Case Report

Single kidney eliciting a search for associated genital tract anomaly

Tzung-Hai Yen¹, Ping-Chin Lai¹, Chiu-Ching Huang¹ and Yu Chen²

¹Department of Nephrology and ²Department of Urology, Chang Gung Memorial Hospital, Taipei, Taiwan

Keywords: double uterus; hemivagina; renal agenesis

Introduction

Double uterus with completely or incompletely obstructed hemivagina and ipsilateral renal agenesis is rare [1]. Generally, the condition is not detected until puberty, after menstrual blood accumulates in the obstructed side. The accumulation of menstrual blood then distends the vagina, uterus and fallopian tubes and can even spill into the pelvic cavity. The clinical presentation varies considerably depending on the extent of the obstruction of the unilateral hemivagina and on the location of the opening. The most common clinical presentation is dysmenorrhoea associated with a pelvic mass resulting from a completely hemi-obstructed vagina. Other presentations include pelvic pain, hypermenorrhoea, menometrorrhagia, intermittent vaginal spotting, malodorous vaginal discharge and urinary symptoms. The variable clinical presentation makes diagnosis difficult. Early and accurate diagnosis and management is important for preventing future fertility problems.

Case

A 23-year-old female had enjoyed good health previously. At a routine examination she was found to have a solitary kidney and was referred to us for further investigation. She had suffered from dysmenorrhoea for several years. Additionally, the patient reported intermittent, foul, mucopurulent vaginal discharge and polymenorrhoea with unpredictable staining between menstrual cycles. Physical examination was unremarkable. Laboratory examinations and urinalysis were normal. Renal ultrasound revealed a solitary right kidney of normal renal size and echogenicity. Magnetic resonance imaging and urography confirmed a solitary right kidney. Additionally, two uterine corpuses and two uterine cervixes were seen, compatible with double uterus (Figure 1). Gynaecological examination under anaesthesia found an opening in the partially obstructed small vaginal pouch. The final diagnosis was a double uterus with an incompletely obstructed left hemivagina and left renal agenesis.

Discussion

Embryologically, this complex anomaly probably results from three different developmental failures. Duplication of the female reproductive tract results from lack of fusion of the paired mullerian ducts appearing at the sixth week of embryonic development [1]. The lack of a caudal opening on one side of the duplicated vagina results from the failure of formation of the uterovaginal canal, mesodermal proliferation or the vaginal canalization [2]. The reason for unilateral occurrence of the anomaly is unclear. Finally, because urinary and genital systems are derived from a common embryonic mesoderm understanding a simultaneous defect is easy. With agenesis of one kidney in females, the incidence of associated genital anomalies has been estimated to be up to 50% [3]. The precise aetiology and pathogenesis of the syndrome and its embryologic origin remain unknown. Conservative surgical treatment, that is excision of the obstructing vaginal septum and marsupialization of the blind hemivagina, is considered the most appropriate treatment [4]. This procedure enables evacuation of the sequestered material and preserves reproductive function.
Fig. 1. Magnetic resonance imaging and urography revealed non-visualization of the left kidney in the presumed left renal fossa location. Non-visualization of the left kidney also was found in the abdomen and pelvic cavity. Additionally, there was visualization of two uterine corpuses and two uterine cervixes.

Conflict of interest statement. None declared.

References


Received for publication: 21.8.03
Accepted in revised form: 14.10.03