Mullerian adenosarcoma of the uterus: a rare neoplasm with a need for onco-fertility

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Summary

A 23-year-old nulliparous woman re-presented with menorrhagia and intermenstrual bleeding two years after her first presentation with a similar history. Her initial symptoms were thought to be due to a removed fibroid polyp with histological confirmation. However at the second presentation, following a polypectomy, a diagnosis of low-grade mullerian adenocarcinoma of the uterine body was made. She had total abdominal hysterectomy and pelvic lymph node dissection, peritoneal fluid for cytology with conservation of ovaries to conserve her fertility. No residual tumour was found and lymph nodes were negative. She remains well under clinical surveillance in a multidisciplinary team setting. Different management options that have been used in past reports have been examined and also fertility sparing surgical techniques available for use in successful management of gynaecological cancer are also being explored to shed more light on potential surgical techniques that may be used in treating such rare tumours, particularly in women wishing to retain their fertility.

Key words: Fertility; Mullerian adenocarcinoma; Fertility-sparing surgical techniques.

Introduction

Mullerian adenosarcoma of the uterus is a rare occurrence, particularly in premenopausal women. It comprises benign mullerian epithelium and sarcomatous stroma. There is little data on the ideal management of the condition.

Due to improved cancer treatment, cancer survivors are living longer. Women wish to have children with the hope of improving their quality of life. In a scenario where the patient in question is keen to retain her fertility, the fertility-sparing surgical options available have to be considered.

Case Report

A 23-year-old nulliparous woman presented for the second time with menorrhagia and intermenstrual bleeding two years after her 1st presentation with a similar history. The initial presentation was attributed to a fibroid polyp (confirmed on histology) which had been removed. Other than resolving bulimia and mixed hyperlipidemia, she had no other significant past medical history.

A hysteroscopy and polypectomy following an ultrasound scan finding of an intrauterine lesion was done. Gross macroscopy revealed a gelatinous flesh-like polyp in the upper third of the uterine body while histology returned as low-grade mullerian adenosarcoma of the uterine body.

Subsequently, total abdominal hysterectomy and pelvic lymph node dissection were carried out. Peritoneal fluid was collected for cytology and the ovaries were conserved to protect her fertility. No residual tumour was found and lymph nodes were negative. There was no evidence of extra-genital lesions following imaging studies. The patient is presently under clinical surveillance following a multidisciplinary team review.

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Mullerian adenosarcoma of the uterus is a rare occurrence particularly in premenopausal women, hence the uniqueness of this case.

Discussion

Uterine mullerian sarcoma is a rare subtype of uterine cancer and few cases have been reported in the literature. It is composed of benign mullerian epithelium and sarcomatous stroma [1]. The incidence is unknown and occurs largely in postmenopausal women [2]. The survival rate is unknown but poor prognostic factors have been suggested to include depth of invasion, sarcomatous overgrowth, high-grade malignant features in the stromal component and extra-uterine genital lesions [1]. It presents typically as vaginal bleeding, polyps (recurrent) or simply as a pelvic mass. Management of the condition has been varied as reported in several studies [1-3] and includes hysterectomy plus or minus bilateral salpingo-oophorectomy plus or minus chemotherapy and radiotherapy. Therapy has been individually tailored. As recurrences may occur locally or after long periods of remission following initial conservative treatments (cone biopsy/ trachelectomy), long-term follow-up has been suggested [1].

With sparse available literature, management of these patients in a multi-disciplinary setting could be at best tricky.

Management of this patient was limited to a total abdominal hysterectomy and long-term surveillance in order to preserve her fertility. This management plan was arrived at by taking into account the low-grade nature of the lesion, lack of invasion and extra-genital lesions, and the patients wish to retain her fertility. The procedure was carried out after due counseling, particularly regarding risks of recurrent disease, need for long-term surveillance

questioned. Due to the risk of ovarian metastases following ovarian transposition in women with bulky disease or lymphovascular space involvement, its safety is questionable [18-20].

Secondly, chances of fertility have to be discussed (some series have reported up to a 70% pregnancy rate following radical trachelectomy [21]. Pregnancies have also been reported in women who had undergone ovarian transposition [22]. There is little information regarding the background fecundity. Therefore an assessment of background fecundity and presence of comorbidities must be made as these will be confounding factors.

Thirdly, the risks of obstetric complications such as miscarriages, preterm labour and its sequelae in the fetus delivered before 34 weeks should be discussed with the woman. As there is a significant risk of preterm labour following certain treatments (incidence of preterm delivery in women who have undergone radical trachelectomy has been reported by Shepherd *et al.* to be about 25% [23], adequate counselling has to be put in place.

Conclusion

Mullerian adenosarcoma is a fairly rare tumour which can present at anytime during a woman's reproductive period. In those women still keen to retain their fertility, fertility-sparing techniques or management need to be considered. Oncofertility is an emerging subspecialty which affords women who have survived their malignancies to achieve their dreams of becoming mothers in order to improve their quality of life. Difficult decisions may have to be made following adequate counseling in a particularly rare tumour such as this.

Howewer it is imperative that surgeons are aware of fertility-sparing surgical techniques currently available, indications and limitations according to tumour type, degree of differentiation and treatment, and also possess the ability of relaying these to the patient in an unbiased manner while providing care in a multi-disciplinary setting of adequate expertise.

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