Case Report

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DOI: 10.4103/pjog.pjog_26_22

Paratubal mass carcinosarcoma: A case report of a rare malignancy in a rare location

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Abstract:

Carcinosarcoma, formerly known as malignant mixed Mullerian tumors (MMMTs) are highly aggressive tumors that include both malignant epithelial and mesenchymal or stromal elements. The most common site of carcinosarcomas in a female reproductive organ is the endometrium with an incidence of 2/100,000 females, whereas carcinosarcomas arising from the paratubal mass are extremely rare malignancies accounting for fewer than 0.1% of MMMTs. Carcinosarcomas of the Fallopian tube usually occur in the fifth to sixth decades in postmenopausal women with the most common presenting symptom of abdominal pain, followed by vaginal bleeding and abdominal distention. There have been limited published cases worldwide, that is, it has been a thing of interest to be analyzed in today's era. A rare case of paratubal carcinosarcoma is highlighted in this paper as it discourses its clinicopathological characteristics and assesses the prognostic factors associated with treatment outcome and survival.

Keywords:

Carcinosarcoma, Malignant mixed Mullerian tumor, paratubal mass carcinosarcoma

Introduction

Carcinosarcomas are a type of tumor that is highly aggressive with a poor prognosis.[1] The incidence is known to be very low to which most often occurs within the female genital tract. The Fallopian tubes are the least common site.[2] Immunohistochemical stains are an important entity in establishing the diagnosis of paratubal carcinosarcoma.[3]

Due to its rarity, no national guidelines have been reported and established for the treatment of paratubal mass carcinosarcoma. Total abdominal hysterectomy with bilateral salpingo-oophorectomy with surgical staging, followed by radiotherapy and/or chemotherapy is the preferred standard option. In cases of metastasis, cytoreductive surgery is highly recommended.[4]

Case Report

A 51-year-old, G2P2 (2002married, presented with a 2-month history of abdominal pain associated with weight loss and hypogastric pain. There were no changes in menstrual patterns, bowel, and urinary habits. The patient was diagnosed with infiltrating ductal carcinoma of breast last 2001 and underwent 29 fractions of radiotherapy. The patient was on tamoxifen for 5 years and is in remission for her breast cancer. Family history, social, sexual, and obstetric history are unremarkable.

On physical examination, a right adnexal mass was palpated measuring 15 cm × 13 cm × 7 cm, solid, slightly movable, and nontender. Another physical examination was unremarkable.

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Submitted: 23-Jun-2022
Revised: 24-Jun-2022
Accepted: 24-Jun-2022
Published: 23-Aug-2022

How to cite this article: Albon IK, Billod JA. Paratubal mass carcinosarcoma: A case report of a rare malignancy in a rare location. Philipp J Obstet Gynecol 2022;46:126-30.

Transvaginal ultrasound showed a normal-sized anteverted uterus measuring 4.5 cm × 4.5 cm × 4 ×, with an intact endometrial stripe measuring 0.4 cm. A right adnexal mass measured 12 cm × 16 cm × 6 cm with solid components with no fluid detected at the posterior cul-de-sac with an impression of ovarian new growth probably malignant.

She underwent an exploratory laparotomy. Intraoperatively, there was a massive hemoperitoneum, a necrotic right adnexal mass measuring 14 cm × 11cm × 6 cm seemingly arising from the right Fallopian tube, ruptured and exuding brainy tissue. A portion of the omentum was adherent to the superior aspect of the mass with multiple subcentimeter implants [Figure 1]. The uterine serosa was smooth, while the endometrium was thin and smooth. Both ovaries and the left Fallopian tube were grossly normal [Figure 2]. Peritoneal fluid cytology, excision of the paratubal mass, total abdominal hysterectomy with bilateral salpingo-oophorectomy, random peritoneal biopsy, and bilateral lymph node dissection were done. In addition, a para-aortic lymph node assessment was made.

Histopathologic diagnosis revealed consistency with carcinosarcoma of the right paratubal mass and random peritoneal biopsy. Lymphovascular invasion is observed. Intramural leiomyomas, atrophic endometrium, bilateral ovaries, and the left Fallopian tube were noted. There was a note of acute and chronic inflammation with reactive mesothelial cell hyperplasia of the omentum and reactive lymphadenitis.

The final diagnosis is carcinosarcoma, paratubal mass, stage IIIB s/p excision of the paratubal mass, total abdominal hysterectomy with bilateral salpingo-oophorectomy, and surgical staging.

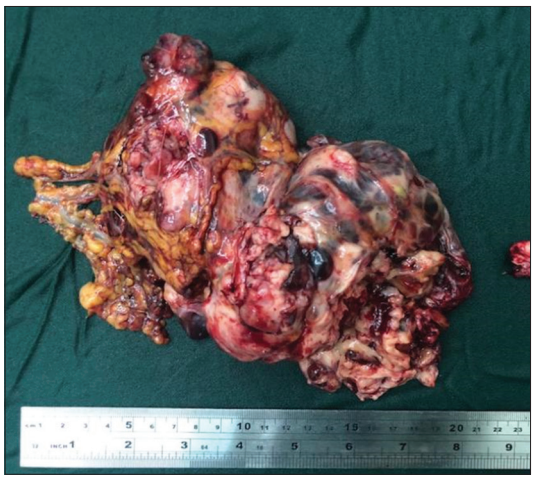

Figure 1: The right paratubal mass that is seemingly arising from the right Fallopian tube measuring 14 cm × 11 cm × 6 cm, that is cream to tan, with irregular, lobulated, variegated, hemorrhagic, and necrotic areas

She received six cycles of chemotherapy (carboplatin–paclitaxel). Surveillance Ca-125 and imaging studies were normal. She is in remission of disease for 1 year.

After 2 years of regular follow-up, a surveillance computed tomography (CT) scan was done revealing an intraperitoneal mass likely metastasis. Ca-125 was elevated prompting admission for chemotherapy (ifosfamide protocol). The patient eventually expired on day 3 of the third cycle of chemotherapy due to pulmonary embolism and acute respiratory failure.

Discussion

There are four theories proposed to explain the histogenesis of malignant mixed Mullerian tumors (MMMTs). These are to following:

1. Collision theory – The theory proposes that two different cellular lines have a biclonal origin where two distinct neoplastic, separate, synchronous proliferations or clones to form one tumor
2. Composition theory – This theory suggests monoclonal origin postulating that the tumor composition originates from the epithelial component during a metaplastic transformation. That is, it hypothesizes that the sarcomatoid component is reactive and not neoplastic, developing as a consequence of paracrine factors secreted by the epithelial component
3. Metaplastic theory – This theory favors a common cell of origin, stating that the sarcomatous component arises from carcinoma by metaplasia of the subclonal population. This theory is favored currently
4. Combination theory – This theory supports all the above mentioned.[4]

Almost 81 cases of carcinosarcoma of the Fallopian tube have been previously reported in the literature; however, reports on paratubal carcinosarcomas are scarce. Carcinosarcoma cells are divided into two categories.

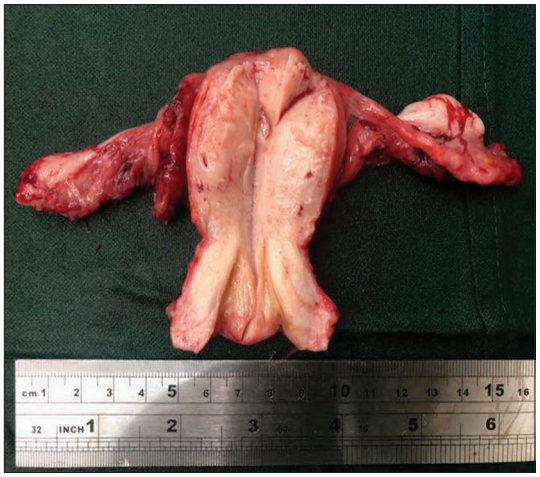

Figure 2: Normal gross section of corpus uteri, cervix, and left Fallopian tube

First is a homologous type that includes endometrial stroma and fibrous and smooth muscle tissue. Moreover, next is a heterologous type which includes cartilage, skeletal muscle, and bone.

Risk factors for MMMTs are nulliparity, obesity, prolonged estrogen exposure, diabetes, and history of pelvic radiation. Cases of tamoxifen-associated carcinosarcoma have also been reported. Our index case has been on tamoxifen therapy for 5 years and had undergone a series of radiotherapy.

Carcinosarcoma approximately affects women in their postmenopausal stage. The patient may become asymptomatic and at the time of examination, they are already at an advanced stage. The usual presentation is abdominal pain due to tubal distention and forced peristalsis in 20%–50% of cases, followed by vaginal bleeding or palpable mass on the adnexa. The patient presented with abdominal pain.

Reports on imaging carcinosarcomas of the paratubal mass are rare in the publications; however, it was reported that compared to carcinoma of the Fallopian tube, carcinosarcoma of paratubal origin has a heterogeneous signal on magnetic resonance imaging (MRI) compared to a homogeneous signal in carcinoma. This could suggest that MRI should be used over CT to not only localize the origin of a pelvic mass but also to determine certain imaging characteristics that may assist in the preoperative diagnosis of a paratubal carcinosarcoma.[5] Sonography is an ineffective investigation for uterine carcinosarcoma. Doppler imaging reveals poor evidence in predicting tumor stage, evaluating the retroperitoneum, and evaluating the deep pelvic node chains. In a reported case, it may be reliable to detect hypervascularity of a uterine carcinosarcoma; however, it has poor sensitivity and specificity to detect areas of neovascularization associated with malignant tumors. These tumors are homogeneously echoic with small cystic spaces.[6]

On gross examination, paratubal tumors are large and predominantly solid with frequent cystic degeneration which may rupture, and with extensive hemorrhage and necrosis[7] to which such description has been consistent with that of the patient [Figure 2].

The most common site of metastasis, besides nonspecifically within the pelvis, are the ovaries and the omentum.[8] With our patient, the omentum was positive for carcinosarcoma cells.

On histopathologic examination, sections from the paratubal mass and random peritoneal biopsy show haphazardly arranged spindled cells with moderate to markedly enlarged, pleomorphic, and hyperchromatic nuclei surrounded by fibromyxoid stroma. Some of the cells appear multinucleated. Admixed is a gland-like formation of atypical epithelioid cells. Interspersed mitoses, necrosis, and hemorrhage are noted. Tumor cells are seen within lymphovascular spaces [Figure 3]. Sections of the omentum proved necrotic tissue with interspersed neutrophils, lymphocytes, and plasma cells, and dilated endocervical glands [Figure 4]. The microscopic descriptions are consistent with that of the index patient.

Immunohistochemistry is used to confirm the diagnosis of carcinosarcoma. It is a unique and very essential tool in determining the biphasic nature and areas of heterologous sections of the tumor which may give a prognostic impact. Cytokeratin and epithelial membrane antigen are positive for epithelial components, whereas, vimentin is for smooth muscle actin. The mesenchymal component is positive in stains such as CD10, desmin, and myoglobin, while S100 is used to detect chondroid or adipose tissue differentiation. CD34 marker distinguishes ovarian carcinosarcomas from the epitheloid component which strongly expresses CD34. It has been suggested that cytokeratin and vimentin or smooth muscle antigen are the gold standard for diagnosis.[9] Immunohistochemical staining done in our case showed strong positivity with cytokeratin, while the spindle areas are positive for vimentin, [Figures 5 and 6] hence confirming the diagnosis of carcinosarcoma of paratubal mass.

Based on the recommendation of the National Oncology of the U.S. , the treatment of Carcinosarcoma of any stage is to follow the guidelines for a group of epithelial origin. The staging is performed according to the criteria of tumor, node, metastasis or (The International Federation of Gynecology and Obstetrics) FIGO as

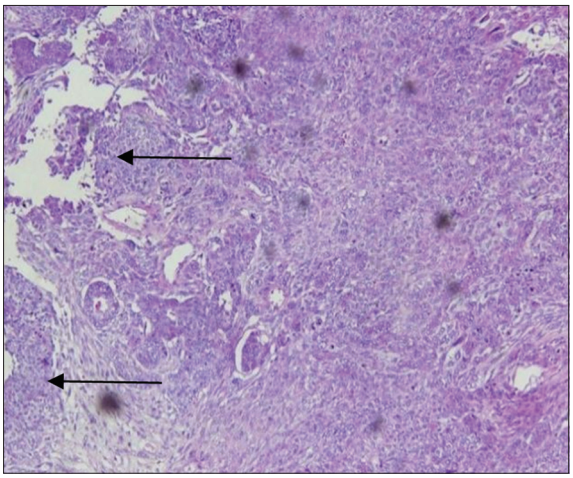

Figure 3: Scanning view (H and E, ×40) and LPO view of the section of the paratubal mass showing haphazardly arranged spindled cells (lower arrow) with moderate to markedly enlarged, pleomorphic, and hyperchromatic nuclei surrounded by fibromyxoid stroma (upper arrow)

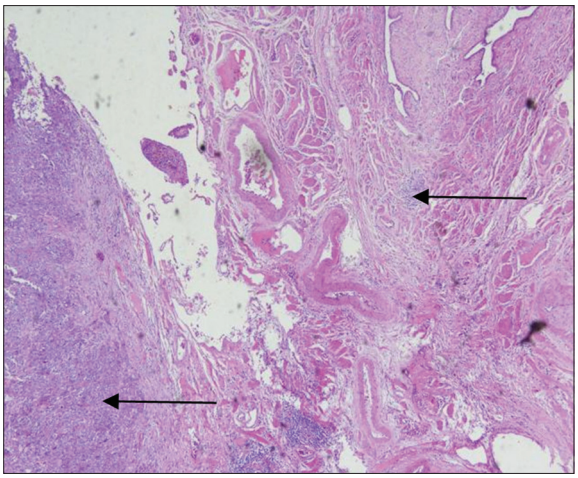

Figure 4: LPO view of the cross-sections of the omentum revealing necrosis with interspersed neutrophils, lymphocytes, and plasma cells (upper arrow) and dilated endocervical glands (lower arrow)

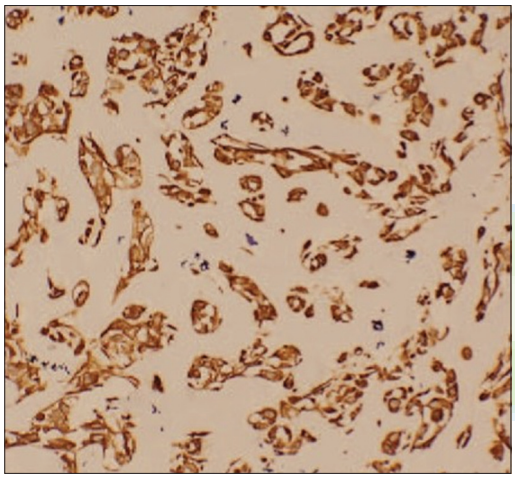

Figure 5: Strong positivity with cytokeratin confirming the diagnosis of carcinosarcoma of paratubal mass

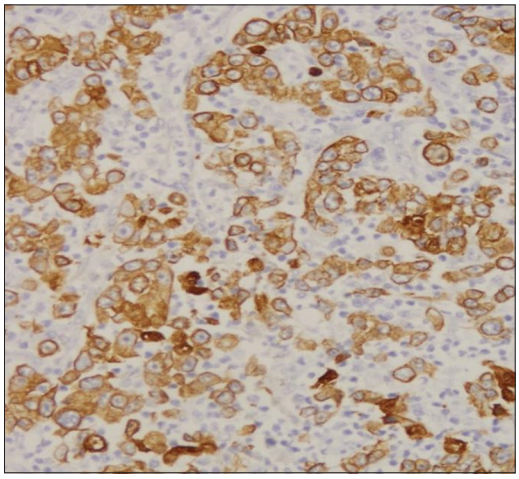

Figure 6: The spindle areas are positive for vimentin hence confirming the diagnosis of carcinosarcoma of paratubal mass

revised in 2009. The initial approach to diagnosis includes history, physical examination, blood tests, tumor markers (CEA, Ca-125), and additionally in the presence of previous histologic confirmation: chest radiography, mammography, gastroscopy, colonoscopy, ultrasound, and/or CT scan of the abdomen. Based on the 2009 FIGO staging of ovarian, fallopian, and primary peritoneal cancer, our patient is Stage IIIB.

The median survival is <24 months; and the 5-year survival is 15%–30%, which compares unfavorably with that of high-grade serous carcinoma. Surgical resection of carcinosarcoma is considered an initial step in its management; however, it may not be suitable in patients with issues of preserving their fertility. The cytoreductive surgery includes the removal of all visible disease masses and complete surgical staging; obtaining ascitic fluid cytology, hysterectomy, bilateral salpingo-oophorectomy, omentectomy, appendectomy, and para-aortic and pelvic lymphadenopathy to the renal vein, especially on the left, peritoneal, paracolic, subdiaphragmatic, and minor pelvic biopsies. In advanced-staged disease (FIGO III-IV), cytoreduction is also necessary. Selected cases may be given preoperatively three or six cycles of chemotherapy to achieve an optimal surgical result. In a series of case studies done by Melissa Brackman *et. al*, she made a comparison of the first-line chemotherapy regimens which are the carboplatin/paclitaxel versus ifosfamide/paclitaxel. Patients that have undergone a carboplatin/paclitaxel regimen have longer progression-free survival than a patient treated with ifosfamide/paclitaxel. Moreover, platinum-containing regimens appear to prolong the progression-free survival when compared to nonplatinum-containing regimens. Due to the advantages such as lower morbidity, lower morbidity, lower cost, and fewer hospital days associated with carboplatin-paclitaxel regimens as well as longer progression-free survival, it is favored as the first-line chemotherapy for carcinosarcomas after surgical debulking. Studies of optimized chemotherapy remains an interest and warranted, including clinical experimental trials and labeling biomolecular description of ovarian carcinosarcomas.

Given the limited data regarding the management of ovarian and tubal carcinosarcoma, patients are usually treated similarly to women diagnosed with other subtypes of epithelial ovarian/Fallopian tube cancer. Most of the existing retrospective studies support the role of cytoreductive surgery in the management of pelvic carcinosarcoma, with optimal debulking associated with improved survival. The current accepted adjuvant treatment is platinum-based chemotherapy.[10] Our index underwent a carboplatin-paclitaxel regimen for 6 cycles.

Summary

This report and review conclude paratubal mass carcinosarcoma as a rare adnexal tumor that possesses an aggressive behavior; however, survival is improved with cytoreductive surgery and adjuvant chemotherapy. To this, it requires a meticulous and wide array of workups to come up with the diagnosis and eventually provide adequate and prompt treatment. Carcinosarcoma of the paratubal origin because of its rarity and of limited record emphasizes the need for collaborative prospective studies targeted to better understand the disease entity, its behavior, and the need to establish new therapeutic regimens to increase the survival rate of patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Shakti BK, Kiranabala D, Deep SA. Malignant mixed Mullerian tumor – Case reports and review article. Apollo Medicine, September 2009;6:227-40.
2. Yang S, Lin L, Peng Z, Yang K, Lou J. Malignant mixed Müllerian tumor of the fallopian tube in a patient with irregular vaginal bleeding. *Lab. Med*. 2009;40:401-3.
3. Imachi M, Tsukamoto N, Shigematsu T, Watanabe T, Uehira K, Amada S, *et al.* Malignant mixed müllerian tumor of the fallopian tube: Report of two cases and review of literature. Gynecol Oncol 1992;47:114-24.
4. Akiki M, Farkhonde F, Fourchotte V, Garau X. Primitive fallopian tube carcinosarcoma: Three cases with immunohistochemical profiling. Int J Cancer Clin Res 2015;2:1.
5. Shen DH, Khoo US, Xue WC, Ngan HY, Wang JL, Liu VW, *et al.* Primary peritoneal malignant mixed müllerian tumors. A clinicopathologic, immunohistochemical, and genetic study. Cancer 2001;91:1052-60.
6. Tanaka YO, Tsunoda H, Minami R, Yoshikawa H, Minami M. Carcinosarcoma of the uterus: MR findings. J Magn Reson Imaging 2008;28:434-9.
7. Kanthan R, Senger JL. Uterine carcinosarcomas (malignant mixed müllerian tumours): A review with special emphasis on the controversies in management. Obstet Gynecol Int 2011;2011:470795.
8. Thawal YA, Tambe SG, Tania A, Chavan RR, Patel JA. A rare case of malignant mixed mullerian tumour of uterine corpus. Int J Med Appl Sci 2014;3:100-2.
9. Yang S, Lin L, Peng Z, Yang K, Lou J. Malignant mixed müllerian tumor of the fallopian tube in a patient with irregular vaginal bleeding. Lab Med 2009;40:401-3.
10. Brackmann M, Stasenko M, Uppal S, Erba J, Reynolds RK, McLean K. Comparison of first-line chemotherapy regimens for ovarian carcinosarcoma: A single institution case series and review of the literature. BMC Cancer 2018;18:172.