Rapid Progression of Undifferentiated Uterine Sarcoma Despite Early Stage at Presentation and Prompt Treatment

Dr Kate Mannolini,[1] Dr Yancey Wilson,[2] Dr Hasthika Ellapola[3]

1 Department of Obstetrics and Gynaecology, Albany Health Campus, WA Country Health Service
2 Department of Histopathology, Pathwest, King Edward Memorial Hospital, Perth, Australia
3 Department of Obstetrics and Gynaecology, Logan Hospital, Queensland

Background:

Undifferentiated Uterine Sarcoma (UUS) is a rare mesenchymal uterine malignancy. It is a diagnosis of exclusion after immunohistochemical and molecular cytogenetic features are examined.[1,2] This entity is aggressive and imposes a poor prognosis, with rapid progression of disease despite treatment.[1-4] Due to its rarity, there is limited quality evidence to guide management, with knowledge extrapolated from small case series and the broader high grade uterine sarcoma group.[3-5]

Case:

A woman in her late 70s was referred with vaginal bleeding, lower abdominal pain and urinary symptoms with pelvic ultrasound showing a uterine mass. Pipelle biopsy suggested leiomyosarcoma. She underwent a laparoscopic hysterectomy, bilateral salphingoopherectomy and omentectomy. The disease was staged as International Federation of Gynaecology and Obstetrics (FIGO) 1B. Immunohistochemical analysis lacked specific staining and FISH studies showed no disruption of BCOR or YWHAE genes, therefore the lesion was classified as grade 3 UUS.

Staging imaging noted a lung nodule shown to be concurrent neuroendocrine spindle cell carcinoid tumour on biopsy. On workup, Octreotide and FDG-PET scans 2 months after diagnosis showed new lung nodules with differing avidity and two bony lesions suspicious of metastatic UUS, confirmed on biopsy. The patient commenced doxorubicin monotherapy and palliative radiation to skeletal metastases for pain management.

Despite systemic treatment her disease progressed with a vaginal recurrence 8 months after initial diagnosis whilst on doxorubicin therapy. She received radiotherapy for bleeding control and completed 6 cycles of doxorubicin. She remains alive with active disease at 13 months of follow-up.

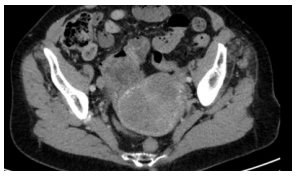

Figure 1: staging CT demonstrating a heterogenous solid lesion in the uterus measuring 8x9x7cm.

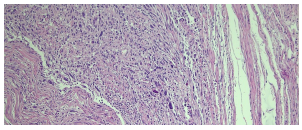

Figure 2: the hysterectomy specimen displayed a myometrial centred pleomorphic spindle cell lesion with infiltrative borders. The tumour was composed of a mixture of spindle and pleomorphic cells with large hyperchromatic nuclei, some showing intranuclear inclusions or prominent nucleoli. Scattered large bizarre forms were noted, along with abundant necrosis and mitoses up to 14 per 10 high powered field.

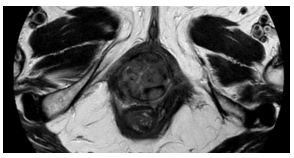

Figure 3: MRI demonstrating 3x4x3cm soft tissue mass with heterogenous diffusion restriction and enhancement.

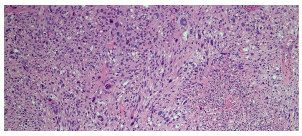

Figure 4: histopathology of vaginal recurrence shares features with the original tumour.

Conclusion:

UUS remains a rare, aggressive, and challenging diagnosis to manage. This case illustrates the rapid, sequential progression of the disease, despite early stage at time of diagnosis and commencement of systemic treatment. Further quality evidence is required to build knowledge around UUS and its treatment to improve poor outcomes.

References:

1. Allanson E, Cummings M, Kuchel A, Wastney T, Nicklin J. Undifferentiated uterine sarcoma: a multidisciplinary challenge. Int J Gynecol Cancer 2022;32:198–202.
2. Mbatani N, Olawaiye AB, Prat J. Uterine sarcomas. Int J Gynaecol Obstet 2018;143(Suppl 2):51–8.
3. Meurer M, Floquet A, Ray-Coquard I, et al. Localized high grade endometrial stromal sarcoma and localized undifferentiated uterine sarcoma: a retrospective series of the French sarcoma group. Int J Gynecol Cancer 2019;29:691–8.
4. Tanner EJ, Garg K, Leitao MM, Soslow RA, Hensley ML. High grade undifferentiated uterine sarcoma: Surgery, treatment, and survival outcomes. Gynaecologic Oncology 2012;127:27031
5. Rizzo A, Pantaleo MA, Saponara M, et al. Current status of the adjuvant therapy in uterine sarcoma: a literature review. World J Clin Cases 2019;7:1753–63.