A Rare Case of Pyometrocolpos With Obstructive Uropathy: Sequelae of Distal Vaginal Atresia

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ABSTRACT

Hydrometrocolpos is a rare abnormality in female newborns. It follows congenital vaginal obstruction which may present as a midline abdominal mass causing either urinary tract or gastrointestinal tract obstruction without a co-existing bowel anomaly. Our experience was that of an infant with obstructive uropathy from an infected hydrometrocolpos.

Keywords: Hydrometrocolpos, Vaginal obstruction, Obstructive uropathy

INTRODUCTION

Hydrometrocolpos is an uncommon congenital disorder,characterised by the cystic dilatation of the vagina and uterus occurs as a result of accumulated secretions from the reproductive tract following a distal genital tract obstruction. Obstruction of the vagina with accumulated secretion was first described and reported in 1856.[1]When only the vaginal is obstructed, it is termed hydrocolpos; but if there is also uterine enlargement, the term hydrometrocolpos is more appropriate. The anomaly results from failure of development of the distal mullerian duct and/or a part of urogenital sinus that forms the distal vagina.[2,3] In the newborn, it usually presents as a lower midline abdominal mass with regional compression.[4]Compression of the lower urinary tract is reported to cause hydronephrosis.[5] Secondary infection may also occur, resulting in pyometrocolpos, a potentially lethal disease. Children with pyometrocolpos due to distal vaginal atresia may present as acutely ill, with severe obstructive uropathy and septicaemia.[6,7]

This case report highlights the importance of a thorough clinical and radiological evaluation of a newborn with abdominal mass to achieve early diagnosis and appropriate treatment. Rarely, hydrometrocolpos due to distal vaginal atresia can lead to acute renal injury due to pressure effects on the ureters and/or the urinary bladder.[7] The rarity and variability in presentation of this condition can lead to delayed diagnosis and erroneous management.

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CASE REPORT

A 6-month-old female infant, the second of a set of twins born via spontaneous vertex delivery presented at the paediatric out-patient department of the Lagos State University Teaching Hospital (LASUTH) on account of poor weight gain, occasional diarrhoea, vaginal discharge and progressive abdominal swelling noticed about three months after birth. The vaginal discharge was cream-coloured and occasionally purulent and foul-smelling. The abdominal swelling was progressive in size, initially in the lower abdomen and increased up to the umbilicus at presentation. There was no associated vomiting or change in bowel habit. She also came with an abdominopelvic ultrasound scan report, done elsewhere prior to presentation at LASUTH, which showed bilateral hydronephrosis.

Physical examination revealed an infant who, ill-looking and irritable. She was pale and febrile. Her chest was clinically clear. The abdomen was distended with a suprapubic mass which was firm and tender on palpation. The liver was not palpable and the spleen was tipped. Digital rectal examination was normal. Vaginal examination revealed an oedematous and hyperaemic vulva with areas of excoriation. Purulent discharge was seen trickling from the vagina. A working diagnosis of sepsis with obstructive uropathy was made.

Her packed cell volume was 20% and she was transfused. The white blood cell count showed leukocytosis (22,000 per microliter) while the renal function test was normal. Urine culture yielded growth of *Acinetobacter baumannii* while the vaginal swab culture yielded growth of *Klebsiella aerogenes*.

A kidney,ureter and bladder (KUB) scout film of the abdomen showed a bilobed soft tissue opacity in the abdomen and pelvis, displacing bowel loops superiorly and laterally (Fig.1a). The lateral radiograph of the pelvis showed an elongated soft tissue mass in the pelvis, displacing rectal gas posteriorly, extending from the perineum to the lower lumbar vertebrae (Fig.1b).

Intravenous urogram (IVU) revealed bilateral calyceal clubbing and ureteral dilatation with tortuosity. There was also a lateral displacement of the lower third of both ureters (Fig.2). The urinary bladder was however normal.

A repeat abdominopelvic ultrasound scan revealed a well-defined pear-shaped complex cystic mass with medium to low internal echoes and posterior acoustic enhancement. It measured 14x12x9cm in size and rose from the pelvis, in-between the urinary bladder anteriorly and the rectum posteriorly. The mass extended inferiorly towards the perineum. A radiological assessment of 'a distended uterus and vagina filled with complex materials, most likely pus' was made. The kidneys showed bilateral grade II hydronephrosis and there was no free peritoneal fluid. The spleen was also enlarged.

Drainage by suctioning under anaesthesia was done. Findings during the procedure were a suprapubic mass extending up to the umbilicus; eodematous vulva; de-epithelized perineum and about 380ml of foul smelling, putrid, greenish

pus was drained using a size 5 nasogastric tube. A distal vaginal atresia was found as the cause of the pyometrocolpos. Thorough saline irrigation was done and a paediatric Foleys catheter was left in-situ for continuous drainage. An urethral catheter was passed for continuous drainage of the urinary bladder. Post-operatively, daily dilatation with a size 8 Hager's dilator was commenced. There was dramatic improvement in clinical and laboratory findings and she was discharged home and planned for a definitive procedure to correct the vaginal atresia.

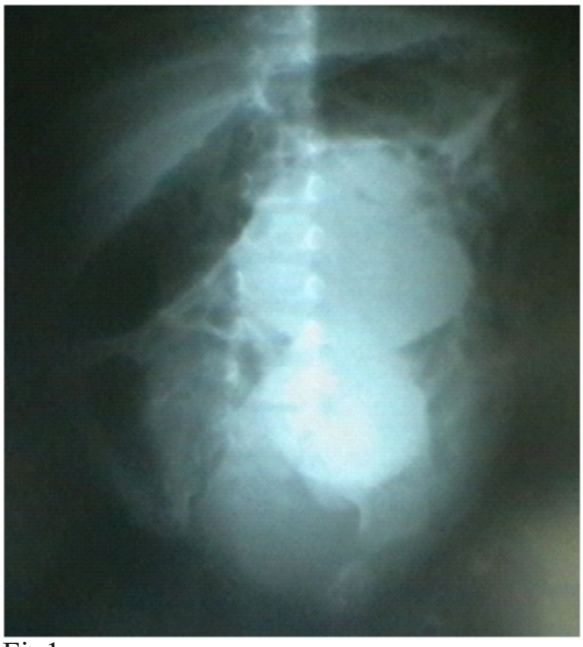

Fig1a

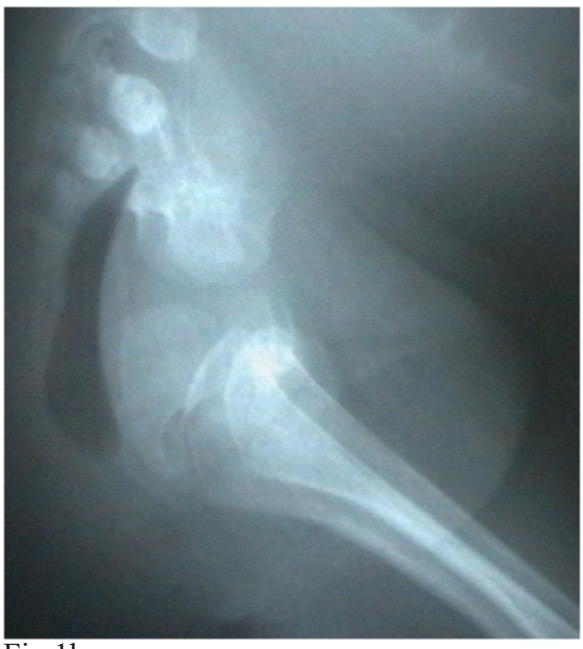

Fig.1b

Figures 1a: Scout film of the KUB showing a bilobed soft tissue opacity in the abdomen displacing bowel loops superiorly and laterally.

1b: Lateral pelvic radiograph showing soft tissue opacity extending from the perineum to the lower lumbar vertebrae. Note the rectal gas shadow displaced posteriorly.

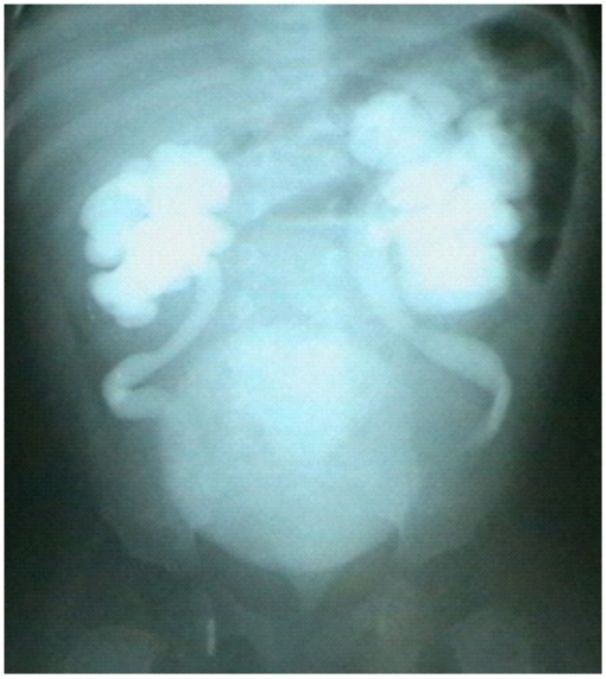

Figure 2 Intravenous Urogram showing bilateral calyceal clubbing, hydroureters with lateral displacement of its distal third from soft tissue opacity in the lower abdomen and pelvis. Note the columnisation of the

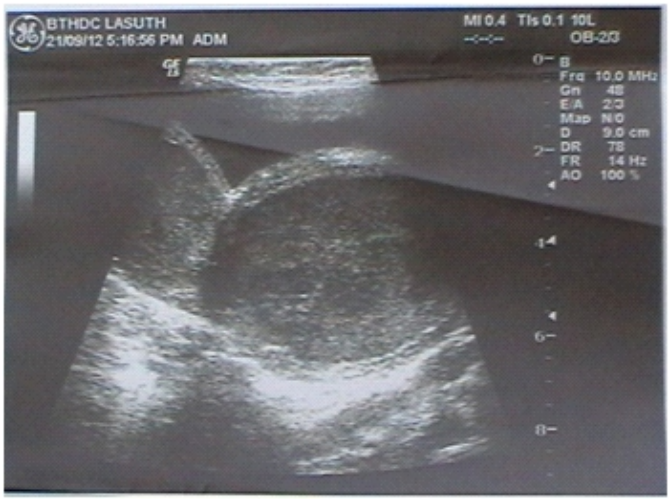

Figure 3: A sonogram showing a thick-walled bilobed structure with low to medium level internal echoes and posterior acoustic shadowing arising from the pelvis to the lower abdomen. The urinary bladder is anterior to this structure

DISCUSSION

Hydrometrocolpos is a distension of the uterus (metro) and vagina (colpos) caused by obstruction to drainage of genital secretions. Causes include imperforate hymen (commonest), vaginal stenosis, transverse vaginal septum, lower or distal vagina atresia and cervical stenosis.[8] Cases of distal vaginal atresia and anorectal malformations have also been reported in literature.[9] The spectrum of hydrometrocolpos is broad, ranging from cases undetected until menarche in adolescents to those cases associated with congenital conditions in newborns or infants. In the latter, agenesis or atresia of the vagina or cervix is responsible for the obstruction. Lower vaginal atresia is a type in which the lower third of the vagina fails to develop. It occurs from failure of recanalisation of the urogenital sinus. Vaginal atresia is estimated to occur in 1 in 4000–5000 live female births. It is often unnoticed until adolescence, when pain and a lack of menstrual flow may point to the condition.[10]

By 8 weeks of gestation, the paramesonephric (mullerian) ducts normally meet in the midline to form the uterovaginal canal and this develops into the uterus, cervix and approximately three fourths of the proximal vagina. The sinovaginal bulbs of the primitive urogenital sinus (vaginal cord) form the distal vagina. Atresia of the lower vagina is attributed to failure of recanalization of the vaginal cord at the 150 mm crown-rump stage. It may take the form of a septum in any plane and at any level, including the site of the hymen, or it may take the form of a stenosis.[3,11] The term vaginal atresia is used for cases in which there is more than just a simple thin imperforated hymen.[12] Vaginal atresia and agenesis are congenital anomalies of the female genitourinary tract and may occur as an isolated developmental defect or as part of a complex of anomalies. Examples of some of these associations include the Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome, Bardet-Biedl syndrome, McKusick-Kaufman syndrome (MKS), Fraser syndrome and Winters syndromes.[8,13] Mayer-Rokitansky-Kauster-Hauster syndrome is an autosomal recessive disorder characterised by hydrometrocolpos, post-axial polydactyl, congenital heart disease, primary amenorrhea and normal ovarian function. In this patient, there were no sign suggesting these syndromes. Isolated vaginal atresia such as this case is an extremely rare finding. Hydrometrocolpos and its association with the syndrome are well described in the literature. Occurrences of pyometrocolpos secondary to distal vaginal atresia in children with Ellis Van Creveld (EVC) syndrome have been reported[14], though this is very rare. MKS must be differentiated from the EVC syndrome, which also includes polydactyly and congenital heart disease, but is associated with chondrodyplasia and ectodermal dysplasia.[1]

[5]

CONCLUSION

The diagnosis of pyometrocolpos in this patient was based on clinical and radiological findings. Prompt diagnosis by imaging studies and recognition of the underlying condition prevented further complication of spillage of the infected contents of the uterus and vagina into the peritoneal cavity which could have resulted in increased mortality and morbidity in this infant.

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