IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 14, Issue 9 Ver. VI (Sep. 2015), PP 06-08
www.iosrjournals.org

Primary Peritoneal Serous Carcinoma: A rare entity masquerading as ovarian carcinoma

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Abstract:*Primary papillary serous carcinoma of peritoneum (PPSCP) is a rare type of primary peritoneal adenocarcinoma which is clinically and histologically similar to primary ovarian papillary carcinoma which displays multicentric peritoneal and omentalinvolvement.Diagnostic criteria to distinguish it from primary ovarian cancer include normal sized ovaries, extraovarian site involvement greater than surface involvement of the ovary, the ovarian component less than 5×5 mm within the ovary and otherwise confined to the surface of the ovary, and the histological characteristics must be predominantly of the serous type. The treatment of a PPSCP is essentially based on cytoreductive surgery and platinum based chemotherapy. We encountered this rare case of PPSCP in a 55 year postmenopausal female who presented with abdominal distension and pain. Her CA-125 was markedly elevated and CECT showed bilateral ovarian masses and peritoneal implants. She underwent cytoreductive surgery followed by chemotherapy.*

Keywords:*Primary peritoneal Serous Carcinoma,Ovarian carcinoma,CA-125,Platinum based chemotherapy*

I. Introduction:

Primary papillary serous carcinoma of peritoneum (PPSCP) is a rare type of primary peritoneal carcinoma which is clinically and histopathologicallysimilar to primary ovarian papillary carcinoma. Originally classified as "carcinoma of unknown primary", several terms have been used for this disease, including serous surface peritoneal papillary carcinoma, multiple foci extra ovarian serous carcinoma and normal-size ovarian carcinoma syndrome.[1] Though uncommon; this tumor may be encountered by Gynecological oncologists and hence is being reported owing to its rarity.

II. Case summary:

A 55 years old multiparous postmenopausal female presented to us with the complaints of increasing abdominal distension, pain abdomen and loss of appetite since 1 month. Her general physical examination was normal. On abdominal examination, distension was present, more in lower abdomen. A vague cystic mass was felt in lower abdomen with ill-defined margins along with presence of fluid thrill. On bimanual pelvic examination, cervix was flushed with vagina. No mass was felt through any fornix. On rectal examination, fullness was felt in pouch of Douglas anduterosacral ligaments were nodular. Her routine investigations were found to be normal. CA 125 was more than 600U/ml; CEA and CA-19.9 being normal. On transvaginalsonography,uterus was normal with endometrial thickness of 4.8mm. There was presence of 8.2x3.5 cm homogenous mass in right ovary and 4.4x3.1 cms lesion in left ovary with presence of free fluid in pouch of Douglas (POD). On CECT (figure 1), heterogenous solid cystic masses were seen with infiltrating lobulated margins in both adnexa (7.4x6.2x4 cm on right side and 5.9x3.5x4.2cm on left) suggestive of bilateral ovarian tumors abutting the parametrium. Uterus was normal. Moderate amount of free fluid was present. Peritoneal implants of 5.8x4.4 cm and 3x3 cm in right lower abdomen were also reported. There was no evidence of retroperitoneal and pelvic lymphadenopathy and perirectal fat planes were normal. Upper GI endoscopy and colonoscopy were found normal. With these findings, the patient underwent staging laparotomy with total abdominal hysterectomy with bilateral salpingo-oophorectomy with debulking with infracolicomentectomy. Intra-operatively, 2200 mlascitic fluid was present which was sent for cytology. Parietal peritoneum was thickened. There was presence of bilateral solid-cystic ovarian tumor;3x3 cm on left side and 5x4 cm on right side(figure 2). Tumor was adherent to bowel on right side. A7X7 cm irregular, friable and extremely vascular tumor was present in POD which was removed. Omental nodular masses were seen with caking.Pelvic and Para-aortic nodes were not involved. The patient had an uneventful post-operative period. The peritoneal fluid cytology did not show any abnormal or malignant cells. On histopathological examination, 7x7x2 cm tumor tissue in POD and 10x9x4 cmomental mass revealed stage III moderately differentiated primary serous

peritoneal carcinoma (figure 3). Both ovaries and tubes were free of tumor. The patient then underwent chemotherapy.

III. Discussion:

PPSCP is defined as primary tumor of the peritoneum that diffusely involves the peritoneal surface but spares or only superficially involves the ovaries.[2]Primary peritoneal carcinoma was first described by Swerdlow in 1959 as a pelvic peritoneum mesothelioma.[3]Patients usually have advanced disease (stage III or IV) at the time of presentation. The mean age at the time of clinical presentation is 64 years of age. Abdominal pain and distension with ascites are the most common complaints and so the symptoms also mimic those of patients with ovarian carcinoma with peritoneal metastasis. However, PPSCP may also occur in women whose ovaries are normal or even previously excised.

As described by Bloss et al, the Gynecologic Oncology Group (GOG) developed the diagnostic criteria for primary peritoneal carcinoma [4]:

- Both ovaries physiologically normal in size or enlarged by benign process.
- Ovarian involvement is absent or limited to the surface and/or superficial cortex with no tumor nodule within ovarian cortex exceeding 5x5 mm.
- Volume of extra ovarian disease significantly exceeds that of ovarian disease.
- Histologic characteristics are primarily serous type, similar to ovarian serous papillary adenocarcinoma.

At present, the CA-125 antigen is considered the most effective tumor marker for PPSCP. HER-2/neu, p53, Wilm's tumor suppressor protein (WT1), estrogen and progesterone receptor have also been found to be involved in the tumorigenesis of PPSCP [5,6]. CT scan findings that suggest primary papillary serous carcinoma of the peritoneum include ascites, omental caking, diffuse enhancement with nodular thickening of the parietal peritoneum of the pelvis, normal-sized ovaries, with or without a fine enhancing surface nodularity of the ovary.[7]

The treatment of a PPSCP is essentially based on cytoreductive surgery and chemotherapy. Optimal cytoreduction is the primary goal of the surgery with excision of all visible implants. In a series reported by Fromm et al., the rate of successful debulking surgery was only 41% [8]. First line chemotherapy is a platinum-based combination with a taxane. The median survival in these patients is usually 12-18 months. However, the use of platinum-based chemotherapeutic regimens improves long term patient survival. Roh et al reported that the use of platinum-based chemotherapy increases the survival; the estimated 3-year survival rate being 36% [9]. Choi et al [10] reported that patients with PPSCP have higher levels of CA-125, more omental involvement, and less effect on response to chemotherapy than that with epithelial ovarian cancer.

IV. Conclusion:

Though clinically and histologically is similar to ovarian carcinoma, Primary papillary serous carcinoma of the peritoneum is a distinct clinicopathologic entity. Hence, whenever a patient presents as advanced ovarian tumor, the diagnosis of PPSCP should be kept in mind as a differential diagnosis.

References:

[1]. Álvarez JV, Gómez MM, Prats MDG, Agorreta MA, Cajal JMR, López JIL, Goyanes JPR. Extraovarian primary peritoneal carcinoma. A case report. Rev EspPatol 2007; 40: 47-52.

[2]. Morita H, Aokil J, Taketomi A, Sato N, Endo K. Serous surface papillary carcinoma of the peritoneum: clinical, radiologic and pathologic findings in 11 patients. Am J Roentgenol 2004;183:923–8.

[3]. Swerdlow M. Mesothelioma of the pelvic peritoneum resembling papillary cystadenocarcinoma of the ovary: Case report. Am J ObstetGynecol 1959; 77: 197-200.

[4]. Bloss JD, Liao SY, Buller RE, et al. extraovarian peritoneal serous papillary carcinoma: a case-control retrospective comparison to papillary adenocarcinoma of ovary. GynecolOncol 1993;50:347–51.

[5]. vonRiedenauer WB, Janjua SA, Kwon DS,Zhang Z and Velanovich V. Immunohistochemicalidentification of primary peritoneal serouscystadenocarcinoma mimicking advancedcolorectal carcinoma: a case report. J MedCase Reports 2007; 1: 150.

[6]. Cross SN, Cocco E, Bellone S, AnagnostouVK,Brower SL, Richter CE, Siegel ER, Schwartz PE,Rutherford TJ and Santin AD. Differential sensitivityto platinum-based chemotherapy in primaryuterine serous papillary carcinoma celllines with high vs low HER-2/neu expression invitro. Am J ObstetGynecol 2010; 203: 161-168.

[7]. Park JY, Kim KW, Kwon HJ, Park MS, Kwon GY, Jun SY, et al. Peritoneal mesotheliomas: clinicopathologic features, CT findings, and differential diagnosis. AJR Am J Roentgenol. 2008 Sep. 191(3):814-25.

[8]. Fromm GL, Gershenson DM, Silva EG. Papillary serouscarcinoma of the peritoneum. Obstet Gynecol. 1990;75:89-95.

[9]. Roh SY, Hong SH, Ko YH, Kim TH, Lee MA, Shim BY, et al. Clinical characteristics of primary peritoneal carcinoma. Cancer Res Treat. 2007;39:65–8.

[10]. Choi CH, Kim TJ, Kim WY, Ahn GH, Lee JW, Kim BG, Lee JH, Bae DS. Papillary serouscarcinoma in ovaries of normal size: a clinicopathologic study of 20 cases and comparison withextraovarian peritoneal papillary serous carcinoma. GynecolOncol. 2007; 105:762–8.

Legends to the Figures:

Figure 1: CECT abdomen showing heterogenous solid cystic masses in both adnexa. Peritoneal implants of 5.8x4.4 cm and 3x3 cm in right lower abdomen.

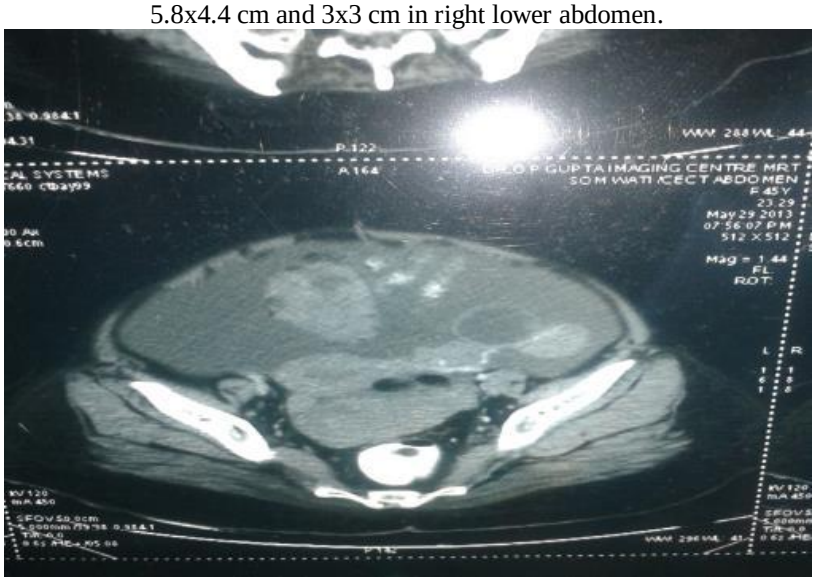

Figure 2: Bilateral solid-cystic ovarian tumor; 3x3 cm on left side and 5x4 cm on right side

Figure 3: Histopathology that revealed moderately differentiated Primary serous peritoneal carcinoma