

Case Report

Primary Fallopian Tube Cancer : A Rare Gynecological Malignancy

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Abstract: Primary fallopian tube carcinoma is a rare gynecological cancer. Loco regionally advanced disease is clinically difficult to differentiate from similar stage ovarian and primary peritoneal carcinomas. Early stage fallopian tube tumors, despite having unique clinical presentation are provisionally diagnosed as an ovarian carcinoma due to rarity and symptom overlap with these more common gynecological cancers. We present a case of early fallopian tube cancer provisionally diagnosed as an ovarian carcinoma and review the current literature regarding etiology and treatment.

Keywords: Fallopian tube; Primary tumor; Carcinoma; Clinical presentation; Treatment

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Consent: Consent was taken from the patient for publication of this case report.

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Introduction

Primary fallopian tube cancer is a rare pelvic malignancy with less than 1500 cases reported in literature [1]. It accounts for less than 1% of all gynecological cancers and has a reported incidence of 0.41 per 100,000 women in the United states [2,3]. It is speculated that the true incidence may be higher as meticulous pathological evaluation of the fallopian tubes has long been ignored and has been standardized (SEE-FIM protocol) only recently after the adoption of prophylactic salphingoophrectomy for hereditary breast and ovarian cancers. Primary fallopian tube cancers share certain similarities with ovarian and peritoneal carcinomas and this has prompted FIGO to publish a unified staging system for these cancers. Understandably these distinct pathological entities are reported, evaluated and treated as a collective clinical entity.

Case Report

A 61-year-old lady presented with postmenopausal bleeding and watery discharge per vaginum. Clinical examination revealed a normal abdomen, adnexa and cervix. A CT scan (figure1) confirmed a left complex adnexal mass with no ascites or retroperitoneal nodal enlargement. A pap smear and an endometrial biopsy was unremarkable. Tumor marker evaluation showed an elevated CA- 125 level of 903 U/ml. A provisional diagnosis of early ovarian carcinoma made and a staging laparotomy was planned. At laparotomy a pleasant surprise as both ovaries were normal on gross examination, however a complex mass arising from left fallopian tube was diagnosed (figure 2), a full surgical staging was completed. Post operative histological examination confirmed both ovaries, endometrial cavity and cervix to be normal and the complex fallopian tube mass as a high grade serous primary fallopian tube cancer (figure 3).The greater omentum, pelvic nodes, peritoneal biopsies and cytology were negative for disease. The patient is currently progressing on a course of carboplatin /Paclitaxel chemotherapy uneventfully.

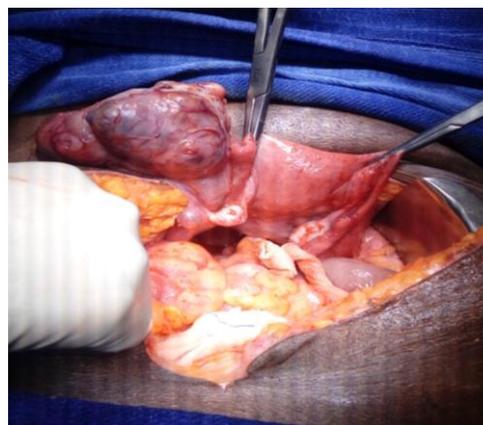
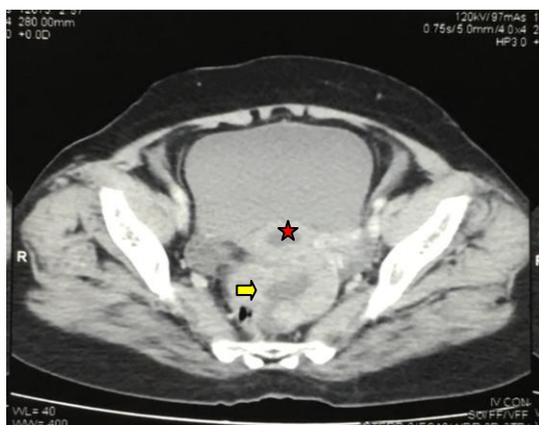


Fig. 1 (left) CT scan showing left complex adnexal mass (arrow) and endometrial collection (star)

Fig. 2 (right) Staging laparotomy – Both ovaries normal with Left fallopian tube complex mass

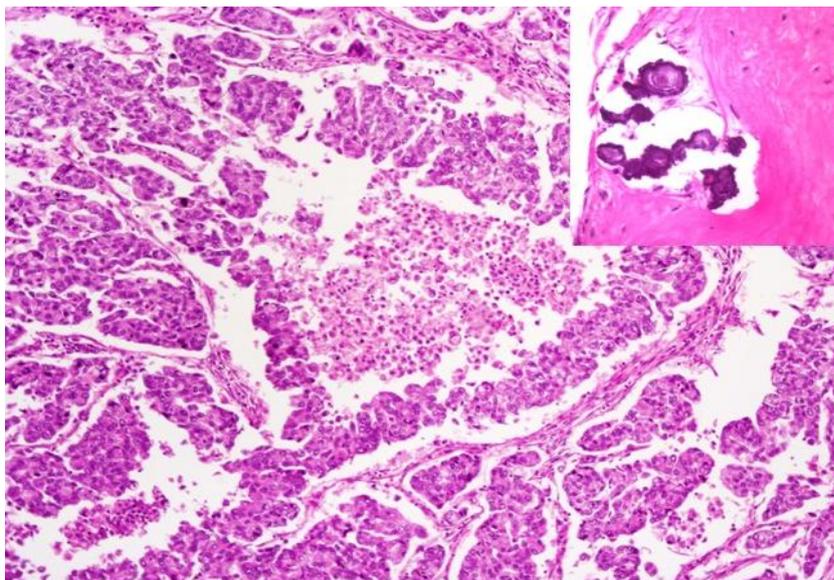


Fig. 3 Showing high grade serous tumor and psammoma bodies (inset)

Discussion

The exact etiopathogenesis remains elusive. Parity and hormonal factors have been evaluated as possible etiology with conflicting evidence, nevertheless the evidence for specific genetic mutations such as the BRCA-1 and 2 for conferring increased susceptibility is robust[4]. The role of serous tubal intraepithelial carcinoma (STIC) as precursor lesions of high grade serous primary fallopian tube is based on identical genetic signatures (P53 and Ki 67) exhibited by these lesions and by the observation of high prevalence of these lesions in prophylactic salpingectomy specimens from high risk women. Their (STIC) role in sporadic fallopian tube cancers is inconclusive as their clinical significance and diagnostic criteria continue to evolve[4].

High grade serous histology is the commonest followed by endometrioid type. Pattern of metastatic spread is identical to ovarian cancers with transcelomic spread (80%), direct invasion, lymphatic and hematological dissemination reported. A higher propensity for nodal involvement than ovarian cancer is explained by the abundant lymphatic network of the tubes that drain through the infundibulopelvic and round ligaments[5]. Lymphadenectomy as a part of staging yields a nodal involvement rate of 40-59 % with equal pelvic and retroperitoneal nodal positivity hence surgical staging including a full para aortic nodal dissection is recommended [6]. The extent of radial tumor spread, the site of origin within the fallopian tube and intra operative rupture has been reported as prognostically significant in early stage disease. Stage at diagnosis and extent of residual disease at surgery are independent prognostic factors. In 2014 FIGO has adopted the revised epithelial ovarian cancer staging to be adapted to primary fallopian tube cancers [5]. Immunohistochemistry expression pattern is variable with P53 and her2 neu commonly expressed but no diagnostic or prognostic correlation has been documented [7].

Primary fallopian tube carcinoma commonly presents in the fourth to sixth decade. The characteristic latzko's triad of colicky abdominal pain relieved by profuse watery vaginal and a pelvic mass is rather uncommon. The pathognomonic hydrops tubae profluens which describes a shrinking adnexal mass secondary to intermittent vaginal discharge is a rare presentation nevertheless the most common pattern is a vaginal discharge or post menopausal bleeding with a complex adnexal mass [4]. Locoregionally advanced

disease is clinically indistinguishable from similar stage primary ovarian carcinoma. The diagnostic criteria developed by Hu and later modified by Sedlis continues to be endorsed by FIGO, however a disease distribution based criterion lacks accuracy in advanced disease.

A preoperative diagnosis is an exception. Trans vaginal ultrasound is the initial investigation attempted and is nonspecific for anatomical origin of the lesion though characteristic features like cog and wheel mass with low impedance flow on Doppler has been described [8]. CT and MRI are reliable for delineating associated features like ascites, abdominal disease or metastasis [9]. The practical utility of CA 125 as tumor marker follows the principles applied in ovarian cancer such as an independent prognostic factor, in response assessment and during surveillance for detection of recurrence. Isolated reports of cytology being diagnostic has been reported however this modality has very low sensitivity (0-23 %) to recommend as a diagnostic or screening test [10].

The principles of treatment follow epithelial ovarian cancer and the same is endorsed by major guidelines. Surgery with adjuvant platinum based chemotherapy is considered standard of care. The sequencing of surgery and chemotherapy follow ovarian cancer treatment philosophy, as diagnosis, reporting and appropriate staging has long been ignored in literature hence current recommendations are extrapolated from ovarian cancer trials. The role of radiotherapy, second look laparotomy and hormonal therapy in the treatment scheme of primary and recurrent disease is unclear as good quality evidence is deficient [5,11].

Overall 5-year survival has been reported as 65%, however small sample size precludes reliable interpretation. Most data are retrospective single institution reports, a series of 101 patients treated with surgery and carboplatin / Taxol chemotherapy with a median follow up of 64 months was recently reported. Despite optimal cytoreduction rate of 90 % recurrence was noted in 43 % with a median progression free survival of 20 months. The five year overall survival and disease free survival were 67 % and 57 % respectively. Multivariate analysis revealed FIGO stage, pelvic lymphadenectomy and chemotherapy as predictive of overall and disease free survival [12]. This contrasts with a prior report that demonstrated grade, FIGO stage, optimal cytoreduction and CA -125 nadir as prognostic variables of overall and disease free survival. This study also revealed a significant benefit to paclitaxel based chemotherapy and to optimal cytoreduction [13]. Important differences in clinical behavior and survival have been reported between fallopian tube and ovarian cancer, however a systematic review noted no differences. The published data are often conflicting and the retrospective nature prevents reliable interpretation [14]. A well designed prospective study is required to clarify several important issues regarding behavior and therapy.

Conclusion

Primary fallopian tube cancer is a rare malignancy. A preoperative misdiagnosis is common due to rarity and symptom overlap with other common gynecological cancers. The natural history and pathogenesis is still unclear and in the absence of dedicated clinical trials management guidelines as applicable to ovarian cancers have been recommended. With the new FIGO 2014 staging and reporting revision document this scenario is likely to change in the future.

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