Female adnexal tumour of probable Wolffian origin: a rare entity with challenging histopathological diagnosis and unpredictable behaviour

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DESCRIPTION

A 39-year-old healthy woman, gravida 2 para 2, was referred to our gynaecological outpatient clinic after an incidental sonographic diagnosis of a right adnexal solid mass. Transvaginal ultrasound revealed an enlarged fibroid uterus and a 51 mm nodular paraovarian mass suggesting either a pedunculated or a broad ligament fibroid. Blood tests showed normal serum cancer markers. At that time, the patient was considering getting pregnant again. Consequently, expectant management was decided.

After 2 years, the patient no longer expressed desire to preserve fertility. On physical examination, the enlarged uterus and a solid nodular mass on the right adnexal area were palpable. On transvaginal ultrasound, the right adnexal mass appeared complex, measuring 115 mm×66 mm, with both solid and cystic components, and low vascularity in colour Doppler evaluation (figure 1). Serum cancer antigen 125 (CA-125) level was 67.3 U/mL.

A solid and cystic structure and increased serum CA-125 raised concern for an ovarian cancer, but hypovascularity of the solid aspects suggested a fibroid or another benign tumour, with degenerative cystic changes. In this context, an exploratory laparotomy with intraoperative pathology consultation was decided, confirming a 15 cm×8 cm×8 cm solid and cystic mass. The frozen section revealed a solid spindle cell pattern, favouring a fibrothecoma. Right salpingo-oophorectomy, total hysterectomy and left salpingectomy were performed. The tumour's various morphologic patterns (predominantly solid, tubular and sieve-like) and Wolffian immunophenotype were consistent with a female adnexal tumour of probable Wolffian origin (FATWO): see figure 2 for details.

After discussing management options with the patient at follow-up, and in the absence of established prognostic criteria, no further intervention was planned. The patient is clinically well (4 years post operation).

Female adnexal tumours of probable Wolffian origin, first described in 1973, presumably originate in the mesonephric (or Wolffian) duct.[1] These epithelial tumours are extremely rare, with only around 100 cases published worldwide.[2] FATWOs are most frequently found in the broad ligament, but can also appear in the mesosalpinx, Fallopian tube, ovary, paravaginal region or peritoneum.[3] The median age at diagnosis is 50 years (range 15–87 years).[2 3]

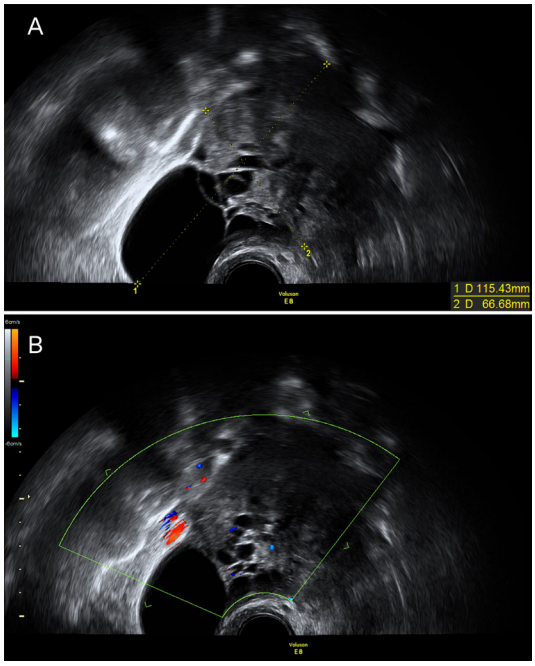

Figure 1 Transvaginal ultrasound showing a complex mass in the right adnexal area measuring 115 mm×66 mm, with both solid and cystic components (A) and low vascularity in colour Doppler evaluation (B).

FATWOs can resemble ovarian endometrioid adenocarcinomas (tubular and sieve-like patterns), Sertoli-Leydig cell tumours (tubular or retiform patterns) or granulosa stromal cell tumours, namely of the fibroma–thecoma group (solid pattern), since they show variable intertumoural and intratumoural morphology (mostly solid, tubular and sieve-like). This challenging diagnosis requires thorough morphologic and immunohistochemical evaluation, since there are no typical morphologic features nor a specific immunostain.[3 4]

These tumours are usually benign, but almost one-fifth of cases show a more aggressive behaviour, with distant metastases and recurrence even long after initial diagnosis.[5] Due to the rarity of this condition—and consequent scarcity of data—its prognostic factors are not clearly established.[6] From the limited information available, prognosis does not seem to correlate unequivocally with either clinical presentation or histopathological features.[3] Consequently, treatment is individualised. Generally, the most adequate treatment is considered to be total surgical resection, with

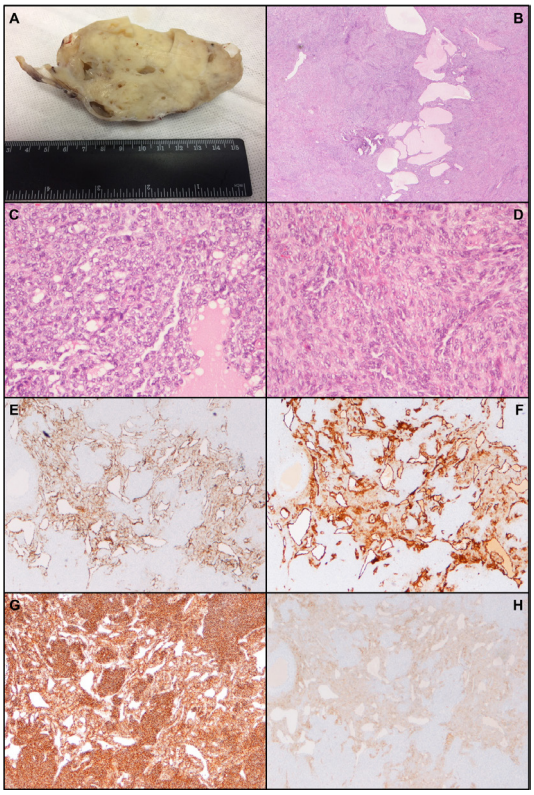

Figure 2 Predominantly solid mass, with cystic areas, yellowish cutting surface and firm consistency (A). The tumour consisted of focal cystic spaces filled with eosinophilic secretions and solid sheets of spindle or oval cells, also lining the cysts; cells had vesicular chromatin, no significant pleomorphism and absent nucleoli (B,C—haematoxylin and eosin, 40× and haematoxylin and eosin, 200×). Occasionally, a sieve-like pattern and packed tubules were found, lined by the same cell types (D—haematoxylin and eosin, 200×). The neoplastic cells were positive for keratins, highlighting areas of retiform pattern (E—Cam5.2, 40×). The cells lining the spaces were positive for calretinin and Wilms' tumour 1 (WT1) (F—calretinin, 40×). Vimentin was diffusely positive (G—vimentin, 40×) and inhibin was positive in areas surrounding the spaces (H—inhibin, 40×). There was no immunostaining for desmin or smooth muscle actin, nor for epithelial membrane antigen (EMA), cluster of differentiation 10 (CD10), paired box 8 (PAX8), or keratin 5 (not shown).

tumour debulking, total hysterectomy and bilateral salpingo-oophorectomy.[3] The role of complementary treatments is controversial.[3] Due to its malignant potential, regular and long-term follow-up is recommended.[6]

Learning points

- Female adnexal tumours of probable Wolffian origin are a rare entity that poses difficulties in terms of histopathological diagnosis and clinical management.
- Despite having a benign behaviour in most cases, they do have some malignant potential. Consequently, long-term follow-up is recommended.
- The diagnosis is extremely difficult in frozen section, given that evaluation of multiple sections and immunohistochemical studies are not possible in that context. Hence, misdiagnosis as a malignant tumour, especially endometrioid adenocarcinoma of the ovary, or as other common tumours can be a pitfall.

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