Anorectal Malformation with Mayer-Rokitansky Syndrome – A Rare Association

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Introduction

The association between Anorectal malformation and MRKH is rare. The review of electronic media and literature shows scanty references regarding this association. Since this association is rare, management is a challenge. Here is a case report where in, this anomaly is managed in a single stage in early infancy with vaginal reconstruction and anterior sagittal ano rectoplasty. In our case we converted the rectum into neo vagina and anastomosed it to fundus of uterine cavity along with pull through of bowel as neoanus.

Case Report

A 3 month old, 5 kg female infant was admitted to Department of Paediatric Surgery for complain of passage of stools from introitus and straining. Examination of perineum revealed, two openings, in introitus. Thus patient was suspected as a case of recto vaginal fistula, through which she was passing stools.

About 2.5 cm posterior to the vestibule a mid line pigmented area was present. Ano cutaneous reflux was present.

The routine blood profile was within normal range. X-ray spine revealed presence of only two pieces of sacrum. Ultrasonography revealed no abnormality of kidneys, ureters and bladder with normal ovaries and uterus. However, fallopian tubes could not be identified. Genitoscopy was performed revealing no vagina and instead the scope entered directly into the rectum.

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After bowel preparation patient was subjected to anterior sagittal anorectoplasty. Having confirmed that vagina was absent and rectum was opening into the introitus, sphincter muscle complex was divided. The bowel was pulled down from sphincter muscle complex in midline and anoplasty was performed at proposed anal site (Fig. 1).

Laparotomy was performed through oblique right lower hockey stick incision. Both overies and tubes were present and appeared grossly normal. The body of uterus was rudimentary, while the fundus of uterus appeared to be normal. The uterine fundus was opened on inferior aspect and uterine cavity was examined and was found obliterated in its lower part. Rectum transected at peritoneal reflection. Rectum, which was opening into the vestibule, was preserved as vaginal orifice and the proximal end of distal pouch of rectum was anastomosed with the opened up uterine cavity (Fig. 2).

In postoperative course, the patient had recession of neoanal opening, for which reanoplasty and sigmoid colostomy was done.

Discussion

In cases of female anorectal malformations, where there are two openings in the introitus, differential diagnosis includes either, a rectovaginal fistula (low or high) or a rectovestibular fistula with utero vaginal agenesis. The incidence of imperforate anus with rectovaginal fistula has been reported to be less than 1% in large series of patients.[1] As compared, Pena reports 8 cases of rectovestibular fistula with uterovaginal agenesis in a large series of 1007 patients with anorectal malformations[2] i.e. incidence of 0.79%. In our case we suspected a rectovestibular fistula with uterovaginal agenesis for which she was subjected to sonography which revealed normal uterus

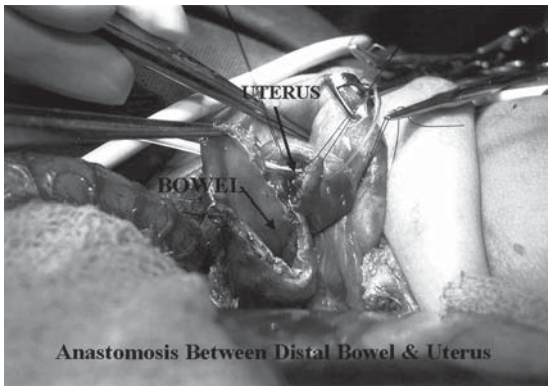

Fig. 1 : Operative photograph showing anastomosis of fundus of uterus to the rectum, which was left as neo vagina.

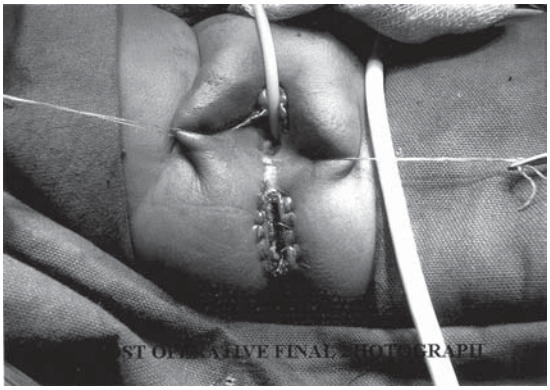

Fig. 2 : Postoperative photograph showing neo anus and neo vagina.

and ovaries.

Sarin *et al*[3] recommends that in all these cases it is mandatory to do an endoscopy or an MRI to distinguish between the rectovaginal fistula and rectovestibular fistula with uterovaginal agenesis. In centres that do not have an access to an MRI scanner or endoscopy, the differentiation can be done by dye study through the introitus. In our case we performed an endoscopy on table which confirmed scope directly entering into rectum with absence of vagina. After establishing diagnosis, the challenge in management is vaginal and anorectal reconstruction. Cohns reports[4] that in rectovestibular fistula with utero vaginal agenesis both rectal and vaginal anomalies could be corrected by an abdomino perineal dissection of the rectum with pull through repair of the anus and utilization of the terminal bowel as an artificial vagina with successful results i.e. the vagina had shown no tendency to stricture formation, anal sphincter tone is good and the patient is continent of faeces. We advocate anterior sagittal anorectoplasty with the simultaneous, reconstruction of vagina and anorectum in single stage as it has definite advantages. Using anterior sagittal approach the sphincter muscle complex is not divided completely, thereby continence is not compromised and simultaneous abdomen and perineum can be approached. No vaginoplasty is required as the terminal portion of the bowel is retained at introitus as vagina and tedious, dissection of separating the terminal bowel from urethra is avoided, thus lessening chances of neurogenic bladder, trauma to urethra, and interference with continence. Simultaneous vaginal reconstruction and anoplasty in infancy offers added advantages-firstly prevents psychological trauma, secondly avoids need of vaginoplasty through scarred perineum, after anorectoplasty.

Coran[5] advocates use of sigmoid colon for vaginal reconstruction in children as against ileum with advantages that thicker wall of sigmoid bowel tolerates trauma with lesser reaction and bleeding and excessive mucus production by sigmoid neovagina gradually tapers off over 1 to 2 months. Griffin[6] *et al* have also reported a case of MRKH where the artifical vagina was attached to uterine cavity. Both ovulation and menstruation has been documented in this case.

Pregnancy has been reported in a Nigerian girl, case of MRKH with ARM in which reconstruction was done at the age of 15 years by using distal rectum in situ to serve as a

neovagina and the rectal stump was anastomosed to upper vaginal pouch.[7]

In our case single staged operation was performed for case of congenital imperforate anus, with rectovestibular fistula and absence of vagina, with rudimentary lower portion of uterus, and normal upper uterine cavity which was anastomosed to proximal portion of rectum keeping distal rectum in situ, which would serve as neo vagina and simultaneous Anterior Sagittal Anorectoplasty being performed. In this case however, a long term follow up is required to document ovulation, and subsequent pregnancy, as operation has been performed in infancy.

Though the operation was done in single stage but due to sacral agenesis, poor development of sphincter muscle complex, post operative infection, child had recession of neo anal opening which was managed by reanoplasty and sigmoid colostomy.

References

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HPV AND CERVICAL CANCER

'Much of the cervical cancer problem can be solved with existing or soon-to-be available technology, sufficient will, and modest resources. [...] We hope to see a major decrease in the numbers of women affected with this cancer within our lifetimes'

Cervical cancer is the second most common cancer in women worldwide, and in 2002 caused about a tenth of all deaths in women due to cancer. Almost all cases of cervical cancer are now thought to result from persistent infection with human papillomavirus (HPV). Mark Schiffman and colleagues review the evidence about the pathogenesis of cervical cancer, and describe strategies for prevention and clinical management, including improved screening tests and vaccines.

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