Case Report: Uterine didelphys with a non-communicating left uterus in a 15 year-old girl presenting with dysmenorrhea and a pelvic mass

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Introduction

Müllerian anomalies represent a range of disorders caused by differing degrees of disruption in embryogenesis.[1] Whilst Müllerian anomalies are relatively common, affecting 7% of reproductive-aged women, they can be misdiagnosed as other gynaecologic conditions leading to inappropriate treatment.[1] Didelphys uterus is a class III Müllerian anomaly that occurs when there is complete failure of fusion of the Müllerian ducts. This often results in the formation two uterine cavities, two cervices, and two vaginas separated by a longitudinal septum and with normal menstrual flow.[2] In isolation uterine didelphys does not require treatment but, if associated with cervical agenesis, hysterectomy is usually required.[2]

Case

A 15-year-old female patient was referred to a regional hospital gynaecology clinic with progressively worsening dysmenorrhea for 6 months. She had a history of menarche at 14 years of age with regular 28-day cycles and 5 days of menses. She was not sexually active, and had a history of left renal agenesis. Pelvic ultrasound showed a normal anteverted uterus, a normal right ovary, a bulky left ovary with two complex areas measuring 41 x 30mm and 18 x 16mm respectively, and a thick walled fluid filled structure in the left adnexa 3cm in diameter. Diagnostic laparoscopy was recommended following discussion with gynae-oncology at a tertiary hospital. Laparoscopy at the regional hospital diagnosed a didelphys uterus and non-communicating left uterus, left hydrosalpinx, and a left ovarian endometrioma. Speculum and bimanual examination under anaesthesia found a single vaginal canal, the hymenal remnant present, and a single normal cervix. Post-operatively, the patient was referred for a pelvic MRI and to a tertiary hospital adolescent gynaecology service.

MRI Pelvis revealed dual and divergent uterine horns consistent with uterine didelphys configuration. The right uterus and cervix were normal and functional draining into a patent vagina. There was a functional but obstructed left uterine horn, seemingly due to cervical agenesis, resulting in a left haematosalpinx or endometrioma. Following MRI a laparoscopic left hemi-hysterectomy, left salpingectomy, and left ovarian cystectomy was performed at a tertiary centre. Histopathology confirmed a benign left hemi-uterus with an absent cervix, and the left ovarian cyst was consistent with an endometrioma.

Discussion and Conclusion

Although Müllerian anomalies should be considered in all patients with known renal tract anomalies and dysmenorrhea, they often pose a diagnostic challenge. Additionally, whilst uncommon, the absence of a cervix completely changes management. Cases with unilateral obstruction often are misdiagnosed or have delay to diagnosis because regular menses occurs from the non-obstructed uterus.[3] Non-obstructive anomalies in adolescent patients' do not require surgical intervention, as the benefits of surgical resection do not clearly outweigh the risks.[2,5] In contrast, an obstructive uterine horn, hemi-uterus, or septum should be excised due to menstrual retention and the resulting cyclical pelvic pain, amenorrhea, hematometra, and/or hematosalpinx.[3] As seen in this case, patients also can develop endometriosis from retrograde menstruation. Surgical resection of the obstructed uterus improves endometriosis in most cases.[4]

This case raises the additional considerations needed when managing patients in regional Australia, including recommendations for the most appropriate location to perform definitive surgery. Surgery for obstructive anomalies should be planned utilising a minimally invasive approach.[1] Whilst there is no data specifically pertaining to Müllerian anomalies, it is known that laparoscopic surgical management of advanced endometriosis is effective but must be performed in tertiary centres with multidisciplinary teams readily available.[5] This patient was appropriately referred to a tertiary centre for definitive surgery, allowing for adequate surgical planning and minimally invasive surgery. This case highlights the importance of thorough history and examination, pre operative work up, and multidisciplinary care in managing a rare case of uterine didelphys with unilateral cervical agenesis.

References

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