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Case Report

A Case of Anorectal Malformation in 10 Years Old Male Child

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Abstract:

Introduction: Anorectal malformations comprise a wide spectrum of diseases, which can affect boys and girls, and involve the distal anus and rectum as well as the urinary and genital tracts. With early diagnosis, management of associated anomalies and efficient meticulous surgical repair, patients have the best chance for a good functional outcome.

Case History: The present case was a 10 years old male child with chief complaints of inability to pass stools for 6 months. On local examination perineal fistulous opening at 5^{o} clock and 9^{o} clock position was noted. On MRI PELVIS – splaying of posterior element of visualised sacral vertebrae s/o spina bifida and complex fistulous tract extending in prostatic urethra was found. Our patent presented with low position of the blind rectal pouch in relation to the muscle complex of the pelvic floor. Patient was managed by Transposition Anoplasty and advised serial dilation.

Conclusion: Atypical presentations may not necessarily follow conventional treatment approaches, and a high index of suspicion should remain in cases with previous urinary tract infection despite normal imaging. Careful planning is also needed with consideration of a possible need for urethral repair during anoplasty, which was needed in both our cases.

Keywords: Anorectal malformations, cloaca, continence, imperforate anus, Anoplasty.

Introduction

Anorectal malformations comprise a wide spectrum of diseases, which can affect boys and girls, and involve the distal anus and rectum as well as the urinary and genital tracts. They occur in approximately 1 in 5000 live births. [1] It is more common among Asians, as well as males compared to females [2]. The spectrum of anorectal malformation (ARM) ranges from simple anal stenosis to the persistence of a cloaca. Complete /partial failure of the anal membrane to resorb results in an anal membrane or stenosis. [3] The perineal body is formed by fusion of cloacal folds between the anal and urogenital membrane. Breakdown of cloaca causes external anal opening being anterior to the external sphincter (anteriorly displaced anus). If an anocutaneous fistula is observed anywhere on the perineal skin of a boy or an external to the hymen of a girl, a low lesion of ARM can be assumed. The main concerns for the surgeon in correcting these anomalies are bowel control, urinary control, and sexual function. With early diagnosis, management of associated anomalies and efficient meticulous surgical repair, patients have the best chance for a good functional outcome. Fecal and urinary incontinence can occur even with an excellent anatomic repair, due mainly to associated problems such as a poorly developed sacrum, deficient nerve supply, and spinal cord anomalies. [1]

Case History:

A 10 years old male child was brought by parents to surgery OPD with chief complaints of inability to pass stools for 6 months. Distention of abdomen for 4 days. No history of abdominal pain, vomiting, burning micturition, faeces in urine, frothy urine, urine through rectum, shortness of breath, cyanotic spells.

Past History – patient was admitted in GMCH Aurangabad 5 years back with complaints of difficulty in passing stools. Patient was advised surgery but was lost to follow up.

On examination- general condition was moderate, afebrile, pulse rate- 110/min, blood pressure – 128/84, spo2 – 98% on room air.

Systemic examination – CVS – s1s2 heard, no murmur. Per abdomen –abdomen distended.

Palpation – soft, no guarding, rigidity, Auscultation - Bowel sounds present.

On local examination perineal fistulous opening at 5º clock and 9º clock position was noted. No e/o hypospadias, crypto orchidism.

MRI PELVIS – splaying of posterior element of visualised sacral vertebrae s/o spina bifida. Complex fistulous tract extending in prostatic urethra.

USG a+p was within normal limits.

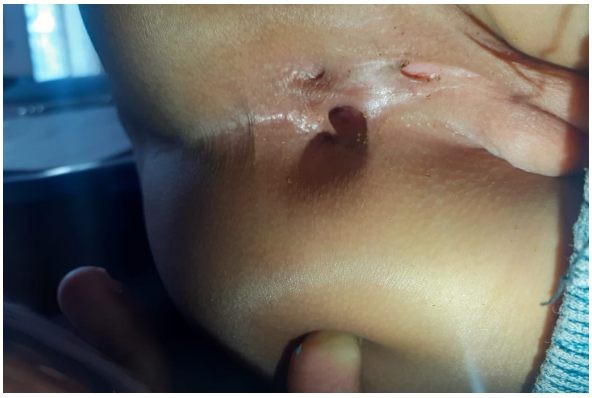
Discussion

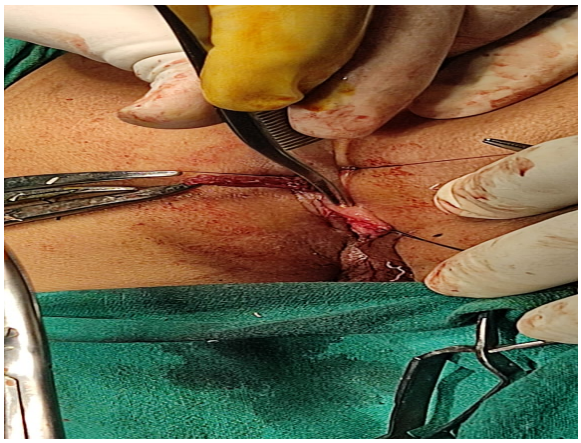
ARMs have been reported from 1 in 2,000 to 5,000 live births, with an inclination toward males. They demonstrate a range of abnormalities that have been reportedly derived from deficient partition of the cloaca and maldevelopment of the perineal mound and genital folds. In male patients, the quick and extensive growth of the perineum usually prevents fistulas from developing at the front of the scrotum, defining low-type anomalies in males, which can present with an enteroperineal or enterourinary tract fistula.[4]

This is a case of late presentation of Ectopic anus with perineal fistula without rectourethral fistula with spina bifida. Patient was evaluated for VACTERAL (vertebral defects, anal atresia,

cardiac defects, tracheoesophageal fistula, renal anomalies, and limb abnormalities) anomalies. ARMs are termed as high, intermediate, or low according to the position of the blind rectal pouch in relation to the muscle complex of the pelvic floor. Our patent presented with low position of the blind rectal pouch in relation to the muscle complex of the pelvic floor. Patient was managed by Transposition Anoplasty and advised serial dilation.

The management of ARM has evolved from classical procedures to PSARP and even minimal invasive approaches with laparoscopic abdominoperineal rectoplasty techniques. [5] Together with these refinements, there has been a paradigm shift in approach to these patients, which involves holistic approach to management of ARM with a goal to achieve complete fecal and urinary continence in conjunction with a favourable quality of life. In terms of the long-term function outcome of males with ARM, it carries a favourable outlook where a study reported a low 15% impaired urinary and bowel function outcome. [6]





Conclusion

ARMs are uncommon but complex congenital anomalies that require an individualized treatment strategy. These treatment strategies are unique to each patient and essential to improve functional outcomes. Rare-variant ARM with rectopenile or

rectoscrotal fistula can be associated with high-type anomalies in contrary to classical rectoperineal fistula. These rare anomalies should be highlighted when a fistula is seen in the perineum, and a diverting colostomy is recommended as the initial surgical procedure prior to definitive operation. Atypical presentations may not necessarily follow conventional treatment approaches, and a high index of suspicion should remain in cases with previous urinary tract infection despite normal imaging. Careful planning is also needed with consideration of a possible need for urethral repair during anoplasty, which was needed in both our cases.

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