ORIGINAL RESEARCH PAPER

Gynaecology

KEY WORDS: Uterus, didelphys, mullerian, anomaly, imperforate hymen

UTERUS DIDELPHYS WITH BILATERAL IMPERFORATE HYMEN

Dr Ashok Kumar Todani

Dr. Mrs. Kiranlata Todani*

*Corresponding Author

ABSTRACT

Rare coexistence of uterus didelphys [5 to 7% of Mullerian defects] and imperforate hymen in each hemi-vagina is reported in this case in a female adolescent. It can be diagnosed through clinical history and physical examination. Further investigation by ultrasonography clinches the diagnosis.

INTRODUCTION:

Uterus didelphys bicollis is a fusion defect of Mullerian ducts, while imperforate hymen, a congenital resorptive defect, represents the extreme in the spectrum of embryological variations in hymenal configuration, with reported incidence ranging from 0.014% to 0.1%. The very rare coexistence of these two entities is described in a 13 year old girl presenting with pelvic pain and double haemato-colpos.

CASE REPORT:

Miss H.T., 13 year old girl, had persistent pain lower abdomen for last one year with periodical exaggeration. Her general condition was fairly normal with all stable vitals.

On ultrasound examination, upper abdomen scan was normal having normal abdominal viscera with normal urinary tree. Pelvic ultrasound revealed single urinary bladder but broad uterus with double endometrial cavities, sitting at the top of dilated longitudinally septed sac representing double vagina containing echogenic turbulent material inside suggestive of haematocolpos. Part of cervix could be identified at lower end of uterus on each side. Partition in vagina was longitudinal situated in antero-posterior plane having vertical and complete vaginal septum. Patient was very poor and unwilling for further investigations.

On physical examination, we found normal secondary sex characters. Abdomen was soft and tender in lower part without any palpable mass.

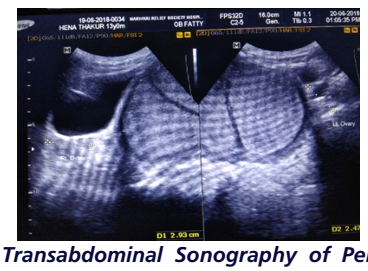

Figure 1: Transabdominal Sonography of Pelvis (double vagina)

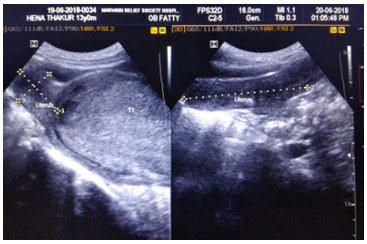

Figure 2: Transabdominal pelvic ultrasound (longitudinal and transverse scan showing broad uterus and haematocolpos)

Gynaecological examination revealed single midline urethral orifice. Two bluish bulzing membranes representing imperforate hymen were visible on either side of lower end of vaginal partition. We arrived at diagnosis of uterus didelphys bicollis with imperforate hemi-vagina on each side.

Photography of private parts was not permitted by patient and party under any circumstances. After proper consent and prerequisite formalities, patient was shifted to operating theatre and bilateral hymenotomy was done under general anaesthesia. Large quantity of chocolate coloured fluid/blood was drained. Intraoperative examination revealed a complete longitudinal vaginal septum. Post-operative period was uneventful.

DISCUSSION:

Co-existence of uterus didelphys bicollis and concomitant imperforate hymen in each hemi-vagina is likely to be coincidental. These two portions of female genital tract are derived from two distinct gynaecological structures and these defects occur at different stages of embryo development. Complete arrest of midline fusion of Mullerian elements leads to two hemiuteri having one tube at each lateral end, two cervices separated or fused at lower uterine segments. Double vagina manifests as a longitudinal septum that extends either completely or partially from the cervices to the vaginal introitus. Patient with uterus didelphys usually become symptomatic in presence of outlet obstruction after menarche.

The hymen is derived from the endoderm of the urogenital sinus epithelium and represents the junction of the sino-vaginal bulbs with the urogenital sinus, it becomes perforate before or shortly after birth. An imperforate hymen is the result of failure of canalization of the vaginal plate, it's reason is not known yet. The fused Mullerian ducts develop into the whole of uterus, Fallopian tubes and upper part of vagina. If this development does not proceed normally, abnormalities usually occur in the female genital tract that are referred to as either 'fusion defects' or occasionally as 'duplication defects'.

Because the external genitalia appear normal in such cases, the condition is usually not diagnosed at birth. Uterus didelphys with outlet obstruction is usually and commonly diagnosed at puberty only after menarche. Although menstruation is normal in these girls, menstrual blood will build up in the blocked vagina, leading to distension and stretching pain with or without pelvic mass.

Hematocolpos should be suspected in adolescent girls in the age group of 12-16 years who present with primary amenorrhea, a cyclic waxing and waning pattern of lower abdominal or pelvic pain, with or without associated symptoms like back pain (38%–40%), urine retention (37%–60%) or constipation (27%). The accumulation of menstrual blood in the vagina and or uterus may result in a mechanical effect on the urethra, bladder or intestines and lead to the obstructive urinary or intestinal

symptoms. Irritation of the sacral plexus or nerve roots is the mechanism for referred low back pain.

Women with a uterus didelphys bicollis more or less have a good reproductive outcome without suirgical intervention. Resection of the intervening vaginal septum is easy and commonly performed. Performing metroplasty or unification operation should be individualized; only selective patients with a relevant history of recurrent abortions or preterm births may benefit from it.

In summary, imperforate hymen is an important cause of abdominal pain in female adolescents and physicians must be aware of this more so when amenorrhoea is associated with it. Treatment is always surgical and adequately performed hymenotomy usually leads to an excellent outcome.

CONCLUSION

Uterus didelphys bicollis with hemivagina and imperforate hymen on each side is an extremely rare condition to be seen clinically in lifetime of a gynaecologist. It is an important aetiology for pain lower abdomen in adolescents girls which can be clinically diagnosed by prompt awareness of physician. Confirmation needs help of imaging modalities. Late diagnosis and treatment of an imperforate hymen may lead to serious complications. Treatment is surgical and it leads to prompt relief and good outcome. Further investigations may be necessary to exclude other uro-genital anomalies.

REFERENCES:

1. Ayaz UY, Dilli A, Api A. Ultrasonographic diagnosis of congenital hydrometrocolpos in prenatal and newborn period: a case report. Medical ultrasonography. 2011; 13(3): 234-236.
2. Eksioglu AS, Maden HA, Cinar G, Yildiz IT. Imperforate hymen causing bilateral hydroureteronephrosis in an infant with bicornuate uterus. Urology Case Reports 2012;2012:102683.
3. Fedele L, Frontino G, Motta F, and Restelli L. A uterovaginal septum and imperforate hymen with a double pyocolpos. Hum Reprod. 2012;27(6):1637-1639.
4. Nagai K, Murakami Y, Nagatani K, Nakahashi N, Hayashi M, Higaki T, Ishii E. Life threatening acute renal failure due to imperforate hymen in an infant. Pediatrics International. 2012;54(2):280-282.