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Female adnexal tumour of Wolffian origin (FATWO) arising from broad ligament: A rare case report with mutation analysis and review of literature

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Abstract

Female adnexal tumour of Wolffian origin (FATWO) is a rare tumour arising from persisting remnants of mesonephric duct. World-wide less than 100 cases have been reported. Diagnosis of this tumour is mainly post-operative based on histopathological examination and Immunohistochemistry (IHC) markers. We report this rare case in a 48 year old woman. She was diagnosed, pre-operatively, to have left ovarian mucinous cystadenoma by radiology imaging. Post-operatively, histopathological examination reported this tumour as FATWO arising from the left broad ligament. The diagnosis was confirmed by IHC markers. Further mutational studies were also carried out. According to the literature search, this is the third case reported in INDIA, until now. Rarity and varied presentation of this tumour, makes the pre-operative diagnosis difficult. If sufficient numbers of these cases are reported, pre-operative investigations, treatment and follow-up protocols for this tumour can be formulated.

Keywords: Wolffian origin, adnexal tumour, broad ligament, immunohistochemistry, genome study

Introduction

FATWO is a tumour of presumptive Wolffian origin. It mainly occurs within the leaves of broad ligament. It can also occur within the areas where mesonephric remnants are distributed mainly in mesosalpinx, serosa of fallopian tube, ovary and retroperitoneum. Rarity of this tumour and lack of strong recommendation about its work-up renders it difficult to diagnose pre-operatively [1]. Neither definitive treatment nor follow-up protocol is available because of the varied presentation, different age-groups of patients, size of tumour and ranging malignant potentiality.

Case report: 48 year old perimenopausal lady presented to us with history of left sided intermittent abdominal pain since 1 year. Her pain used to subside with analgesics. She had some weight loss which she was not able to quantify. She also had some loss of appetite for past 5 months. Her previous menstrual cycles were regular. Her LMP was 9 months prior to her consultation. She had no other significant clinical history. Clinical examination revealed a thinly built and moderately nourished adult lady with mild pallor. Her abdominal examination showed a large pelvic mass extending to umbilicus occupying more onto right side of the abdomen which was mildly tender on palpation. Pervaginal examination revealed an anteverted bulky uterus, right adnexal fullness and pelvic mass felt upto the umbilicus. Ultrasound showed features suggestive of large pelvi-abdominal cyst likely representing left ovarian mucinous cystadenoma and uterine fibroids. Pelvic MRI revealed features suggestive of Mucinous cystadenoma probably arising from left ovary with enhancing solid components and bulky uterus with multiple fibroids (Fig 1). Ca-125 was mildly elevated. She underwent an uneventful Total Laparoscopic Hysterectomy with Bilateral Salpingo-Oophorectomy and ovarian cystectomy. Intraoperatively, 20x16x8cm complex cyst arising from left broad ligament was noted. Omental adhesions were noted to the cyst. Both tubes and ovaries were found to be normal. Macroscopically, cut section of the tumour showed multilocular cyst containing serous fluid, solid areas, cysts filled with blood. No papillary projections were noted. Solid areas were grayish in color with no evidence of hemorrhage or necrosis (Fig 2). Microscopic examination of the tumor revealed histomorphological features of Female adnexal tumor of probable

Wolffian origin arising from Left broad ligament with no cellular atypia (Fig 3). The pathology blocks were subjected to totally 15 IHC markers. ER, PR, WT-1, CD56, Desmin, SMA showed diffuse strong positivity in tumour cells. CK, Melan A and Calretinin showed moderate positivity in tumour cells. EMA, S-100, CK7, Inhibin, CD10 markers were negative. All these suggested that the tumour is of Wolffian origin (Female Adnexal Tumor of Probable Wolffian origin) arising from left broad ligament. In addition we carried out mutation analysis in paraffin tissue blocks. PDGFRA gene Mutation analysis showed mutation in Exon 12 and no mutation in Exon 14 and Exon 18. KIT Mutation analysis based upon PCR and Gene sequencing did not show any mutation in Exon 8 and Exon 11. Her post-operative recovery was uneventful. She was reviewed at regular intervals. Ultrasound scan of abdomen and pelvis was normal on 45th post-operative day.

Discussion: FATWO also called retiform Wolffian adenoma is rare tumour. World-wide about 80-90 cases have been reported till now [1]. To the best of our knowledge, this is third case report and also first case with mutation study from India.

FATWOs are usually tumours with benign clinical behaviour. However cases of malignancy with extremely invasive patterns are described in the literature. The average age of presentation is approximately 50 years (Range 15-83 years). Clinically often patients are asymptomatic or they may be incidentally detected as an abdominal mass or adnexal mass [1, 3]. Ultrasound is the first imaging tool often used which may reveal an ill-defined mass. The next imaging tool is MRI which may reveal hyperintense mass with cystic degeneration in the adnexa. Differential diagnoses include subserosal leiomyoma or an ovarian tumour and it may be difficult to differentiate these lesions from FATWO by MRI findings. Studies have shown normal preoperative tumour marker levels such as Ca-125 [1, 3]. Accurate diagnosis of FATWO may be made depending on its anatomical site, morphological features and IHC pattern [3]. Microscopic features of well-differentiated epithelial cells with tubular, sieve-like pattern and absence of leydig cells help to distinguish Wolffian tumours [2].

Current scientific literature shows the following IHC pattern in FATWO. Commonly positivity is detected for CD10, cytokeratins. Vimentin, calretinin and inhibin. Uncommonly oestrogen and progesterone receptor positivity is noted. Studies have shown that the proliferative activity (Ki67 IHC) is very low (<5%). Immunohistochemistry performed for CD117/c-kit shows negativity in the tumour cells [4].

Literature review on FATWO shows that a very less number of authors have conducted mutational analysis in their studies. Most of them have studied c-kit gene mutation. Cossu *et al* has carried out mutational studies based on NGS approach on FATWOs. These mutational gene studies have been added as another diagnostic tool along with HPE and IHC. All these mutational studies especially C-kit gene study are done as there is a description of usage of tyrosine kinase inhibitors in the treatment of FATWO in the literature. However, further larger studies are needed for us to incorporate the same in the workup and thereby provide appropriate chemotherapeutic treatment regimens to the patients with invasive tumour [3].

Among the treatment options for FATWO, highly effective option seems to be complete surgical resection, including a hysterectomy and bilateral salpingo-oophorectomy. Surgery through laparotomy has been described widely in literature. Surgery via laparoscopy is also feasible by experienced faculty. Fewer facts are known regarding the tumorigenesis of FATWO

as it is a rare disease entity. Both genetic and epigenetic changes might contribute to FATWO tumorigenesis [2]. FATWOs generally are tumours of benign nature. However, scientific literature reveals cases which are malignant with very high invasive patterns.

Chemotherapy and radiation therapy have been described in these cases with minor advantage. Further studies are required for assessment of role of chemotherapy and radiotherapy in these cases. Literature suggests more benefit in prescribing platinum-based and taxane-based chemotherapeutic agents. Tumour recurrence and metastasis have been reported. Recurrences have been noted as early as 13 months and the median time of recurrence is 4 years. In some cases recurrences have been noted after a long interval following diagnosis [4].

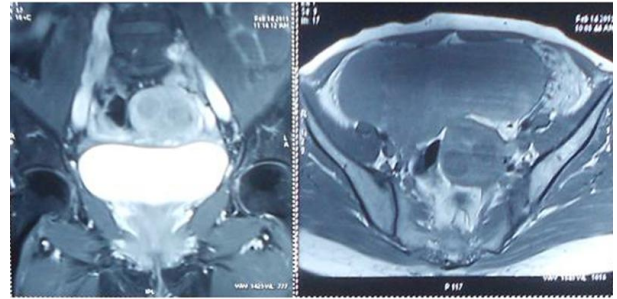


Fig 1: Note the MRI pelvis showing the pelvic mass



Fig 2: Gross specimen of uterus and tumour consisting of cystic and solid areas arising from left broad ligament

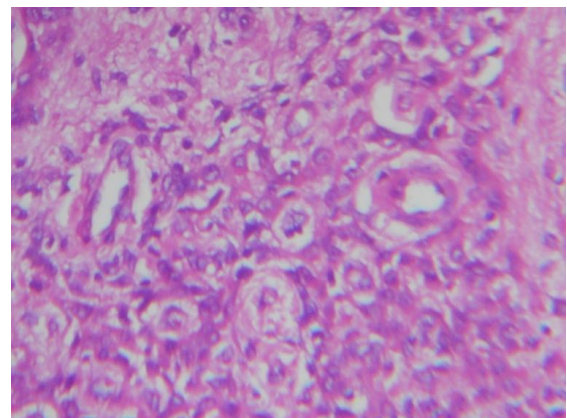


Fig 3: Note the microscopic features of FATWO. H & E Stain. Magnification x 400

Conclusion: FATWO are rare tumours arising from Wolffian duct remnants. With less than 100 cases reported world-wide and only 3 cases in India (including our case), it's difficult to

formulate a definitive diagnostic pre-operative work-up. Complete surgical resection, including hysterectomy and bilateral salpingo-oophorectomy appears to be the most effective treatment modality to prevent further recurrence. Mutation analysis adds as a diagnostic modality. Long term follow-up and further studies are needed in these cases.

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