Rare Urogenital Malformation Associated with Complex Vascular Malformation – Case Report

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Abstract
Background: Class 3 urogenital malformations are a rarely encountered in the clinical setting. The association with complex vascular malformations represent a challenge in diagnosis and intraoperative management with an enhanced degree of the complexity.

Case presentation: Young female presented to the emergency department with pain in the left lower quadrant and unspecific abdominal symptoms that appear regularly one week prior and during her menstruation. Diagnostic studies and intraoperative findings lead to the diagnosis of a rare urogenital malformation associated with complex vascular malformations. In conclusion the most helpful diagnostic study was the abdominal MRI. Regarding the functional status of the post-operative urogenital tract no clear assessment can be made yet.

Key words: urogenital malformation, vascular malformation, didelphys uterus

Introduction

If we include minor malformations (arcuate and hypoplastic uterus), uterine malformations are very common. They can be observed in 7-10% of all women (1). Complex malformations are often incorrectly identified, treated and reported. Their true incidence is not accurately known. The discrepancy between reports is due to inaccurate diagnostic methods, the lack of a uniform system of classification and because many of these defects are asymptomatic and remain undiagnosed (2). Uterine congenital anomalies are linked with infertility, recurrent pregnancy loss, prematurity and other obstetric complications.
which increase perinatal morbidity and mortality. The poorest viability results are found in the, in order, bicornuate, arcuate and septate uterus (3). We present the case a young woman of child-bearing age with a type 3 urogenital malformation coupled with a complex vascular malformation.

Case presentation

SMI, 18 years old female, without any significant comorbidities presents to the Emergency Department on October 12th 2010, with five days history of appetite loss, nausea, bilious and alimentary emesis, asthenia and pain in the left lower quadrant. The symptoms appeared regularly one week prior and during her menstruation. Significant history: menarche at age 13, regular menstrual cycle (28 days), no prior pregnancies or miscarriages.

Physical examination is inconclusive, soft abdomen with normal respiratory movements, slightly tender in the left lower quadrant, no signs of peritonitis, normal bowel movements, painless loins and normal urine output.

Blood count: WBC 7200/mm³; Hb 13 g/dl; Ht 39.8%; PLT 270000/mm³.

Biochemistry: glucose 99 mg/dl; BUN 32 mg/dl; serum creatinine 0.56 mg/dl; ALT 14 U/l; AST 6.6 U/l.

Abdominal ultrasound: homogenous, isoechoic liver with normal size; normal gallbladder with thin walls, folded and free of stones; portal vein 12 mm; common bile duct 4 mm; long axis of right kidney is 140 mm, with normal shape and echostructure; homogenous pancreas of normal size; spleen 120 mm, homogenous; the left kidney cannot be visualized; normal bladder with thin walls; homogenous uterus of normal size, deviated laterally to the right; a 5 cm diameter round-oval tumor to the left of the uterus, homogenous structure with echogenicity close to that of the uterus, with a central, round, 26 mm anechogenic area. The possibility of an ectopic, malformed kidney is taken into account.

Based on these findings, the patient is admitted to the Department of General Surgery for further diagnostic tests.

Intravenous urography: normal right nephrogram with adequate excretion; left renal agenesis, with large compensatory, right kidney. (Fig. 1)

The patient undergoes a magnetic resonance imaging study (October 18th 2010), that shows:

- Uterus deviated laterally towards the right side, with long axis of approximately 70 mm and 34/38 mm in the transverse and anterior-posterior axes; 11 mm thick endometrium, with homogenous signal; heterogeneous myometrium, presenting small, numerous fibroid nodules of at most 8 mm, at the level of the body and the fundus of the uterus;
- Right ovary with follicular structure (largest of approximately 16 mm), 21/22/40 mm;
- Left ovary is 41/33/28 mm, with multiple small cysts; a cystic tumor of 50/48/46 mm is noted close to the left ovary, caudally and posteriorly, and apparently connected to it by a pedicle; it has with a well defined, smooth and thick wall (largest thickness of 14 mm) with a content suggesting a subacute hemorrhage (T2, T1 hyper signal); contiguously to the described tumor we find a tubular, irregular lesion, with a hypo signal T1, T2 (similar to a fibrous tract) and heterogeneous galindophyia heading caudally and posteriorly from the cystic tumor (towards the vagina); all of the previously described lesions, looks closely related to the left iliopsoas muscle and the left external iliac bundle;
noteworthy we found a large vein emerging from the left femoral vein (right above the left inguinal canal), travelling on top of the left psoas muscle and then parallel to the vertebral column to the level of the renal hilum (L1), where it merges with a similar vein from the right (that collects the left common iliac vein too at the L4 level) (Fig. 2);
- Abdominal aorta has a high division (at the level of the renal hilum – L1) (Fig. 3);
- No enlarged lymph nodes were noticed in the pelvis;
- Small amount of ascitic fluid in the pelvis;
- The absence of the left kidney;
- Large, compensatory right kidney (53/54/148 mm), with normal nephrogram and excretion;

Conclusions:
- Uterine malformation, unicornuate uterus with a left rudimentary horn, a non-communicating cavity and hematocolpos at the level of the rudimentary horn;
- Complex vascular malformation with a high division of the abdominal aorta and a double abdominal venous collector that unites at the level of the renal hilum;
- Congenital, unique, right kidney.

The intravaginal ultrasound and the gynecological examination report the same findings.

On the 18th of October 2010, the patient is discharged, planning to return to the Gynecological Department for a scheduled surgical operation, after her menstrual phase. She is admitted on the 2nd November 2010, and on the 3rd she undergoes surgery following spinal anesthesia (log no. 47/3rd of November 2010). Following a Pfannenstiel incision and access into the peritoneal cavity we found: functional right hemi-uterus with a normal looking right adnexa, left hemi-uterus with hematocolpos, hemoperitoneum, and polycystic left ovary; a needle-biopsy of the left ovary is performed. Because of the long distance between the two hemi-uteri and the impossibility of drainage of the left hemi-uterus the reconstructive procedure was impossible. As a consequence, the ablation of the left hemi-uterus is decided with preservation of the adnexa and drainage of the Douglas pouch (Fig. 4). The post-operative evolution is favorable.

Sectioning of the excised mass showed bloody, black content of about 50 ml.

**Histopathological examination**

Macroscopical description – left hemi-uterus, 7/5/5 cm, thickened walls, fibroids.

Microscopical description – the myometrium consists of hypertrophic smooth muscle cells, some of which present hyaline degeneration with a good blood flow to the stroma and a discrete lymphocytic-plasmocytic infiltration; endometrium presenting glands in the proliferative phase, stasis in the blood vessels of the stroma and focal inflammatory infiltrate of lymphocytic-plasmocytic and granulocytic in nature.

Histopathological diagnosis: diffuse uterine leiomyomatosis; proliferative phase endometrium.

**Discussions**

According to the American Fertility Society (1988) classification (4) the described malformation is a class III malformation. Some rare anomalies reported in the literature (5,6,7) are inconsistent with the generally accepted understanding of Mullerian development and do not fit into the current classification system suggesting an update of the classification could be necessary. The
didelphys uterus results from complete failure of the Mullerian ducts to fuse in the midline.

The case presented has the following particular elements (8):
- The absence of a double vagina;
- The absence of the cervix of the left hemi-uterus (the absence of a double cervix), the left hemi-uterus presenting a rudimentary cervix included in the left parametrium;
- The associated presence of complex vascular malformations, a high division of the abdominal aorta and the double abdominal venous collector that unites at the level of the renal hilum.

The patient’s complaints were specifically related to her menstrual cycle, the increasing amount of blood in the left hemi-uterus (hematocolpos) determined premenstrual (about a week before menstruation) and menstrual pain.

The abdominal ultrasound could not determine precisely if the tumor located left of the uterus had myometrial structure, the diagnoses being determined by MRI studies and confirmed during the surgical procedure. The transvaginal ultrasound was more specific, coming close to the MRI study, without properly determining the relationship with the adnexa.

The prognosis of the case is favorable from the surgical point of view. Regarding the functional prognosis of the remaining hemi-uterus, considering that the adnexa was left in place, gestation can occur, but the outcome of the pregnancy depends upon the functionality of the uterus and is difficult to be assessed at the moment. Raga et all (1997) (2) found a term delivery rate, in the setting of a didelphys uterus, of 20% and the possibility of having a living child at home of 40%. These results showed a poor reproductive performance similar to that of the unicorunate uterus. In fact, fertility depends on early diagnosis and treatment (9).

All lesions of the mesonephric duct are associated to unilateral renal agenesia and utero-vaginal anomalies. If the lesion is distal, blind hemivagina with hematocolpos can be observed (10,11,12); if it is higher, a Gartner pseudocyst can be observed (10,11,12); if it is higher, a Gartner pseudocyst can be observed. If it is higher, a Gartner pseudocyst can be observed. If it is higher, a Gartner pseudocyst can be observed. If it is higher, a Gartner pseudocyst can be observed. If it is higher, a Gartner pseudocyst can be observed.

It would be of great interest to study the potential familial related findings, but, unfortunately there was a lack of data in this field. It would also of interest to study association between anatomical variants of inferior vena cava, renal arteries and urogenital malformation (13).

The associated presence of complex vascular malformations (a high division of the abdominal aorta and the double abdominal venous collector that unites at the level of the renal hilum) is not a cited association with urogenital malformation in the literature.

Conclusions

Patients with uterine malformations have higher rates of reproductive loss, pre-term delivery, breech presentation and complications that increase obstetric intervention and perinatal mortality. Reporting Mullerian anomalies can improve the current classification system and improve the general understanding of embryological development. Clinicians should be aware of every clinical suggestion of genital malformation. Attention to long lasting intra- and postmenstrual dysmenorrhea which does not improve with medication, to metrorrhagia or persistent postmenstrual spotting (malodorous), to endometriosis, especially in youth, to obstetric problems and abnormal presentations.

It is worth mentioning the relatively “old” age of diagnosis (18 years of age) and the difficulties of ultrasound differential diagnosis between a tumour, hematocolpos, ectopic kidney and didelphys uterus (intravaginal ultrasound was more specific). It is the abdominal MRI which established an accurate diagnosis, confirmed intraoperatively.

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References


