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Case Report

Recurrent female adnexal tumor of probably Wolffian origin: A case report

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ABSTRACT

Objective: Wolffian adnexal tumors (WATs) are rare, and metastasis is uncommon. We present the case of a 53-year-old female with a recurrent WAT.

Case Report: The patient presented with abdominal distension, and ultrasound revealed heterogeneous echoes (130 × 93 × 116 mm^3) around the uterus and ascites. Her cancer antigen 125 (CA125) and CA19-9 levels were elevated. Hysterectomy, bilateral oophorectomy, and tumor resection were performed. The histopathological diagnosis was a WAT. Two years later, multiple abdominal and pelvic masses were found on ultrasonography and computed tomography. Laparotomy revealed nodules in the omentum, mesentery, and pelvic peritoneum. Resection of the pelvic masses and partial resection of the omentum were performed. Immunohistochemistry revealed that the lesions were inhibin A, calretinin, estrogen receptor, progesterone receptor, cluster of differentiation 99, and Pax2 positive. Despite postoperative chemotherapy, she developed liver and renal failure 2 months after surgery, and died of the disease.

Conclusion: This case further suggests that WATs have malignant potential, and close follow-up is necessary.

Introduction

An adnexal tumor of probably Wolffian origin is a rare lesion, with approximately 70 cases reported in the literature since it was first described in 1973 by Kariminejad and Scully [1]. The tumor is found in tissues of Wolffian origin and is primarily seen in the broad ligament; however, it has also been reported in the mesosalpinx, fallopian tube, ovary, and peritoneum [2,3]. The ultrastructure of the tumor is similar to that of the Wolffian duct [4]. In 2003, the World Health Organization officially named the tumor Wolffian adnexal tumor (WAT) [5]. Although WATs have generally been considered benign, cases of aggressive behavior and distant metastasis have been reported, and a recent review has suggested that WATs cannot be considered benign [3,6]. Herein, we report a recurrent WAT in a 53-year-old female who ultimately died of the disease.

Case Report

A 53-year-old female was seen with a complaint of abdominal distension. Her medical history was significant for diabetes mellitus, hypertension, and thrombocytopenia. Ultrasonography showed heterogeneous echoes (130 × 93 × 116 mm^3) around the uterus, ascites, and an effusion in the rectouterine fossa. Her cancer antigen 125 (CA125) level was 181.4 kU/L and CA19-9 was 15.64 U/mL. Exploratory laparotomy revealed an 8 × 10 cm^2 dark purple cystic tumor in the left mesosalpinx. The liver capsule, spleen, omentum, and bowel appeared normal. An intraoperative frozen section of the tumor showed a low-grade malignant epithelial tumor, probably of Wolffian origin. Hysterectomy, bilateral oophorectomy, and resection of the left mesosalpinx tumor and omentum were performed.

Postoperative pathological examination revealed a gray, nodular, and fragile lesion on cross section. Light microscopy revealed various sizes and shapes of glandular ducts that were curved with the lumen covered by stratified columnar and cuboidal epithelium (Figure 1A). The tumor cells had minimal cytoplasm, the nuclei were round or oval, small nucleoli were observed, the chromatin distribution was even, and a mild allotype and nuclear division were noted (Figures 1B and 1C). The final pathological

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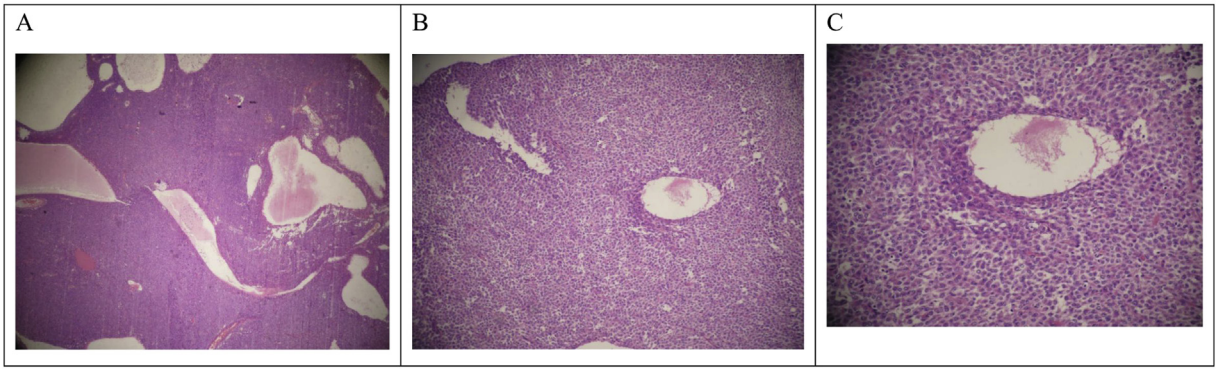

Figure 1. Hematoxylin and eosin staining of the surgical specimen of the first surgery. (A) Glandular structures were observed. The size and shape of glandular ducts varied, they were curved, and the lumens were covered by stratified columnar and cuboidal epithelium (magnification 40×). (B and C) Tumor cells had little cytoplasm with light staining. The nuclei were round or oval, small nucleoli were observed, chromatin distribution was even, and mild allotype and nuclear division were noted (magnification 200× and 400×, respectively).

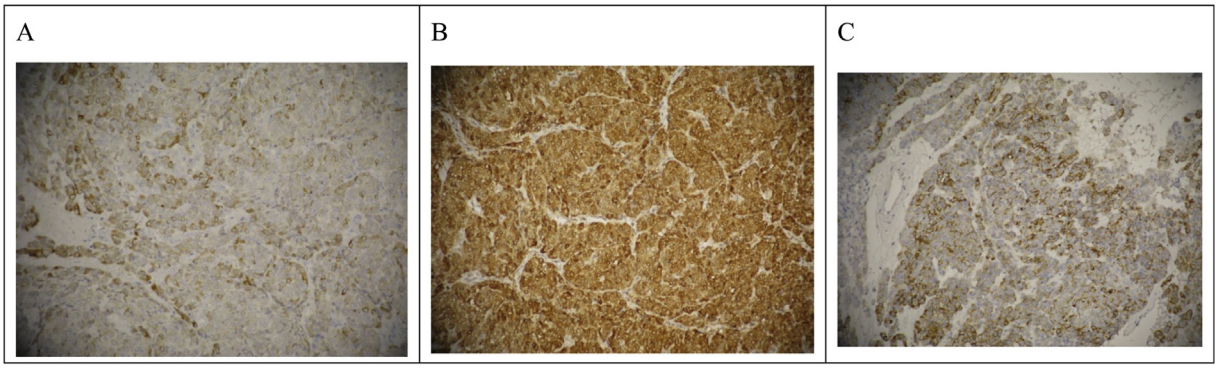

Figure 2. Immunohistochemical staining revealed that the recurrent lesions were positive for (A) inhibin A and (B) calretinin, and (C) focally positive for CK (all 400×).

diagnosis was a tumor of probable Wolffian origin with a low malignant potential. The patient's postoperative course was uneventful, and she was discharged in good condition. However, after discharge she was lost to follow-up.

Two years later, she again presented with abdominal distension. Ultrasonography revealed multiple solid and cystic masses in the abdominal and pelvic cavity. The largest mass measured 66 × 60 mm^2 and had a rich blood flow. A pelvic effusion was also observed. Computed tomography revealed irregular soft masses in the lower abdomen and pelvic cavity, left adrenal nodules, small lesions in the spleen, and mild bilateral pleural effusions. Positron emission thermography–computed tomography showed mildly elevated glucose metabolism in the multiple masses, ascites, and a pelvic effusion. Laboratory testing showed a CA125 level of 118.9 kU/L and a CA199 level of 15.64 U/mL. Laparotomy revealed 2000 mL of bloody ascites and multiple nodules in the omentum, mesentery, and pelvic peritoneum, ranging in size from 1 cm to 12 cm. No lesions were observed in the liver or spleen. Resection of the pelvic masses and partial resection of the omentum were performed.

Postoperative pathological examination revealed the tumors to have a gray-red color with soft edema-like consistency. Light microscopy showed that the characteristics of the lesions were similar to those of the first surgery. Immunohistochemistry evaluation of the lesions revealed that they were positive for inhibin A (Figure 2A), calretinin (Figure 2B), estrogen receptors (ERs), and progesterone receptors, cluster of differentiation (CD)99, and Pax2. Focal cytokeratin (CK) expression was also observed (Figure 2C). The Ki67 proliferation index was 10%. The lesions were negative for epithelial membrane antigen, CD10, caldesmon, p53, S-100, CA125, high-molecular-weight keratin, and Pax8. The final pathological diagnosis was a recurrent/metastatic adnexal tumor of probable Wolffian origin.

At 10 days after surgery, chemotherapy was performed with 70 mg of cisplatin intraperitoneally and 120 mg of docetaxel intravenously. At 33 days after surgery, 150 mg of oxaliplatin was administered intraperitoneally and 120 mg docetaxel was given intravenously.

Two months after the second surgery, the patient developed a body temperature of 38°C. She complained of itching, and her skin, mucosa, and sclera were yellow. Tests of liver and kidney function were abnormal. Magnetic resonance imaging and computed tomography showed a small nodule in the right lower lobe of the lung, which was above the diaphragm and hard to differentiate from the liver. Mild bilateral plural effusions were noted, and a mass was seen in the right lobe of the liver close to the diaphragm, spleen, and hilus hepatis. A nodule was also observed in the left adrenal gland. Magnetic resonance cholangiopancreatography did not reveal obstructive jaundice.

The patient was treated with continuous renal replacement therapy and hepatoprotection therapy. However, her condition continued to deteriorate, and she ultimately died at 83 days after the second surgery.

Discussion

The median age of patients with WATs is around 50 years (range, 13–83 years) [7]. Patients may present with abdominal discomfort,

pain, a mass, and vaginal bleeding, and increased frequency of urination has been reported in some cases [7]. However, in many cases, the tumor is found as an incidental finding [7]. WATs are unilateral in most cases and most often found on the right side [7,8]. They generally are found along the broad ligament or fallopian tube and, in rare cases, have been noted to arise from the rete ovarii and retroperitoneum, and paravaginally [8]. The differential diagnosis of WATs includes Sertoli–Leydig cell tumors, clear cell tumors, granulosa cell tumors, endometrioid ovarian adenocarcinomas, and endometrioid adenocarcinomas of the fallopian tube [5,7,8]. Imaging studies can identify tumors in the abdomen and pelvis, but cannot differentiate WATs from other tumors. However, a recent study suggested that magnetic resonance imaging findings of a low-signal intensity rim around the tumor on T2-weighted magnetic resonance images might be a characteristic feature of WATs [9]. While some lesions can be differentiated based on clinical and morphological characteristics, immunohistochemical analysis is required in most cases for a definitive diagnosis [7,8,10].

Immunohistochemically, WATs are usually positive for CK, vimentin, inhibin A, and calretinin and negative for epithelial membrane antigen, progesterone receptor, and estrogen receptor [3,5]. It has also been reported that WATs can be positive for WT1 and CD99 [11]. Pan-cytokeratin (AE1/3, CK1), CAM5.2, CK7, and vimentin positivity is helpful for the diagnosis of WATs [12]. In recent years, a study has reported that WATs exhibit c-kit expression, and polymerase chain reaction has shown no mutations at the ninth, 11^{th}, 13^{th} and 17^{th} exons of c-kit, and no mutations in the 12^{th} and 18^{th} exons of platelet-derived growth factor receptor [13].

The potential for malignant transformation and metastasis of WATs varies markedly between individuals. Although WATs have been regarded as benign tumors, there is increasing evidence that WATs have the potential for malignant transformation, especially in patients with evident nuclear division in tumor cells [7]. While the prognosis does not appear to have a significant relationship with clinical manifestations and cytopathological features, capsular involvement usually predicts a poor prognosis, especially in cases with pleomorphic nuclei and nuclear division in the tumor cells [6]. Metastasis and recurrence have been noted in some cases where the tumor cells have benign features, and the disease has remained stable for a long period of time [8,14]. Overall, recurrence and metastasis have been reported to occur in 5–10% of cases [8,15]. Lesin et al [12] reviewed 72 cases of WATs and found that the median time to recurrence was 48 months (range: 13–96 months). Recurrence has been reported in the pelvic cavity, abdominal cavity, liver, and lung [10,13,16]. In one case, a patient developed disseminated intraperitoneal recurrence after pregnancy [14]. Heatley [3] reviewed 31 studies of WATs in which 63 patients had complete follow-up data. Heatley's results showed that 50 patients survived and were healthy, seven developed recurrence, and three died, leading the author to conclude that WATs cannot be considered benign. Sivridis et al [6] proposed criteria for malignant WATs: a tumor size >100 mm in diameter, the tumor is rich in cells, the capsule is ruptured, and implantation metastasis or other types of metastasis are present. Of note, the current patient met the criteria proposed by Sivridis et al [6] for malignancy. A recent study of three cases of WATs, of which two were benign and one was malignant, found that the malignant tumor was CD56 positive and the benign tumors were CD56 negative, suggesting that CD56 may be a marker of malignancy [17].

Surgery is the primary treatment for patients with WATs, although radiotherapy with or without chemotherapy is performed in some cases [7,8,15]. There is, however, a lack of evidence supporting any specific therapy. Surgical treatment varies. Usually, hysterectomy and bilateral oophorectomy are performed for patients without the requirement of fertility. Tumor cytoreductive surgery, unilateral oophorectomy, and bilateral oophorectomy are also performed in some cases [7,12]. Whether postoperative radiotherapy/chemotherapy is necessary is unclear, but it is certain that long-term follow-up is necessary because of the risk of recurrence. For patients with recurrence and metastasis, platinum-based chemotherapy and/or radiotherapy are commonly administered [12].

In summary, WATs are rare, and although the initial presentation and histopathological features generally suggest a benign disease, malignant transformation, recurrence, and metastasis can occur. Diagnosis generally requires immunohistochemical analysis. Although there is no consensus on treatment, hysterectomy and bilateral oophorectomy are the primary therapies. The efficacy of chemotherapy and radiotherapy as postoperative adjuvant treatment for recurrence is unclear. Long-term follow-up after initial treatment is necessary due to the risk of recurrence.

Conflicts of interest

The authors have no conflicts of interest relevant to this article.

References

[1] Kariminejad MH, Scully RE. Female adnexal tumor of probable Wolffian origin: a distinctive pathologic entity. Cancer 1973;31:671–7.
[2] Devouassoux-Shisheboran M, Silver SA, Tavassoli FA. Wolffian adnexal tumor, so-called female adnexal tumor of probable Wolffian origin (FATWO): evidence in support of a Wolffian origin. Hum Pathol 1999;30:856–63.
[3] Heatley MK. Is female adnexal tumor of probable Wolffian origin a benign lesion? A systematic review of the English literature. Pathology 2009;41:645–8.
[4] Demopoulos RI, Sitelman A, Flotte T, Bigelow B. Ultrastructural study of a female adnexal tumor of probable Wolffian origin. Cancer 1980;46:2273–80.
[5] Tumours of the fallopian tube and uterine ligaments. In: Tavassoli FA, Develee P, editors. World health classification of tumours: tumors of the breast and female genital organs. Lyon, France: IRAC Press; 2003.
[6] Sivridis E, Giatromanolaki A, Koutlaki N, Anastasiadis P. Malignant female adnexal tumor of probable Wolffian origin: criteria of malignancy. Histopathology 2005;46:716–8.
[7] Ramirez PT, Wolf JK, Malpica A, Deavers MT, Liu J, Broaddus R. Wolffian duct tumors: case reports and review of the literature. Gynecol Oncol 2002;86:225–30.
[8] Sheyn I, Mira JL, Bejarano PA, Husseinzadeh N. Metastatic female adnexal tumor of probable Wolffian origin: a case report and review of the literature. Arch Pathol Lab Med 2000;124:431–4.
[9] Sakai M, Abiko K, Matsumura N, Kondoh E, Yamaguchi K, Minamiguchi S, et al. Two cases of Wolffian tumor with novel magnetic resonance imaging findings reflecting characteristic pathology. J Obstet Gynaecol Res 2016;42:1046–51.
[10] Young RH, Scully RE. Ovarian tumors of probable Wolffian origin: a report of 11 cases. Am J Surg Pathol 1983;7:125–35.
[11] Yang WT, Lu HF, Zhang TQ, Zhang RY, Jiang WC. Wolffian adnexal tumor: a pathological study of 3 cases. Chin J Clin Exp Pathol 2006;22:659–62.
[12] Lesin J, Forko-Ilić J, Plavec A, Planinić P. Management of Wolffian duct tumor recurrence without chemotherapy. Arch Gynecol Obstet 2009;280:855–7.
[13] Syriac S, Durie N, Kesterson J, Lele S, Mhawech-Fauceglia P. Female adnexal tumor of possible Wolffian origin (FATWO) with recurrence 3 years post-surgery. Int J Gynecol Pathol 2011;30:231–5.
[14] Atallah D, Rouzier R, Voutsadakis I, Sader-Ghorra C, Azoury J, Camatte S, et al. Malignant female adnexal tumor of probable Wolffian origin relapsing after pregnancy. Gynecol Oncol 2004;95:402–4.
[15] Daya D. Malignant female adnexal tumor of probable Wolffian origin with review of the literature. Arch Pathol Lab Med 1999;118:310–2.
[16] Taxy JB, Battifora H. Female adnexal tumor of probable Wolffian origin: evidence for a low-grade malignancy. Cancer 1976;37:2349–54.
[17] Nakamura K, Nakayama K, Miura H, Fujiwaki R, Manabe A, Teshima S, et al. Malignant female adnexal tumor of Wolffian origin (FATWO) positive for CD56: a possible diagnostic role for the biomarker. Eur J Gynaecol Oncol 2014;35:580–3.