Unilateral hematometrocolpos associated with double uterus

N. LANG - H. STARK - G. OHEL

Summary: The case of twelve year old girl with double uterus, unilateral vaginal obstruction and ipsilateral renal agenesis is described. Excision of the imperforate vaginal septum resulted in drainage of the hematometrosalpingocolpos and complete disappearance of the large pelvic-abdominal mass. Such a mass in normally menstruating women may detract from the true diagnosis. Awareness of this relatively rare condition is the mainstay of prompt diagnosis which will prevent unnecessary and destructive surgery.

Key words: Double uterus; Hematometrocolpos; Unilateral vaginal obstruction.

INTRODUCTION

The presence of a double uterus does not necessarily cause clinical symptoms. However, an additional vaginal septum gives rise to symptoms of menstrual outflow obstruction. This combination of vaginal septum with failure of fusion of the Müllerian duct may obscure a diagnosis which otherwise would have been obvious. The following report describes such a case.

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Department of Obstetrics & Gynecology
Poriya Government Hospital
M. P. Hagalil Hatachton
Code 15208, Israel

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CASE REPORT

A healthy 12 year old girl presented with abdominal pain and rectal pressure of three weeks duration. Menarche had begun six months earlier. Menstruation in the last three months had been accompanied by severe dysmenorrhea.

Physical examination revealed a cystic abdominal mass, arising from the pelvis and slightly to the right, reaching some 7 cm above the symphysis pubis.

Preliminary inspection of the genitalia demonstrated normal vulva and intact hymen. Rectal examination disclosed a large paravaginal mass in continuation with the abdominally palpable mass. Pelvic sonography demonstrated a large cystic mass, 6.1×7.2 cm with turbid content (Fig. 1). The ovaries seemed normal.

Transrectal sonography confirmed the presence of the cystic mass (Fig. 2). Its contents described as fibrinolytic blood, pus or some other mucinous fluid. Computerized Tomography defined two masses, the lower and larger one communicating with a smaller mass, above and to its right (Fig. 3).

A diagnosis of double uterus and one sided obstruction of the vagina was strongly suspected on clinical grounds. Despite re-evaluation of the tomography films, the diagnosis could not be definitely confirmed. Nevertheless, the larger

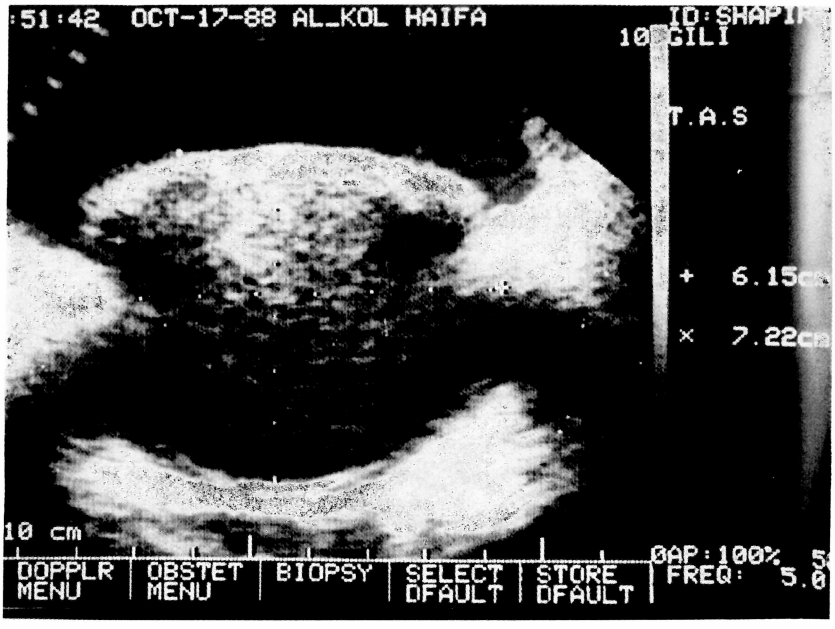

Fig. 1. — Transabdominal sonography showing a large cystic mass – hematometra.

mass was interpreted as possibly being a large hematocolpos and the smaller mass a hematometra and hematosalpinx.

Intravenous pyelography disclosed right renal agenesis and normal left kidney and ureter.

In the theater and under general anesthesia a vaginal examination disclosed cystic bulging of the right vaginal wall. The bulging was continuous with the abdominal mass, its lower border being 3 cm above the introitus.

Puncture and aspiration through the right vaginal wall, confirmed the diagnosis of hematocolpos. The vaginal septum was excised leaving a wide margin. About half a liter of old blood was drained. Following that, two cervices were clearly observed and two uteri palpated. The right uterus was slightly enlarged.

The post operative course was uneventful. Repeat sonography one month later showed two normal uteri.

DISCUSSION

Müllerian anomalies are not often encountered by the gynecologist. More specifically, the unique syndrome of double uterus (a unilaterally obstructed vagina and ipsilateral renal agenesis) is quite rare.

Previous reports ([1, 3, 5, 6, 8, 10]) have described the clinical features of this entity: the mean age at diagnosis is 18, with a range of 11 to 43 years. The diagnosis is often made within a few months of menarche, but may be delayed a few years. The presenting symptoms vary and include progressive dysmenorrhea, rectal pressure, abdominal or back pain and dyspareunia. Menstruation is regular and normal. In various cases of incomplete obstruction there may be intermenstrual brown or foul discharge, the passage being either through a small defect in the septal vaginal wall or through a lateral communication between the double uteri ([8]).

The importance of correct preoperative diagnosis was stressed long ago. A mistake in evaluation might lead to unnecessary operative procedure, which may impair future fertility ([7]). The difficulty in diagnosis arises mainly because of the normal menstruation. The associated abdo-

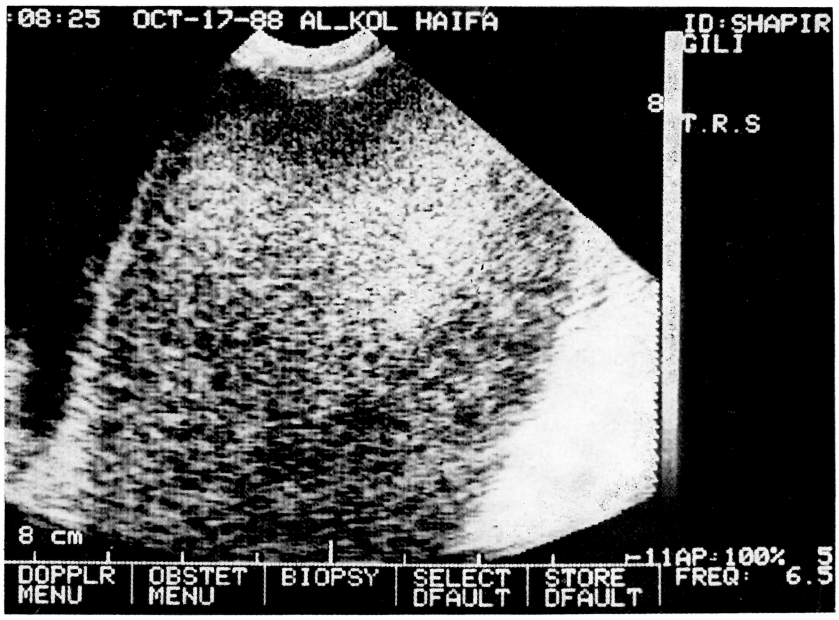

Fig. 2. — Transrectal sonography showing the mass between the rectum and bladder.

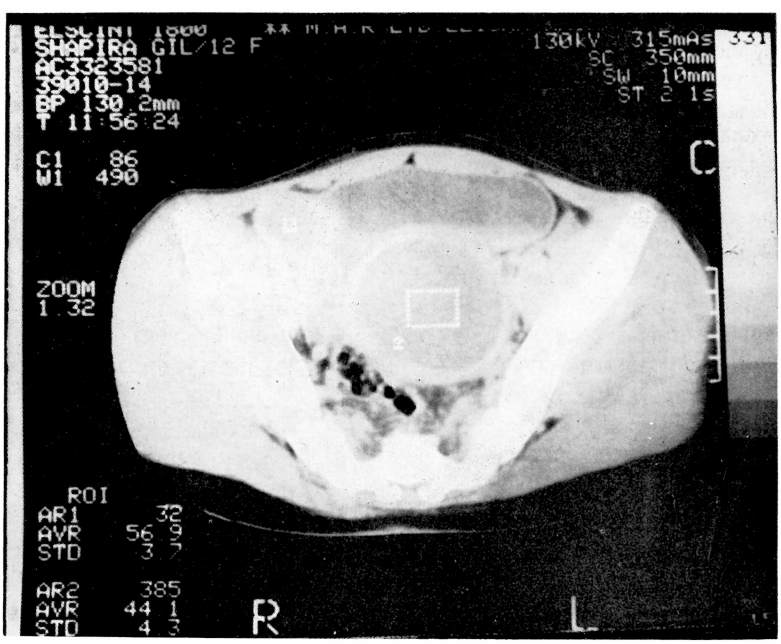

Fig. 3. — CT of pelvis disclosing two masses. The lower is the hematometra and the smaller, to the right, is the hematosalpinx.

minal mass is easily mistaken for a large myoma or ovarian tumor.

Awareness of the condition is the most important aspect in making the correct diagnosis. Suspicion should arise whenever a unilateral pelvic mass terminates in a bulge of the lateral vaginal wall. When a high level of suspicion is entertained, this bulge of the lateral vagina should make the diagnosis quite obvious. In young girls where careful pelvic examination is difficult, an early examination under general anesthesia will hasten the proper diagnosis and may avoid unnecessary procedures. Under general anesthesia the diagnosis can be confirmed by needle aspiration of the characteristic tarlike material from the bulging hematocolpos. Additional investigations are complementary but rarely diagnostic.

Hysterosalpingography is not helpful in this condition of noncommunicating abnormality. Ultrasound findings may help by confirming the cystic nature of the pelvic abdominal mass.

Recently, the use of computerised tomography has been reported to assist diagnosis since it has identified both uteri and accurately characterized the fluid component in the endometrial, cervical and vaginal components of the occluded tract ([2]). In our case, computed tomography was less helpul. Like the ultrasound findings it did confirm the cystic nature of the tumor, but despite an already strong clinical suspicion it was not diagnostic of the exact müllerian abnormality.

Intravenous pyelography or ultrasound examination may demonstrate an absent kidney, ipsilateral to the obstructed hemivagina. Nevertheless the presence of both kidney does not rule out the diagnosis since such a combination has been previously described ([4]).

Once the proper diagnosis of double uterus and unilateral imperferate vagina is made, treatment is simple. Excision of the vaginal septum readily evacuates the hematometrocolpos. The most suitable prognosis following such treatment, in terms of future fertility, has been previously documented ([8]).

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Address reprint requests to:
N. LANG
Dept. Ob./Gyn.
Poriya Government Hospital
M. P. Hagalil Hatachton 15208 (Israel)