



Case Report:

Uterine Papillary Serous Carcinoma with Mature Cystic Teratoma of Left Ovary

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Abstract:

Uterine papillary serous carcinoma (UPSC) is an uncommon histologic variant of endometrial carcinoma that typically arises in post menopausal women, that may present with extrauterine spread, resulting in high relapse rate and poor prognosis. Mature cystic teratomas (MCT) are common tumors that occur during the reproductive years. We report a case of a 60 years old female with UPSC with MCT of left ovary. To our knowledge, this is the second report of UPSC combined with ovarian MCT.

Key Words: Uterine papillary serous carcinoma (UPSC); Mature cystic teratoma; CA 125

Case Report:

A 60 year old female came with history of distention of abdomen and bleeding per vagina since 1 year. Physical examination revealed abdominal distension with abdominal tenderness. X-ray chest showed right pleural effusion. Abdominal sonography revealed solid cystic left adnexal mass measuring 8.9x6.5cms, with moderate ascitis of approximately 700-800ml, with right lobe of liver showing a cystic lesion measuring 4x2cms, suspected to be metastatic. Her serum CA125 level was high >400IU/ml (normal 0-35IU/ml). A diagnostic ascitic fluid tap was performed and subjected for analysis. Ascitic fluid cytology revealed tumor cells in sheets and 3D clusters favoring a diagnosis of metastatic adenocarcinoma. (Figure 1)

A preoperative diagnosis of left ovarian carcinoma was made and a course of neoadjuvant chemotherapy was administered over 3 months comprising of 3 cycles of Carboplatin 600mg/cycle and subsequently an exploratory laparotomy with total abdominal hysterectomy and bilateral salpingo-oophorectomy, pelvic and aortic lymphnodes, peritoneal deposit, liver deposit sampling and omentectomy was performed. Grossly, uterus with cervix was measuring 8.5x5.5x4cms, cut surface endometrium was 5mm thick with a gray white and friable growth measuring 1.5x1.4cms in the isthmic region. (Figure 2)

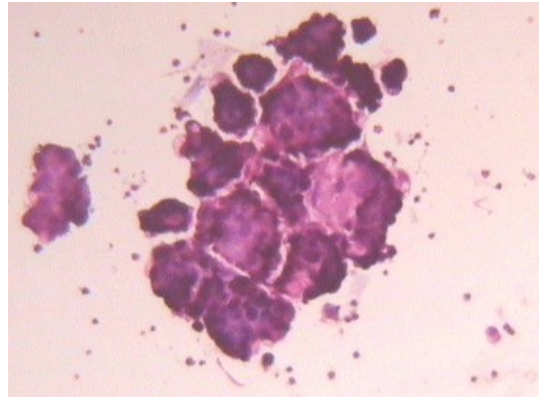


Figure 1: 40x high power view of ascitic fluid cytology shows tumor cells in 3D cluster, cells have pleomorphic and hyperchromatic nucleus with clumped chromatin and prominent nucleoli.



Figure 2: Gross photograph of uterus with cervix with a gray white tumor in the isthmic region.



Figure 3: Gross photograph of left ovary with solid cystic areas filled with cheesy material.

Right ovary was unremarkable. Left ovary was cystically enlarged and measured 7.5x5.5cms, cut surface was predominantly cystic filled with cheesy material mixed with mucinous material and hairs.(Figure 3)

Omentum on dissection revealed multiple gray white nodules with largest being 2.5cms in diameter.

On microscopy endometrium showed tumor cells arranged in papillary architecture with tufting and secondary papillae. Papillae had broad based and hyalinized stalks, lining tumor cells were stratified columnar, individual tumor cells showed nucleomegaly with prominent nucleoli, clumped chromatin and atypical mitotic figures, also seen were psammoma bodies, endometrium adjacent to the tumor was atrophic, tumor was infiltrating >50% myometrium with vascular invasion. (Figure 4)

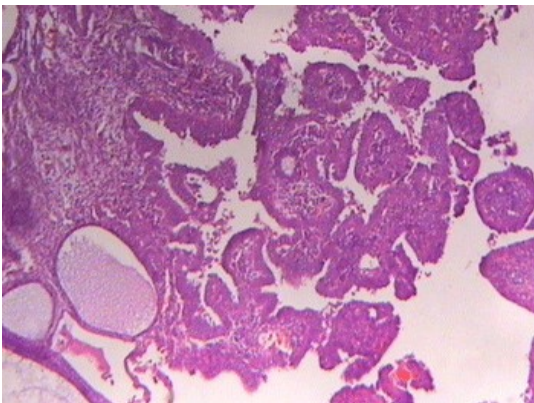


Figure 4: Uterine wall with papillary serous carcinoma in a background of atrophic endometrium

Peritoneal sample, pelvic and aortic lymphnodes and liver tissue showed metastatic deposits. Left ovary showed mature cystic teratoma (MCT).

Based on these findings a diagnosis of Uterine papillary serous carcinoma (UPSC) stage IVA according to international federation of gynecology and obstetric cancer staging system (FIGO), with left ovarian MCT was made. Postoperative stay was uneventful. Patient was advised to undergo 3 cycles of chemotherapy.

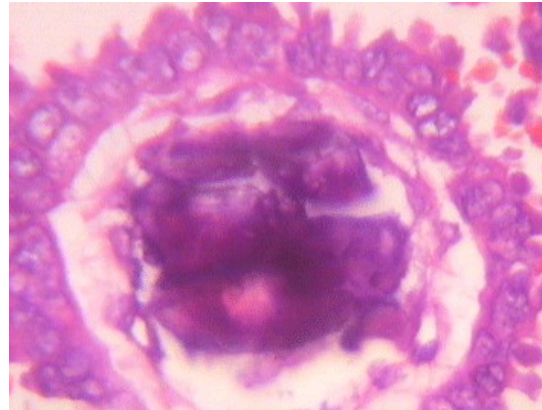


Figure 5: Papillae lined by tumor cells with psammoma body in the stalk of the papillae

Discussion:

Endometrial cancer is one of the common gynecologic malignancy and accounts for 13% of all cancers in women. The most common endometrial cancer is endometrioid carcinoma and accounts for 75-80% and is associated with endometrial hyperplasia. UPSC on the other hand is known to be a rare type of endometrial tumor that comprises only 5-10% of endometrial carcinoma which usually arises in an background of atrophic endometrium and psammoma bodies are found in one third of cases.^{3,5}

UPSC is a distinct type of endometrial carcinoma which resembles Papillary serous carcinoma of ovary, clinically, it has an aggressive biologic behavior with a propensity for lymphatic invasion, myometrial invasion and extrauterine spread, a tumor behavior similar to ovarian carcinoma, instead of typical endometrial carcinoma.^{2,6} Unlike the typical endometrial carcinoma, it has a poor clinical survival and even a small focus of UPSC can be life threatening.^{7,8}

UPSC, which was formerly known as tubal carcinoma, was shown to be more aggressive than conventional endometrial carcinoma and was established as a distinct entity by Lauchlan and Hendrickson et al in 1982.^{1,2} It has long been recognized that excessive estrogens and obesity are associated with endometrial cancers. Bokhman proposed a hypothesis that there were two distinct types of endometrial cancers. Type I is the commonest of the two and is associated with obesity with excess estrogen as a result of aromatase activity in peripheral adipose tissue. Type II tumors are thought to develop through a separate pathway of tumorigenesis and not associated with excess estrogen, clinically there are much more aggressive and often spread outside the uterus by the time of diagnosis.⁹ UPSC is the most common type of type II tumors, it is always diagnosed in postmenopausal women with 10 Yrs older than usual endometrioid carcinoma.^{7,9}

Based on aggressive nature associated with an advanced stage at initial presentation, Geisler et al. concluded that patients with UPSC should undergo a staging laparotomy including lymphadenectomy and omentectomy similar to the procedure undertaken for patients with ovarian carcinoma.¹⁰

In the presented case, although pre-operatively it was misdiagnosed as ovarian carcinoma due to left ovarian mass (MCT) and raised serum CA 125 levels (which is also raised in UPSC), a preoperative chemotherapy and staging laparotomy of TAH+BSO, pelvic lymphnode dissection and peritoneal cytology were performed as per the protocol for Ovarian carcinoma.¹¹

MCT make upto 25% of all ovarian neoplasms and is commonly found in reproductive age group. It is unilateral in 88%

of the cases. Tumors are usually uniloculated with average size of 7-8cms and can undergo malignant transformation in 1% of cases.¹² MCT of ovary is frequently associated with mucinous tumors ovary.¹³ On reviewing the literature we found only one case associated with UPSC.¹⁴

In conclusion, UPSC is very rarely associated with MCT and UPSC by itself is an uncommon type of endometrial carcinoma which is highly aggressive and usually would have disseminated by the time of clinical presentation, it is associated with high CA125, and the treatment protocol is as that of a ovarian carcinoma.

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