Case report:

Management of transverse vaginal septum in a neonate Case report and review of literature

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ABSTRACT

A 2 days old female neonate was admitted to the Pediatric Surgery Center at Al- Kanssa teaching hospital on the third of June 2009 with abdominal mass. After clinical examination and investigations, operation was carried out on the sixth of June 2009 and intra operative diagnosis of vaginal atresia caused by transverse vaginal septum was carried out, which is a rare condition occurring 1 in 70,000 – 100,000 live births.

Cruciate incision of the septum with marsupialization was done without vaginostomy and latter vaginal reconstruction.

الخلاصة

نعرض في هذه المقالة عن حالة طفلة مصابة برتق المهبل الجزئي حيث أدخلت طفلة عمرها يومان الى ردهة جراحة الأطفال في مستشفى الخنساء التعليمي بالموصل بتاريخ الثالث من حزيران ٢٠٠٩ وكانت تعاني من ورم البطن وبعد إجراء الفحص السريري عليها مدعوما بالفحوصات المختبرية والأشعة والسونار تم إجراء عملية جراحية لها بتاريخ السادس من حزيران ٢٠٠٩ وتبين أثناء العملية بأنها تعاني من استسقاء الرحم والمهبل الولادي نتيجة لانسداد المهبل الولادي بحاجز عرضي في الثلث الأسفل من المهبل وهي حالة نادرة جدا تحصل حالة واحدة لكل ٧٠٠٠٠– ١٠٠٠٠٠ حالة. وتمت معالجة هذا الانسداد وفتح الحاجز العرضي بطريقة التجييب بمرحلة واحدة دون الحاجة الى تفويه المهبل وتصليح الانسداد المهبلي على مراحل. وتمت مناقشة هذه الظاهرة مع مراجعة ما نشر حول الموضوع في الأدبيات الطبية.

Vaginal atresia is an uncommon congenital anomaly resulting in uterovaginal tract obstruction [1]. It is estimated to occur in 1 in 4000-5000 live female births. Often the anomaly is undetected until adolescence, when primary amenorrhea or abdominal pain from an obstructed uterovaginal tract prompts a diagnostic evaluation [2].

Transverse vaginal septum , or partial vaginal agenesis, is less common which occurs at an incidence of 1 in 70,000 females [3]. Variants of vaginal atresia, formerly termed partial vaginal agenesis currently are classified as variants of transverse vaginal septum. So, failure of vaginal canalization at various levels of the vaginal plate results in the transverse septum, the most common location of transverse septum is the upper vagina, followed by middle, with least common location being the lower third [4]. The most common clinical presentation of vaginal atresia occurs in conjunction with an abscent uterus, which is termed Mayar-Rokitansky-kuster-Hauser syndrome. Type I Rokitansky syndrome is characterized by an isolated absence of the proximal two thirds of the vagina, where as type II is marked by other malformations, these

include vertebral, cardiac, urologic and otologic anomalies [5].

There are two kinds of vaginal septa, transverse and longitudinal. A transverse vaginal septum is one of the most common congenital anomalies of the female genital tract, it is thought to result from faulty canalization of the embryonic vagina[8] .

Transverse vaginal septum may be complete resulting in retained secretion of genital tract in neonate, or partial with pin point opening allowing for menstrual flow. They are usually found in mid vagina, but may occur at any level, when the septum is in the upper vagina it is more likely to be incomplete. If it is located in lower part of vagina, it is more likely to be complete [8].

Longitudinal vaginal septum is usually benign, though obstruction can occur when septum is associated with other mullerian anomalies and surgical excision may be required [8].

Case report

A 2 days old female neonate was referred on the 3rd of June 2009 from Al-Batool Hospital because of abdominal mass. The baby was a full term, delivered by Cesarean section to a multiparous woman, her weight was 3500 gm. On examination, the baby was active with physiological jaundice and polydactyly of both hands and feet. Systemic examination revealed normal heart and normal vesicular breathing, with abdominal distention and mass-like lesion occupying most of the abdominal cavity. On examination of genitalia, the baby had normal perineum, normal introitus and normal anus. The baby had normal passage of meconium without vomiting inspite of the huge abdominal distention. The laboratory investigations revealed that: Hb= 16 gm/dl, TSB = 6.1mg% (normal range = 0.6- 1.2 mg %), Blood urea =4.2 mmol/l (normal range = 3.5 – 7.5 m mol / liter).

Chest x – ray was normal. First ultrasound of abdomen revealed a cystic mass connecting to the stomach mostly of gastric diverticulum or duplication cyst, and the second ultrasound report indicated a mesenteric or omental cyst, in addition to moderate hydronephrosis of both kidneys. Plain x- ray of abdomen and upper gastro intestinal tract contrast study revealed normal stomach, duodenum with deviation of the most of the small bowel to the left caused by the mass effect.

After stabilization of the general condition of the baby in the incubator with intravenous fluid, antibiotics and vitamin K prophylaxis, operation was carried out on the 6th of June 2009.

Examination under general anesthesia revealed a huge pear-like mass extending from the pelvis to the epigastric region (fig. 1) and it was mobile from side to side. Through transverse upper abdominal incision, the abdomen opened and revealed huge hydrometrocolpus (fig. 2), with normal fallopian tubes and normal ovaries. Vaginal atresia was diagnosed, then vaginostomy was performed to evacuate the fluid and mucus material from the vagina (fig. 3). A Hegar dilator No. 11 was introduced through the vaginostomy to determine the level of the vaginal atresia and on putting the patient in the lithotomy position the dilator was reaching the lower third of the vagina, so vaginoplasty with cruciate incision and deroofing of the edges of the transverse vaginal septum was done with marsupialization of the edges using a Vicryl 4/0 suture (fig .4) and Foley's catheter No.8 inserted in the bladder and Foley's catheter No. 12 kept inside the vagina to keep it open during healing phase (fig. 5). Both catheters removed after 7 days. Repeated vaginal dilation every week for about 3 months was done to prevent vaginal stenosis. The patient had uneventful recovery during the last 6 months.

Discussion

Vaginal atresia is a developmental defect resulting in uterovaginal obstruction. Variants of vaginal atresia, formerly termed partial vaginal agenesis, currently are classified more correctly as variants of transverse vaginal septum [5]. Although controversy exists regarding the development of a patent genital tract, canalization of the uterovaginal canal is believed to occur from the caudal to the cephalic aspect, with an epithelial lining derived from the urogenital sinus. Failure at the vaginal plate level may explain the

transverse vaginal septum variants. The presence of transverse vaginal septum is a rare finding, occurring in an estimated 1 to 2 per 70,000-100,000 female births[5]. The commonest presentations were related to genitorurinary obstruction, followed equally by abdominal pain and pelviabdominal mass. Physical examination is fundamental component of workup but often is not adequate to establish a definitive diagnosis. On evaluation of introitus an isolated vaginal dimple or small vaginal pouch with a normal hymenal ring may be seen, these features do not allow the examiner to distinguish between the myriad of internal variants. In other patients, features of ambiguous genitalia are evident in neonates and infants, and a complete radiographic evaluation is warranted[9]. One of the largest series of patients with uterovaginal obstruction was reported by Spencer and Levy in 1962[6]. They described 62 cases of hydro or hematometrocolpos, with 42 secondary to imperforate hymen, 13 to vaginal atresia, and only 7 resulting from transverse vaginal septum. Another series of patients with uterovaginal obstruction was reported by Omar Mansour and Hani Morsi in 2008[7]. They described 11 cases of hydrmetrocolpos resulting from transverse vaginal septum and managed by incision of septum with marsupialization.

The diagnosis can be made at any time between the perinatal period and adolescence. Often the anomaly is undetected until adolescence, when primary amenorrhea or abdominal pain from obstructed uterovaginal tract prompts a diagnostic evaluation.

The reasons for reporting this case are its rarity, it's huge hydrometrocolpos which was reaching epigastric region in a 2 days old neonate, and because of the error in reaching the diagnosis by U/S examination; and lastly because vaginoplasty was carried out in the neonatal period without vaginostomy and later vaginal reconstruction.

Conclusion

Transverse vaginal septum is a rare genital problem that may present early in infants. Proper diagnosis is difficult to achieve despite meticulous physical examination. Incision of the septum together with marsupialization is the standard treatment. Whenever anatomical evaluation is not precisely done, vaginostomy, and staged repair is recommended.

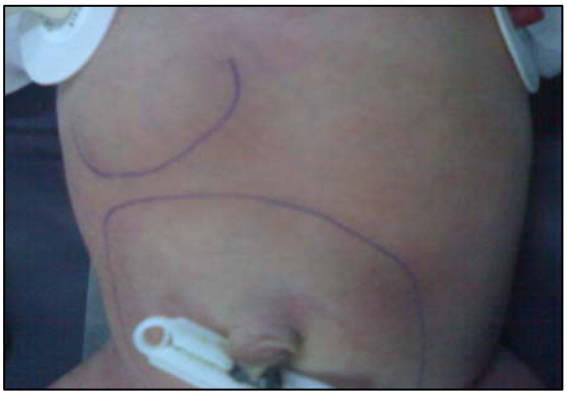

Figure (1): Mass extending from pelvis to epigastric region.

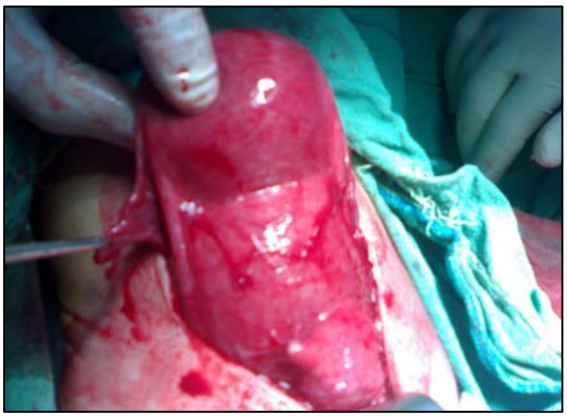

Figure (2): Huge hydrometrocolpos.

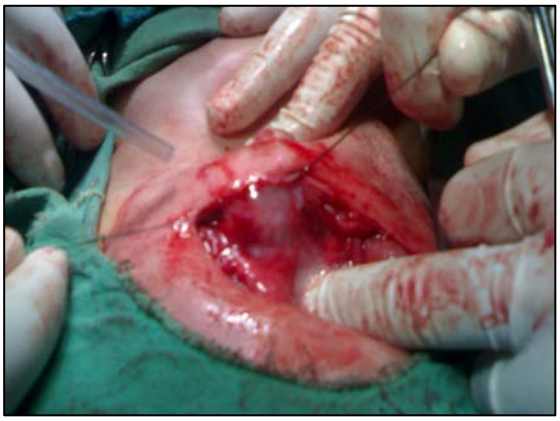

Figure (3): Vaginostomy to evacuate the fluid and mucus material from the vagina.

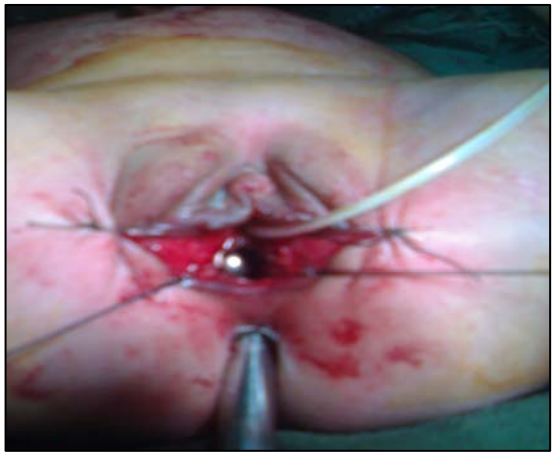

Figure (4): Marsupialization of the septum with two Hegar dilators; the upper one appears through the vaginostomy, and the lower one is in the anus.

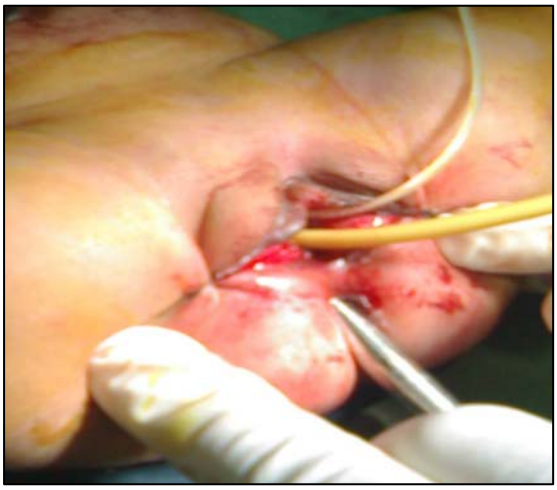

Figure (5): Foley's catheters in the bladder and vagina.

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