Undifferentiated Sarcoma of Fallopian Tube Managed with Robotic-Assisted Surgery

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About the Author

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Introduction

Primary fallopian tube malignancy is one of the rarest gynecological cancers, accounting for 0.3% of all malignancies of the female genital tract. Sarcomas arising from fallopian tube are even rarer. In the present case report, we report a case of undifferentiated fallopian tube sarcoma after obtaining permission from the institutional ethical committee.

Case Report

A 66-year-old postmenopausal lady presented with a 1-month history of left lower quadrant abdominal pain. On abdominopelvic examination, she was found to have a non-tender left adnexal mass with restricted mobility about

6 weeks in size. Ultrasound scan and magnetic resonance imaging (MRI) confirmed the presence of a well-defined $5 \times 5.5 \times 6.2$ cm solid mass in left adnexa, suggestive of left ovarian mass lesion. In MRI, the mass was T1 isointense, T2 hyperintense with diffuse STIR bright signal and diffuse restriction. Post-contrast dynamic LAVA sequence showed a progressively enhancing mural nodule blooming in gradient sequence. Right ovary, uterus, and rest of the abdomen and pelvis including nodal basins appeared normal. Tumor markers—serum LDH, CA125 (28), CA19-9, and CEA (0.98)—were within normal limits.

Provisional diagnosis of ovarian neoplasm was made, and patient was taken up for robotic-assisted staging surgery as for carcinoma ovary. Intraoperatively, an ill-defined 6x6 cm ruptured, necrotic mass was seen arising from the left fallopian tube infiltrating the sigmoid epiplocae. The tumor comprised of mainly solid components with large area of necrosis and hemorrhage (Fig. 1). Normal left ovary was seen lying by the side of mass (Fig. 1). Intraoperative frozen section of the mass was suggestive of spindle cells with necrosis with the possibility of sarcoma. The mass was removed completely, and entire surgery was completed robotically. Complete robotic-assisted staging surgery was performed with hysterectomy, bilateral salpingo-oophorectomy, resection of sigmoid epiplocae deposits and infracolic omentectomy.

Postoperative pathological examination grossly showed a tumor measuring $7 \times 6 \times 2$ cm. On microscopic examination, the neoplasm was characterized by the proliferation of atypical plump oval/spindle cells, with moderate cytoplasm and pleomorphic vesicular nuclei with irregular margins (Fig. 2a). Bizarre cells and sheets of cells with

scanty neoplasm were seen. The neoplasm showed extensive necrosis (Fig. 2b) and mitosis including atypical mitosis. Left ovary was seen separate and was normal. Sigmoid epiplocae showed similar necrotic tumor deposits. Immunohistochemically, the tumor cells showed diffuse positive reaction to smooth muscle actin (including the aberrant nuclear positivity) and focal CD10 positivity (Fig. 2c, d). Other markers CD117, cyclin D1, h-caldesmon, desmin, and EMA were negative ruling out leiomyosarcoma, GIST, endometrial stromal sarcoma or rarely a sarcomatoid mesothelioma. Expression of these 2 markers along with the negativity of other markers of specific differentiation and the absence of epithelial elements led to the diagnosis of undifferentiated sarcoma.

Except for the direct invasion into sigmoid epiplocae, there was neither direct invasion nor metastatic spread into the uterus, bilateral ovaries, omentum and retroperitoneum. According to the current surgical staging for tubal cancer, this tumor was staged as FIGO IIb. Metastatic workup with whole body PET CT showed no evidence of any residual intrapelvic primary malignancy and any abdomino pelvic/other lymph nodal and distant metastasis.

Case was discussed in multidisciplinary tumor board, decided for adjuvant treatment with chemotherapy. Patient completed the 4 courses of adjuvant chemotherapy with intravenous Adriamycin (50 mg/m^2 , day 1) and ifosfamide (1.5 g/m^2 , day 1–4) without any grade 3&4 complications. Currently, after 8 months of treatment completion she is on follow-up with no evidence of disease recurrence.

Discussion

Primary fallopian tube carcinoma (PFTC) accounts for approximately 0.14–1.8% of all female genital malignancies. Even more uncommon are the soft tissue tumors of fallopian tube, and in most of the cases it is leiomyosarcoma. To the best of our knowledge, till date 39 cases of primary tubal sarcoma have been reported [1].

First case of pure sarcoma was described in 1886 by Senger et al. Jacoby et al. reported the 34th case of tubal sarcoma in 1993 and reviewed the previous reports [2]. The 39th case of primary tubal leiomyosarcoma was described by Ueda et al. in 2009 [1].

Based on published case series, soft tissue tumors of fallopian tube show hemorrhage and necrosis; were microscopically characterized by spindle cells arranged in fascicles with nuclear pleomorphism; and increased mitotic activity in most of the cases [3]. In the current case report, the tumor exhibited all the histopathological characteristics.

Immunohistochemically, these tumors need to be differentiated from extra-gastrointestinal stromal tumors

Fig. 1 Intraoperative findings showing ill-defined, ruptured, necrotic tumor arising from left fallopian tube, the tumor has undergone torsion and is seen filling the pouch of Douglas

Fig. 2 Histopathological findings of the left fallopian tube tumor. (A) The tumor shows atypical plump oval/spindle cells with irregular margins, moderate cytoplasm, and pleomorphic nuclei (magnification x 100, staining Hematoxylin and Eosin (H&E)). (B) The tumor has

areas of extensive necrosis (X400, H&E). (C) Immunohistochemistry (IHC) findings of left fallopian tumor showing diffuse positive reaction to alpha smooth muscle actin (SMA) including the aberrant nuclear positivity. (D) Tumor cells show focal positivity to CD 10

(EGIST). Gastrointestinal stromal tumors (GISTs) are mesenchymal tumors of the gastrointestinal tract, generally CD117 positive and KIT or PDGFRA mutation driven. CD117 is a c-Kit proto-oncogene product and is currently considered the most sensitive and specific marker for GIST. The tumor in the present case stained negative for CD117.

Primary fallopian tube sarcoma can occur throughout adult life, with a median age of incidence of 47 years. The typical presenting symptoms as seen in other tubal malignancy include abdominal pelvic pain, pressure symptoms and abdominal lump or distension. However, the characteristic PTFC syndrome ‘hydros tubae profluens’ characterized by intermittent colicky lower abdominal pain relieved by a profuse, serous, watery, yellow discharge from the vagina is not seen with sarcoma of fallopian tube. It has been hypothesized that this characteristic symptom is seen in PTFC by filling and emptying of a partially blocked fallopian tube by the tumor arising from the inner lining of the tube. However, this symptom is not observed in

sarcoma maybe because it arises from the muscle layer of fallopian tube and grows outward.

The prognostic factors of fallopian tube sarcoma are difficult to evaluate because of its rarity. In general, sarcoma of fallopian tube is associated with poor prognosis. The primary treatment of tubal sarcoma is surgery, followed by adjuvant radiation and/or chemotherapy.

The role of robotic-assisted staging surgery for endometrial and cervical cancer is now well established. However, the use of robotic surgery for ovarian and fallopian tube malignancies, especially carcinoma, is limited to a few series of cases and comparative studies with laparoscopy or laparotomy. Evidence regarding the use of robotic-assisted surgical treatment for women with ovarian cancer are still scarce, but its use is progressively growing. Gallotta et al. [4] have demonstrated the shorter operative time with robotic-assisted staging surgery as compared to laparoscopy with the similar oncologic outcome in early-stage ovarian cancer. Evidence regarding the management of sarcoma of fallopian tube with robotic-assisted staging

surgery are not present in the literature. To the best of our knowledge, this is the first case report where primary undifferentiated sarcoma of fallopian tube was managed with robotic-assisted staging surgery.

Sarcoma in general has a low sensitivity to radiation therapy. Randomized trial has failed to show survival benefit for adjuvant RT in stage I or II sarcoma. Various chemotherapeutic regimens used for the treatment of fallopian tube sarcoma include vincristine, actinomycin D, cyclophosphamide, doxorubicin, 5-fluorouracil, dacarbazine, and cisplatin. Combination chemotherapy with docetaxel and gemcitabine is the preferred regimen for uterine leiomyosarcoma. In the current case, we have treated the patient with adjuvant chemotherapy with Adriamycin and ifosfamide.

Compliance with Ethical Standards

Conflict of interest Drs. Reshu Agarwal, Anupama Rajanbabu, and Indu R. Nair declare that there are no conflicts of interest.

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