Management of three cases of pyometrocolpos due to distal vaginal atresia in infants.

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الخلاصة :

الأطفال الذين يعانون من الإصابة بالتهاب (خمج) الرحم والمهبل التقيحي نتيجة انسداد المهبل الخلقي يقدمون إلى المستشفيات وهم مصابين بحدة مرض شديد وأهمها اختلاجات انسداد مجرى البول الشديدة وتسمم الدم الجرثومي .اغلب الأعراض السريرية عند هؤلاء المرضى تتحسن بدرجة كبيرة بعد التداخل الجراحي الأولي لإزالة وتصريف الكيس القيحي في العضو المصاب للتغلب على الخمج الناتج من الانسداد الخلقي لقناة المهبل ولإتاحة الفرصة للتداخل الجراحي المستقبلي الجازم لإصلاح قناة المهبل. نقدم ثلاث حالات لإناث بلغت الأولى شهرين من العمر والأخريات أربعة أشهر عند المراجعة صحبة ذويهم إلى العيادة الاستشارية لوحدة جراحة الأطفال في مستشفى النسائية والأطفال التعليمي في محافظة القادسية , اثنين منهن يعانين من اختلاجات انسداد مجرى البول وتسمم الدم الجرثومي والأخرى كانت مصابة بمتلازمة ماير روكيتانسكي والتي ضمت مجموعة تشوهات خلقية الظاهرة منها كانت تشوه القدمين واليدين وانسداد قناة المهبل. جميع هذه الحالات أجريت لها عمليات جراحية عاجلة (فتح البطن) وتصريف كيس القيح في الرحم عن طريق فتح الرحم وتصريف قناة المهبل عن طريق فتح قناة المهبل والتي أنتجت تحسن كبير في جميع هذه الحالات المصابة بعد مرور 4-6 أسابيع على هذه التداخلات والتي تتيح لنا التدخل المستقبلي لإجراء التداخل الجازم والناجح لإصلاح قناة المهبل والرحم ومجرى البول.

ABSTRACT

Children with pyometrocolpos due to distal vaginal atresia may present as acutely ill, with severe obstructive uropathy and septicemia. In such patients the clinical course is markedly improved by urgent drainage of the infected cystic mass, and a temporary drainage procedure is required to allow local findings of infection to subside before definitive surgery. We present a 3 cases one at 2 months and two cases at 4- months of age old females with pyometrocolpos two of them with the above-mentioned clinical course and one of them with typical presentation of Mayer Rokitansky Kuster Houser syndrome (MRKH syndrome) (is characterized by Mullerian duct structures agenesis, vaginal atresia being

the commonest variant) ,presenting and having association of anorectal malformation, Mullerian duct agenesis and renal anomaly , all received urgent laparotomy and drainage through the upper abdominal wall

transverse incision (was performed in spite of the high risks of complex drainage procedures and general anesthesia). This drainage did result in dramatically improved clinical status, and findings of local inflammation resolved within 4-6 weeks, which facilitated a later successful definitive surgical procedure.

INTRODUCTION

Transverse vaginal septum

TVS is formed when the tissue between the vaginal plate and the caudal aspect of the fused Müllerian ducts fails to reabsorb. This anomaly divides the vagina into 2 segments, reducing its functional length. TVS can be perforate or imperforate, and can occur at nearly all levels in the vagina. Most of these septa are located in the superior vagina at the putative junction between the vaginal plate and caudal aspect of the UVP (46%). The next most common locations are the mid vagina, at a rate of 40%, and the inferior vagina, at a rate of 14% (Rock, 1982).

TVS is one of the most rare MDAs, with an approximate frequency of 1 case in 70,000 females (McKusick, 1964; Suidan, 1979). Unlike other MDAs, TVS is only occasionally associated with urologic defects. TVS has been associated with other structural anomalies, including imperforate anus, bicornuate uterus, coarctation of the aorta, atrial septal defect, and malformation of the lumbar spine (Suidan, 1979). No evidence indicates that this disorder is genetically inherited, although a study of an inbred Amish community suggested that hydromucocolpos due to obstructive TVS was the result of a rare autosomal disorder (McKusick, 1964).

Diagnosis of TVS - Fetuses, neonates, and infants

TVS is rarely diagnosed in the neonate or infant unless the obstruction causes a significant hydromucocolpos. In rare cases, copious amounts of fluid may collect in the vagina above the obstructing septum and create a mass effect in which the surrounding organs are compressed. Compression may cause serious consequences if not promptly diagnosed and treated (Jones, 1992). Hydromucocolpos can be diagnosed in utero during third-trimester transabdominal sonography. In these cases, fetal abdominal distension is noted secondary to an abdominal or pelvic mass (Banerjee, 1992).

Diagnosis of hydromucocolpos in the neonate and infant can be challenging. A large mass is often palpated in the lower abdomen. However, unlike an imperforate hymen, the obstruction is well in the

vagina, and a bulging septum is not noted. Initial studies should include an abdominal ultrasonography of the pelvis (Shatzkes, 1991). MRI should also be performed to help make a definitive diagnosis. MRI can also be useful to depict pelvic anatomy and determine the thickness of the vaginal septum. Proper imaging studies frequently eliminate the need for laparoscopy or laparotomy (McKusick, 1964).

Surgical management of TVS - Fetuses, neonates, and infants

When third-trimester ultrasonographic findings lead to the diagnosis, early delivery and drainage of the obstructed vagina and uterus are indicated when other organs are compromised (Banerjee, 1992).In infants, the vaginal septum is usually thin and can be corrected without extensive procedures. Surgical excision of the obstructed septum through a perineal approach is most efficacious. Bilateral incisions may be required to ensure complete removal (Rock, 1992).

Vaginal atresia

Vaginal atresia occurs when the UGS fails to contribute to the inferior portion of the vagina (Shulman, 1988). The Müllerian structures are usually normal, but fibrous tissue completely replaces the inferior segment of the vagina. Although not müllerian in origin, vaginal atresia can clinically mimic vaginal agenesis and imperforate hymen. Surgical management is similar to that of vaginal agenesis.Most cases of vaginal atresia occur sporadically. Some authors describe vaginal atresia as a component of a syndrome characterized by autosomal recessive inheritance, anomalies of the middle ear ossicle, and renal dysgenesis (Winter, 1968; Turner, 1970).

Diagnosis of vaginal atresia

Children with this anomaly may develop pyometrocolpos and present with obstructive uropathy and septicemia (Imamoglu, 2005). Young women with vaginal atresia usually present to the gynecologist with primary amenorrhea. Physical examination reveals age-appropriate developmental milestones, and secondary sexual characteristics are usually normal. A midline pelvic mass can often be palpated during the abdominal-rectal examination. Pelvic examination reveals findings of normal external genitalia; however, a vaginal dimple is present at the introitus. The karyotype is 46,XX, and the results from endocrine studies are normal. Transperineal ultrasonography reveals the presence of ovaries, a uterus, a cervix, and an obstructed blind-ending superior vagina. All of these features distinguish vaginal atresia from vaginal agenesis (Scanlan, 1990).

MRI can aid in detecting the presence of a cervix, which should distinguish this anomaly from cervical agenesis, which is quite rare. This modality can further establish the presence of a functioning endometrial cavity (Tolhurst, 1991). These patients are not candidates for HSG studies.

Preoperative evaluation

Some authorities recommend nonsurgical methods as the first approach in managing vaginal atresia (Tolhurst, 1991). When nonsurgical methods fail, surgical approaches are recommended.

DISCUSSION

Pyometra is the accumulation of pus in the uterine cavity (Muram D 1981) . It is not uncommon in postmenopausal women. In this age group, it is generally caused by the occlusion of the cervical canal secondary to chronic atrophic cervicitis, benign or malignant cervical lesions, puerperal infections, foreign bodies or congenital anomalies (Nuamah NM 2006) . However, it is thought to be relatively rare in the premenopausal age group (Chan LY 2001).

A MEDLINE search showed numerous case reports of pyometra in postmenopausal women. However, there are very few reports of pyometra in children. One article discussed the presentation and management of two children with pyometrocolpos, aged 1 year and 9 years old (Imamoglu M 2005) . It stated that pyometrocolpos can be caused by imperforate hymen or distal vaginal atresia. The typical presentation of such patients can include recurrent urinary tract infections, abdominal mass, findings of obstructive uropathy and, sometimes, secondary infection. The report goes on to discuss the management strategies in this condition. It recommends deferring definitive surgical correction until the local findings of infection resolve. Therefore, a temporary drainage that is both effective and minimally invasive is necessary. The three children in the case report received laparatomy (open drainage) through the upper abdominal wall transverse incision . We could find no case reports of a pyometrium presenting in such a young patient, and therefore believe that these cases are unique.

Children with pyometrocolpos due to distal vaginal atresia may present as acutely ill, with severe obstructive uropathy and septicemia. In such patients the clinical course is markedly improved by urgent drainage of the infected cystic mass, and a temporary drainage procedure is required to allow local findings of infection to subside before definitive surgery. We present three children with pyometrocolpos with the above-mentioned

clinical course, in whom laparotomy (open drainage) was done through the upper abdominal wall transverse incision in spite of the high risks of complex drainage procedures and general anesthesia. This drainage did result in dramatically improved clinical status, and findings of local inflammation resolved within 4-6 weeks, which facilitated a later successful definitive surgical procedure .

Classic finding using radiological study and pelvic US revealed the presence of well-defined cystic round structure posterior to the bladder and anterior to the rectum compressing all these structures .There was rim enhancement and displacement of the uterus superiorly .The uterus was felt to be unremarkable in size but fluid was noted in the endometrial cavity .The diagnosis of hydrometrocolpos was suggested in two patients and just abdominal cyst in the third patient! .

One of the cases presented with an unusual cause (pyometrium) for an acute abdomen in a four-month-old infant , at presentation to the pediatric surgery clinic having been unwell for the past 2 weeks with a gastroenteritis-type illness. Her diarrhea and vomiting (which was non-bilious) had settled in the 24 h before her attendance. Her parents said that she had not been herself; her appetite had decreased and she had cried inconsolably just before her attendance. They had noticed that her abdomen was becoming increasingly swollen and this was their main reason for presenting to the pediatric surgery department. There was no history of fever. In the 24 h before attendance, she had not vomited but had passed four yellow-colored stools. Her medical history showed that she was born at 37 weeks' gestation and required admission to the neonatal unit due to being a small size for dates. She was not ventilated whilst in the special care baby unit. She also had a non-troublesome umbilical hernia and had previously had reflux.

On initial examination, she was clinically dehydrated, with sunken fontanelle. She had a temperature of 37.9°C, a pulse rate of 160 beats/min, a capillary refill time of < 2 s, and her saturations were 98% in room air. She was having intermittent bouts of inconsolable crying, alternating with periods of drowsiness. Her abdomen was grossly distended with overlying veins (picture 1). It was tense and tender on palpation, and minimal bowel sounds were heard. Her chest was clear and heart sounds were normal.

Initial management included an abdominal radiograph, intravenous access, routine blood tests, intravenous bolus fluids and an urgent pediatric review. A nasogastric tube was also inserted, and intravenous ceftriaxone and metronidazole were given. The blood results showed mild hyponatraemia (sodium 129), dehydration (urea 11.6, creatinine 85), and a

raised white cell count (21.2 x 10^9/l) .
The abdominal radiograph showed minimal gas in most parts of the abdomen and no gas on the right side. Seven radio-opaque areas were found on the film. These are "poppers" on the infant's clothing and should be ignored.

The baby was then transferred urgently to the pediatric surgical intensive care unit, and on the second day was taken to theatre. Laparotomy was carried out and showed a large distended pyometrium (picture 2) , which was drained (around 500 cc pus) with a drainage catheter left in situ after preliminary vaginostomy (tube vaginostomy)(picture 3) . A foly catheter was inserted and contrast studies of the urogenital tract were arranged later on. The pyometrium was thought to be secondary to congenital malformation of the urogenital tract—in particular, a fistula between the urinary and genital tracts. She was discharged from hospital 10 days later with a drainage catheter in her uterus and a urethral catheter in situ. She re-attended 1 week later for removal of her urinary catheter, and was started on treatment with trimethoprim antibiotic prophylaxis.

In two cases treated transverse vaginal septum found in the upper vagina (picture 8) , and the other with Mayer-Rokitansky syndrome complete vaginal atresia was found(is best explained by failure of caudal development of Mullerian ducts) (pictures 4 to 7) .In two cases the cyst wall was marsupialized to the introitus , at the end of the procedure the catheter was left inside the cyst for average 7 days (pictures 8 and 9). Preliminary vaginosotomy done by division of the septum in those cases. This was the selected approach whenever proper delineation of structural anatomy was still confusing.

The prognosis of the three infants regarding there renal function was very favorable with normalization of upper tract and serum creatinine level.

Clinical follow-up is necessary because vaginal stenosis with subsequent accumulation of fluid may develop postoperatively. Vaginal reconstruction may be required to allow satisfactory menstruation and coitus .

Patients data

Age at presentation	Presenting symptoms	Associated anomalies	Management
1- 8wks	pelviabdominal mass Septicemia	left vesicouretric reflex	laparotomy, drainage , preliminary vaginostomy
2-16 wks	pelviabdominal mass Obstructive uropathy	congenital heart disease, VSD, polydactyly Mayer-Rokitanskysyndrome	laparotomy, drainage , preliminary vaginostomy
3-16 wks	pelviabdominal mass Septicemia	vesicouretric reflex , Bilateral hydronephrosis	laparotomy, drainage , preliminary vaginostomy

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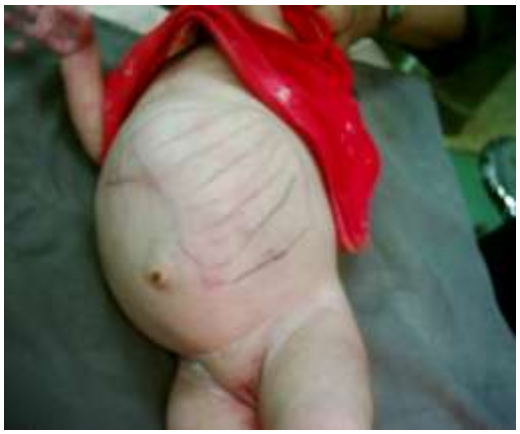
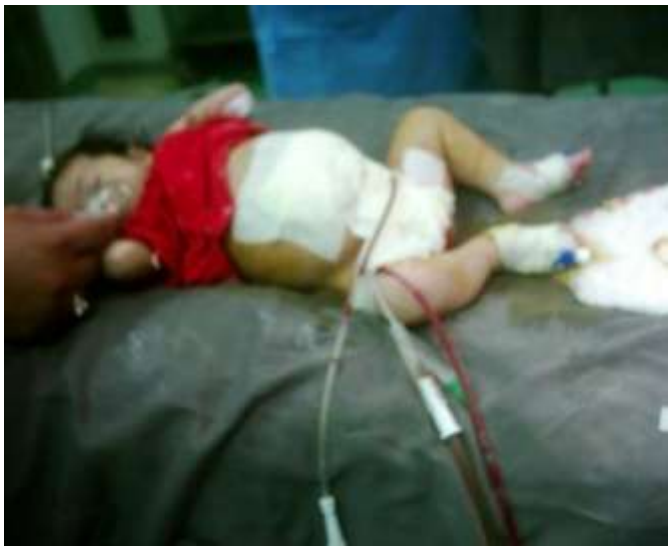
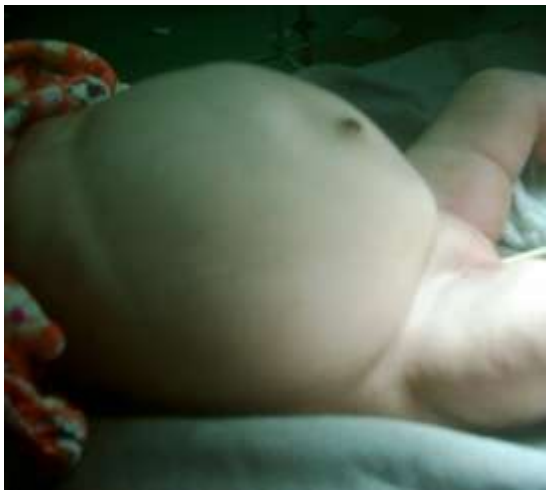


(Picture -1- pelviabdominal mass)

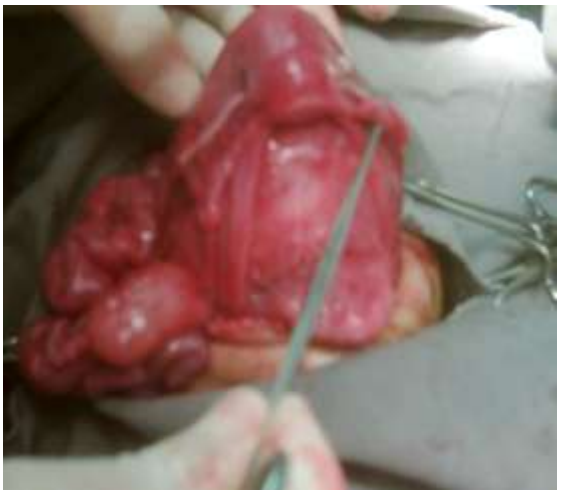
(picture -2- uterus filled with cyst)



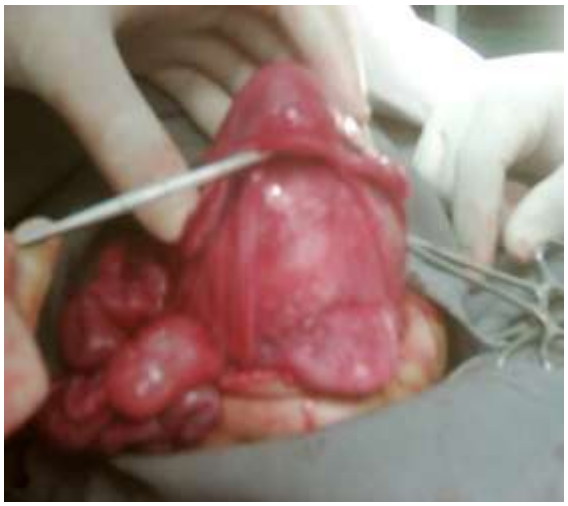
(Picture-3-three tubes drain abdomen ,bladder and site of preliminary vaginostomy)



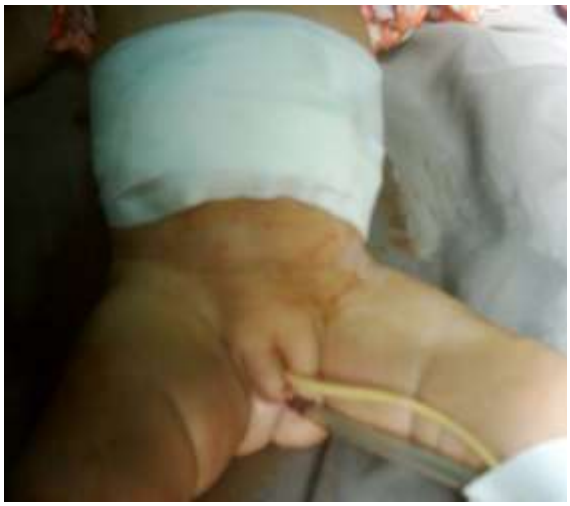
(Picture-4-MR-syndrome abdominal mass)



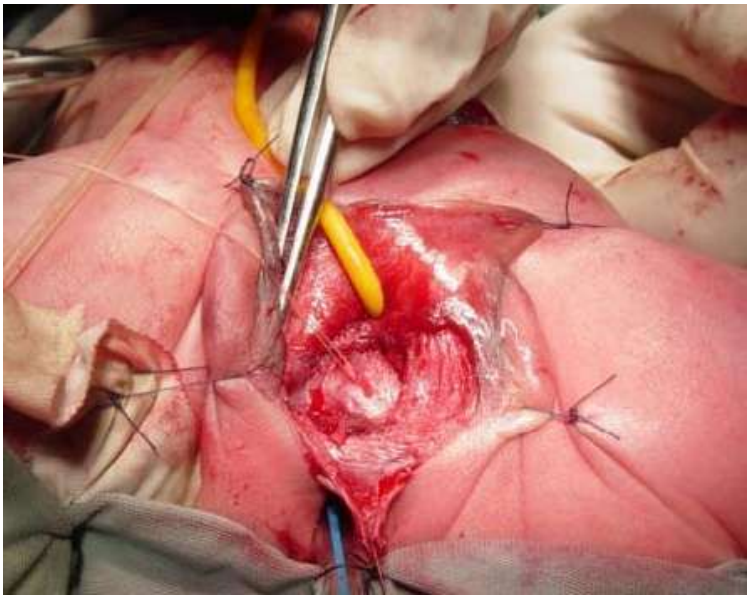
(Picture -5- a hugely distended uterus)



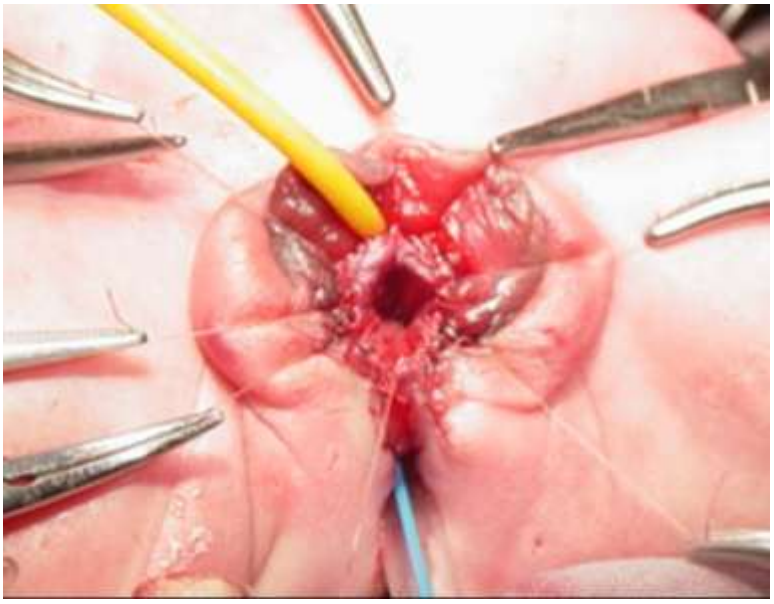
(Picture- 6- a hugely distended uterus)



(Picture -7- tube drain after preliminary Vaginostomy)



(picture -8- intraoperative view showing the transverse vaginal septum)



(Picture -9- marsupialization of vaginal septum)