Case Report

A Case of Multiple Genito-Urinary Malformations in a 6 Years old Girl

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Abstract

A 6 years old girl had a large abdominopelvic mass extending from vagina, due to the presence of hemi-vaginal septum in the middle third of vagina. Excision of septum, drainage of mucopurulent fluid with disappearance of mass confirmed the diagnosis of pyomucocolpos. She also had multiple genito-urinary anomalies in the form of double uterus and ectopic malrotated right kidney.

Key words: Congenital, Genitourinary, Malformation.

Introduction

Transverse vaginal septum is a rare condition, the incidence being 1 in 2100 - 1 in 72,000 women.[1] It may lie at the level of upper, middle or lower third of vagina in the frequencies of 46%, 35% and19% respectively.[2] Six percent of cases of vaginal septum with duplicated cervix and uterus are characterized by unilateral obstruction of one hemi-vagina by the vaginal septum.[3] Adolescent girls with obstructed hemi-vagina and ipsilateral renal anomaly (OHVIRA) syndrome or Herlyn-Werner-Wunderlich syndrome, typically present in late puberty.[1] It is usually diagnosed in adolescence following menarche. The frequent presence of vaginal septum gives rise to unilateral haematocolpos, haematometra and sometimes haematosalpinax, resulting in abdominopelvic pain, especially during menstruation. Very rarely, it can present in early childhood as in our case.

Case Report

A 6 years old girl was brought by her mother in the out- patient department with history of pain in the lower abdomen and high grade fever for last 4 months. For this problem she was seen by many physicians who prescribed various medications, but her symptoms could not be relieved. One

month back she also had surgery. The review of surgical record revealed separation of fused labia and drainage of mucocolpos. She was febrile while rest of general physical examination was unremarkable. On abdominal examination there was cystic, tender mass corresponding to 14 weeks pregnancy size in the hypo gastric region. External genitalia appeared normal. Vagina was a blind-ending pouch. A mass was palpable above the examining finger on recto-abdominal examination. Abdominal ultrasound revealed a large cystic mass 5 x 6 cm in the vagina and normal sized uterus. Right kidney was not visualized at its normal anatomical position. Intravenous-urography showed contrast density shadow resembling pelvi-calyceal system overlying upper part of over distended urinary bladder suggestive of ectopic, atrophic malrotated right kidney. Left Kidney showed compensated hypertrophy. CT scan showed hypo dense area, measuring 4.2 x 4.1cm in the pelvis posterior to the bladder and anterior to the rectum. Findings were suggestive of mucocolpos (Figure 1), and abnormally placed kidney (Figure 2).

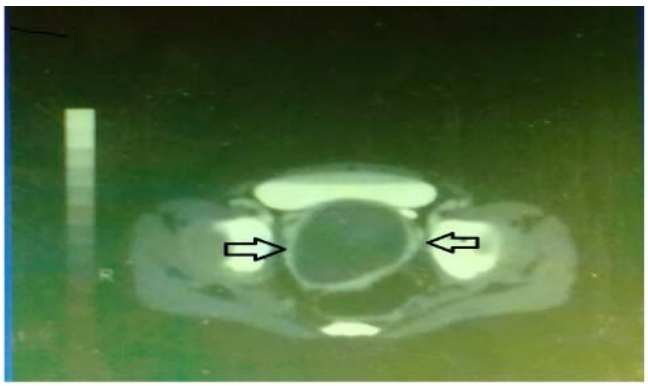

Figure 1. A CT scan of pelvis showing a hypodense cystic area, posterior to bladder and anterior to rectum, about 4.2×4.1cm, suggestive of mucopyocolpus/mucopyometra

Parents were counseled about the nature of the problem, clinical diagnosis and need for surgery. Examination was done under anaesthesia. After separating the labia, the vaginal septum could not be approached because of fibrosis from previous surgery. The abdomen was opened by midline incision. A double uterus was seen. Right uterus was well developed but the left was rudimentary (Figure 3). Upper part of the vagina was distended. After reflecting the bladder down, a transverse incision was given over the upper distended part of the vagina. About 100 ml of mucopurulent fluid was drained. A uterine sound was passed to localize the septum. By combined abdomino-vaginal route the transverse vaginal septum was resected. On post-operative follow-up, patency of vagina was found intact.

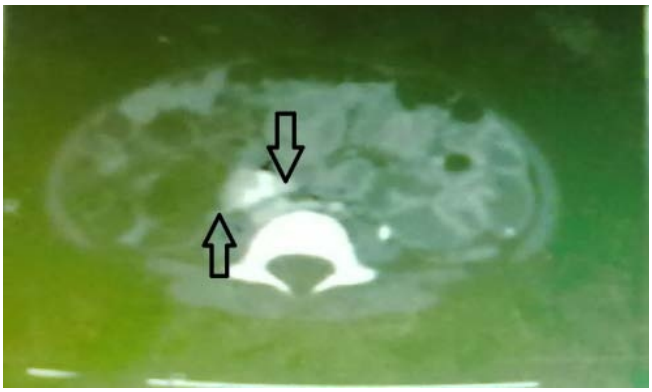

Figure 2. A CT scan of pelvis showing right kidney, lying at an ectopic site at the level of L5 vertebral body

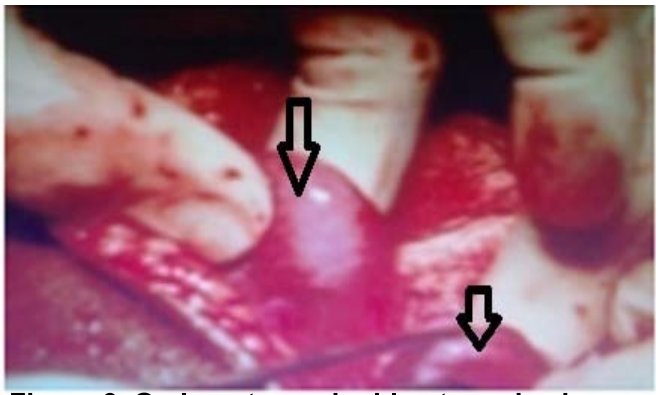

Figure 3. On lapratomy double uterus is visualized

Discussion

The uterus, Fallopian tubes, cervix and upper 2/3rd of the vagina develop from the paired Mullerian ducts while the lower 1/3rd of the vagina develops separately from the urogenital sinus[4]. Because of the close embryological association of the Wollfian and Mullerian ducts, renal anomalies, predominantly renal agenesis are almost always seen in the patients with uterus didelphys with obstructed hemivagina.[5] Incidence of these anomalies is believed to be between 0.5 and 5%. Majority of these cases are diagnosed at menarche. The usual clinical presentation of this syndrome is abdominal pain, that starts right after menarche and is caused by hematocolpos.[6] The children may present with mococolpos or pyomucocolpos due to an ascending infection through a small perforation. Magnetic resonance imaging (MRI) and ultrasound provide an excellent delineation of internal and external uterine contours and septal position and thickness, whereas intravenous pyelography (IVP) is the preoperative requisite to rule out renal tract abnormalities, which are often associated.[7, 8] The surgical resection of transverse vaginal septum is required due to pain, increased risk of infection, and retrograde menstruation. The septum is resected vaginally followed by approximation of upper vagina with lower vagina. In our case vaginal route was not possible because of fibrosis from previous surgery. Hysteroscopic resection of the vaginal septum under transabdominal ultrasound guidance in cases of uterus didelphys with an obstructed hemivagina was found to provide an excellent alternative to conventional treatment.[9] Vaginal stenosis is a postoperative possibility, and may be associated with vaginal adenosis.[9]

References

1. Vallerie Amy M, Beech Lesley L. Update in Mullarian anomalies: diagnosis, management, and outcomes. Current Opinion in Obstetrics and Gynecology 2010, 22:381-386.
2. Rock JA. Transverse vaginal septum. In: Rock JA, Thompson. JD (eds). Telend's Operative Gynecology, 8th edn. Philadelphia: Lippencott-Raven, 1996; 704-706.
3. Heinonen PK. Complete septate uterus with longitudinal vaginal septum. Fertil Steril 2008; 85:700-705.
4. Gindaal G, Kachhawa S, Meena GL, Dhakar G. Uterus didelphys with unilateral obstruted hemivagina with haematometrocolops and haematosalpinax with ipsilateral renal agenesis. Journal of human reproductive sciences Jul-Dec;2(2):87-89.
5. Smith NA, Laufer MR. Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome: management and follow–up. Fertil steril 2007; 87:918-922.
6. ANTONIO J Madureir, Carlos M Mariz, Joao c. Bernardes, Isbel M. Ramos. Case94: Uterus didelphys with obstructing hemivaginal septum and ipsilateral renal agenesis. Radiology J Society of North America May 2006; (293): 602-606
7. Agarawal G, Riherd JM, Busse RF. Evaluation of uterine anomalies: 3D FRFSE cube verses standard 2D FRFSE. AM J Roentgenol 2009; 193(6):558-562.
8. Edmonds DK. Normal and Abnormal development of genital tract. In: Edmonds DK (ed.) Dewhurst's Textbook of Obstetrics and Gynaecology for Postgraduates, 6th edn. Oxford: Blackwell Science, 1996; 6-11.
9. Tae Eun Kim, Gyoung Hoon Lee,* Young Min Choi, Byung Chul JeeSeung-Yup Ku, Chang Suk Suh, Seok Hyun Kim, Jung Gu Kim, and Shin Yong Mo. Hysteroscopic Resection of the Vaginal Septum in Uterus Didelphys with Obstructed Hemivagina: A Case Report J Korean Med Sci. 2007 August; 22(4): 766–769.