

## Case Report

# Female adnexal tumor of probable wolffian origin, FATWO: Report of a rare case

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### Abstract

Female adnexal tumors of probable Wolffian origin (FATWO) arise in the broad ligament from the remnants of the mesonephric duct. The behavior of these tumors is generally benign. However, they can also behave aggressively and exhibit recurrences or metastases. Herein we present a rare case of FATWO that was diagnosed in a premenopausal woman.

### Key words:

Adnexal mass, FATWO, wolffian

## Introduction

Female adnexal tumors of probable Wolffian origin (FATWO) were first documented in 1973 by Kariminejad and Scully [1]. These tumors arise in the broad ligament from the remnants of the mesonephric duct such as epoophoron, paroophoron and Gartner's duct [2]. Approximately 80 cases of FATWO have been previously reported in the literature [3]. The behavior of these tumors is generally benign. However, they can also behave aggressively and exhibit recurrences or metastases [4,5]. Macroscopically, they can be grossly solid, cystic or both. Microscopically, they contain diffuse epithelial cells with sieve-like, tubular patterns [6].

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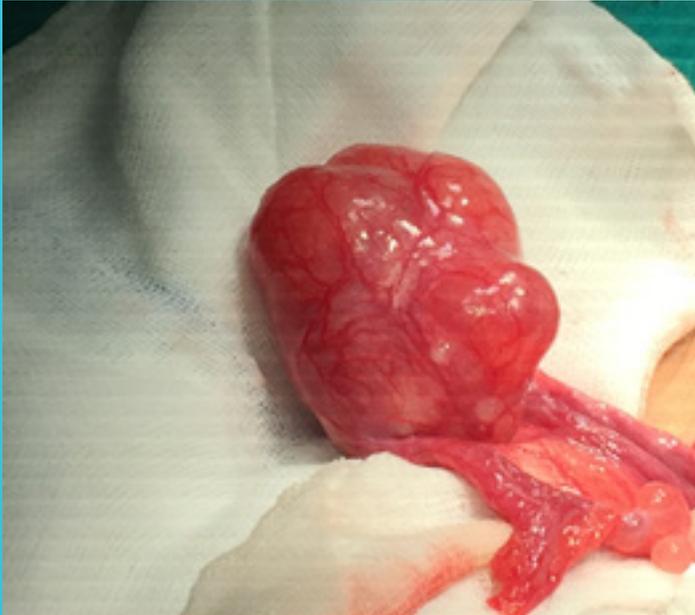
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## Case presentation

A 42-year-old pre-menopausal woman, gravida 2, para 2, was referred to our clinic for evaluation of a left adnexal mass suspected to be malignant. Her previous gynecological history included a cesarean section for dystocia. Pelvic ultrasound showed a normal sized uterus, with a thin and regular endometrial lining. The right ovary was normal. On the left adnexal region, a pure solid mass sized 8x7 cm was noted. There was no ascites in the peritoneal cavity. Serum tumor markers levels (reference range) were as follows: CA-125=15 U/mL (0-35U/mL), CA 15-3=9 U/mL (0-25 U/mL), CA 19-9=7 U/mL (0-27 U/mL) and CEA=0.5 ng/ mL (0-10 ng/mL). The patient underwent exploratory laparotomy with a midline incision. A 8x7 cm solid mass was discovered within the left broad ligament with attachment to the left fallopian tube (Figure 1). Uterus, bilateral ovaries and other intraperitoneal organs were normal. The adnexal mass was totally excised, and was examined via frozen-section (FS). The mass was reported as a tumor highly suspicious for a malignancy.

Hysterectomy, bilateral salpingo-oophorectomy, bilateral pelvic and paraaortic lymphadenectomy, omentectomy and appendectomy were performed for staging purposes.

**Figure 1.**



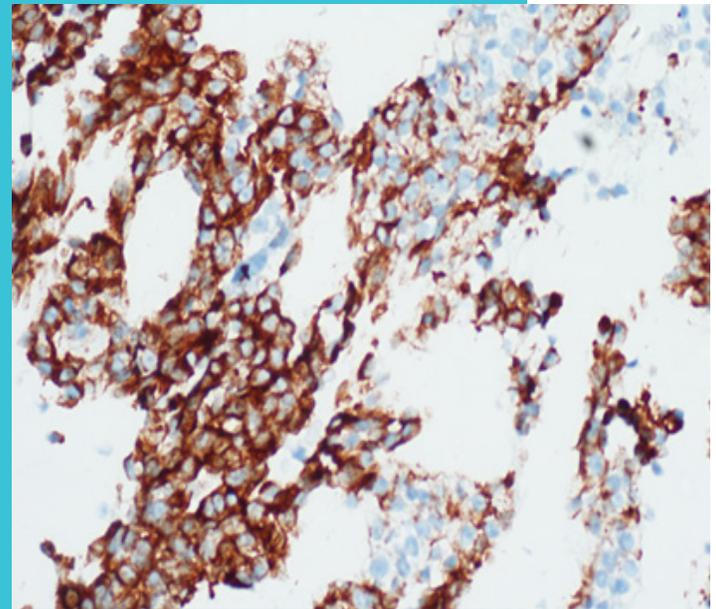
*Solid mass in the broad ligament*

The patient was discharged home on postoperative day 6 without any complications. The final pathology was reported as Female Adnexal Tumor of Probable Wolffian Origin (FATWO). Ki-67 proliferation index was 8-10%. Immunohistochemical staining was positive for pancytokeratin, kalretinin, inhibin and vimentin, and was negative for ER, s100, EMA, CEA, CK20, CD10 and WT-1 (Figure 2,3). The other excised tissues including uterus, bilateral tubes and ovaries, lymph nodes, omentum and appendix were free of tumoral metastases.

## Discussion

During embryogenesis, the female internal genitalia develop from müllerian ducts, and the wolffian ducts regress consequently. If the wolffian ducts fail to regress, its remnants may be encountered within the ovarian hilum, in the broad ligament or lateral to the uterus or vagina [7]. FATWO develops from the persisting remnant of the mesonephric duct [8]. These tumor may occur at any age [9].

**Figure 2.**

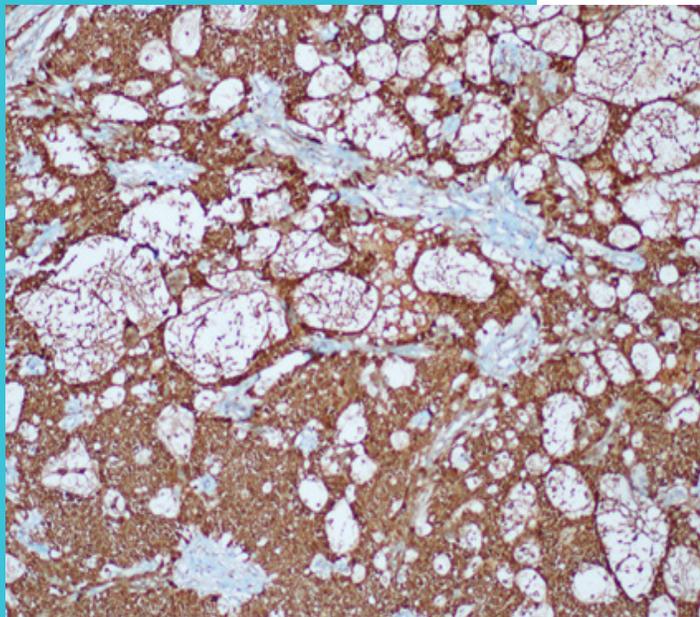


*The positive immunoreactions for vimentin stain (x400)*

Various tumor sizes have been previously reported, ranging between 10 and 25 cm in the literature. Macroscopically, the tumors may be solid, may have a mixed appearance of solid with cystic areas, or more frequently, may have a spongy appearance. These tumors are generally associated with an indolent clinical course, although malignant forms have also been previously described. A review of the literature indicates that metastases and recurrences may develop in these patients, which are characteristics of malignant behavior [10, 11]. The main histological feature is epithelial cells arranged in packed strands with slit-like tubular structures. Cellular pleomorphism and an increased number of mitotic figures are indicators of malignancy, but cases with minimal nuclear

atypia and very low mitotic rates may also occur [10,12].

**Figure 3.**

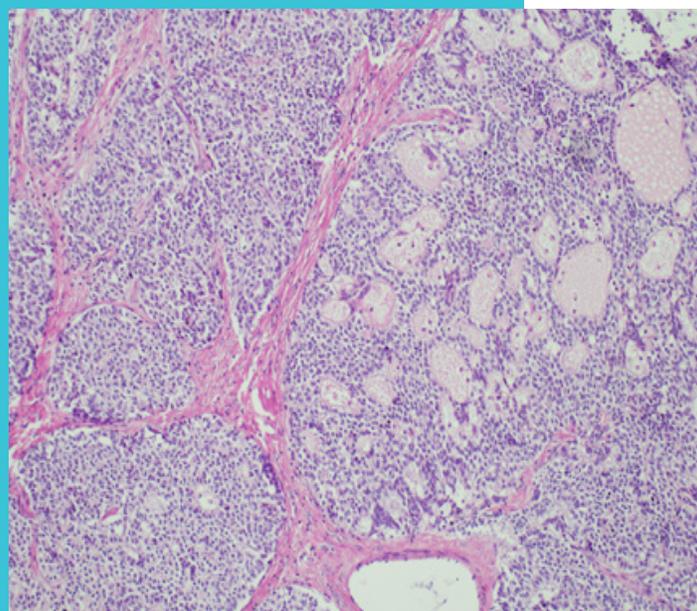


*The positive immunoreactions for calretinin stain (x100)*

The FATWO should be distinguished from granulosa cell tumors, Sertoli-Leydig cell tumors, adenomatoid tumors and endometrioid carcinomas of the Fallopian tubes. Endometrioid carcinoma of the fallopian tube usually occurs as an intraluminal mass and has abnormal hyperchromatic nuclei and high mitotic activity [13]. Sertoli-Leydig cell tumours more commonly occur in young females, generally in women aged 20–30 years, and develop in the ovaries, rather than the broad ligaments or the mesosalpinx. More importantly, they contain typical Leydig cells with abundant eosinophilic granular cytoplasm, and may be associated with endocrine manifestations due to androgen secretion, such as hirsutism. In addition, Sertoli-Leydig cell tumours often express inhibin-A [14]. The main differential diagnosis of broad ligament granulosa cell tumor (GCT) should be made with FATWO [15]. The diagnosis of FATWO is based mainly on its topography and its morphologic appearance of a sieve-like retiform pattern of hollow tubules and cysts, closely packed tubules, and diffuse solid sheets of cells (Figure 4). In these tumors, limited data on their exact nature and outcome are available, thus therapeutic recommendations are mainly

based upon expert opinions derived from case reports. In the previously published literature, complete surgical resection of the mass with hysterectomy and bilateral salpingo-oophorectomy was recommended [16, 17]. Most of the tumor relapses have occurred in patients initially treated with only tumor resection [16]. The role of adjuvant chemotherapy or radiation therapy is controversial. Also, there are limited options in treating recurrent or metastatic disease [18]. In summary, FATWO are rarely encountered tumors in the gynecology practice. The optimal management strategy for these tumors should be investigated in future studies.

**Figure 4.**



*Photomicrograph showing tumor cells arranged in tubular pattern (Hematoxylin and Eosin, x100).*

#### **Acknowledgement**

None

#### **Conflict of Interest**

Authors declare no conflict of interest

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