Female adnexal tumor of probable Wolffian origin: A case report
Gowardhan Vidula, Patrikar Anjali, Bothale Kalpana, Mahore Sadhana

Introduction:
Female adnexal tumour of probable Wolffian origin (FATWO) were first described by Kariminejad and Scully in 1973 and were reported to arise within the leaves of the broad ligament or suspended from it or the fallopian tube. Less than eighty cases of Wolffian tumors of the ovary and female adnexa have been reported in English publications. The behavior is generally benign, but a few have recurred or metastasized. Therefore, a close follow up after surgical intervention should be undertaken.

Case History:
A 38-year-old woman, P2L2, presented with 8 year history of oligomenorrhea and pain in lower abdomen since 4-5 months. Physical examination showed normal secondary sex characteristics. On vaginal examination, a mass of 12 weeks size with restricted mobility was felt separately from the uterus. Routine biochemical and hematological tests showed no abnormal findings. Ultrasound showed a hypoechoic, solid, well defined mass measuring 10.4 cm x 7.2 cm and arising from the left ovary. Serum levels of CA-125 were within normal range. At laparotomy, left paraovarian mass was found. The uterus, both fallopian tubes and ovaries were macroscopically normal. Therefore total abdominal hysterectomy with bilateral salpingoopherectomy was performed along with excision of the mass.

Abstract:
Female adnexal tumour of probable Wolffian origin (FATWO) is a rare tumour that arises from remnants of the Wolffian duct. Rarity and variable location of FATWO make the diagnosis difficult. We report a case of a 38-year-old woman with FATWO which arose within the broad ligament.

Keywords: FATWO, Wolffian tumor, ovary, female adnexa

Figure 1: Gross photograph of encapsulated tumour with smooth external surface

The excised tumour was measuring 9 cm x 9 cm x 7 cm. Grossly, the tumour was encapsulated with smooth external surface (Fig. 1).

The cut surface was homogenous, solid, yellowish white (Fig. 2).

Figure 2: Photograph of tumor showing homogenous, solid, yellowish white cut surface

Microscopically the tumor cells were arranged indiffuse solid, tubular and hollow tubular pattern, separated by scanty stroma (Fig. 3-5).

The diffuse solid areas show compact proliferation of ovoid to spindle cells. The hollow tubules had sertoliform appearance. The tumour cells had uniform round to oval nuclei with fine chromatin. Nuclear pleomorphism, mitoses or tumornecroses were not found. Leydig cells were not seen in the stroma. Therefore histopathological diagnosis of Female adnexal tumor of probable Wolffian origin was given.

The tumor cells were strongly positive for pan-cytokeratin and Cytokeratin (CK-7) and were weakly positive for calretinin, inhibin.
Discussion:

The female wolfian duct after giving a ureteric component completely regresses in the third embryonic month. Only remnants may be found in adults such as epoophoron, paraepoophoron and Gartner's duct. They occur mainly in the broad ligament but also in the mesosalpinx, the serosa of the fallopian tube, the ovary and the retroperitoneum.

In the original report describing tumors of this type, all the tumors were located within the leaves of the broad ligament or were attached to it or to the fallopian tube(1). Since then, less than eight pathologically documented cases of probable wolfian tumor of the female adnexal origin and ovary have been reported in English literature(2).

The age of the patients ranged from 18 to 83 years with mean of 50 years. Most tumors were unilateral originated from the broad ligament and some from the ovary or paravaginal tissue. They typically have non-specific clinical manifestations, such as abdominal pain or swelling or asymptomatic masses that are discovered incidentally(1).

In imaging studies, computer tomography shows a well demarcated and multi-lobulated tumor. Ultrasonography shows a hypoechoic and homogenous tumor with a thick capsule(2). MRI findings were reported in a case which showed a well-defined, ovoid mass as well as cyst in the adnexa and no normal ovary was detected(3). Most of these lesions behaved in a benign fashion, but recurrences and metastases have been described and reported(4-5).

Morphologically, the most important differential diagnosis is sertoli-leydig cell tumor. In our case, the lack of leydig cells, absence of hormonal manifestations and characteristic sieve like pattern of tumor cells favor the diagnosis of FATPWO. In addition, sertolileydig cell tumors have never been reported in the broad ligament or other locations where FATPWO is seen. The differential diagnoses of ovarian tumor of wolfian origin (OTWO) tumors include sertoli cell tumor, sertoli-leydig cell tumor, endometrioid adenocarcinoma, carcinoid tumor, yolk sac tumor and metastatic cancers(6-7). In immunohistochemical studies, the tumor cells are positive for cytokeratin (AE1+AE3), alpha-inhibin, calretinin, CD10 and vimentin, but negative for EMA, CK7, CK20, alpha-fetoprotein, ER, PR and chromogranin A10(8). In our case the tumor was strongly immune-reactive for CK-7 and weakly positive for calretinin and inhibin. Although immunoprofile of sertoli-leydig cell tumor and wolfian tumor are quite overlapping, typical histological features of wolfian tumor are helpful to rule out other diagnoses(2). There is limited knowledge about the optimal treatment for this neoplasm. The most common treatment is total abdominal hysterectomy with bilateral salpingo-oophorectomy. Targeted molecular therapy is proposed in patients with C-kit positive FATWO tumors(6, 9). The histological criteria for recurrence or malignancy are not absolute. Patients with FATPWO should be carefully monitored and followed up. The current case had complete surgery performed and has been advised close follow up.
References: