Case Report

Primary peritoneal serous carcinoma: A diagnostic dilemma of pelvic epithelial neoplasms

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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. 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

Key words: Ovary, peritoneum, primary peritoneal serous carcinoma

INTRODUCTION

Pelvic epithelial carcinomas are high-grade lesions as they were discovered only after extensive spread into the peritoneal surfaces. They arise from the female genital tract that includes serous carcinoma from the endometrium, fallopian tube, ovary, and peritoneum.[1] Primary peritoneal serous carcinoma (PPSC) is a rare malignant epithelial tumor that is indistinguishable histologically from serous carcinoma of the ovary and also tubal carcinoma arising from fimbrial end of the fallopian tube. The origin of this entity is from embryonic nests of mullerian cells from the peritoneum.

CASE REPORT

A 55-year-old postmenopausal female presented with lower abdominal pain since a year and abdominal distension since a week. On per abdominal examination, the abdomen was distended and ultrasonography revealed moderate ascites with absent organomegaly. Upper and lower gastrointestinal endoscopy were normal.

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Computed tomography abdomen showed multiple peritoneal deposits with ovarian involvement. Hence, a preoperative diagnosis of metastatic deposits to the pelvic peritoneum, probably from ovarian carcinoma was made. Preoperative levels of serum cancer antigen-125 (CA-125) were found to be elevated to 450 U/mL (reference range 0–30 U/mL).

Exploratory laparotomy revealed moderate ascites with extensive peritoneal deposits and diffuse thickening of lower abdominal peritoneum. Patient underwent hysterectomy, bilateral salpingo-oophorectomy along with pelvic omentectomy and a specimen was sent for histopathological examination.

On gross examination, a specimen of uterocervix showed unremarkable changes. Both the ovaries with fallopian tubes were examined extensively. On examination, right and left ovaries were normal in size measuring 2.5 cm × 2 cm × 1 cm and 2 cm × 1.5 cm × 1 cm respectively. Lower pelvic peritoneum attached to ovaries was measuring 19 cm × 9 cm × 4 cm. It showed diffuse thickening with irregular surface studded with multiple nodules and on serial sectioning it showed grey-white to grey-yellow uniformly solid, firm to hard peritoneal flap of variable thickness [Figure 1]. Lymph nodes were not identified.

Microscopy of pelvic peritoneum showed an invasive tumor arranged in solid, complex irregular tubulo-glandular structures and forming occasional

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papillae at places surrounded by extensive desmoplastic stroma with intervening adipocytes. Numerous glandular structures showed comedo-type of necrosis [Figure 2]. The solid and papillary structures were lined by multilayered cells with atypical nuclei with high N/C ratio and coarse vesicular chromatin and moderate eosinophilic cytoplasm. Furthermore, seen are few bizarre cells with irregular hyperchromatic nuclei and atypical mitosis (6–8/10 hpf). Numerous psammoma bodies were also seen [Figure 3].

Right ovary showed tumor deposits within the cortex measuring <5 mm × 5 mm. The surface epithelium of left ovary and also the surface of both fallopian tubes also showed tumor deposits. Both the fallopian tubes and the uterus showed no significant pathology.

On immunohistochemistry (IHC), the tumor cells showed diffuse nuclear positivity for estrogen receptor (ER) [Figure 4a] and both cytoplasmic with membranous positivity for cytokeratin 7 (CK7) [Figure 4b] with negative status for progesterone receptor (PR) and CK20.

A final diagnosis of "PPSC" of the pelvic peritoneum with surface involvement of both tubes and ovaries were considered based on histopathological and IHC examination.

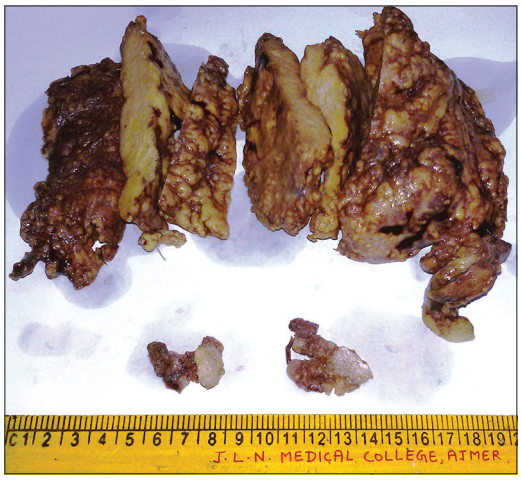

Figure 1: Gross picture showing diffuse thickened peritoneal flap with multiple tiny nodules studded over the surface

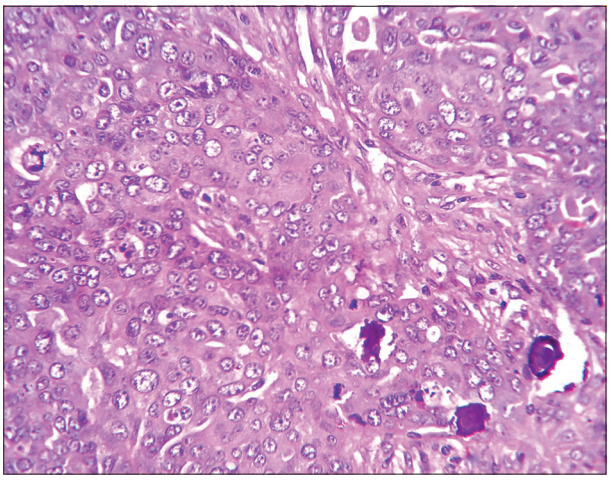

Figure 3: Photomicrograph showing atypical nuclei with high N/C ratio and coarse vesicular chromatin, atypical mitosis, and numerous psammoma bodies are seen (H and E, ×40)

DISCUSSION

In 1959, Swerdlow[2] described primary peritoneal carcinoma as "mesothelioma of the pelvic peritoneum" and in 1972 Lauchlan[3] defined secondary mullerian system of female peritoneum which includes pelvic and lower abdominal mesothelium. The müllerian potential of this layer arises by the invagination of the coelomic epithelium, and it may undergo malignant transformation independently. The coelomic epithelial lining of the ovary, fimbrial end of the fallopian tube, and also the pelvic peritoneum all share a common embryonic origin.

A multifocal origin of PPSC was mentioned by Muto *et al*.[4] whereas others identified the unifocal origin. Hereditary

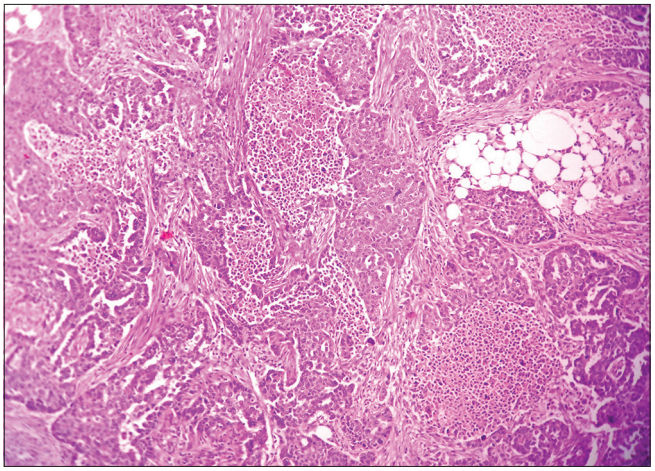

Figure 2: Photomicrograph of peritoneum showing irregular glands with multilayered tumor cells along with comedo type necrosis (H and E, ×10)

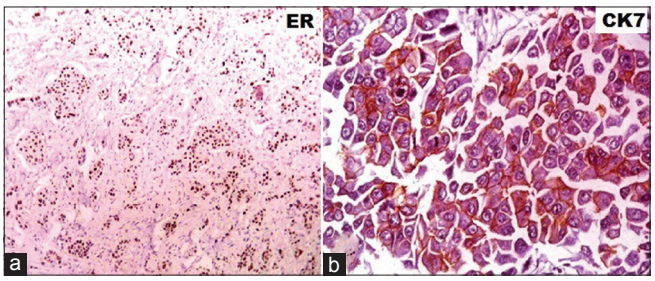

Figure 4: (a) Photomicrograph showing diffuse nuclear positivity of the tumor cells for estrogen receptor (IHC, ×10). (b) Cytoplasmic with membranous positivity for cytokeratin 7 (IHC, ×40)

predisposition may play a role in primary peritoneal carcinoma with the *BRCA1* mutation comparable with that seen in ovarian carcinoma. Recently, fimbrial end of the tube has been identified as the site of origin for early serous carcinoma in *BRCA* positive women suggesting the plausible origin of many pelvic serous carcinoma.[1]

Most cases of PPSC were reported in elderly postmenopausal women, with a median age range of 57–66 years and observed lifetime risk of 1 case/500 women.[5] PPSC are about a tenth as common as their ovarian counterparts. Very rarely few cases were also reported in males. Better recognition of this entity has contributed to an increasing diagnostic frequency approaching 18% of laparotomies performed for ovarian carcinoma.[6]

Most PPSC s are high-grade, invasive with rapid progression and extensive extra-ovarian spread at the time of diagnosis in the absence of a known precursor lesion. These were traditionally considered as primary ovarian carcinomas, with a few designated as fallopian tube carcinoma, but recently primary peritoneal carcinoma is recognized as a distinct entity with little or no ovarian and fallopian tube disease.

Preoperative diagnosis of this entity (PPSC) is often not made, as it cannot be morphologically differentiated from malignant mesothelioma of peritoneum, metastatic peritoneal carcinomatosis, and peritoneal psammocarcinoma. Diagnosis is possible only after laparotomy and extensive histopathological assessment.

Peritoneal malignant mesothelioma has a close relation to long-term asbestos exposure; affecting predominantly males.[7] Histologically this shows absence of high-grade nuclear features with occasional mitosis and psammoma bodies to compared to PPSC. ER is positive in serous carcinomas thus differentiates mesothelioma which is negative for this receptor.[8]

Peritoneal serous psammocarcinoma has more numerous psammoma bodies >80% of epithelial nests, less aggressive cytology, absent or moderate nuclear atypia, and rare mitosis, compared to PPSC.[9] Less frequently metastatic peritoneal carcinomatosis occurs from the breast, gastrointestinal tract, lungs, and thyroid gland.

Primary peritoneal serous carcinoma is very similar to advanced epithelial ovarian carcinoma that 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. Therefore, these are also termed as normal sized ovary carcinoma syndrome.

Immunohistochemistry used in diagnosing 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 excluding peritoneal metastasis and mesothelioma.

The staging, prognosis, and treatment are very similar to high-stage (stage III or IV) serous ovarian carcinomas. however, few studies suggest significantly worse prognosis in PPSC with median survival between 7 and 27.8 months, while 5 years survival rate from 0% to 26.5%.[10]

The treatment of choice includes debulking surgery with hysterectomy, salpingo-oophorectomy, omentectomy, followed by platinum-based chemotherapy.

CONCLUSION

Primary peritoneal serous carcinoma is considered as a distinct entity seen commonly in elderly postmenopausal women, the diagnosis typically made after exclusion of other pelvic epithelial neoplasms. They are rare primary malignant epithelial neoplasm of peritoneum, histologically similar to serous carcinoma arising from distal end of the fallopian tube and ovaries validating thorough sampling of both the tubes and ovaries to reach the final diagnosis. It is also considered in the differential diagnosis of metastatic malignancies to the peritoneum. As definitive preoperative diagnosis is not possible, histopathological assessment with IHC studies becomes mandatory to reach the final diagnosis. PPSC have a more indolent disease course, a higher response rate to systemic therapy, and a chance for long-term, disease-free survival after therapy.

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