Report of Two Complicated Anorectal and Genital Malformation: Total Mobilization of Pelvic Organ and Prinea to Midline

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Abstract

We had two cases of female complicated anorectal and genital malformations which underwent total mobilization of all structures of perinea to midline.

Case1: A 6 month old female with skin covered bladder, pubic diastasis, ectopic anus, vagina, and urethra to the right side of median cleft and lipoma on the left side. Closure of pubic diastasis with iliac osteotomy and double barrel sigmoid colostomy was done at 8 months age. In the second operation, right displaced anus, vagina and urethra and the entire muscle complex underwent total mobilization to the midline.

Case2: An 8 Month old female with complicated anorectal malformation who underwent double barrel colostomy on day 8 of birth. Median cleft lipoma was excised and total mobilization of anus, urethra, right hemivagina and muscle complex to the midline was done and the two hemivaginas were sutured together.

Unusual and bizarre anatomic arrangements can be seen in this group. Each case represents a unique challenge for the surgeon, with a different prognosis and therapeutic implications. No general guidelines can be drawn for the management of these patients. Each case must be cared for individually.

Keywords: Anorectal malformation, Genital malformation

Case presentation:

Complex anorectal malformation is a group of unusual heterogonous defects.

In this type of defect, the surgeon may encounter bizarre cases with complex anatomy. Complicated anorectal malformations are correctable but each of them represents a peculiar challenge and will require a great deal of ingenuity and imagination in order to reconstruct them.

We had two cases of female complicated anorectal and genital malformations which underwent total mobilization of all structures of perinea to midline.

Case I: 6 month old female with skin covered bladder, pubic diastasis, ectopic anus, vagina, and urethra to the right side of median cleft and lipoma on the left side. She also had cerebral palsy.

VCUG showed the bladder in the right pelvic with a low capacity and trabeculation with no reflux. Ultrasound and DMSA showed left kidney agenesis.

Endo-anal sonography reported normal internal sphincter and weak external sphincter on the right side. Pubo-rectalis was not seen.

MRI of pelvic without contrast media showed: Partial sacrococcygeal agenesis with fibrolipomatous tissue in the distal part of canal (S2-S3). Deficiency in pelvic muscular floor was seen associated with deformity of the pubic bones since they had almost a parallel course with symphysis pubis. Lower abdominal anterior wall muscles were thin.

Muscle stimulation around the ectopic tight displaced anus was acceptable but there was no muscle in the median cleft and left side. Closure of pubic diastasis with iliac osteotomy and double barrel sigmoid colostomy was done at 8 months age.

In the second operation, right displaced anus, vagina and urethra and the entire muscle complex underwent total mobilization to the midline.

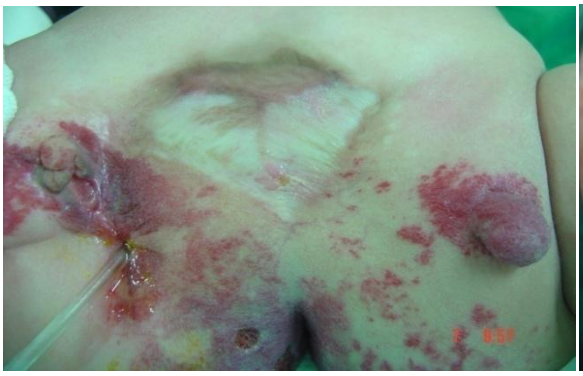

Fig1. Pre operation-Case I

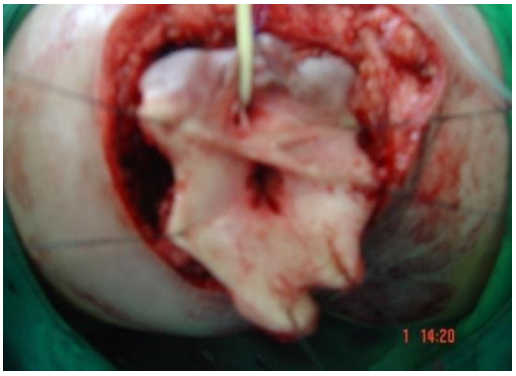

Fig2. During operation-Case I

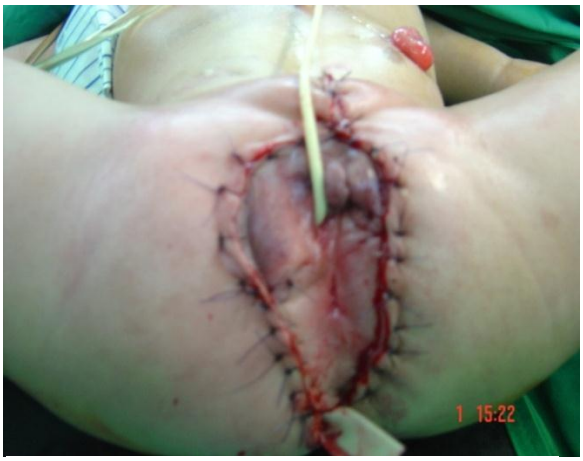

Fig3. Postoperation-Case I

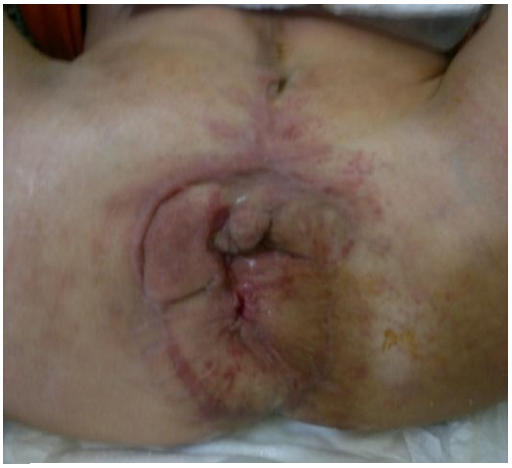

Fig4. Postoperation-Case I

Case II: 8 Month old female with complicated anorectal malformation that underwent double barrel colostomy on day 8 of birth. On examination she had hemivagina, urethra and anus at the right side of median cleft and another hemivagina at the left and lipoma between the two vaginas.

Ultrasound and explorative laparatomy showed left side pelvic kidney and bicorn uterus and duplicated vagina. In the second operation, median cleft lipoma was excised and total mobilization of anus, urethra, right hemivagina and muscle complex to the midline was done and the two hemivaginas were sutured together.

In the third operation the wall between the two hemivaginas was divided by ligature bipolar.

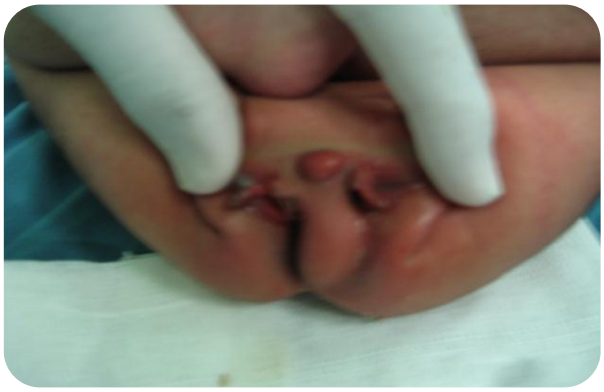

Fig5. Pre Operation-CaseII

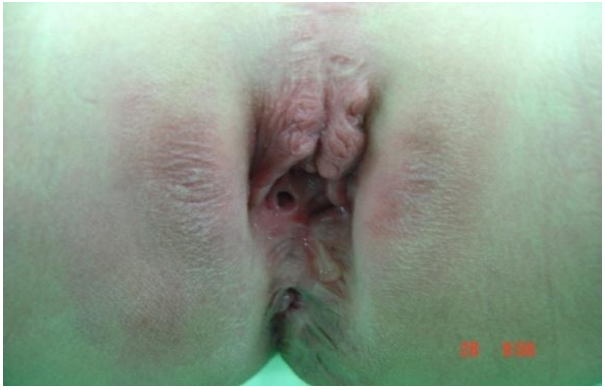

Fig6. Post Operation-CaseII

Discussion

The term anorectal malformation has come to stand for a broad variety of irregularities engaging the termination of the hindgut. Infants with anorectal malformations have a broad range of sickness with conflicting degrees of complexity. Anorectal malformations represent a spectrum of congenital abnormalities where the anus fails to open normally onto the perineum[1]and it occurs in approximately 1 in 5000 live births[2]. The most frequent defect in male patients is imperforate anus, with a rectourethral fistula. The most frequent defect in female patients is rectovestibular fistula[3].

Genital abnormalitiesare very frequent in both male and female anorectal malformation patients. In one report 52% of boys had anomalies of the genitalia [4]. Mullerian abnormalities occur in 30–45 % of girls with anorectal malformation [5]. Cloacal anomalies are characterized by a convergence of the urethra, vagina, and rectum into a solitary conduit, or constant cloaca with a lonely opening on the perineum [6].various amount of septation of the uterus and vagina, varieties from an incomplete septum in a large vagina with single cervix and uterus to an entirely divided double vagina with double cervix and uteri, is seen in 60% of cloaca patients [7].

The management of anorectal malformation patients has been a challenge for paediatric surgeons and the most common outcome is faecal incontinence. As more lasting data is obtained, it becomes clear that urinary incontinence, sexual dysfunction and fertility troubles may also affect patients born with anorectal defects[8].

Management of anorectal malformations necessitates a precise diagnosis at the time of diagnosis, accurate neonatal management with either primary anoplasty or defunctioning colostomy together with treatment of associated malformations. An anatomical rebuilding at the suitable time is fundamental. Postoperatively the management should be multidisciplinary. If the child does not have normal continence then early intervention by bowel and bladder management programs will lead to a child that is clean and dry with an outstanding quality of life, because he or she either possesses the ability for continence or can be kept unnaturally clean. Long-term follow up of these patients into adolescence and adulthood is vital to care for continuing troubles and hold up the patient psychologically as he/she grows.

Unusual and bizarre anatomic arrangements can be seen in this group. Each case represents a unique challenge for the surgeon, with a different prognosis and therapeutic implications. No general guidelines can be drawn for the management of these patients. Each case must be cared for individually.

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