UTERUS DIDELPHYS, OBSTRUCTED HEMIVAGINA AND IPSILATERAL RENAL AGENESIS, 2-CASE REPORT.

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ABSTRACT

Two female teenagers with progressive dysmenorrhea were sent to tertiary care unit for further management of the pelvic masses which erroneously given preliminary diagnosis to be endometriotic disease and pelvic abscess. Following thoroughly review of clinical data and imaging studies, the patients showed to have double uterus, obstructed hemivigina and absent right kidney. The pelvic mass, thus, represented hematocolpos and hematometra. Resected vaginal septum is the treatment of choice and should be performed earlier, to prevent gynaecologic complications and improve reproductive outcome. Non invasive imaging techniques may have the important roles in the diagnosis of intrapelvic mass in female adolescence with absent kidney which could be the consequence of developmental uterine anomalies.

INTRODUCTION

The definite association of uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis was documented in the literature since 1922 by Purslow.[1] Rock and Jones found a more common occurrence of this complex anomalies on the right side.[2] Uterus didelphys with obstructed hemivagina is the extreme spectrum of defective fusion of paramesonephric (Mullerian) duct and derivative thereof. Because of accompanying growth with the mesonephric (Wolffian) duct, renal anomaly always comes about together. Both cases had been misdiagnosed by referring physicians to be endometriotic diseases and pelvic abscess and were sent for further management.

2-CASES REPORT

CASE1: 15 year old female with lower abdominal mass

This female adolescent experienced dysmenorrhea since her first menstruation. The symptom became progressive and brought her to a private hospital. Initial transabdominal US exhibited two right adnexal masses with absence of right kidney at the native site. She was further investigated by intravenous urography (fig.1) and abdominal CT scan (fig.2,3). These were concluded to be right endometriotic cyst and right pelvic kidney. The patient was sent to tertiary care hospital for further treatment. Following transabdominal US by gynecologic ultrasonologist, it was found that these two right adnexal masses could be hematocolpos and hematometra (fig.5,6). No right kidney was found. Subsequent vaginal exam during narcosis noted that she had obstructed right hemivagina. The patient received surgical treatment by vaginal septum resection for ample drainage of hematocolpos and exposing the cervix. Final diagnosis was uterus didelphys with obstructed right hemivagina and ipsilateral renal agenesis.

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CASE 2: 12 year old female with right sided abdominal pain

The patient was referred from a general practioner clinic for a suspicion of pelvic abscess. She had a history of severe dysmenorrhea since the menarche and last more than one day. On her initial transabdominal US, a well demarcated hyperechoic mass was shown. This was contiguous with uterus which appeared to be hydrometra and markedly deviated to the right side. A smaller left sided uterine-like structure was also found. Further lower abdominal CT scan was performed and obviously showed the obstructed right vagina causing hematocolpos and hematometra (fig.6,7). This patient also had absent right kidney as well (fig.8). All findings were supportive for the diagnosis of uterus didelphys with obstructed right hemivagina.

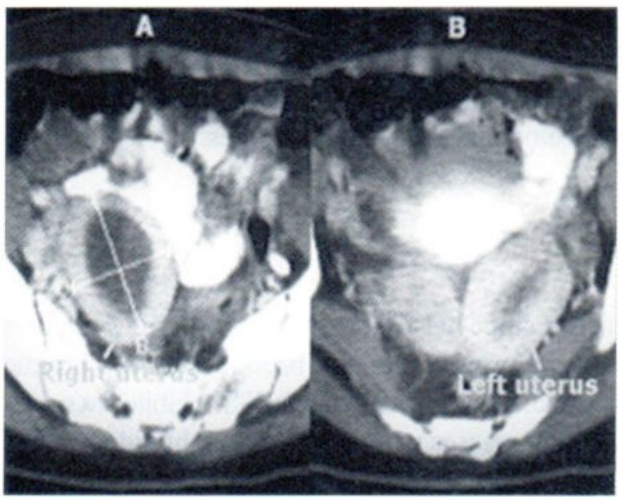

Fig.1

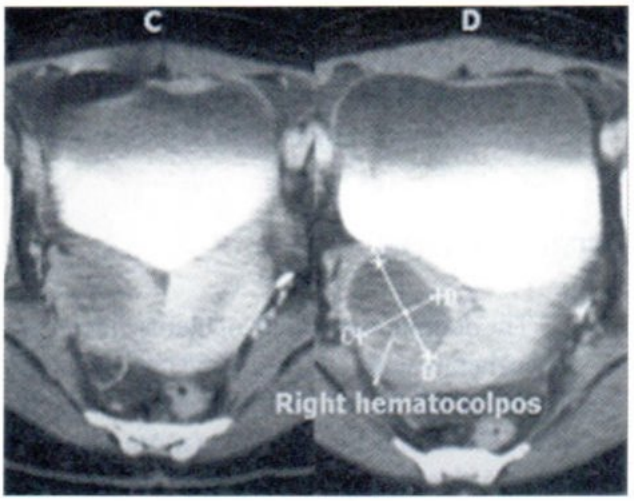

Fig.2

Fig.1-2 Sequential slices of the 15-year-old female with uterus didelphys and obstructed right hemivagina clearly unveil hydrometra of right sided uterus(A), smaller nonobstructed left sided uterus(B) and right sided hematocolpos(C).

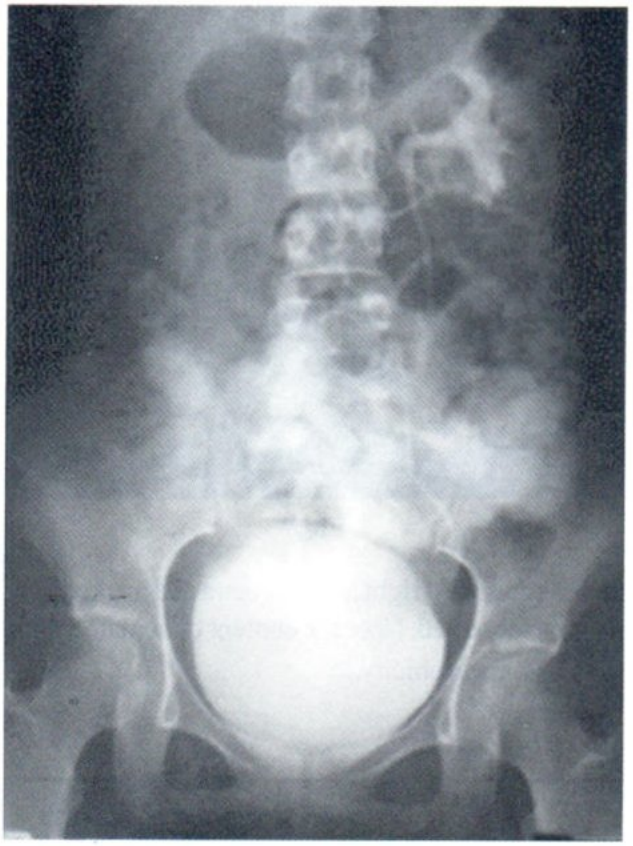

Fig.3 Intravenous urography of the 15-years-old female shows no right renal pyelogram. Opacity in lower abdomen from previous oral contrast administration for previous abdominal CT is noted.

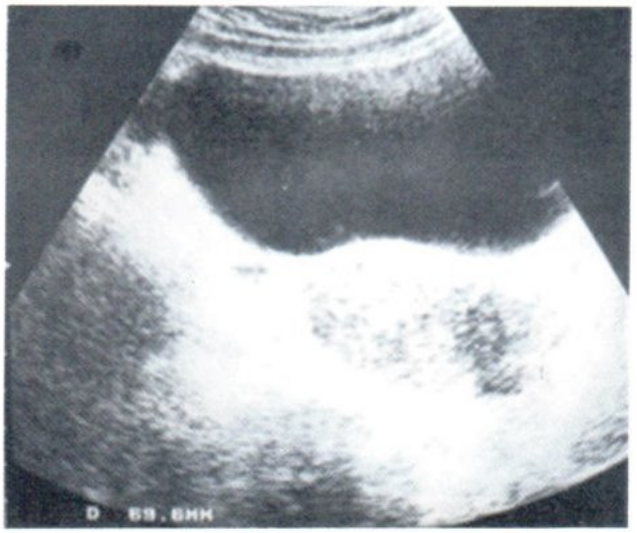

Fig.4 Left oblique sagittal scan shows left sided uterus.

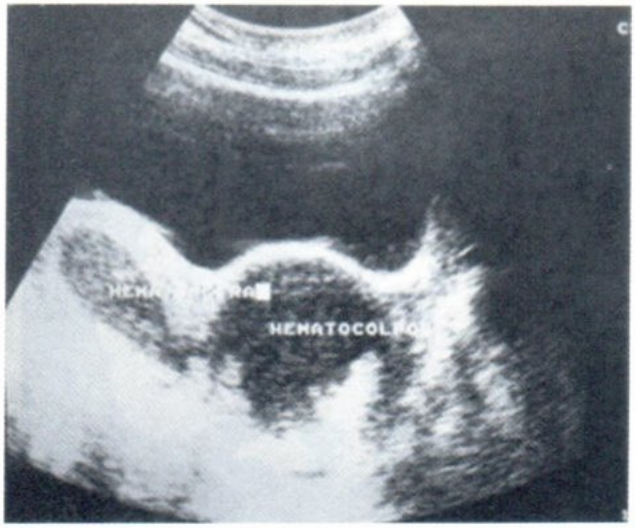

Fig.5 Right oblique sagittal scan shows hematometra and hematocolpos of right sided uterus and obstructed right hemivagina.

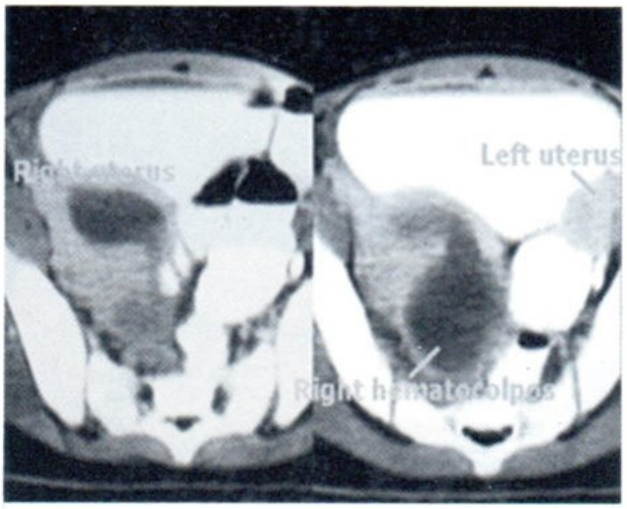

Fig.6

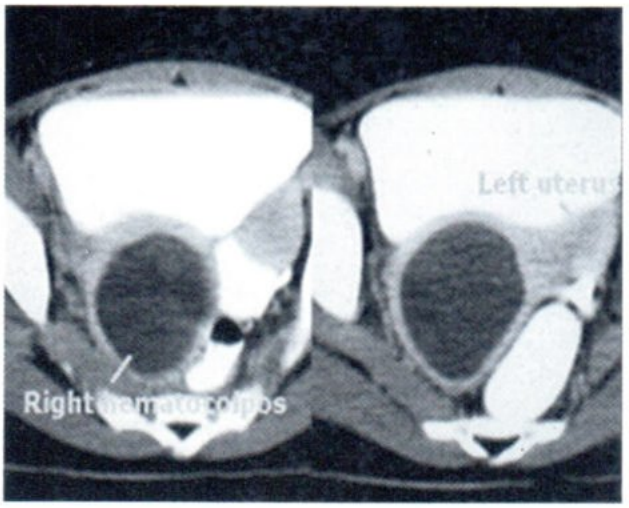

Fig.7

Fig.6-7 Lower abdominal cuts of the 12-years-old female shows mild right hematometra and marked right hematocolpos. Smaller left uterine-like structure is also observed. Note the content of hematocolpos appeared clearer than the first patient, suggestive of less chronicity.

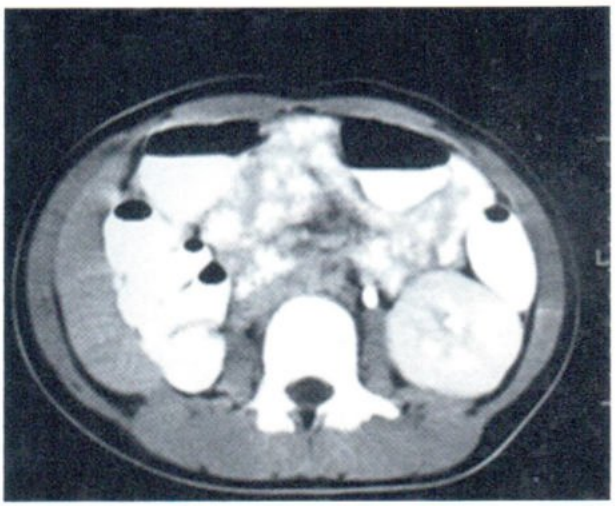

Fig.8 Higher level cut of the 12-years-old female shows absence of the right kidney in the right renal fossa.

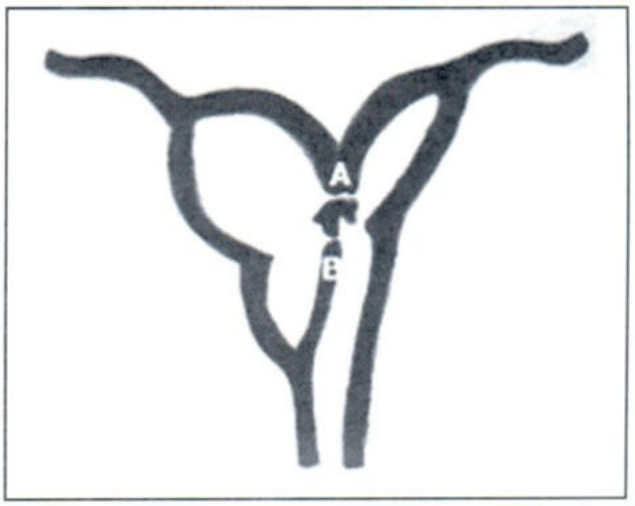

Fig.9 Diagrammatic illustration for both patients' mullerian anomalies shows association of uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis. Communicated sites between the two uteri may occur either above the internal cervical ora (A), or anywhere at the level of the vagina (B).

DISCUSSION

The clinical syndrome comprising of double uterus, obstructed hemivagina and ipsilateral renal agenesis, is suggestive of an embryonic arrest occurring between 8th and 12th week of pregnancy that simultaneously affect the Mullerian and metanephric ducts.[3] Patients may be asymptomatic until menarche. Obstructive symptoms may present with progressive dysmenorrhea, lower abdominal mass or chronic vaginal discharge depending on degree of obstruction and presence of communication connection the right and left sides of Mullerian system at the level of vagina or above internal cervical ora (fig.9).[4]

Manifestations of the fusion defect ranges from uterus didelphys to arcuate uterus which uterine fundus is somewhat indented in the center. The most common form is bicornuate uterus in which the uterus has two cornua connected at their caudal ends.[5] Fusion failure also results in longitudinal vaginal septa which can cause a complete double vagina and found in up to 57% of cases.[6,7,8] If one of the vaginal outlet is not utterly developed, obstructive symptoms of hematocolpos and hematometra will occur. Longitudinal vaginal septum is very common in the case of uterus didelphys as we encountered in these reported cases.

Double uterus with blind hemivagina may be the consequence of damage to the caudal portion of the Wolffian duct. From embryologic viewpoint, while the cephalic portions of Mullerian and Wolffian ducts are divided by mesenchyma, more caudally the anatomic relations become closer; however they continue to present different morphologic pictures of interactions between epithelium and interstitial tissue which brought about the differentiation of the two ducts.[9] These findings, according to which development of the Mullerian duct may be inhibited by a defective Wolffian duct, support evidence that the former duct uses the latter duct to lead its descent toward urogenital sinus.[10] Hence, the frequent associated urinary tract anomaly is detected.

Awareness of this syndrome should be lead to its prompt diagnosis. This in turn should allow for early and suitable surgical management for decreasing the complications such as hematosalpinx, pyosalpinx, pyocolpos,[4,11] endometriosis,[12] intrapelvic adhesions[4] and can improve reproductive outcome.13 If proper treatment has been considerably delayed, hemihysterectomy with or without salpingo-oophorectomy may become necessary.

In view of the fact that the age group of the presented patients was teenage. Vaginal exam or other invasive methods e.g. hysterosalphingography may be prohibited. Non invasive imagings are more favoring in abnormality detections. Nowadays, transabdominal US is frequently the method of choice. If this provides insufficient anatomical details, CT may be more informative. Appropriate imaging procedures would save the patient from unnecessary further mostly invasive examinations and/ or surgical interventions e.g. laparoscopy or hysteroscopy. Even though, no matter what imaging performed, basic knowledge of developmental urogenital system is needed.

SUMMARY

Radiologists ought to concern hematocolpos or hematometra from developmental Mullerian anomaly in the differentials diagnosis of the young adult female pelvic masses. And if this is inconspicuity, additional detection of the associated anomalies, in particular, the urinary tract must be done. One must know that there may have been other Mullerian anomalies that did not come to light; that is because of many woman have no leading symptoms as mentioned above.

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