[7].

The PPSC is significantly difficult to diagnose as the imaging modalities such as ultrasound, CT scan and MRI could not visualize the large omental malignant infiltration. Often at the early-stage serous adenocarcinoma in 50 % cases, may not significant raise the tumor marker CA 125 which can fail to diagnose these cases at early stage thus jeopardizing prognosis. Omental infiltrate imaging in ascitic abdomen is a radiological challenge. As the omentum is completely floating in the ascites fluid it imbibe someascitic fluid leading to no significant attenuation change/ signal change in comparison to ascitic fluid especially in cases where there is early cellular infiltration. Delayed post contrast enhancement / diffusion imaging are helpful if pre study suspicion is there.[8]

Serous carcinoma of the ovary, fallopian tube, and peritoneum are almost similar on histopathology. Microscopically, the architecture could vary from glandular to complex papillary to solid pattern, with the tumor cells infiltrating or replacing the surrounding normal tissues.

The diagnostic criteria of this entity to differentiate it from primary serous carcinoma of ovary has been defined by the Gynaecology Oncology Group includes: - (a) Ovaries must be normal in size or enlarged as a result of benign process. (b) Extraovarian involvement must be greater than the surface involvement of either ovary (c) ovarian involvement must be absent, confined to the ovarian surface epithelium without stromal invasion, or involve the cortical stroma with a maximal tumour dimension of less than 5x5 mm. (d)The tumor in the extra-ovarian site should be serosal both histologically and cytologically[4].

In our case all the four GOG criteria were fulfilled .Although the PPSA is staged and treated in a similar to epithelial ovarian tumors ,we must differentiate them because these tumors present with an advanced stage and have a shorter survival when compared to their ovarian counterpart.

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