Management of Partial Vaginal Atresia in Infancy: Early Experience

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Background/ Purpose: Transverse vaginal septum or partial vaginal agenesis is a subtype of vaginal atresia characterized by uterovaginal obstruction. This study was carried out to highlight the various patterns of clinical presentations, management options and outcome of this rare congenital anomaly.

Materials & Methods: Eleven female patients presenting with manifestations related to uterovaginal obstruction during infancy were studied. Proper clinical evaluation and imaging studies including ultrasonography and computerized tomography were done. Cystovaginoscopy and laparoscopy were used for better delineation of the genitalia, adnexa and the uterus besides guiding for drainage and incision of the vaginal septum. Each patient was evaluated as regard to the age at presentation, presenting symptoms, associated malformations, imaging studies, operative details, and outcome.

Results: The main presenting symptoms were related to genitourinary obstruction. Endoscopic guided laparoscopic assisted division of the septum was done in 6 patients. Preliminary vaginostomy followed by vaginal pullthrough was done in 3 patients, and only endoscopic incision of the septum was done in 2 patients. No reported major complications were encountered during 1 year follow up.

Conclusion: Diagnosis of transverse vaginal septum is still a challenge during early infancy. Division of the septum together with vaginal pull-through was found to facilitate marsupialization of the incised septum to the edges of the introitus. Incision of the septum guided by laparoscopy and vaginoendoscopy can minimize accidental injuries to the bladder, rectum and urethra. Vaginostomy and staged repair is advisable in confusing cases.

Index Word: Vaginal atresia, hydrometrocolpos, septum, marsupialization.

INTRODUCTION

Vaginal atresia is an uncommon congenital anomaly resulting in uterovaginal tract obstruction. According to the classification of uterovaginal anomalies by the American Fertility Society, complete vaginal atresia is categorized under agenesis or dysgenesis of the Müllerian ductal system. Consequently, vaginal atresia occasionally is termed Müllerian agenesis.²

Between the sixth and eighth weeks of fetal development, the caudal aspects of the bilateral Müllerian ducts fuse at the midline. A single midline tubular structure forms as a consequence of cell proliferation (i.e., uterovaginal canal). This midline
structure extends to the Müllerian tubercles, where it encounters the urogenital sinus. Bilateral endodermal invaginations (i.e., sinovaginal bulbs) form as the Müllerian tubercles regress. 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. Failures at the vaginal plate level may explain the transverse vaginal septum variants. 2

Vaginal atresia 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. 3

Transverse vaginal septum, or partial vaginal agenesis, is less common, with a reported incidence of 1 in 70,000 females. Variants of vaginal atresia, formerly termed partial vaginal agenesis, currently are classified more correctly 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 the transverse septum is the upper vagina, followed by the middle, with the least common location being the lower third. 3

The most common clinical presentation of vaginal atresia occurs in conjunction with an absent uterus, which is termed Rokitansky-Küster-Maier-Hauser syndrome (RMKH) syndrome. Uterovaginal atresia in those patients with RMKH syndrome is best explained by the failure of the caudal development of the Müllerian ducts. Vaginal atresia occurs also in conjunction with renal anomalies, such as (ectopic kidney(s), horseshoe kidney, and crossed-fused ectopia), occur in 30% of these cases. 5

Obstructed uterovaginal anomalies typically present either in the neonatal period or at menarche. Hydrometrocolpos is the accumulation of secretions in the vagina and uterus caused by the combination of intrauterine stimulation of the infant’s mucus glands by circulating maternal estrogen and an obstruction of the genital tract by an intact hymen, vaginal septum or vaginal atresia.

If the accumulation of secretions is limited to the vagina, the condition is termed hydrocolpos. The typical presentation of these conditions in a newborn is as a lower midline abdominal mass or a bulging perineal mass. Dysuria or urinary retention also may be present. 6

PATIENTS AND METHODS

In this study, we present 11 female infants that presented with symptoms related to uterovaginal tract obstruction, upper or lower urinary tract obstruction (LUTS). Clinical examination, endoscopic cystovaginoscopy and radiological investigations revealed that they all had partial vaginal atresia. Data collected from patients records included the age at presentation, presenting symptoms, and associated malformations.

The clinical records and radiological findings together with laboratory findings were reviewed. Structural anatomy was determined by clinical examination, ultrasonography, and computerized tomography. Cystovaginoscopy and laparoscopy was used to further delineate the genitalia, adnexa and uterus and facilitate the operative technique. Timing and type of surgical intervention were studied. Findings during a period of one-year follow up were discussed.

RESULTS

Age at presentation ranged between 2 weeks and 4 years but the majority were below 1 year. Mean age was 7.5 months. Presenting symptoms were mainly related to genitourinary obstruction. Urinary outlet obstruction was reported in 6 patients (54.5%), recurrent lower abdominal pain in 5 patients (45.5%), pelviabdominal mass in 5 patients (45.5%) and 3 infants presented by upper urinary tract obstruction (hydroureteronephrosis) secondary to compression of both ureters by the pelviabdominal mass, with elevated serum creatinine level (Table.1).

The selection of operative interventional technique was dictated by the clinical presentation of the patient, genitourinary obstruction and depth of the vaginal canal.
Operative details:

Laparoscopic assisted endoscopic guided division of the septum was done in the last 6 patients in this study. The procedure started by endoscopic examination of the urethra and vagina. Then laparoscopic exploration was followed by incision and evacuation of the distended proximal vagina. A dilator was inserted inside the proximal vagina to guide the site of endoscopic guided incision of the vaginal septum avoiding injury to the urethra and rectum in particular. No patients with uterine or adnexal agenesis were detected in this study.

In 6 patients (55.5%), the vaginal septum was found to be in the upper third of the vagina with distal hypoplastic vagina. Incision of the vaginal septum was initially done followed by mobilization of the proximal part of the vagina down to the introitus. This pullthrough procedure was facilitated by transabdominal or transperineal dissection both anterior and posterior to the distended proximal vagina.

Table 1. Patient's Data

<table>
<thead>
<tr>
<th>Age at Presentation</th>
<th>Presenting symptom</th>
<th>Associated anomalies</th>
<th>Management</th>
</tr>
</thead>
<tbody>
<tr>
<td>1   6 weeks</td>
<td>Pelviabdominal mass</td>
<td>Bilateral hydroureteronephrosis Congenital heart disease, VSD</td>
<td>Endoscopic incision of the vaginal septum. Vaginostomy, Later transperineal vaginal pullthrough mobilization.</td>
</tr>
<tr>
<td>2   8 weeks</td>
<td>Genitourinary obstruction,</td>
<td>Congenital heart disease, ASD, Haemangima.</td>
<td>Vaginostomy, Later transperineal vaginal pullthrough mobilization.</td>
</tr>
<tr>
<td>3   23 weeks</td>
<td>Pelvi abdominal mass.</td>
<td></td>
<td>Endoscopic incision of the vaginal septum.</td>
</tr>
<tr>
<td>4   21 weeks</td>
<td>Genitourinary obstruction,</td>
<td>Left vesicoureteric reflux disease.</td>
<td>Laparoscopically assisted endoscopic incision of the septum + Marsupialization.</td>
</tr>
<tr>
<td>5   14 weeks</td>
<td>Dysuria, UT infection.</td>
<td></td>
<td>Vaginostomy, Later transperineal vaginal pullthrough</td>
</tr>
<tr>
<td>6   14 months</td>
<td>Genitourinary obstruction</td>
<td>Left hydronephrosis, Down’s Syndrome.</td>
<td>Laparoscopic Assisted, Endoscopic incision of the septum + Marsupialization.</td>
</tr>
<tr>
<td>7   23 months</td>
<td>Pelvi abdominal mass, UT infection</td>
<td></td>
<td>Laparoscopically assisted endoscopic incision of the septum + Marsupialization.</td>
</tr>
<tr>
<td>8   2 years</td>
<td>Genitourinary obstruction,</td>
<td>-</td>
<td>Laparoscopically assisted endoscopic incision of the septum + Marsupialization.</td>
</tr>
<tr>
<td>9   6 weeks</td>
<td>Pelvi abdominal mass</td>
<td>Syndactyly</td>
<td>Laparoscopically assisted endoscopic incision of the septum + Marsupialization.</td>
</tr>
<tr>
<td>10  12 weeks</td>
<td>Pelviabdominal mass</td>
<td>-</td>
<td>Laparoscopically assisted endoscopic incision of the septum + Marsupialization.</td>
</tr>
<tr>
<td>11  4 years</td>
<td>Genitourinary obstruction</td>
<td>Horse shoe kidneys.</td>
<td>Laparoscopically assisted endoscopic incision of the septum + Marsupialization.</td>
</tr>
</tbody>
</table>
The principle of the technique of the transabdominal mobilization of the vagina was the same both laparoscopically and by open approach. It started by mobilizing the rectum first anteroposteriorly then the space between the rectum and vagina was further dissected to allow further mobilization of the vagina. Anteriorly no trial for separation of the vagina and the urethra was done. Instead both were mobilized and dissected along the retopubic space. Transperineal mobilization was facilitated by traction sutures applied to the vagina. (Fig 4)

Marsupialization of the edges of the incised septum was done to the edges of introitus to avoid possibility of stenosis.
Preliminary vaginostomy was done in 3 patients (27%) followed by endoscopic guided division of the septum and vaginal pullthrough. This was the selected approach whenever proper delineation of the structural anatomy was still confusing. Only endoscopic incision of the septum was required in 2 patients (18%) with low septum.

The mean duration of tube drainage of the vagina was 7 days while for urinary diversion 8.5 days. Follow up included regular weekly calibration and dilatation of the opening in the vaginal canal for one month.

The prognosis of the 3 infants referred by the nephrology unit due associated upper urinary tract obstruction, with elevated serum creatinine level was very favorable with normalization of the upper tract & serum creatinine level.

An average follow up for about 12 months could be achieved in studied patients. No reported major complications were encountered. Only one patient had wound infection and was treated medically, and another one needed repeated dilatation for 2 months.

DISCUSSION

Vaginal atresia is a developmental defect resulting in uterovaginal obstruction. The vaginal atresia occasionally is termed Müllerian agenesis. Variants of vaginal atresia, formerly termed partial vaginal agenesis, currently are classified more correctly as variants of transverse vaginal septum. 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. Failures at the vaginal plate level may explain the transverse vaginal septum variants. The presence of a transverse vaginal septum is a rare finding, occurring in an estimated 1 to 2 per 100,000 female births. One of the largest series of patients with uterovaginal obstruction was reported by Spencer and Levy in 1962. They described 62 cases of hydro- or hematometrocolpos, with 42 secondary to imperforate hymen, 13 to vaginal atresia, and only 7 resulting from a transverse vaginal septum. 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 an obstructed uterovaginal tract prompts a diagnostic evaluation.

Vaginal atresia is reported to be the second most common cause of primary amenorrhea in tertiary care centers. In this study which was performed only on children, all presented below the age of 4 years. The commonest presentations were related to genitourinary obstruction (54.4%) followed equally by abdominal pain and pelviabdominal mass (45.5% for each). Physical examination is a fundamental component of the workup but often was not adequate to establish a definitive diagnosis. On evaluation of the introitus, an isolated vaginal dimple or a 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. A complete radiographic evaluation is warranted.

In this study we depended on the use of perineal ultrasound and computed tomography for all patients. Findings were suggestive in all cases of the presence of an uterovaginal obstruction with presence of a cystic mass of variable size between the bladder and the rectum in addition to trace of fluid inside the uterus. Structural anatomy was further delineated following endoscopic examination of the genitalia and urethra. The vaginal septum was found in the upper third of the vagina in 55.5% of patients. Precise structural anatomy of the genitalia could not be identified in 3 patients.

An additional assessment could be further accomplished using laparoscopy for both diagnostic and therapeutic purposes. Paediatric surgeon and urology surgeon have contributed to the treatment in these patients. Choice of the technique was variable in different patients. Whenever the anatomy of the perineum was not precisely done, vaginostomy was the procedure of choice. This was done in 3 patients. The distal vaginal pit was found to be almost rudimentary in most of patients. Simple division of the septum was combined by a vaginal pullthrough procedure getting the proximal part of the vagina more distally to allow its anastomosis to the introitus following division and marsupialization of the septum. Only endoscopic incision of the septum was done in 2 (18%) patients with low septum.
For the last 6 patients in this study, following endoscopic and laparoscopic delineation of the anatomy of the perineum and pelvis, the anterior wall of the cyst was incised, through a laparoscopic cannula, evacuating the contents, followed by the passage of a dilator into the cyst, stretching the septum at the perineal part. Guided by the endoscope, the septum was incised avoiding injury to the bladder, urethra and rectum. Then the cyst wall was marsupialized to the introitus. At the end of the procedure a catheter was left inside the cyst for an average of 7 days. Regular dilatation was done under sedation during the follow up period weekly for 4 weeks. No significant complications were encountered during the postoperative period.

Incomplete drainage and failure of marsupialization may result in recurrent obstruction and, potentially, in ascending pelvic infection. Although prophylactic antibiotics are not recommended, development of fever and abdominal pain in the postoperative phase was evaluated and treated promptly. Potential complications include endometritis, salpingitis, or tubo-ovarian abscess—any of which can affect subsequent fertility. All such complications were not encountered in studied patients during the follow up period. Still long term follow up is needed for further assessment of such patients.

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. Radiographic studies and endoscopic evaluation are necessary to achieve diagnosis. Whenever anatomical evaluation is not precisely done, vaginostomy and staged repair is recommended. Incision of the septum together with marsupialization is the standard treatment. Combined endoscopic and laparoscopic drainage of the cyst can further safely facilitate the technique. Follow up and regular vaginal dilatation is essential to avoid complications involving restenosis and ascending genital infection. Long term follow up is still needed for further better functional and cosmetic assessment.

REFERENCES