Diagnosis and management of female adnexal tumor of probable Wolffian origin (FATWO) arising from ovary: a case report

Overlerden gelişen olası Wolffian kökenli adnexal tümörün tanı ve tedavisi: Olgu sunumu

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Abstract

Female adnexal tumor of probable Wolffian origin (FATWO) is a rare neoplasm which is usually considered as benign, although in some cases metastasis or recurrences have been reported even after a long interval following the initial diagnosis. Preoperative diagnosis of FATWO is very difficult because of the rarity of the disease and the limited literature available. In this case report, we present a case of FATWO arising from the ovary and review the literature based on the clinical characteristics and management of this rare condition. A 51- year- old postmenopausal woman was referred to our clinic for evaluation of an adnexal mass. After diagnostic evaluation, the patient underwent explorative laparotomy. Intra-operatively, a solid- cystic mass was found in the right ovary, the rest of the abdomen and the pelvis were normal. The ovarian mass was removed and examined with frozen-section (FS). When the frozen section proved negative for malignancy, total abdominal hysterectomy and bilateral adnexectomy were performed. The anatomic study revealed a well-capsulated mass which was 3.5×1.5 cm in diameter. Based on pathological and immunohistochemical results, the final diagnosis was concluded to be FATWO. Adjuvant therapy was not administered. Te patient was followed up after discharge from the hospital. One year after surgery she was asymptomatic. No evidences of recurrence were observed throughout this period. Although FATWOs are rare tumors, they should be kept in mind in women with an abdominal mass. They can present diagnostic difficulties and the diagnosis is based on the exclusion of other neoplasms. FATWO has malignant potential, after the initial surgical treatment patients should be appropriately followed up for possible recurrence and metastasis.

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Özet

Olası wollfian kökenli kadın adneksal tümörü (FATWO) genellikle benign doğada olan nadir bir neoplazmdır. Bununla beraber ilk tanı konulmasından uzun zaman sonra bile bazı vakalarda metastaz ve yinelemeler bildirilmiştir. Preoperatif olarak FATWO tanısı konulması bu hastalığın çok nadir ve literatürün sınırlı olması nedeniyle çok güçtür. Bu yazıda overden gelişen FATWO tanısı almış bir kadın olgu sunulacak, ve bu nadir görülen durumun tanı ve tedavisiyle ilgili literatür gözden geçirilecektir. Elli bir yaşındaki postmenapozal kadın hasta kliniğimize adneksal kitlenin değerlendirilmesi için gönderildi. Tanısal incelemeden sonra, hastaya eksploratif laparotomi uygulandı. Operasyon esnasında sağ overde solid kistik bir kitle bulundu, bunun dışında abdomen içinde herhangi bir başka anomali saptanmadı. Ovaryan kitle cerrahi olarak çıkarıldı ve frozen kesi ile incelendi. Frozen kesilerde malignansi negatif olarak gelince, total abdominal histerektomi ve bilateral adneksektomi gerçekleştirildi. Anatomik olarak iyi kapsüle olmuş 3.5x1.5 cm çapında bir kitle idi. Patolojik ve immüno histokimyasal tetkik sonucunda nihai tanı FATWO olarak teyid edildi. Adjuvan terapi uygulanmadı. Hasta daha sonra taburculuk sonrası klinik izleme alındı. Halihazırda bir yıldır takipte olan hastada herhangi bir yineleme gözlenmedi. FATWO nadir bir tümör olmasına rağmen kadınlarda karın içi kitlede akılda bulundurulması gereken bir durumdur. Tanısı diğer diğer neoplasm nedenlerinin ekarte edilmesini gerektirdiği için zorluk taşıyabilir. FATWO malign olma potansiyeline sahip bir neoplasm olduğu için olası tekrarlama ve malignansi nedeniyle uygun bir biçimde takip edilmesi gereklidir.

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Anahtar kelimeler: Neoplazm, Wollfian adneksal tümör, FATWO, over, benign

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Introduction

Female adnexal tumor of probable Wolffian origin (FATWO) is a rare, poorly understood lesion first described in 1973 (1). It is defined as a tumor of presumptive mesonephric (Wolffian duct) origin characterized by a variety of epithelial patterns and occurs most commonly in the broad ligament, but is also known to occur in mesosalpynx, fallopian tube, ovary, and peritoneum (2, 3). FATWO is usually regarded as a benign lesion, although in some instances more aggressive behavior is encountered with a tendency to distant metastases and recurrences (4, 5).

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Preoperative diagnosis of FATWO is very difficult because of the rarity of the disease and the limited literature available. There are no comprehensible recommendations regarding its preoperative workup and optimal treatment (6). Here, we report a case of FATWO arising from the ovary and review the literature based on the clinical characteristics and management of this rare situation.

Case Report

A 51-year-old postmenopausal woman gravida 5, para 3, was referred to our clinic for evaluation of a right adnexal mass suspected to be malignant. Her previous gynecological history included cesarean section and appendectomy. Pelvic transvaginal ultrasound revealed a normal sized uterus, with a thin and regular endometrial layer. The left ovary was normal, but the right ovary was enlarged and there were two masses, 2.5 and 2 cm, which contained solid and cystic areas. No traces of ascites were found. In the color Doppler examination of these solid- cystic masses, it was found that Resistance Index (RI) value was 0.55 and Pulsatility Index (PI) value was 0.85. Because the sonographic findings suggested an ovarian malignancy, magnetic resonance imaging (MRI) was performed. The T_1 weighted images showed a hypointense bi-lobule solid mass probably originating from the right ovary. On T_2 weighted images, the mass was heterogenic and hyperintense. A chest X-ray was unremarkable. Serum tumor markers were CA-125=5.8 U/mL (nL<35U/mL), CA 15-3=4.1 U/mL (nL<38.6 U/mL), CA 19-9=7.25 U/mL (nL<37 U/mL) and CEA=0.76 ng/mL (nL<10 ng/mL). The patient underwent explorative laparotomy. Intra-operatively a 3×4 cm solid- cystic mass was found in the right ovary. The rest of the findings in the abdomen and the pelvis were normal. The ovarian mass was removed and examined with frozen-section (FS). When the FS proved negative for malignancy, total abdominal hysterectomy and bilateral salpingo-oopherectomy were done. The postoperative period was uneventful and the patient was discharged on day 5 after the surgery.

The anatomic study revealed a well-capsulated mass which was 3.5×1.5 cm in diameter. The cut section of the tumoral mass contained solid and microcystic parts and had a sponge-like, yellow appearance. Microscopically, the lesion was well distinguished from ovarian tissue. The tumor, composed of sheets of epithelial like cells, contained oval, polygonal nuclei with regular chromatin and thin- pale cytoplasm (Figure 1a). There wereseparate retiform areas like tubules (Figure 1b). In the stromal tissue some hyalinization and hyalinized vasculature were seen. There were no nuclear atypia or necrosis. The mitotic activity was 0-1 per 10 high-power fields.
Immunohistochemically, the tumor cells were positive for pancytokeratine (Figure 2a), vimentin (Figure 2b) and calretinin (Figure 2c) but negative for inhibin, Factor 8 and epithelial membrane antigen (EMA). Based on pathological and immunohistochemical results, the diagnosis of FATWO was confirmed. The patient was informed about the malignancy risk of this tumor and was recommended close follow up which included clinical examination, tumor markers, abdominal and pelvic ultrasound and MRI. One year after surgery she was asymptomatic. No evidence of recurrence was observed throughout this period.

Discussion

Female adnexal tumor of probable Wolffian origin (FATWO) arises by the rare persisting remnants of the mesonephric duct. FATWO is an uncommon lesion and approximately 80 cases have been reported in the literature including case reports (3). The age at diagnosis ranged from 18 to 81 years, with a mean of 50 years (7). These tumors usually express a benign behavior. However they have recently been considered to have malignant potential. Some of the tumors follow a malignant course with metastasis and recurrence (4, 5). In the review of the literature, it has been reported that almost one-fifth of the cases is associated with an adverse outcome (3). The prognosis of this tumor does not correlate with their clinical presentation and their cytology (4). However, the presence of necrosis, capsular

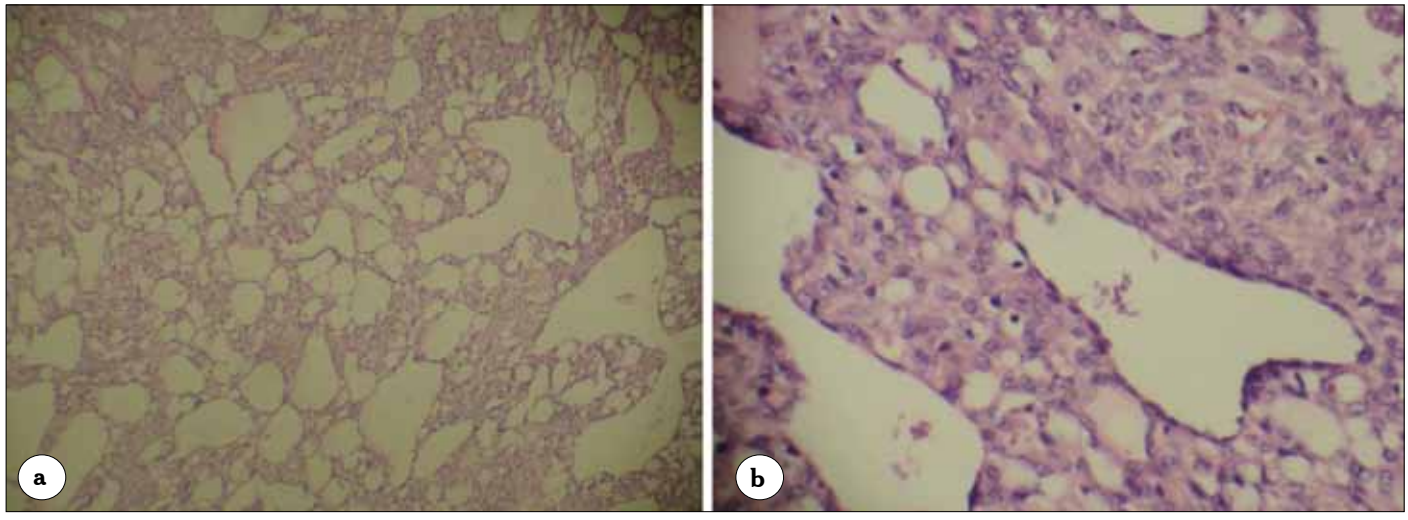

Figure 1. Histological section showing a) solid pattern with sieve-like cystic areas, b) the presence of tumor cells with oval- polygonal nuclei, small nucleoli and scanty- pale cytoplasm (Hematoxylin and eosin, x20)

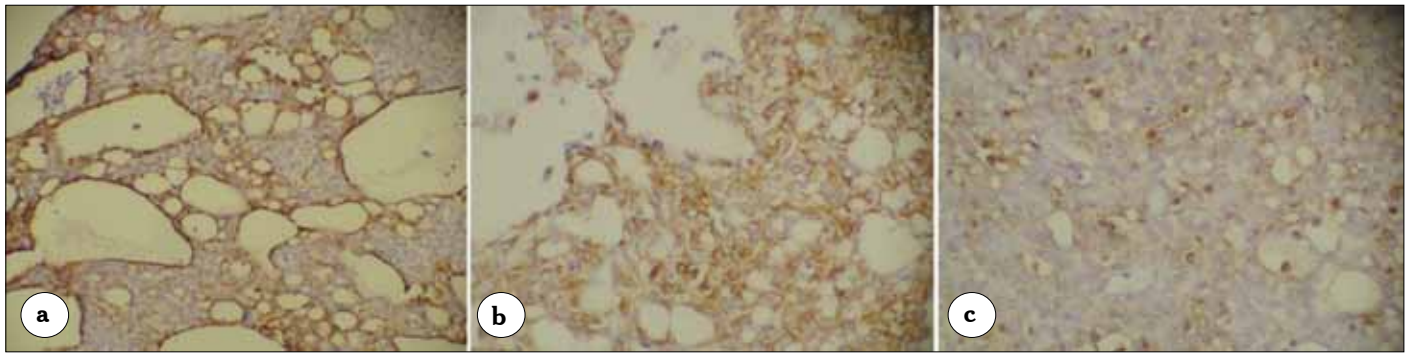

Figure 2. Immunohistochemical finding showing the positive immunoreactions for a) pan- cytokeratin, b) vimentin and c) calretinin (x40)

invasion and especially a high number of mitoses are elements justifying caution regarding their prognosis (4, 8, 9). However, recurrence could occur in the absence of the aggressive histological character and even after several years following the initial diagnosis (8, 10). In the literature, recurrence median time was 48 months with a range from 13 to 96 months, and in some cases recurrences occurred even after a long interval following the diagnosis (4, 9).

The rarity and variable location of FATWO make the diagnosis difficult. Its diagnosis depends largely on histological features which are characterized by a tubular pattern with either closely packed tubules or even solid cords, a sieve-like growth pattern produced by cysts of varying sizes lined by flattened cells and a diffuse growth composed of spindle or polygonal cells (11).

Usually, an ultrasound examination shows a pelvic semisolid mass with high vascularization in these cases. The MRI findings of this tumor were described by Matsuki et al. (7) as a slightly hyperintense mass with cystic degeneration in the adnexa, which were difficult to differentiate from a subserosal leiomyoma or an ovarian tumor. According to the Ramirez et al. (9) review, it has been found that pre-operatively the serum CA-125 levels were normal in all of the patients.

The main differential diagnosis includes well differentiated endometrioid ovarian adenocarcinoma, endometrioid adenocarcinoma of the fallopian tube and Sertoli Leydig cell tumor (3, 12, 13). Endometrioid adenocarcinoma arises from the fallopian tube, whereas FATWO usually arises outside the tube within the broad ligament and ovary (1). The degree of nuclear atypia and mitotic activity is more impressive in endometrioid adenocarcinoma (12). Sertoli-Leydig cell tumors may bear a strong morphological resemblance to FATWO, but FATWO tends not to be associated with the endocrine symptoms that are features of Sertoli Leydig cell tumors (13). Also, Sertoli–Leydig cell tumors have not been reported in the paratubal site or in the broad ligament and the presence of a sieve-like pattern, the absence of Leydig cells, may be useful for the diagnosis of FATWO (10). It is not always possible to differentiate between the broad ligament GCT and female adnexal tumor of probable Wolffian origin (FATWO). Nuclear grooving is not an exclusive feature of GCT and can be seen in a variety of other neoplasms, in the context of the differential diagnosis between broad ligament GCT and FATWO, but the presence of this feature may be very useful in establishing the diagnosis of broad ligament GCT (14).

Specific antibodies for use in immunohistochemistry have become available in the last few decades and immunohistochemical investigations have been performed for the diagnosis of these tumors (11). Positive immunoreactions to pan-cytokeratin, CAM 5.2, cytokeratin 7 (CK7) and vimentin support diagnosis of FATWO (4) .These tumors are generally EMA negative (11, 12). Positive staining for α-inhibin in nine of 10 tumors of probable Wolffian origin has been demonstrated by Kommos et al. (15), but they have also reported that positive staining for α-inhibin is not a useful marker to distinguish between Sertoli cell and Wolffian tumors.

Therapeutic options in these tumors remain ill defined due to the rarity of the disease. It has been considered that the most effective therapy is complete surgical resection with hysterectomy and bilateral adnexectomy (4, 7). Most of the tumor relapses have occurred in patients initially treated with only tumor resection (4). Some authors asserted that the most suitable initial treatment for FATWO was surgical debulking with hysterectomy and bilateral adnexectomy (16). The role of adjuvant chemotherapy or radiation therapy is controversial (7). Also, there are limited options in treating recurrent or metastatic disease. In recurrent or metastatic FATWO, molecular targeted therapy, such as tyrosine kinase inhibitor, could be considered. However, to determine the effectiveness of this option, collective data are needed from multiple centers (17).

In our case, ovarian malignancy was suspected preoperatively. Pathological and immunohistochemical analysis revealed the lesion to be a FATWO. Immunohistochemical findings of the case reported are similar to those described by other authors except for inhibin which has not been detected by us (11, 12, 15). After the surgery, chemotherapy and radiation therapy were not administered and she was scheduled for clinical follow-up.

In conclusion, FATWOs are rare tumors arising from the remnants of the mesonephric duct. Although most of them behave as benign lesions, some cases have the potential for malignant behavior. They can present diagnostic difficulties and the diagnosis is based on the exclusion of other neoplasms. It should be differentiated from other possibilities with careful pathological and immunohistochemical examination. Because the role of adjuvant therapies is questionable, they are not routinely administered. After the initial surgical treatment, patients should be appropriately followed up during a long-term period.

Conflict of interest

No conflict of interest was declared by the authors.

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