Management of a Female Adnexal Tumor of Probable Wolffian Origin Complicated by Intercurrent Disseminated Intravascular Coagulation

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Clinical Practice Points

- Female adnexal tumors of probable wolffian origin are very rare neoplasms that coincide with abdominal pain and swelling.
- These lesions are typically considered to be of low malignant potential, but they are periodically aggressive and can metastasize.
- Immunohistochemistry is not always effective in determining the clinical diagnosis of these tumors, and standard treatment is indeterminate.
- This report presents a patient diagnosed with a female adnexal tumor of probable wolffian origin who intraoperatively developed incidental disseminated intravascular coagulation that was successfully addressed with pressure, microfibrillar collagen hemostat, and blood products. Regrettably, she died of disease progression 4 weeks postoperatively.
- Despite the uncharacteristic coexistence of a female adnexal tumor of probable wolffian origin and disseminated intravascular coagulation, oncology surgeons should be prepared for these 2 conditions in an attempt to confer an optimal patient outcome.

Introduction

Female adnexal tumors of probable wolffian origin were originally documented in 1973, and since then approximately 86 cases have been reported in the literature.1,2 Clinically, these neoplasms are predominantly of low malignant potential, but they can be aggressive, with a proclivity for distant metastases and disease progression.3

Grossly, female adnexal tumors of probable wolffian origin are solid, cystic, or both, and histologically, they contain diffuse epithelial cells with sieve-like, tubular patterns; the tumor cells grow in solid sheets, and may also exhibit a mesenchymal, spindle cell appearance.5,6 These lesions are immunoreactive for cytokeratin, inhibin, and vimentin.1,5,6 Nevertheless, because immunohistochemistry may not necessarily differentiate these malignancies from sex cord–stromal tumors or adenocarcinoma,3,4 patient diagnosis and corresponding therapeutic management remain indeterminate.

Case Report

A 67-year-old (gravid 1, para 1) woman presented with a pelvic mass in June 2013. She was taken to the operating room, whereupon the mass was found to be adherent to the anterior abdominal wall, omentum, sigmoid colon, left pelvic sidewall, and small intestine mesentery. There was discernible metastatic disease in the pelvis; thus, the decision was made to bisect the friable tumor to enhance visibility and facilitate resection.

Suddenly, the patient experienced massive intraperitoneal hemorrhaging, indicative of disseminated intravascular coagulation (DIC). Initially, direct abdominal pressure was used to staunch the hemorrhaging. Her international normalized ratio (1.9) and activated partial thromboplastin time (50 seconds) were significantly prolonged. Moreover, the patient’s fibrinogen level (70 mg/dL)
was markedly diminished, and she was thrombocytopenic (74,000 cells/mm³).

The patient received 7 units of packed red blood cells, 2 units of fresh frozen plasma, and 15 units of cryoprecipitate to address the coagulopathy. Finally, microfibrillar collagen hemostat topical powder and sheets (C.R. Bard Inc, Murray Hill, NJ) were placed on the bleeding peritoneal surfaces and liberally positioned in the pelvic; hemostasis was ultimately achieved.

In consideration of the DIC, the omentum and rectal nodules were not resected. Postoperatively, a 2.5-cm nodule on the anterior rectum represented the largest amount of residual disease; the estimated blood loss for the procedure was 2600 mL. Once the surgery was concluded, hemostasis was excellent and the patient was in stable condition.

Immunostaining found that the epithelial cells were positive for PAX8 (paired box 8) (strong, diffuse), CD10 (multifocal), calretinin (multifocal), pankeratin, vimentin, CD56, and cytokeratin 7 (CK7). The neoplastic tissue was morphologically biphasic, comprising uniform nuclear morphology with epithelial differentiation and an admixture of glandular and prominent spindle components. Initially, endometrial carcinoma and mesothelioma were considered, but the CD10, calretinin, and PAX8 findings were more indicative of a female adnexal tumor of probable wolfian origin (Figures 1 to 5).

The patient was discharged and slated to undergo adjuvant paclitaxel and carboplatin chemotherapy. However, 4 weeks
postoperatively, she experienced acute decompensation, upon which she was immediately referred to the emergency room and underwent a laparotomy; the surgical findings revealed profound, recurrent malignant tissue throughout the small intestine. In consideration of the cancer’s recrudescent nature, further surgical resection was contraindicated; shortly thereafter, the patient died of her disease.

Discussion

The genital ducts effectuate the transportation of reproductive cells to a location amenable to fertilization. Embryologically in males, the emerging gonad contributes to the wolfian ducts’ development and regression of the müllerian ducts.7 Alternatively, in a female embryo, the müllerian ducts develop and the wolfian ducts regress. When the female wolfian ducts fail to regress, remnants may be encountered throughout several regions therein (eg, in the ovarian hilum, in the broad ligament, and lateral to the uterus or vagina).7

This report describes a patient with a female adnexal tumor of probable wolfian origin whose intraoperative management was complicated by DIC. DIC is characterized by systemic activation of blood coagulation, which can severely diminish coagulation proteins and platelets, resulting in intractable bleeding.8 Clinically, patients with ascites who undergo cytoreductive surgery and have a preoperative serum albumin less than 3.5 g/dL or metastases greater than 10 cm may be at increased risk for the development of a perioperative coagulopathy.9 Interestingly, the present authors were unable to identify any cases involving these 2 intercurrent conditions, and it is presumed that the DIC was attributable to the resection and bisection of the large, friable tumor.

Optimal therapy in the management of female adnexal tumors of probable wolfian origin remains indeterminate (Table 1).1,2,4-6,10 The disease can be focally hemorrhagic and necrotic, and thus an attempt should be made to preserve the lesion’s integrity3; although the data are very limited, surgical management should resemble the approach to epithelial ovarian malignancies (ie, staging for disease clinically confined to the ovary and tumor debulking in patients with advanced-stage disease).1,2 However, because these malignancies can be aggressive and associated with progressive disease,3 surgery, adjuvant therapy, and surveillance are recommended.

Regarding cytotoxic therapy, Steed et al2 reported a 2-year progression-free interval after cisplatin (75 mg/m²) and cyclophosphamide (450 mg/m²) chemotherapy in a patient diagnosed with a female adnexal tumor of probable wolfian origin. When the patient developed progressive disease, she was treated with several lines of chemotherapies, of which imatinib (300 mg) conferred quiescent disease with 10 months of follow-up.

The reported combinations of cisplatin/cyclophosphamide and cisplatin/paclitaxel have been associated with a recurrence-free interval ranging from 4 to 61 months,2,5,10 and the present authors

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Abbreviations: AUC = area under the curve; AWD = alive with disease; DOD = died of disease; NED = no evidence of disease; NS = not specified.
elected to treat this patient with the latter combination. One may also consider radiotherapy, as this treatment modality has been associated with a favorable response.  

Immunohistochemical analysis is reasonable if the tumor over-expresses c-kit (Mast/stem cell growth factor receptor Kit), particularly because imatinib has reportedly conferred a beneficial, clinical outcome. Conversely, if the tumor does not overexpress c-kit, cisplatin and either paclitaxel or cyclophosphamide chemotherapy should be a consideration. The strategy would ultimately be modified based on the patient’s response, toxicity, and comorbid history.

**Conclusion**

Despite the uncharacteristic coexistence of a female adnexal tumour of probable wolfian origin and DIC, oncology surgeons should be prepared for them in an attempt to confer an optimal patient outcome.

**Disclosure**

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**References**