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Case Report

Surgery Section

Anteposed Anus with Scrotal Hypospadias in a Three-Year-Old Child: A Case Report

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ABSTRACT

Anteposed anus and hypospadias are both multifactorial and complex diseases that exist on a spectrum. Hypospadias occurs as a result of abnormal penile development in the foetus between 8 to 14 weeks, while anorectal malformations are often associated with other congenital conditions. The co-existence of these two anomalies is rarely documented, and many cases remain inadequately treated due to a lack of understanding of their causes. Here, the author presented a rare case of a three-year-old male child with anteposed anus and penoscrotal hypospadias. The patient's primary complaints included foecal incontinence, dysuria, irritability, and intermittent abdominal pain. Physical deformities of anteposed anus with scrotal hypospadias were noted since birth. The patient underwent a step-wise approach to management, which involved anorectoplasty followed by Stage-I and Stage-II repair of the hypospadias, with a subsequent colostomy. The patient experienced no adverse events and achieved successful outcomes, including normal passage of urine and stools through the neo-meatus and neo-anus, respectively.

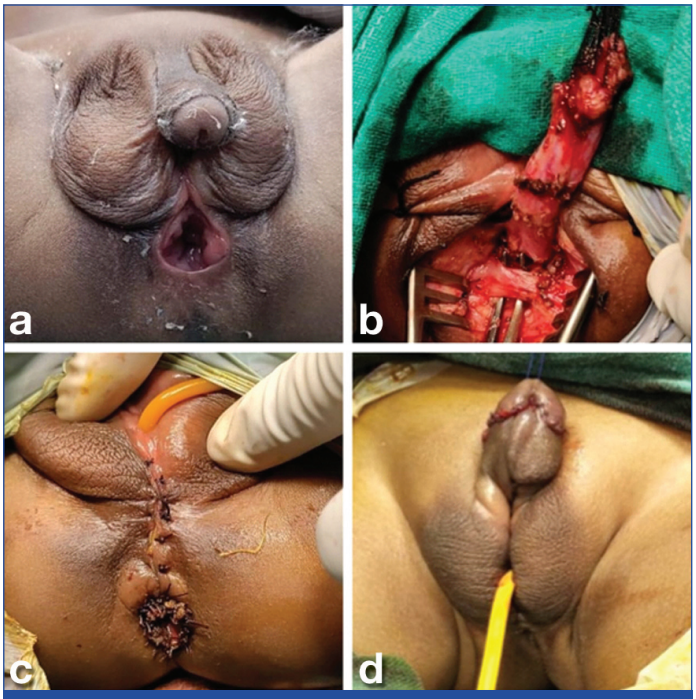
Keywords: Anal malformation, Anorectoplasty, Colostomy, Neo-meatus

CASE REPORT

A three-year-old male child presented with complaints of foecal incontinence, dysuria, irritability, and intermittent abdominal pain, with urine passing from the scrotal opening and no other active complaints since birth. The patient was born to a G1P1A0 mother with no antenatal scan depicting the mentioned condition. Bilateral inguinoscrotal ultrasonography suggested bilateral testes visualised in the bilateral inguinal region. On physical examination, bilateral descended testes, bifid scrotum, and severe chordee with scrotal opening meatus were observed. The history of deformity was reported since birth but had not been treated earlier.

After a physical examination, the patient was planned for elective colostomy surgery. The distal colon was divided into two parts: the proximal part for passing stools, and the distal part remained the same, followed by distal loop washes. After the colostomy was performed, the patient underwent repair of the anorectal malformation after three weeks [Table/Fig-1]. An interposed anus was identified with the help of a muscle stimulator marking the anterior/posterior borders of the sphincter muscles. A circumferential incision was made around the anus and extended posteriorly in the midline upto the posterior limit of the sphincter muscle complex. The incision was deepened in layers, identifying the endorectal fascia and opening the wall of the rectum. The rectum was mobilised circumferentially by dividing fibromuscular bands to an adequate length. The rectum was fixed within the limits of the sphincter muscle complex, which had been identified earlier. Anorectoplasty was performed with Vicryl 3-0. The position was rechecked with a muscle stimulator. The Anterior Sagittal Anorectoplasty Technique (ASARP) was performed in a manner similar to that done in female children, and it was a straightforward surgery without any difficulty. Anal dilatation was started on the 14th postoperative day. Anal dilatation was performed daily using a Hegar dilator, gradually increasing the size up to 12 mm. The excised specimen of the rectum was sent for histopathology, which reported no tissue abnormality [Table/Fig-2].

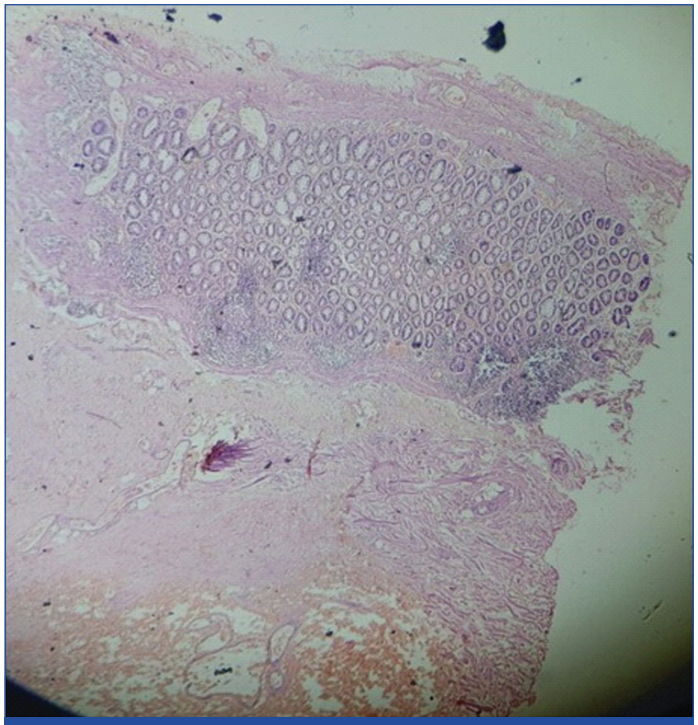
The patient underwent Stage-I hypospadias repair following the anorectal malformation repair. The patient was followed-up for hypospadias Stage-II repair six months after Stage-I repair. The patient did not have undescended testes, which is clearly depicted in the intraoperative photos. Clinically, both testes were present in the scrotum. The patient also underwent colostomy closure and was on laxatives, with a thorough explanation of the high-risk of constipation. Postoperatively, bowel training was initiated for the patient [Table/Fig-3]. The outcome can be reported as positive, as the patient started passing urine through the tip of the neo-meatus and passing stools normally through the neo-anus until the last follow-up at 15 months postoperatively.



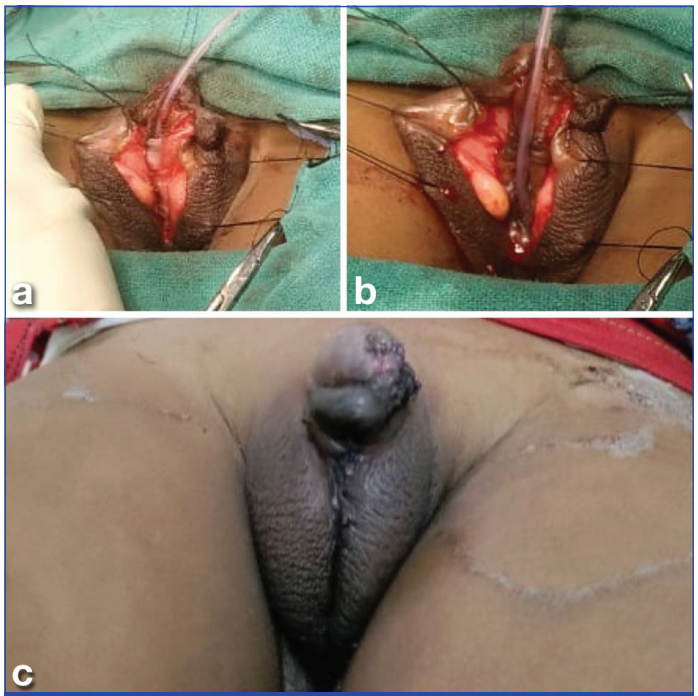
[Table/Fig-1]: a) UL-Preoperative image showing antiposedanal opening; b) UR-External muscle sphincter with the rectum pulled up; c) LL-Postoperative positioned anus after anterior sagittal anorectoplasty; d) LR-Postoperative hypospadias Stage-I repair.

DISCUSSION

Anteposed anus is a common congenital abnormality of the anorectal region, with a significantly higher incidence in females compared to males [1]. This abnormality can be easily diagnosed



[Table/Fig-2]: Histopathology of excised rectum specimen showing unremarkable mucosa, underlying submucosa and glands, deeper tissue-fibrocollagenous muscular tissue with normal ganglion cells {Haematoxylin and eosin (H&E) stain, 4x}.



[Table/Fig-3]: a,b) Intraoperative images of the hypospadias Stage-II repair; c) Postoperative image of hypospadias repair with neo-meatus.

through physical examination. Hypospadias refers to malformations of the ventral opening of the urethral meatus, curvature of the penis (chordee), and distribution of the foreskin, which can vary in severity [1]. The treatment and management of these rare clinical presentations can be approached in a staged manner.

Anteposed anus associated with scrotal hypospadias is a rare anomaly. Most of the available research on hypospadias and anorectal malformations is based on animal models, and there are differences in the mechanisms between animal models and humans [2,3]. This anomaly cannot be classified as a cloacal malformation, as previously reported. However, the incidence of anorectal malformations with hypospadias has been reported to be 8% [4,5]. Since the treatment of hypospadias with anal malformations is poorly described, there is an urgent need for more case presentations in order to develop a customised management approach. In this case, a male with two hypospadias and an anteposed anus with a strip of mucosa joining the urethra and anus was observed. Although, it does not fit the absolute definition of cloaca, it closely resembles a variant of Cloaca-Forme Fruste Cloaca [4]. Chang J et al., have reported some genes and their polymorphisms associated with penoscrotal hypospadias in humans, such as ATF3, BMP4, CDH11, CTNNB1, EMX2, ESR1, ESR2, FGF8, FGFR2, HOXD13, HOXA4, HSD17B3, INSL3, LSM1, MID1, MYRF, NR5A1/SF1, SMADIP1, SRD5A2, SRY, STARD3, STS, TGFBR2 [2].

The urogenital and anorectal canals develop as two separate tubes starting from the 7th week of gestation. The activation of the Sex-Determining Region Y (SRY) gene initiates the differentiation of the genitalia into either testes or ovaries from the 8th week of gestation [6]. Proximal hypospadias can cause micturition problems and limit sexual intercourse and fertility, requiring correction. Hypospadias is related to both the cosmetic and functional aspects of the penis. A 20-year single-center study reported that 10% of cases had a combined presence of anorectal malformation and hypospadias. The study mainly focused on the correlation between the complexity of anorectal malformations and the occurrence of hypospadias, as well as its management [7]. Patients were assessed for occult spinal dysraphism and neurologic lower urinary tract dysfunction before repair. Anorectal malformations with proximal hypospadias were managed using a two-stage repair approach, similar to the present case [7,8].

Patients with a combined presence of anorectal malformations and hypospadias need to undergo extensive evaluation for the presence of related risk factors before any repair is attempted, as it can impact the expected outcomes. Despite extensive research, the basis of penoscrotal hypospadias with an anteposed anus has not been described in the available literature to date. Therefore, the author reported this case as a stepwise approach to management. The repair of both congenital malformations, anteposed anus, and hypospadias, was performed with positive outcomes using a stepwise approach. This approach included colostomy followed by anorectal malformation repair (Anterior Sagittal Anorectoplasty) based on the closest available literature reference and correlation with the current clinical conditions [8].

CONCLUSION(S)

The reported case of rarely found congenital anorectal malformation and its presentation is significant. As more cases are presented and diagnosed, better management approaches and outcomes can be expected. In cases where there are combined clinical presentations of penoscrotal hypospadias with an anteposed anus, early treatment and follow-up may lead to improved living standards and primary functioning. This can be achieved through a stepwise approach with a systematic plan that includes a two-staged hypospadias repair, resolving any urinary complaints.

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