This is a case report of two patients who have uterus didelphys with obstructed hemivagina and ipsilateral renal agenesis with different clinical manifestations. Progressive abdominal pain after menarche, anuria or obstipation with the existing paravaginal tumor indicates this rare anomaly. Initially, the anomaly remains unrecognized, while patients most frequently referred to surgeons for assistance. The method of choice for diagnosis is clinical examination, ultrasonography and magnetic resonance (MR) imaging. Transvaginal excision of the septum is appropriate mode of treatment.

Key words: uterus didelphys, obstructed hemivagina, renal agenesis, clinical picture

INTRODUCTION

Uterus didelphys with obstructed hemivagina and ipsilateral renal agenesis is a rare congenital anomaly, which is diagnosed in adolescence following the menarche. Due to functional patency of hemivagina and commonly normal menstrual cycles, this type of obstructive anomaly initially remains unrecognized, and patients most often refer to surgeons for consultation and treatment.

Atresia of hemivagina gives rise to unilateral hemocolpos, hematometra and sometimes hematosalpinx. Progressive abdominal pain may be associated with urinary and gastrointestinal tract symptoms.1

Case report

A 17 years old female patient. Menarche occurred at the age of 16. Menstrual cycles were irregular at 30-90 day intervals, with marked dysmenorrheic difficulties. Due to abdominal pain and obstipation lasting for 10 days, she was hospitalized at the General Surgical department of a regional hospital. Ultrasonography showed a mass growth in the pelvis and she was referred to our Institute. On admission gynecological examination revealed a mass on the right side, located paravaginally close to the upper third of the left, patent hemivagina which was painfully sensitive on palpation. Cystoscopy showed no orifice of the right ureter, while intravenous pyelography showed the agenesis of the right kidney. MR imaging of the pelvis verified the anomaly, i.e. uterus didelphys with obstructed hemivagina and ipsilateral agenesis of the right kidney (Fig. 1). Transvaginal excision of the upper third of vaginal septum was performed and about 400 ml of hematic content was evacuated. Postoperative course and long-term follow-up was uneventful.

A 17 years old female patient. She had menarche at the age of 15, with irregular menstrual cycles at 28-60 day intervals, and marked dysmenorrhea. She was admitted to our institution due to anuria, lower abdominal pain, occasionally very intensive, and a pelvic mass diagnosed by clinical examination. Her medical history revealed that, at the age of 6, she was examined by an urologist for frequent urinary infections when agenesis of the right kidney was detected. On admission, clinical examination confirmed paravaginal mass located close to the right upper half of patent vagina. Given the clinical findings and history of agenesis of the right kidney, developmental anomaly of uterus and vagina was suspected. Ultrasonography verified hemihematometra and hemihematocolpos (Figure 2).

Transvaginal excision of vaginal septum was performed and approximately 500 ml of hematic content was evacuated. Postoperative course was uneventful.

DISCUSSION

Uterus didelphys and obstructed hemivagina is an anomaly which usually remains unrecognized at first because of regular menstrual cycles, and due to frequent association with urinary and gastrointestinal difficulties. In the event of paravaginal tumor growth along with ipsilateral renal agenesis and described symptomatology, further
studies are indicated for additional investigation of this rare anomaly.

Diagnosis is based on history, clinical examination, ultrasonography. In some cases MR imaging are required for definite diagnosis. Jurkovic et al. reported significance of 3D ultrasonography for detection of developmental anomaly of Müller’s canals. Good knowledge of normal embryology and sexual differentiation is important for timely diagnosis and appropriate management. The fact that our patients with this type of anomaly had menstrual cycles did not initially direct us to suspect of obstructive developmental anomaly of genital organs. Two patients were initially hospitalized at surgical departments. This type of anomaly is diagnosed most frequently in adolescence. Yet, a case of 10-month old infant with anuria and purulent vaginal secretion has been reported.

Congenital developmental anomalies of Müller’s canals are associated with menstrual difficulties; they are significant etiological factor of infertility and increase the risk of obstetrical complications. Long-term follow-up of reproductive characteristics of 38 patients with this type of developmental anomaly of Müller’s canals within Haddad’s study reported 20 pregnancies in 9 patients were reported. In four patients, pregnancy developed in ipsilateral hemiusuterus on the side of atretic vagina following the resection of vaginal septum. These results suggest that transvaginal excision of vaginal septum is the appropriate therapeutic procedure. It should be emphasized that early diagnostics and surgical treatment are the best prevention of complications that may significantly diminish already reduced reproductive capacity, and sometimes even cause severe psychological problems.

REFERENCES
