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COMMUNICATION

Laparoscopic excision of fibroid uterine remnants in a patient with Mayer-Rokitansky-Kuster-Hauser syndrome

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Abstract We report a case of laparoscopic excision of fibroid uterine remnants in a patient with Mayer-Rokitansky-Kustner-Hauser syndrome, in whom a previous laparoscopy had confirmed the absence of a uterus.

Keywords Laparoscopy · Rokitansky syndrome · Fibroids

Introduction

We report a case of laparoscopic excision of fibroid uterine remnants in a patient with Mayer-Rokitansky-Kustner-Hauser syndrome, in whom a previous laparoscopy had confirmed the absence of a uterus.

Case report

A 41-year-old woman presented with Mayer-Rokitansky-Kustner-Hauser syndrome. This had been confirmed at the age of 18 by a laparoscopy, which showed an absent uterus and short vagina. She presented to the urologists with the problem of long-standing urinary incontinence. As a part of the investigation an ultrasound scan of the renal tract and abdomen showed a pelvic mass, which was followed up with a CT scan that showed a right pelvic mass measuring 5.8×3.3 cm , suspected to be an ovarian tumour. Tumour markers, including Ca125,CEA,AFP and HCG, were normal. A repeat CT scan done after 2 months showed no change in the pelvic mass and tumour markers remained normal. She remained asymptomatic except for cyclical abdominal pain, which was likely to be related to ovarian function. In view of this, conservative management with regular ultrasound follow-up of the mass was planned at the request of the patient. A repeat CT scan 4 months later showed an increase in the pelvic mass to 6.5×3.9 cm . In view of the increase in size of the mass a laparoscopy was performed, revealing bilateral uterine remnants with fibroids, which were excised laparoscopically. The ovaries were preserved (Fig. 1). At the time of laparoscopy the fibroid remnants were easily mobilised and removed from the pelvic sidewall using the bipolar diathermy (Fig. 2). The histology report has confirmed a diagnosis of leiomyoma. The patient had a satisfactory clinical outcome.

Discussion

Congenital absence of the upper vagina and uterus is the prime feature of Mayer-Rokitansky-Kustner-Hauser syndrome, which, in addition, is often found associated with unilateral renal agenesis or dysplasia as well as skeletal malformations (MURCS association). The pathophysiology is due to a failure of fusion of the two mullerian ducts in the seventh week of embryological development. The two laterally situated, solid, muscular, rudimentary uterine buds connected by midline fibrous bands are formed instead of the uterus. The endometrium is missing. The fallopian tubes and ovaries are normally developed and external genitalia, karyotype, secondary sex characteristics and hormonal function normal. As ovarian function is normal estrogen-dependent pathological conditions can develop, including myomata, neoplasms and endometriosis. The existence of

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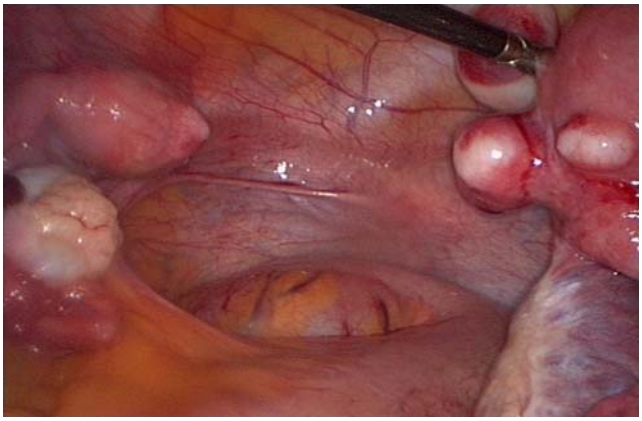

Fig. 1 Multiple leiomyoma arising from bilateral uterine remnants

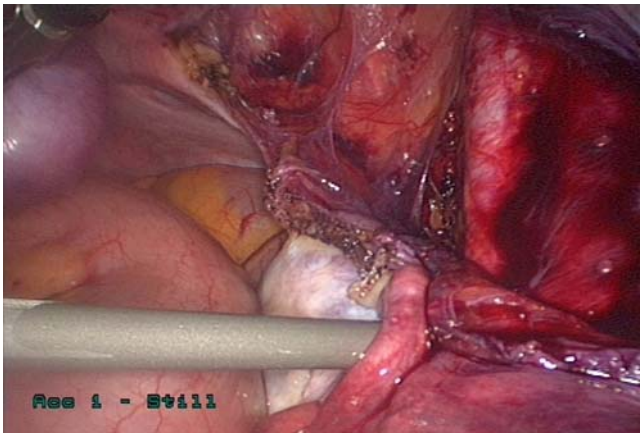

Fig. 2 After laparoscopic excision of the leiomyoma, leaving a normal ovary

fibroids in uterine remnants has been documented in the literature; however, all cases have been managed using the open laparotomy technique [1–6]. Clinicians faced with similar clinical scenarios should be aware that fibroid remnants are a possibility in the differential diagnosis when the patient has a pelvic mass and that the laparoscopic route for removal should be considered.

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