

Case report

Uterine Sex Cord Tumour- Management Dilemma

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Abstract

Uterine sex cord tumour is a very rare tumour with uncertain management strategies and prognosis. A 61-year-old, nulliparous, who was not on hormone replacement therapy, presented with first episode of postmenopausal bleeding. A transvaginal scan revealed an enlarged uterus with thick endometrial lining and features of multiple degenerated fibroid. Endometrial biopsy was negative for malignancy. Computed tomography of the abdomen and pelvis confirmed the mass, with atrophic ovaries and incidental finding of bilateral hydronephrosis requiring stentings. Otherwise, there were no pelvic lymph nodes enlargement. Our impression was a uterine sarcoma and we decided for total abdominal hysterectomy with bilateral salpingo-oophorectomy. Surprisingly, the histology report confirmed uterine sex cord tumour. There are less cases of recurrence and there is no general consensus on the management. However, we decided for adjuvant chemotherapy (BEP regime) as the malignant cells infiltrated more than half of myometrial thickness, with good outcome.

Keywords: Bleomycin; postmenopausal bleeding; sex cord tumour; uterine tumour.

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Introduction

Uterine malignancy of sex-cord-like elements is extremely rare. It accounts for 0.25% of all uterine neoplasm (1). There were about 48 cases being reported to date (2). Due to its rarity, it possess diagnostic difficulties, treatment options and limited data on the recurrence and disease outcome.

The tumour was first discovered in 1945 by Morehead and Bowman and further evaluated by Clement and Scully in 1976. It is described as epithelial-like or sex cord like elements, often with epithelioid appearance, arranged in nests, cords, trabeculae, solid, or tubular structures. If this element predominates, the tumour is considered to be a uterine tumour resembling ovarian sex cord tumour (UTROSCTs) (1, 3).

Case report

A case of 61-year-old woman, nulliparous, who was not on hormone replacement therapy presented with first episode of post-menopausal bleeding. Otherwise, there was no history of vaginal discharge, constitutional or compressive symptoms. She was not known significant medical or family history. She had an appropriate body mass index, no palpable lymph nodes. A sixteen weeks size uterine mass felt with multilobulated surface, firm in consistency and it was not fixed. Ovaries were not palpable and there were no ascites nor hepatosplenomegaly.

Ultrasound scan revealed an enlarged uterus with evidence of multiple degenerated fibroids, 6 mm endometrial thickness and atrophic ovaries. No ascites

or swelling was noted. Doppler study was not suggestive of malignancy as sarcoma was one of our differential diagnosis. Endometrial biopsy excluded neoplasia. Computed tomography of abdomen and pelvis were performed as uterine sarcoma was highly suspected in this case, and incidentally she was found to have bilateral hydronephrosis, and required stentings.

Total abdominal hysterectomy with bilateral salpingo-oophorectomy were performed. Intraoperatively, no ascites was found with healthy omentum. Multilobulatedly enlarged uterus with presence of cystic areas at 16 weeks size was found. A few small, calcified subserosal fibroids were seen and both ovaries were atrophic.

Microscopically, there was highly cellular tumour forming focal tubular and glandular-like-structures with diffused multiple hemorrhagic and necrotic areas. The tumour composed of generally-uniform spindle shaped cells with vesicular nuclei and prominent nucleoli. The malignant cells infiltrated more than half of the myometrial wall thickness but did not penetrate the serosal layer. Immunohistochemical staining for CK, CK 7, Vimentin and EMA was positive but inconclusive or CD 99. There was no invasion to cervix, parametrial or paracervical tissues. Both fallopian tubes and ovaries were normal. The findings were consistent with malignant sex-cord tumour of the uterus.

A course of four cycles adjuvant chemotherapy consisting bleomycin, etoposide and cisplatin (BEP regime) was given four weeks postoperatively with good result and no evidence of tumour recurrence.

Discussion

Endometrial stromal tumour has two variants which is combined smooth muscle stromal tumour and uterine neoplasm resembling an ovarian sex cord tumour (UTROSCT) (4). The later is extremely uncommon variant and due to its rarity. There is no established protocol, management consensus and prognostication on the disease.

The WHO publication classifies UTROSCT as miscellaneous tumour rather than endometrial stromal tumour group, yet the management strategies remains debatable. The role of radical hysterectomy and bilateral salpingo-oophorectomy in this case was not known but in this case we performed less morbidity approach-non radical surgery, which had promising results.

There is a published case of potential tumour recurrence or even a metastasis especially if the tumours are invasive (5). Therefore, adjuvant treatment namely chemotherapy would be an acceptable modality especially poorly circumscribed borders with possible focal vascular invasion. Even though in some publications reported no recurrence, the time frame was limited and no conclusion could be drawn (5). As this case was extremely rare and there is no established data on this issue, we opted for BEP chemotherapy which she agreed upon, and it was with good acceptance.

Biochemical monitoring for disease recurrence is not established. However, we hope serum inhibin would be sensitive and specific with low false positive value

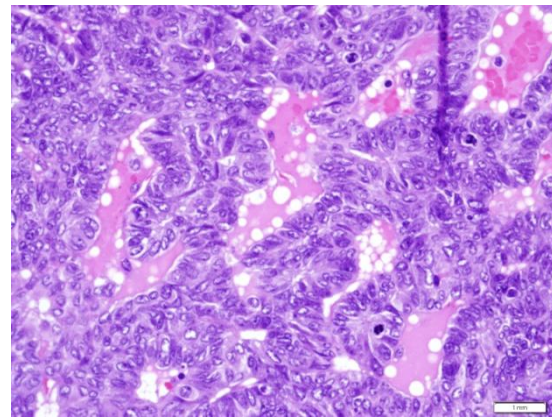
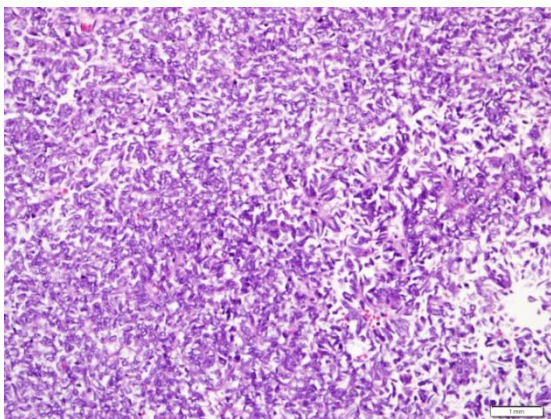


Figure 1: Histopathologic appearance of the tumour; (a) Tubules and cord like epithelial structures of the tumour (b) Foamy tumour cells

albeit no supportive evidence of proof as the variant of the disease unrevealed.

Conclusion

The present case was rare and assumes much importance because of absence of unreported cases of recurrence and no general consensus on the further management and prognosis. We believe adjuvant chemotherapy has a role to reduce risk of tumour recurrence as observed in this case.

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