Case Report

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Female Adnexal Tumour of Wolffian Origin: A Rare Case Report

Pathology Section

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ABSTRACT

Female Adnexal Tumour of Wolffian Origin (FATWO) is a rare tumour occurring in adnexal region, arising from the remnants of the mesonephric duct, in places like the broad ligament, fallopian tube, ovarian hilum, peritoneum. Here the authors have reported a case of FATWO in a 47-year-old woman occurring in the wall of fallopian tube and confirmed with immunohistochemistry. Most of these tumours behave in a benign fashion but certain histological features like hypercellularity, cellular pleomorphism and nuclear atypia cause significant confusion in histopathological diagnosis. Multiple differential diagnoses were considered. After careful histopathological examination and thorough investigation with multiple immunohistochemical stains, the diagnosis was ultimately established.

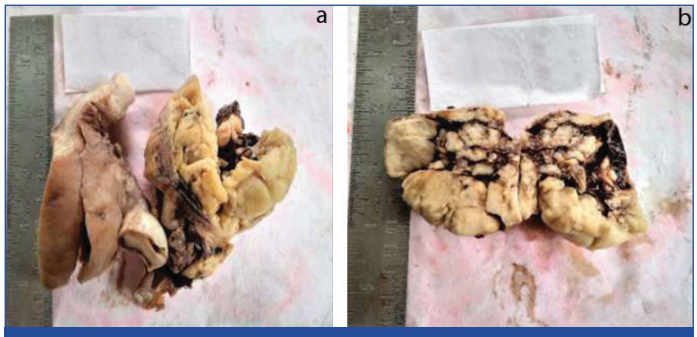
Keywords: Mesonephric duct, Paratubal tumour, Periadnexal tumour

CASE REPORT

A 47-year-old, primigravida, presented with abdominal pain with a lump in right lower abdomen for last six months. There was no significant past medical history. Subsequent pelvic examination and ultrasound reports revealed a 7×5×4 cm solid mass located in the right fallopian tube (picture plate could not be presented, as it was misplaced by the subject and also could not be recovered from the outside laboratory despite best of efforts).

During open hysterectomy a large adnexal mass was noted which was completely separated from her right tube and ovary. There was no vascular pedicle. This was excised along with uterus and cervix. Thereafter, it was sent for pathological examination. Patients's postoperative period was uneventful.

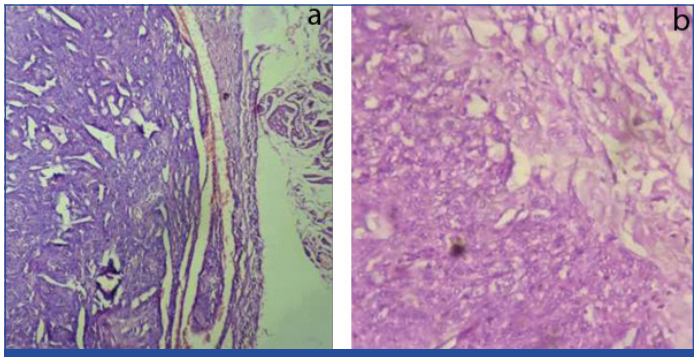
Gross Examination: On gross examination, the size of the mass was 7×5×4 cm, which revealed a well encapsulated smooth solid mass and the cut section of the mass appeared greyish yellow and homogeneous in appearance [Table/Fig-1a,b].



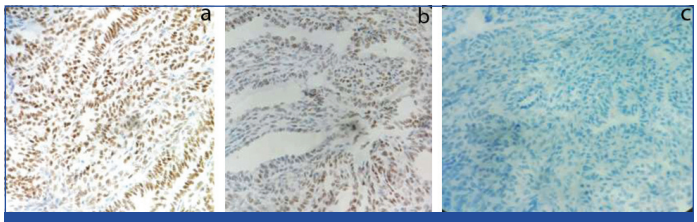
[Table/Fig-1]: a) Uterus, cervix with right adnexal mass; b) Cut surface of the mass.

Microscopic examination: On microscopic examination with periodic acid schiff (PAS) stain, revealed an admixture of solid, diffuse area with a packed tubular pattern [Table/ Fig-2a]. The nuclei of the tumour were uniform and ovoid [Table/Fig-2b]. No significant nuclear pleomorphism and mitoses were noted. In the diffuse area, small tubular structures were revealed by Periodic Acid Schiff (PAS) stain. In histopathological examination differential diagnosis were Adenomatoid tumour, serous and endometroid tumour.

Immunohistochemical staining was performed on formalin fixed paraffin embedded tumour tissue by using standard avidin biotin complex peroxidase technique. The tumour showed diffuse nuclear positivity for Wolffian tumor (WT) [Table/Fig-3a] and focal positivity in CD99 [Table/Fig-3b]. Epithelial Membrane Antigen (EMA), Cytokeratin (CK) 20 and Calretinin were negative. The cells are negative for Paired Box Gene 8 (PAX8) immunomarker, thus differentiating from the commoner serous and endometrial tumours [Table/Fig-3c].



[Table/Fig-2]: a) Solid area with packed tubular pattern (H&E, X100); b) Uniform and ovoid nuclei in the solid area (H&E, X400).



[Table/Fig-3]: a) Diffuse nuclear positivity for WT1 (IHC X100); b) Focal positivity for CD99 (IHC X100); c) Negative for PAX 8 immunomarker (IHC X100).

DISCUSSION

Female adnexal tumour of wolffian origin was first described in 1973 by Kariminejad and Scully as a single tumour [1]. It is a rare tumour occurring in adnexal region, arising from the remnants of the mesonephric duct [2]. The Wolffian duct regresses in the female during embryogenesis. It's remnant can be traced to the ovarian hilum, the broad ligament and the lateral aspect of the uterus and vagina as the Gartner's duct, paroophoron and epoophoron. FATWO is presumed to be arising from these relics of the Wolffian duct. The clinical behaviour of FATWO is generally benign but malignant cases do exist [3,4].

The age of the patients diagnosed with FATWO have a wide age range from 15 to 83 years and most often present with a complaint

of lower abdominal pain and abnormal genital bleeding [5, 6]. Most of the tumours are unilateral. Size of the tumour differs in cases ranging from 0.8 to 25 cm in diameter.

Devouassoux-Shisheboran M et al., described four histological patterns: 1) diffuse solid nest of spindle cells; 2) epithelioid tubular pattern packed, winding, branching and anastomosing tubules; 3) cribriform-sieve like pattern of epithelioid cells; 4) multicystic [7]. The present case corresponds mostly with epithelioid tubular pattern. The main differential diagnoses in histology included broad ligament Granulosa Cell Tumour (GCT), endometrioid carcinoma of the fallopian tube and Sertoli-Leydig cell tumour [8,9].

Endometrioid carcinomas arise from the fallopian tube, whereas FATWO is extratubal and usually arise within the broad ligament. Endometrioid carcinoma frequently presents focal squamous cell differentiation as squamoid morules, which is not present in FATWO. Nuclear atypia is more common in endometrioid carcinoma and has an infiltrating pattern. The above features correspond well with the present case.

In general, FATWO resembles Sertoli-Leydig cell tumours in morphology. However, Leydig cells are absent in FATWO and tends not to have the endocrine symptoms that are features of Sertoli-Leydig cell tumours. Moreover, Sertoli-Leydig cell tumours have not been demonstrated in the paratubal site or in the broad ligament.

It is not always possible to differentiate GCT from FATWO. Both the morphology and immunophenotype of GCT are similar to those observed in FATWO. Nuclear grooving is not an exclusive feature of GCT and can be seen in a variety of other neoplasms and have not been described in FATWO. In the context, of the differential diagnosis between broad ligament GCT and FATWO, the presence of this feature may help to establish the diagnosis of broad ligament GCT. The present tumour lacked the nuclear grooves.

The role of immunohistochemistry in clinching the diagnosis of FATWO has also been studied. FATWO are diffusely immunoreactive for CAM5.2, CK8, CK18, CD10, CK7 and vimentin, while it was negative for Epithelial Membrane Antigen (EMA), S100, Actin, CD 15, Human Bone Marrow Endothelium Marker-1 (HBME-1) and CK20. The present case showed diffuse nuclear positivity for WT1 and focal positivity of CD99 [10,11]. PAX-8 and Calretinin were negative in this case [12,13]. There is currently no entity specific immunomarker for FATWO [14].

CONCLUSION(S)

Although most FATWO behave in a benign fashion, there have been reports of malignant tumours. It is difficult to characterise malignant tumours due to the extremely low number of reported cases. In benign FATWO, the most common treatment modality is total abdominal hysterectomy with bilateral salpingo-oophorectomy unless the patient is young and requiring preservation of fertility. After initial treatment the patient should be appropriately followed-up over a long period. Further research on this tumour is recommended and the long-term prognosis of this entity needs to be determined. Awareness regarding this entity needs to be established.

REFERENCES

[1] Kariminejad MH, Scully RE. Female adnexal tumour of probable Wolffian origin. A distinctive pathologic entity. Cancer. 1973;31(3):671-77.
[2] Ramirez PT, Wolf JK, Malpica A, Deavers MT, Liu J, Broaddus R. Woffian duct tumours: Case reports and review of the literature. Gynecol Oncol. 2002;86(2):225-30.
[3] Lesin J, Forko-Ili J, Plavec A, Planini_P. Management of Wolffian duct tumour recurrence without chemotherapy. Arch Gynecol Obstet. 2009;280(5):855-57.
[4] Wakayama A, Matsumoto H, Aoyama H, Saio M, Kumagai A, Ooyama T, et al. Recurrent female adnexal tumour of probable Wolffian origin treated with debulking surgery, imatinib and paclitaxel/carboplatin combination chemotherapy: A case report. Oncol Lett. 2017;13(5):3403-08.
[5] Steed H, Oza A, Chapman WB, Yaron M, De Petrillo D. Female adnexal tumour of probable wolffian origin: A clinicopathological case report and a possible new treatment. Int J Gynecol Cancer. 2004;14(3):546-50.
[6] Fanghong L, Szallasi A, Young RH. Wolffian tumour of the ovary with a prominent spindle cell component: Report of a case with brief discussion of unusual problems in differential diagnosis, and literature review. Int J Surg Pathol. 2008;16(2):222-25.
[7] Devouassoux-Shisheboran M, Silver SA, Tavassoli FA. Wolffian adnexal tumour, so-called female adnexal tumour of probable Wolffian origin (FATWO): Immunohistochemical evidence in support of a Wolffian origin. Hum Pathol. 1999;30(7):856-63.
[8] Heatley MK. Is female adnexal tumour of probable Wolffian origin a benign lesion? A systematic review of the English literature. Pathology. 2009;41(7):645-48.
[9] Fukunaga M, Bisceglia M, Dimitri L. Endometrioid carcinoma of the fallopian tube resembling a female adnexal tumour of probable Wolffian origin. Adv Anat Pathol. 2004;11(5):269-72.
[10] Seixas A, Tente D, Pinto E, Gonçalves J. Female adnexal tumour of probable Wolffian origin: A rare entity with challenging histopathological diagnosis and unpredictable behaviour. BMJ Case Reports CP. 2020;13(10):e237127.
[11] Nakamura K, Nakayama K, Miura H, Fujiwaki R, Manabe A, Teshima S, et al. Malignant female adnexal tumour of Wolffian origin (FATWO) positive for CD56: A possible diagnostic role for the biomarker. Eur J Gynaecol Oncol. 2014;35(5):580-83.
[12] Tiltman AJ, Allard U. Female adnexal tumours of probable Wolffian origin: An immunohistochemical study comparing tumours, mesonephric remnants and paramesonephric derivatives. Histopathology. 2001;38(3):237-42.
[13] Goyal A, Masand RP, Roma AA. Value of PAX-8 and SF-1 immunohistochemistry in the distinction between Female Adnexal Tumour of Probable Wolffian origin and its Mimics. Int J Gynecol Pathol. 2016;35(2):167-75.
[14] Shalaby A, Shenoy V. Female adnexal tumour of probable wolffian origin: A review. Arch Pathol Lab Med. 2020;144(1):24-28.

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