2021 RANZCOG Virtual Annual Scientific Meeting 15–18 February

A rare case and surgical innovation; Hereditary leiomyomatosis and renal cell carcinoma

Shafiezadeh, Saman[1], Hong Lee[2]

Background

Hereditary leiomyomatosis and renal cell carcinoma (HLRCC) are autosomal dominant syndromes caused by fumarate hydratase gene mutation.

Patients may develop benign uterine leiomyoma, leiomyosarcoma (LMS) and type 2 papillary renal cell carcinoma.

Inadvertent disruption of occult uterine LMS during hysterectomy or myomectomy may result in peritoneal seeding and consequently subject patients to unnecessary adjuvant treatment.

Epidemiology

Hereditary Leiomyomatosis and Renal Cell Cancer affect approximately 200 to 300 families worldwide. The lifetime renal cancer risk in HLRCC is 15%.

Lifelong renal cancer surveillance with annual MRI is recommended by current guidelines.

Case Introduction

A 47-year-old female presented with a history of a new onset stress and urge urinary incontinence, in addition to a sensation of rectal pressure worsening rapidly over a period of 6 months.

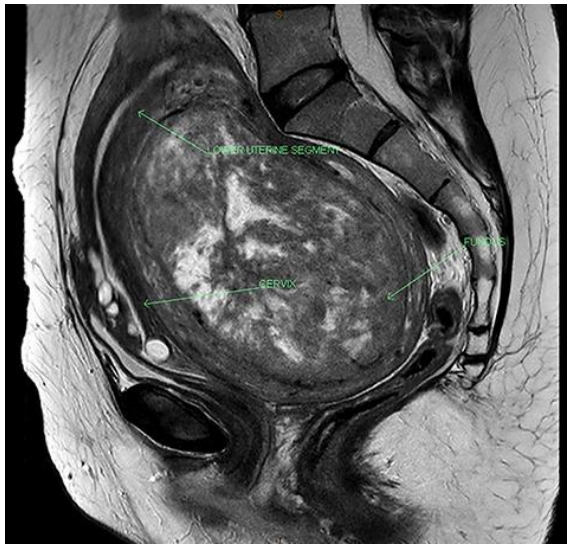

Figure 1; Magnetic resonance imaging of the leiomyosarcoma

Case Details

Pelvic ultrasound revealed a posterior leiomyoma measuring 112 x 72 x 89 mm.

The MRI pelvis showed no evidence of parametrical infiltration nor regional lymphadenopathy.

The patient underwent an uncomplicated total abdominal hysterectomy and bilateral salpingectomy, with ovarian conservation.

A Kiwi® OmniCup was used instead of sharp instruments for uterine manipulation.

The histopathology ruled out LMS but revealed leiomyomata showing a loss of fumarate hydrates expression.

Conclusion

Uterine leiomyosarcoma still remains a rare but possible manifestation of HLRCC.

The obstetric vacuum cup may be used for uterine manipulation to avoid a breach of the uterine serosa and preventing intra-abdominal dissemination of potentially malignant intrauterine tissue.

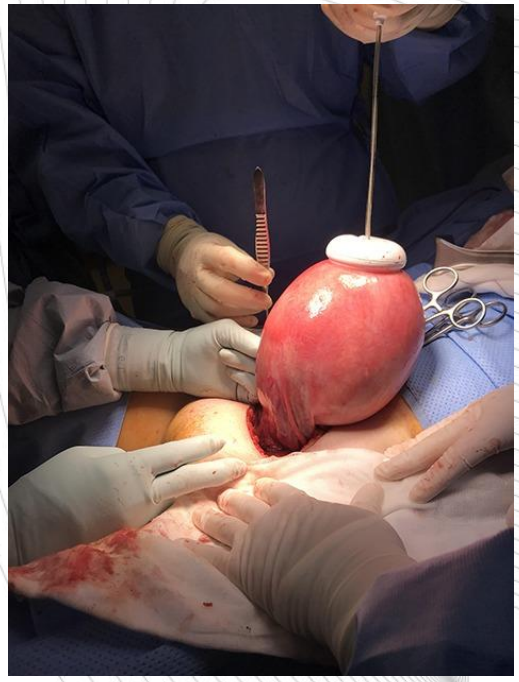

Figure 2; Intraoperative image showing uterine manipulation with Kiwi Omnicup

The Royal Australian and New Zealand College of Obstetricians and Gynaecologists
Excellence in Women's Health

[1] Service Registrar, King Edward Memorial Hospital, Western Australia
[2] Gynaecological oncology fellow, Chris O' Brien Lifehouse

Contact: saman.shafiezadeh@health.wa.gov.au