International Journal on Obstetrics and Gynecology

Female Adnexal Tumors of Probable Wolffian Origin (FATWO); Challenges and Diagnostic Clues on Intra-operative Frozen Section Diagnosis

Case Report

Volume 2 Issue 2- 2022

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Article History

Received: August 05, 2022 Accepted: August 23, 2022 Published: August 24, 2022

Abstract

Female adnexal tumors of probable Wolffian origin (FATWO) or Wolffian adnexal tumors (WAT) are rare neoplasms that arise from the remnants of the mesonephric duct. The tumor is usually benign but can histologically look similar to some other malignant tumors. FATWO is characterized by diffuse and tubular patterns. The intra-operative diagnosis of FATWO is challenging because of the tumor's rarity and its morphologic overlap with these other entities, and FATWO should be considered in the differential diagnoses of sex-cord stromal tumors and ovarian carcinomas based on the exclusion of other neoplasms. The case was presented at College of American Pathologists (CAP) annual conference in Florida (September 2019), and the abstract was published in Archives as an online supplement only.

Abbreviations: FATWO: Female Adnexal Tumors of Probable Wolffian Origin; WAT: Wolffian Adnexal Tumors; CAP: College of American Pathologists; IHC: Immunohistochemistry; EAC: Endometrioid Adenocarcinoma; GCT: Granulosa Cell Tumors

Introduction

An adnexal tumor arising from remnants of the Wolffian (mesonephric) duct, also known as Female adnexal tumor of probable Wolffian origin, (FATWO) or Wolffian adnexal tumor, (WAT) are rare neoplasms with variable histologic appearance [1]. The World Health Organization (WHO) classifies them as borderline tumors with low malignant potential. Although FATWO usually shows a benign behavior, recurrence and metastasis are reported in the literature, though rarely. Patients with FATWO often present with nonspecific symptoms such as abdominal pain and/or a mass with abdominal distension, ascites, and vaginal bleeding [2] Some of these tumors secrete hormones and cause endometrial hyperplasia and abnormal uterine bleeding [3]. Most reported cases are unilateral and have a solid or solid-cystic structure, yet bilateral tumors have also been reported [4]. To date, no mutations or genetic alterations are specifically identified for this entity. Considering the rarity of this FATWO, preoperative diagnosis may present a challenge to surgical pathologists.

Case Presentation

Our patient was a 79-year-old woman with no known comorbidities, who presented to our hospital with abdominal discomfort. Imaging revealed a left adnexal mass. The basic laboratory results and tumor markers, such as CEA, CA19-9 and CA125, were within normal ranges. She underwent laparoscopic left salpingo-oophorectomy and an intraoperative consultation was requested. The ovary showed a cystic appearance with sub-centimeter solid nodules and a yellow cut surface. Microscopic examination of the nodule revealed a proliferation of epithelioid to spindled cells (Figure 1A), arranged in a sieve-like and tubular pattern with foci of eosinophilic secretions and occasional grooves (Figure 1B). Immunohistochemistry (IHC) on permanent sections showed the tumor to be positive for Pan-cytokeratin, CD10 and CD117 (c-kit), whereas PAX8, CK20, CEA and SF1 were negative. Based on its overall features, the tumor was categorized as WAT and the clinical team decided not to proceed with hysterectomy. The patient performed well after surgery and was kept on regular follow-ups. The patient is alive currently with no recurrence

or residual disease after two years from this initial diagnosis.

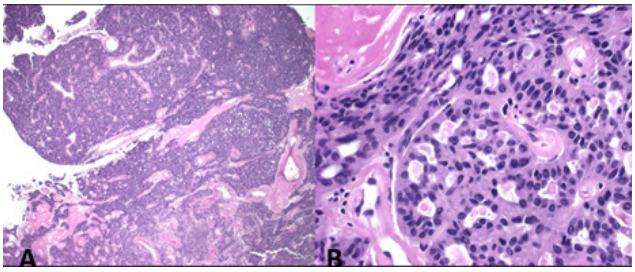

Figure 1: Hematoxylin and eosin stain shows proliferation of epithelioid to spindled cells arranged in a sieve-like and tubular pattern (A: 100x) with foci of eosinophilic secretions, and occasional grooves (B: 400x).

Discussion

FATWO can show different morphologies and should be diagnosed based on the exclusion of other entities. This makes the intra-operative consults challenging because IHC cannot be used to confirm a diagnosis. While some cases of FATWO have mixed patterns including tubular or cord-like, cribriform, cystic, tubulocystic, sieve-like, and diffuse in most cases usually a single pattern predominates [1-5]. Sometimes unusual patterns, such as signet ring cell-like areas with large intra-cytoplasmic vacuoles, are present [5]. The cells are usually round and uniform with pale chromatin. However, the nuclei often have grooves which can mimic granulosa cell tumors. There is minimal nuclear atypia and mitotic activity is usually low (up to 1-2 cells per HPF). On the other hand, malignant FATWO can have as many as 16 mitoses per HPF [4,5].

IHC facilitates concluding a final diagnosis, but it cannot be utilized during frozen section evaluation. It is recommended to have IHC test specifically for FATWO, such as CD10 and cytokeratin but also use other stains to exclude the other histologically similar entities, such as metastatic lesions, sex-cord tumors and mesothelioma. IHC, such as calretinin, inhibin, Melan-A, CD99, WT1, ER, PR, androgen receptor, EMA, GATA3, CD117, vimentin, and SMA (in spindle cell areas if present) show variable expression [1,5,6]. Although no disease-specific mutations or molecular alterations have been identified yet, some reported mutations include CTNNB1, MET, PIK3CA, BRAF, CDKN2A, STK11, and ARID1B [7,8].

Overall, the differential diagnosis of FATWO is broad and includes endometrioid adencarcinoma (EAC) of the ovary, granulosa cell tumors (GCT), and to a lesser extent, metastatic adenocarcinoma [4-6]. The EAC in the ovary is mostly seen in patient with endometriosis and the history or histologic clues of endometriosis can be helpful in ruling out this diagnosis. The presence of uniform and pale nuclei with little or no mitotic activity can facilitate the exclusion of EAC. Because of the overlap among morphologic characteristics, it is not always possible to differentiate between GCT and FATWO on frozen section evaluations. Although nuclear grooves are not an exclusive feature of GCT, the differential diagnosis between GCT and FATWO may be facilitated with the presence of this feature. Sertoli-Leydig cell tumors may also some histologic similarities with FATWO [4-6]. However, the absence of Leydig cells and hormone release help to exclude this diagnosis. Other serological tumor markers are also helpful in ruling out other ovarian tumors, given that the IHC studies are not possible during the intraoperative frozen section evaluation, however the serologic markers are usually measured during the the pre-operative evaluation [7-10].

Conclusion

In summary, FATWO is a rare tumor that may be diagnostically challenging because of its vast histological patterns. It usually shows a benign pattern, and it must be differentiated from more aggressive tumors in order to prevent unnecessary and aggressive measurements. Considering that some of these tumors may recur, sometimes with more aggressive behavior, long-term follow up is recommended.

Disclosures: Parts of this case were presented at College of American Pathologists (CAP) annual conference in Florida (September 2019), and the abstract was published in Archives as an online supplement only.

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