Pediatr Surg Int (2003) 19: 559–561
DOI 10.1007/s00383-003-0994-5

CASE REPORT

Amar A. Shah · Anirudh V. Shah

Imperforate anus with rectopenile fistula

Accepted: 25 June 2002 / Published online: 5 September 2003

Abstract We report a 9-month-old child with anorectal anomaly which was surgically treated in the neonatal period for a colostomy and was referred to us for a definitive procedure. The child had features suggestive of low anorectal abnormality; however, investigations indicated that he had a supralevator type of imperforate anus associated with a rectopenile subcutaneous fistula. The surgical treatment of this infant is discussed.

Keywords Rectopenile fistula · Anorectal anomaly

Introduction

Anorectal anomalies is a complex subject, which though commonly encountered, encompasses multiple congenital defects of diverse severity. It is further complicated by the occurrence of rare variants, which are sometimes difficult to classify. An imperforate anus with a fistula in the perineum of a boy is usually treated as a low variety of anorectal malformation, which would comprise of an anocutaneous fistula with a well-formed and normally positioned anorectum. An anoplasty with laying open of the anocutaneous fistula is recommended for such malformations, without any further investigations to locate the rectum [1]. A 9-month-old boy who had been surgically treated for a loop transverse colostomy in the neonatal period was referred to us for a definitive procedure. The child was posted for limited posterior sagittal anorectoplasty (PSARP) where we found that the blind-ending anorectum was located above the levator muscle. The boy turned out to have a rectopenile urethral fistula rather than the usual anocutaneous fistula. We present herein this rare form of anorectal malformation.

A. A. Shah · A. V. Shah
Department of Pediatric Surgery,
K.M. School of Postgraduate Medicine and Research,
N.H.L. Municipal Medical College,
V.S. Hospital, 380006 Ahmedabad, India

A. V. Shah (✉)
"Anicare", 13, Shantisadan Society,
Nr. Parimal Garden, Nr. Doctor House,
Ellisbridge, 380006 Ahmedabad, India
E-mail: anirudhshah@icenet.net
Tel.: +91-79-6442478
Fax: +91-79-6466855

Case report

A 9-month-old boy with imperforate anus was referred to us for a definitive procedure. The child had been surgically treated at the age of 2 days for a transverse loop colostomy. The child had a solitary kidney and no other associated malformations. The child was admitted and distal loop washes were started. On giving washes, some amount of saline was seen escaping through a small opening over the ventral aspect of the penis in the midline (Fig. 1) On reviewing the records, there was no obvious fistula that was noted at the time of the transverse colostomy, which was done by a different surgeon. The invertogram was not available. The child had recovered well post-operatively. Since then, he had been passing stools regularly through the colostomy. The mother also had not noticed any discharge through any abnormal opening below the penis.

Thinking of the possibility of a low anorectal malformation, a distal colostogram was performed which showed a faint opacification of the fistula which was communicating with the opening located over the ventral aspect of the penis, and a very low-placed anorectum (Fig. 2). A cystourethrogram showed a normal urethra and bladder and no communication with the rectum could be demonstrated. A contrast study through the fistulous tract on the undersurface of the penis showed it to be communicating with the blind-ending rectal pouch.

We performed a limited PSARP, thinking of the possibility of a low anorectal malformation with an anocutaneous fistula to be more likely. On exploration, we found the blind anorectum to be supralevator. A formal posterior sagittal anorectoplasty was then carried out with ligation of the rectopenile fistula. During the procedure it was confirmed that the rectum was above the pelvic floor and the rectopenile fistula was in the centre of the sphincter complex. The distal part of the fistula was kept undisturbed. This persisted as a mucous fistula, and was eventually laid open at the time of colostomy closure, following which it healed. The perurethral catheter was kept for 7 days and removed. The colostomy was closed after 4 weeks. The result of the procedure was satisfactory anatomically. Functional results, though hard to assess, appear acceptable for his age. A regular follow-up is being maintained to assess the long-term functional results. The patient is gaining weight and thriving, 3 months after the last surgery.

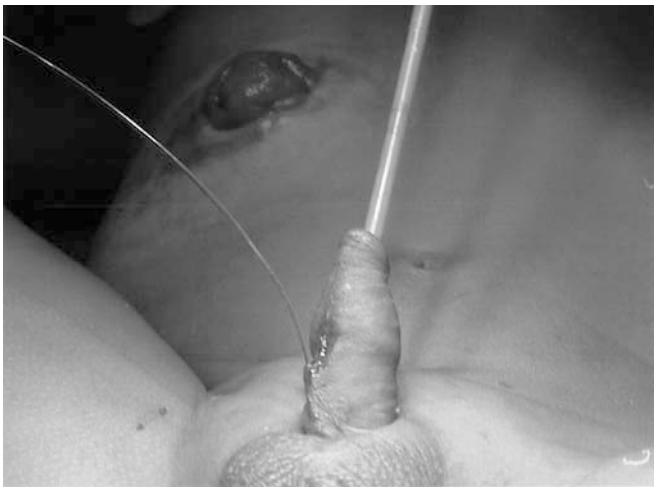

Fig. 1 The probe is placed into the fistulous tract over the ventral aspect of the penis. The catheter is placed perurethrally

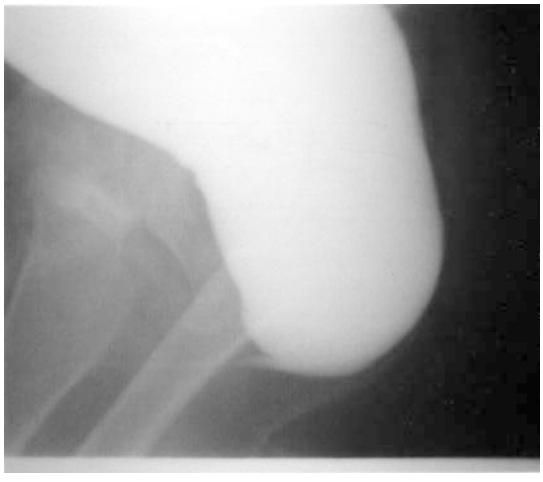

Fig. 2 Distal cologram shows a blind-ending anoectum with a rectopenile cutaneous fistula

Discussion

Most boys presenting with imperforate anus with a perineal fistula are considered to have a low anorectal malformation, and are treated by local anoplasty only. However, as is our case, they can also present as intermediate or high anomaly. The Japan Study Group of Anorectal Malformations in 1982 reported a case with a fistulous opening at the tip of the penis with a rectum ending above the puborectalis sling [2]. The Study group recommended that this case be considered an intermediate type despite the external fistulous orifice.

Various types of rectobulbar cutaneous fistulas have been described [3, 4, 5]. In the first type, i.e., anal agenesis with rectobulbar urethral fistula, a short fistula connects the rectal pouch with the bulb of the urethra just below the membranous urethra. In the second type, which is anal agenesis with anobulbar urethral fistula, a long thin fistulous tract originates from the rectal pouch, passes distally within the corpus spongiosum, and ends in the ventral surface of the bulbar urethra anterior to the openings of the Cowper ducts. In the third type, as is with the case reported herein, a long filiform rectocutaneous fistula contiguous with the corpus spongiosum and extending deep into the scrotum opens in the skin of the ventral surface of the penis without connection with the urethra [6, 7, 8, 9, 10, 11]. The first of this rare anomaly was first described by Nixon in 1970. In a case reported by Stringer [12], the fistula ended in the ventral surface of the penis, near the glans.

Stephens [10] attributed these three anomalies to a maldevelopment of the external cloaca, specifically a defect in the more caudal part of the embryonic urorectal septum and/or genital folds. He also postulated that the anal membrane in the affected fetuses perforates normally, but when the perineal mound is defective, or when the genital folds bordering the cloaca are more posteriorly placed or are hypertrophied, the anus does not migrate backward to the normal anal site; it remains contiguous with the urethral orifice, and both urethral and anal orifices are covered by the fusion of the inner genital folds, projecting the anus forward in a common urethral canal, resulting in a rectobulbar or anobulbar fistula. In cases in which the perineal mound is present but rudimentary, the rectum is rolled even more anteriorly, between the mound and the fused inner genital folds, to form a long slender rectocutaneous fistula running deep into the scrotum and terminating on the undersurface of the penis.

Suspected low anorectal anomalies in boys should be approached with caution. If the perineal fistula can be cannulated easily and dilated, the possibility of a low anomaly would be likely and a local anoplasty would suffice; however, it should be kept in mind that a perineal fistula does not always mean a low anomaly, and in case of any difficulty in identifying or locating the blind-ending rectal pouch, the procedure should be abandoned and either a colostomy or a posterior sagittal approach should be performed rather than risking damage to the sphincteric musculature.

References

1. Cook RCM (1988) Anoectal malformations: neonatal management. In: Spitz L, Nixon H (eds) Rob and Smith's operative surgery. Paediatric surgery, 4th edn. Butterworths, London, pp 348–355
2. Japan Study Group of Anorectal Malformations (1982) A group study for the classification of anoectal anomalies in Japan with comments to the international classification. J Pediatr Surg 17:302–308
3. Stephens FD, Smith ED (1971) Anorectal malformations in children. Year-Book, Chicago
4. Stephens FD, Smith ED (1988) Anoectal malformations in children: update 1988. Liss, New York
5. Asano S, Kitatani H, Konuma K (1983) A newborn with a covered anus complicated by two concomitant unique fistulas. Z Kinderchir 38:258–261

6. Stephens FD, Smith ED (1971) Individual deformities in the male in anorectal malformations in children. Year-Book, Chicago, p 80
7. Smith ED (1988) Incidence, frequency of types and etiology of anoectal malformations. In: Stephens RD, Smith ED (eds) Anoectal malformations in children: update 1988. Liss, New York, p 224
8. Stephens FD (1988) Wingspread anomalies, rarities and super rarities of the anoectum and cloaca. In: Stephens RD, Smith ED (eds) Anoectal malformations in children: update 1988. Liss, New York, p 581
9. Stephens FD, Smith ED (1971) Classifications in anorectal malformations in children. Year-Book, Chicago, p 142
10. Stephens FD (1988) Embryology of the cloaca and embryogenesis of anoectal malformations. In: Stephens RD, Smith ED (eds) Anoectal malformations in children: update 1988. Liss, New York, pp 186–187
11. Stephens FD (1988) Wingspread anomalies, rarities, and super rarities of the anoectum and cloaca. In: Stephens RD, Smith ED (eds) Anoectal malformations in children: update 1988. Liss, New York, p 583
12. Stringer DA (1989) Pediatric gastrointestinal imaging. Decker, Philadelphia, p 375