Uterus Didelphys with Obstructed Hemivagina and Ipsilateral Renal Agenesis: Ultrasound Findings

UTERUS DİDELFİS, OBSTRÜKTE HEMİVAJİNA VE İPSİLATERAL RENAL AGENEZİ: ULTRASON BULGULARI

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Abstract

To define the sonographic features in uterus didelphys with obstructed hemivagina and ipsilateral renal agenesis.

A 11 year-old girl admitted to our hospital complaining of lower abdominal pain. Physical examination and pelvic ultrasound findings established a diagnosis of hematocolpometra secondary to uterus didelphys with unilateral imperforated hemivagina. Laparoscopy revealed a typical appearance of uterus didelphys, right hematosalpinx and normal ovaries. An incision in the vaginal septum allowed drainage of the hematocolpos, providing relief of the patient symptoms.

This anomaly can be diagnosed with ultrasound preoperatively. Surgical intervention is unnecessary when used only for diagnostic purpose.

Key Words: Uterine anomalies, uterus didelphys, diagnosis, sonography

Özet

Uterus didelfis, obstrükte hemivajina ve ipsilateral renal agenezide sonografik bulguların tanımlanması.

On bir yaşında kız pelvik ağrı şikayeti ile hastanemize başvurdu. Klinik ve ultrason bulguları ile uterus didelfis, obstrükte hemivajina ve ipsilateral renal agenezi tanısı konuldu. Laparoskopide uterus didelfis, sağ hematosalpinks saptandı, overler normaldi. Vajinal septumun insizyonu ile hematokolpos drenajı sağlandı ve hastanın septomları düzeldi.

Bu anomalide tanı preoperatif ultrason ile konulabilir. Cerrahi girişimler sadece tanısal amaçlı kullanıldığında gereksiz olabilir.

Anahtar Kelimeler: Uterus anomalisi, uterus didelfis, tanı, sonografi

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Uterus didelphys with obstructed hemivagina and ipsilateral renal agenesis usually presents after menarche with progressive abdominal pain during menses secondary to hematocolpos. In most cases, diagnosis is reached during surgical intervention although the clinical presentation and imaging findings are characteristic.[1-4] Despite the ultrasonographic (US) findings were defined in many cases, we could find only a few radiological reports about this issue diagnosed by sonography.[1,5-9]

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Our aim to present this case is to define the US findings and discuss the role of the other diagnostic modalities.

Case Report

A 11-year-old girl presented with a history of worsening severe pelvic pain for 2 days of duration. Her menarche had occured 5 months prior to admission with regular menstrual periods. In her past history, dysmenorhea and cyclic abdominal pain were reported. On rectal examination, a large, tender pelvic mass was detected.

Transabdominal US revealed a large, retrovesical cystic mass containing old blood below the double uteri (Figure 1a, b, c). Left sided uterus was normal appearing but righ sided uterus was enlarged and was containing fluid consistent with hematometra. Another cystic mass containing

echogenic fluid and septations were detected on the right adnexial localization consistent with hematosalpinx. Both ovaries appeared to be normal. There was solitary normal left kidney, and the right kidney was absent.

Clinical presentation and US findings established a diagnosis of hematocolpometra secondary to uterus didelphys with unilateral imperforated hemivagina. Sonography guided drainage and vaginal septum excision were planned. However, patient preferred to attend another hospital for social security reasons. We were informed about those vaginoscopy and laparoscopy. Drainage of the obstructed hemivagina was performed by transverse incision and the septum was excised under vaginoscopy. Laparoscopy revealed a typical appearance of uterus didelphys, right hematosalpinx and normal ovaries. Pelvic endometriosis was diagnosed and endometriotic implants of 1-3 mm diameter were fulgurated by electrocautery on laparoscopy. Now the patient is free of symptoms and has no further problems with normal periods for a year.

Discussion

Mullerian duct anomalies affect between 0.1% and 3% of women.[8] In the most extreme form of the mullerian duct nonfusion, uterus didelphys results, with complete duplication of the uterus, cervix and vagina. This anomaly accounts for 11% of uterine malformations and is typically asymptomatic. Uterus didelphys is most often recognized as part of a syndrome associated with an obstructed hemivagina and ipsilateral renal agenesis. The obstruction of one hemivagina will block outflow and the creation of a cystic mass, leading to complications such as hydrocolpos, hematocolpos, hematosalpinx. There is often a delay in diagnosis compared to the complete obstruction (like imperforated hymen or transverse vaginal septum) due to the fact that the patients with the obstructed hemivagina have periods.[10] The right side is 2 times more likely to be affected than the left.[6]

The triad of common symptoms seen in these patients are:

1- Dysmenorrhea that begins shortly after menarche, 2- Increasing severity of dysmenorrhea

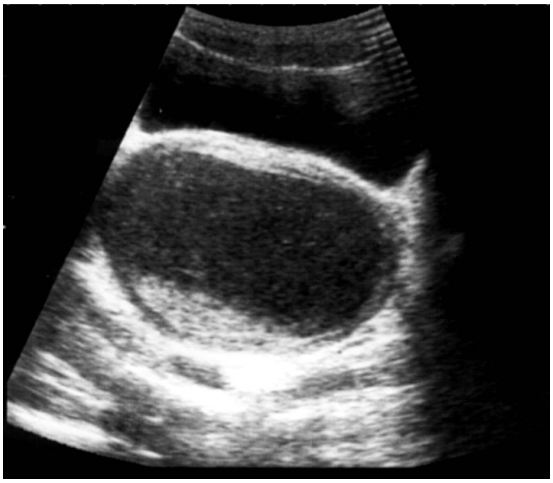

Figure 1a. Coronal image from a transabdominal pelvic sonogram demonstrating a large, retrovesical cystic mass representing the hematocolpos.

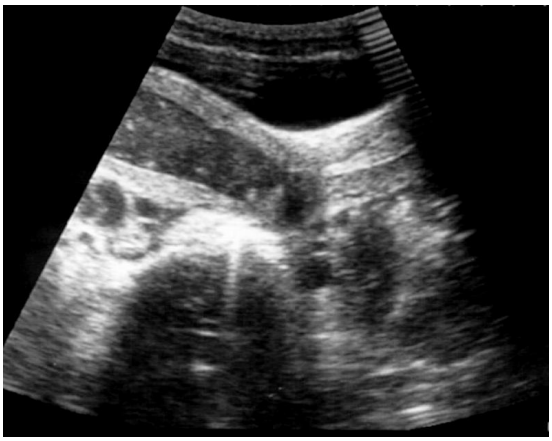

Figure 1b. Coronal image from a transabdominal pelvic sonogram demonstrating the double uterus with enlarged, fluid-filled right uterus and normal left uterus.

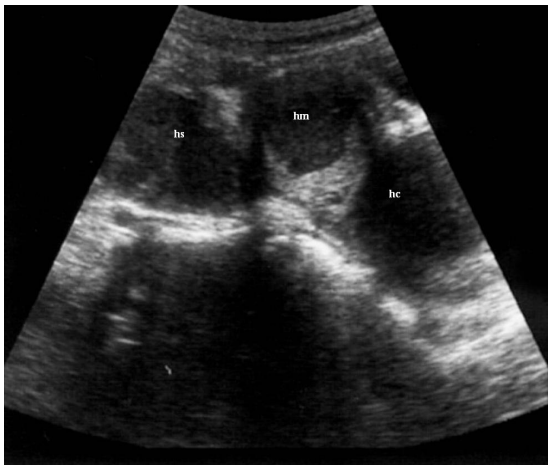

Figure 1c. Coronal image from a transabdominal pelvic sonogram demonstrating structures representing hematometra (hm), hematocolpos (hc), hematosalpinx (hs).

with each subsequent period and 3- A unilateral pelvic mass.[3,4] Occasionally, patients present with symptoms from the urinary tract, such as acute urinary retention, dysuria, pollacuria, gastrointestinal complaints such as rectal pain and constipation, signs of acute abdomen.[4,5,8]

Diagnostic findings of US examination are double uterus and unilateral pelvic cystic masses. The mass, related to dilated hemivagina, is located in retrovesical region and it is visualized as a big sized, thick and smooth walled, anechoic or echogenic lesion.[7,9] If there is hematometra, related side of uterus becomes larger, myometrium gets thinner and this condition is visualised as a secondary cystic mass. The continuation of those two masses with each other and the bilobed view are typical sonographic findings.[1,8] In some cases, another cystic mass related to dilated tubal structure in ipsilateral adnexial area is seen such as in our case. Ovaries may be in normal or cystic appearance due to hemorrhage. Sometimes myometrium in obstructed side gets extremely thin and it may be impossible to distinguish dilated uterus and cystic adnexial mass from each other.[11]

While the US findings are typical, in some cases the diagnosis may be difficult when the enlarged part of the genital canal compresses the normal part. In this cases, differential diagnosis of other obstructive anomalies like isolated vaginal septum or imperforated hymen could be considered. But the clinical presentation of these particular cases are distinct (primary amenorrhea). Imperforated hymen can be diagnosed by physical examination. Moreover, imperforated hymen and transverse vaginal septum are not associated with other urological anomalies.[12]

Magnetic resonance imaging (MRI) is the most reliable and noninvasive modality for evaluating Mullerian duct anomalies.[10,13,14] It might provide an accurate preoperative evaluation since it describes the length and level of the vaginal septum, structure of the soft tissues more properly than the others. It is also useful in searching coexisting urinary anomalies which include renal agenesis, hypoplasia, dysplasia, ectopic ureters and Gardner's duct cyst. Although MRI is considered the gold standard for diagnosis, many authors agree that the US examination does deliniate what we need to see. MRI is recommended for further evaluation if results of US are not definitive.[12,13]

Computed tomography may be helpful in the diagnosis of the condition and associated urinary anomalies.[7,14] However it is mostly not preferred because of radiation exposure.

Laparoscopy and laparotomy are expensive, invasive procedures associated with risk from anesthesia and surgery. These reveal not only the type of uterine malformation but also the presence of pelvic endometriosis and adhesions caused by hematometra, hematosalpinx or ascending inflammatory processes.[2] Some surgeons perform laparoscopy with surgical treatment of any endometriosis seen. Others simply relieve the obstruction, avoid an intra-abdominal procedure, and follow the patient clinically.[10]

In our case, the diagnosis was realized in the light of characteristic clinical and US findings. Although laparoscopy had no advantages in the diagnosis, it provided the treatment of endometriosis and adhesions. But, the necessity of this intervention is controversial.

In conclusion, since sonographic imaging is an appropriate, easily available, noninvasive, nonexpensive technique and can provide valuable diagnostic information, surgical interventions may be unnecessary when used only for diagnostic workup. We believe that the ultrasound should have been sufficient in this case.

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