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Wolffian Adnexal Tumour of Broad Ligament Mimicking as Granulosa Cell Tumour

Sathiya Bama¹, Niranjana Kesav², Radhakrishnan³, Arbind Kumar^{4*}

- ¹Associate Professor of Pathology, I.R.T.PMCH, Perundurai-638053, Tamilnadu, India
- ²Research scholar, Virchow Biopsy Centre, Thanjavur-7, Tamilnadu, India
- ³Consultant pathologist, Virchow Biopsy Centre, Thanjavur-7, Tamilnadu, India
- ⁴Assistant Professor of Pharmacology, I.R.T.PMCH, Perundurai-638053, Tamilnadu, India

*Corresponding author

Arbind Kumar

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Abstract: Wolffian tumour of board ligament of uterus is a rare, but distinctive, epilthelial tumour of wolffian (mesonephric) origin. The tumour has wide variety of histolgical patterns which mimics like Sex cord – stromal tumours like Sertoli and Sertoli – Ledig cell tumours, Granulosa cell tumours as well as surface epithelial serous and endometriod cancinomas. The tumour may present at wide age range from midteens to 80 Years of age with median age of 50 Years¹. Over half of the cases were found incidently presenting as pelvic mass lesion. It is important for the pathologist to know the histological features of Wolffian tumours, in case of huge adnexal mass, to avoid over diagnosis of sex cord stromal tumours expecially granulosa cell tumours, which have high chances of recurrences and needs aggressive platinum based chemotherapy. Though the morphology looks benign, some of the tumours are known for its recurrence and some behaved in a malignant fashion.

Keywords: Wolffian tumour, Sex cord, endometriod cancinomas.

INTRODUCTION

Wolffian tumours of adnexnal origin is supposed to have unspecified, borderline or uncertain behavior. These are very rare tumours and about 70 cases had been reported in the literature.

Various synonyms been used previously and most commonly called as "Female Adnexal Tumours Of probable Wolffian origin" (FATWO). About 10% of the tumours behaved aggressively and these tumours are considered tumours of low malignant potential as they rarely metastased to liver and lungs. Large size, capsule rupture, tumour implants, increased cellularity, nuclear pleomorphism & mitotic activity are features of adverse out comes.

The aim of the present study was to relate a rare case of Wolffian tumour, in a 55 Years old female, presenting as broad ligament fibroid and microscopically mimicking like granulosa cell tumour with review of literature[2].

CASE REPORT

Patient, 55 Years old female, who presented to gynaecology Department of IRT,Perundurai Medical College, Perundurai at Tamilnadu with chief complaints of mass descending per-vaginum and abdominal distension for past 3 Years. On abdominal examination a huge pelvic mass was seen extending upto umblicus,

it was mobile and clinically suggestive of right ovarian mass or fibroid.

Laprotomy done and a huge pelvic mass was seen adjacent to uterus and cevix and was removed with capsule intact. Uterus and cervix, both tubes and ovaries were removed vaginally and sent for histopathological Examination.

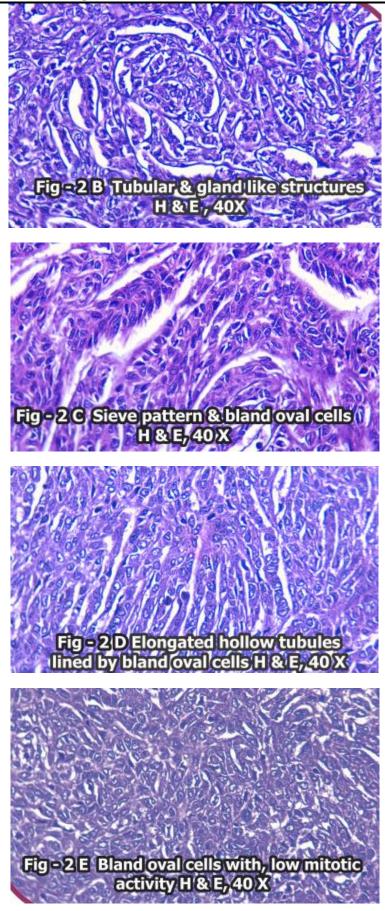
Grossly recieved uterus with cevix, and both tubes and ovaries, with a separetely sent right paraovarian mass. Uterus with cervix measures 8x6x4cm. Cut section endometrium and myometrium were unremarkable, cervix was hypertrophied, elongated and keratotic with evidence of prolapse. Both ovaries measured 3x2x2xcm, cut sections showed tiny cysts, otherwise unremarkable. Both fallopian tube measures 4cm in length and was grossly unremarkable. Right paraovarian mass, received separately with no breach of the capsule. The external surface of the mass showed mild nodularity. On cut section, the tumour was solid and cystic, tan white to tan yellow, with central areas of haemorrhage and cystic change.

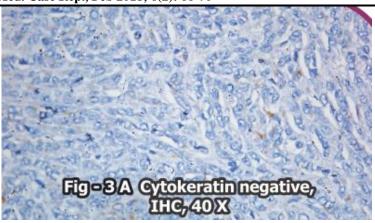
On microscopic examination of the broad ligament tumor revealed a predominantly solid tumor composed of sheets of plump uniform, bland cuboidal epithelial cells growing in sheets, punctured by numerous tubules and gland-like structures with typical "sieve - like" pattern. Cystic dilation was also observed. These cleft like spaces is of varying sizes, resembling well formed hollow tubes and some solid tubules are seen at areas. The tubal lumens and sievelike spaces occasionally contain eosinophilic substance. highly cellular, had The tumour though pleomorphism of cell nuclei . Necrosis haemorrhage is not found. Mitotic activity in cellular areas is <3/10HPF. The tumour was initially diagnosed as Adult granulosa cell tumour as the cells resembled, pale oval cells with occasional nuclear grooves, but characteristic Call-Exner bodies or microfollicles of granulosa cell tumour is not seen.

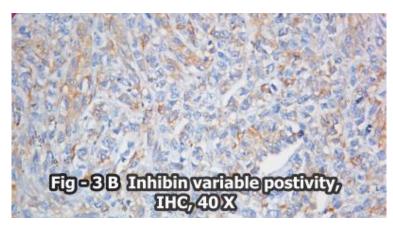
We also had the differential of Sertoli- Leydig cell tumour as it had tubules and cord like structures, but it was ruled out as there are no interglandular Leydig cell like clusters seen. The morphology sieve like/cleftlike spaces resembled the primary serous carcinomas, but the nuclei are bland and because of absent high grade nuclei, absent necrosis and low mitotic activity we rule out serous surface epithelial tumours of ovary. As Wolffian adnexal tumour is a morphological diagnosis and there is a positivity of immunolistochemistry markers of Inhibin, Calnetinin, vimentin, CD 10, Cytokeratin especially CK7 and CK 19 and immunonegativity for EMA, we confirmed the diagnosis of Wolffian tumour based on broad ligament frequent sieve like morphological pattern, diffuse sheets of plump spindle cells with bland cytology and absent classical features of Granulosa cell tumour, Sertoli -Leydig tumour and Serosal surface epithelial tumour.











DISCUSSION

Female adnexal tumor of probable Wolffian origin (FATWO) was first reported in 1973 [6]. Wolffian tumour is a distintive epithelial tumour of Wolffian (meronephric) origin arising from the remnants of the mesonephric duct [3]. It is a rare and has distinctive histological pattern. The tumour is often misdiagnosed to sex cord stromal tumour due to wide histological patterns simulating others pelvic tumours. The tumour was previously called by different names as Female Adnexal Tumour Of probable Wolffian Origin(FATWO), retiform Wolffian adenoma and Wolffian adnexal tumour. The tumour arises in a wide age range from 15 to 80 Years with median age of 50 Years. It is diagnozed as incidental tumours and mostly presenting as abdominal mass.

Most of the female adnexal tumors of wolffian origin behave in a benign fashion. However, there is always a potential risk for recurrence. Surgical excision by total abdominal hysterectomy and bilateral salpingo-oophorectomy at the time of diagnosis may be the best recommended mode of therapy. The role of adjuvant radiation therapy or chemotherapy remains questionable

WATs were unilaterally located in the broad ligament, mesosalpinx, ovarian hilus, and pelvis in areas of rich mesonephric remanats. They showed varying morphologies with solid (spindle cells), tubular (lined by columnar cells), retiform and multicystic (spaces lined by cuboidal and attenuated cells) patterns.

WATs were immunoreactive for wide range of IHC markers such as pan-cytokeratin (AE1/3, CK1), CAM 5.2, cytokeratin 7 (CK7), epithelial membrane antigen, estrogen receptor, progesterone receptor, androgen receptor , inhibin, calretinin, vimentin. Ckit and CD10 and are immune negative to Epithelial mamabrane antigen [4]. Because of immunoreactivity for inhibin and calretinin in a significant number of WATs, our results further show that these immunostains alone do not allow absolute distinction of WATs from sex cord-stromal tumors and adenomatoid tumors, respectively, with which they may be confused.

Our cases did not show significant and widespread nuclear pleomorphic with increased mitotic activity to endorse the diagnosis of a malignant tumor. These tumors are generally thought to have a low malignant potential, since only 20 patients have shown evidence of recurrence. Because few cases have been reported, there are no firm recommendations regarding initial evaluation, treatment, or adjuvant therapy. We report this case of FATWO characterized by diverse features with overlap of morphological features and immuhistochemical markers with sex cord stromal tumor. Our case had bland nuclear features, but owing to large size, the patient needs follow up foe any chance of recurrence and malignant behavior [5].

CONCLUSION

Wolffian tumour of broad ligament of uterus is a rare, but distinctive, epilthelial tumour of wolffian

(mesonephric) origin. The tumour diverse histolgical patterns which mimics like Sex cord – stromal tumours especially Granulosa cell tumours and also as sertoli leydig cell and surface epithelial serous and endometriod cancinomas. These tumors have uncertain malignant potential and known for its recurrence and occasional behave aggressively. The pathologists should be aware of the morphological patterns and avoid over diagnosis of granulose cell tumor and other pelvic tumors.

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