Primary Peritoneal Papillary Serous Carcinoma

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Abstract: Primary peritoneal serous carcinoma (PPSC) is a rare entity that diffusely involves the peritoneum seen predominantly in elderly postmenopausal women which is often gets diagnosis delayed. Exclusion of serous carcinoma arising from the ovary and fimbrial end of fallopian tube is required to diagnose the above entity. Recent studies show an increased incidence of primary peritoneal serous carcinoma as it is better recognized. The inability to detect large omental malignant infiltrate by imaging modalities such as ultra sound, CT scan and MRI due to presence of ascitic fluid which has similar signal quality. The case is presented to highlight the need of high degree of clinical assessment not to solely depend on available imaging modalities which missed to report.

Key words: Primary peritoneal papillary serous carcinoma, malignant ascites, epithelial ovarian cancer.

I. Introduction

Primary peritoneal serous carcinoma (PPSC) also known as serous surface papillary carcinoma, primary peritoneal carcinoma, extra-ovarian serous carcinoma, primary serous papillary carcinoma, psammomacarcinoma, that diffusely involves the peritoneum, indistinguishable from primary serous ovarian carcinoma. It is a rare primary malignancy of abdominal cavity. [1] Primary peritoneal carcinoma was first described in 1959 by Swerdlow as Mesothelioa of the pelvic peritoneum .[2] Clinically and histopathologically, PPSC is similar to serous ovarian papillary carcinoma, and most centers apply the FIGO staging criteria for epithelial ovarian cancer to determine the stage of PPSC. [3] The origin of PPSC has not been well characterized. The epithelial layer of the ovary and the peritoneum shares a common embryonic heritage, deriving from coelomic epithelium early in life. PPSC appears to be a part of the hereditary breast-ovarian cancer syndrome as the frequency of BRCA mutations in peritoneal and ovarian cancer cases is comparable .[4] Patients with gerin line mutation of BRCA1 develop multifocal ori ng.

We present a case of PPSC which had diagnostic dilemma.

Case report:

A 53 years old lady with postmenopausal status for 8 years with co morbidity of essential hypertension and type diabetes mellitus for 2 years on anti hypertensive and OHA medication had reported to ex service clinic with the complaints of irregular pain in both flanks. The pain was insidious in onset for last 1 year which gradually increased in intensity and frequency as well as duration. The discomfort and pain was more while lying in supine and in right lateral side with slight relief of the pain while in left lateral position. She also observed gradual increase in size of the abdomen for similar duration. Due to enlargement of abdomen and pain she used to experience frequent episodes of breathlessness. She had experienced gradual loss of appetite and constipation, bowel habit got disturbed with rectal clearance only once in 3 to 4 days with passage of small hard dark round stool. A feeling of fullness and discomfort at the epigastric region was a constant irritating symptom. There was no pallor, pedal edema, lymphadenopathy , clubbing or koilonychea, palmar erythema and spider naive . Vital parameters were – no pallor, pulse 88/ min regular, BP 130/94 mm of Hg, no raised JVP. Abdomen was enlarged with huge ascites, with percussion dullness and fluid thrill. There was no periubmilical engorgement of vessels. The perineum, vulva, vagina and cervix was normal. Pap smear report was normal . Imaging study of the abdomen consisting of high resolution ultra sound, CT and MRI detected massive ascies and right flural effusion . The ovariess were atrophic, uterus normal size with a central cervical fibroid. The gut, kidneys, liver and biliary system as were essentially normal. No mass was observed in the abdominal cavity. The imaging
Fig. 1 Ultra sound abdomen with ascites no mass was observed

Fig. 2 CT images no abdominal/pelvic mass observed

Fig. 3 MRI scan images normal ovaries with ascites and no intra peritoneal mass.
She was initially admitted and treated as a case of ascites investigation by medical specialist due to negative imaging reports. Ascitic fluid aspirate cytology was positive for malignant cell. CA 125 was raised up to 671 units. CEA was within normal limit. She was prepared for exploratory laparotomy with suspicion of ovarian malignancy. Standard staging laparotomy was performed, ascites was drained and collected and peritoneal cavity was inspected systematically. A large omental cake measuring approximately 25 x 10 x 7 cm lying transversely was observed in the epigastric region. The whole peritoneal surface was studded with small (5 mm x 3 mm x 4 mm) papillary growth, the uterine surface, and pelvic peritoneum were also affected. The hepatic, splenic and gall bladder surfaces were free of the diseases. Standard surgical staging, TAH BSO, complete omentectomy was performed, and sample biopsies were obtained. The ovaries were atrophic and normal in size no gross lesion except a small 5 mmx3 mm lesion on the left ovary. The retro peritoneal and iliac nodes were not palpable.

Post operatively she had experienced excessive accumulation of malignant ascites, and wound sepsis which had healed after secondary suturing. The histopathological study of the specimen was consistent with high grade serous adenocarcinoma with superficial involvement of the ovarian surface. Histopathology report: High grade serous adenocarcinoma, TNM stage v p T3c, FIGO Stage III C.
Subsequently chemotherapy was started with paclitaxol and cisplatin regime, she has tolerated first pulse of chemotherapy with significant improvement of feeling of general wellbeing, improve appetite and cessation of ascites formation.

II. Discussion

Primary peritoneal serous carcinoma a rare malignant neoplasm is considered as a distinct entity commonly affecting elderly postmenopausal women. The diagnosis usually made after exclusion of other pelvic epithelial neoplasm in the abdominal cavity. In 1972 Lauchlan [6] first reported secondary Mullerian system of female peritoneum consist of pelvic and lower abdominal peritoneum.

The Mullerian potential of this layer arises due to the invagination of the coelomic epithelium, which may undergo malignant transformation independent of ovaries, and a male can also suffer from similar malignancy. The coelomic epithelial lining of the ovary, fimbrial end of the fallopian tube, and the pelvic peritoneum has a common embryonic origin. PPSC are about a tenth as common as their ovarian counterparts. With increasing recognition of the entity has also contributed to an increasing diagnostic frequency approaching 18% of laparotomies performed for ovarian carcinoma. [7]

Primary peritoneal serous carcinoma akin to advanced epithelial ovarian carcinoma extensively involves the peritoneal surface, which either spares ovaries or superficially invades them in the absence of an obvious primary site and grossly normal ovaries.

Both PPSC and primary ovarian carcinoma stain positive for ER, CK7, Wilms tumor suppressor gene-1, and CA-125 and negative for CK20, PR thus help in exclusion of the peritoneal metastasis and mesothelioma. Due to early and extensive involvement intra peritoneal structures treatment are similar to high-stage (stage III or IV) serous ovarian carcinomas. However, worse prognosis in PPSC with median survival between 7 and 27.8 months, with a low 5 years survival rate ranging from 0% to 26.5% are reported. [8]

The PPSC is significantly difficult to diagnose as was in this case 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 rise of tumor marker CA 125 which can fail to diagnose these cases at early stage thus jeopardizing prognosis. Omental infiltrate imaging is ascetic abdomen is a radiological challenge As the omentum is completely floating in the ascites fluid it imbibe some
ascitic 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.

The diagnostic criteria of this entity to differentiate it from primary serous carcinoma of ovary has been defined by the Gynecology 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.

[9]

III. Conclusion

A high degree of clinical acumen and suspicion could diagnose the condition and successfully early surgical and chemotherapy treatment could be initiated with improvement of the general well being although the prognosis is not considered favorable due to the aggressive nature of the disease.

References