Case Report

An unusual presentation of rectal atresia with rectal duplication cyst in a neonate: A case report

Prashant S Patil, Paras Kothari, Rahul Gupta, Abhaya Gupta, Geeta Kekre, K Vishesh Dikshit, Ravi Kamble

From, Department of Pediatric Surgery, LTMG Hospital, Sion, Mumbai, Maharashtra, India

Correspondence to: Dr. Prashant S Patil, Ward 1A, Paediatric Surgery Ward, LTMG Hospital, Sion, Mumbai, Maharashtra, India. E-mail: docprashant2010@gmail.com

Received: 07 July 2015 *Initial Review: 20 July 2015* *Accepted: 13 August 2015* *Published Online: 25 August 2015*

ABSTRACT

A 10 days old male baby brought with abdominal distension and cystic swelling in the left buttock. Radiological investigations suggested para-rectal abscess or rectal duplication cyst. Patient underwent transverse loop colostomy and later definitive surgery by posterior sagittal approach. Intra-operative findings were suggestive of small atretic rectal segment with duplication cyst of the rectum. To the best of our knowledge, neonatal rectal atresia with rectal duplication cyst has not been reported in English literature.

Keywords: *Duplication cyst, Meconium, Posterior sagittal anorectoplasty, Rectal atresia,*

Ladd and Gross defined and characterized the alimentary tract duplication as having well-developed muscle coat, epithelial lining and attachment to a portion of gastrointestinal tract [1]. Rectal duplication is a rare congenital anomaly, with less than 100 cases reported in literature [1-8]. Without treatment, a rectal duplication cyst may cause a variety of complications like constipation, bleeding, pressure effect on bladder and rarely transforming into a malignancy. Thus, clinician should be aware of different clinical presentations and the subsequent management so as to avoid undue morbidity and mortality associated with delayed diagnosis and treatment. Excision of rectal duplication cyst is the treatment of choice preferably by standard posterior sagittal approach.

CASE REPORT

A 10 day old male infant presented with increasing constipation for five days and swelling in left buttock since birth. Baby was passing meconium intermittently though not in adequate amount. No other associated gastrointestinal and urinary symptoms were present. The general physical and chest examination revealed no abnormality. The abdominal examination revealed non-tender, distended abdomen with normal bowel sounds. Baby had swelling in the left buttock which was cystic with blue-greenish tinge of overlying skin (fig 1). Anus was in normal location and had normal appearance. Little finger could be passed through anal opening. Sacral examination was normal, all sacral pieces were present.

Routine blood investigations, renal and liver function tests were normal. Abdominal erect X-ray suggested gaseous distension of bowel loops. Ultrasound examination of the gluteal region revealed fluid collection (3.6 X 6.3 X 1.4 cm) with internal echoes on left ischiorectal area in subcutaneous plane extending down upto rectum and a thin tract seen communicating with fluid collection suggestive of Para rectal abscess. To get better idea regarding diagnosis and pelvic anatomy, MRI pelvis was done.

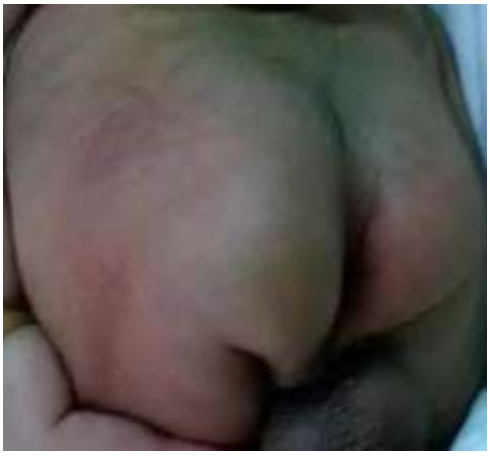

Figure 1 - Showing left buttock swelling with bluish-green tinge over skin.

MRI pelvis revealed 3.6 x 1.8 x 1 cm extrasphincteric thin walled collection looking like abscess in left ischiorectal fossa extending to subcutaneous plane abutting anal canal or it could be rectal duplication cyst. Needle aspiration of cystic swelling was performed which showed the presence of meconium.

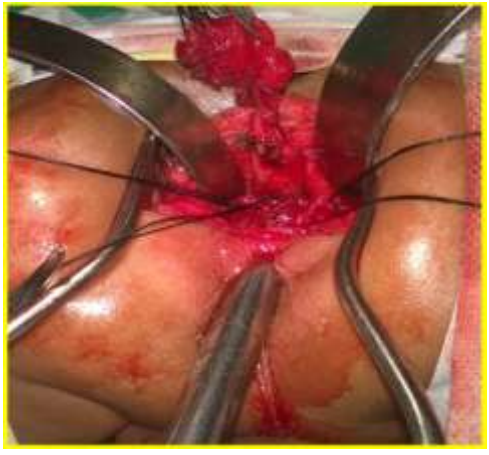

Figure 2 - Intraoperative photo of atretic rectal segment with cyst.

We decided to perform diversion transverse loop colostomy to relieve abdominal distension. Patient's left buttock swelling decreased after surgery. Patient was kept on regular follow-up and distal loop stoma washes were started. Distal loop dye study was done to know the level of distal lumen obstruction. We decided for exploration by posterior sagittal approach. Intra-operatively, a short segment of atretic rectum with duplication cyst posterior to rectum (fig. 2) was found.

Involved segment of atretic rectum with rectal duplication cyst was excised and end to end rectal anastomosis was done over rectal tube. Histo-pathological examination of the specimen revealed colonic type of epithelium with single layer of muscle in the wall of cyst consistent with the duplication cyst of the rectum. No evidence of ectopic mucosa found. Transverse loop stoma was closed after 6 weeks. Patient is on regular 2 monthly follow-up and doing well.

DISCUSSION

Present case is a rare case of rectal duplication cyst presenting in a neonate with atypical presentation as cystic swelling in left buttock and causing subacute intestinal obstruction. Radiological studies including MRI pelvis could suggest presence of rectal duplication cyst and it was confirmed intra-operatively. Rectal atresia is a rare condition with reported incidence of 0.3–1.2% of all anorectal malformations [1]. Rectal atresia is characterized by presence of proximal rectum which ends at or above pubococcygeal line (PC line) and well formed distal anus that is in its normal location and has a normal appearance. Usually, there is no fistula between the rectum and the urethra or vagina [1, 2].

Rectal duplication, as described by Ladd and Gross is an extremely uncommon congenital anomaly in infants constituting about 2 % of all gastrointestinal duplications [3,5,7,9]. The exact etiology of rectal duplication cysts is not known and several theories have been proposed. A split notochord may explain some, but not all duplications [11]. More rare causes include intrauterine trauma or hypoxia. Based on their embryogenesis, rectal duplication cysts are almost always located posterior to the rectum. There are only four reported cases of anterior rectal duplication cysts [12].

Associated congenital genitourinary and vertebral abnormalities which may be present in patients with rectal duplication cysts include double bladder, double external genitals, double uterus or vagina, hemivertebra, separation of the pubis symphysis and cardiac abnormalities [13]. While the patient discussed in this case report did not have any of these associated abnormalities, presence of one or more of these malformations should prompt further evaluation for a potential rectal duplication cyst. The presence of perianal swelling, perineal fistula, presacral mass, intestinal prolapse as 'polyp', bleeding per rectum

and symptoms of lower gastrointestinal obstruction due to mass effect are the common clinical presentations [3,6,10].

Treatment of rectal duplication is primarily surgical and it aims at complete excision with preservation of anorectal functions [3, 6-8]. An early surgical intervention is recommended so as to avoid complications like bleeding, infection and malignant degeneration [3, 6]. The various surgical approaches recommended for rectal duplications include transanal, transcoccygeal (Kraske) and posterior sagittal [3,7,8]. However, as experienced in the present case, the posterior sagittal approach provides the best surgical access, thus facilitating the complete excision of rectal duplications with preservation of anorectal function.

CONCLUSION

The duplication cysts of rectum are rare and presentation in a neonate as in this case underlines the fact that clinician has to be aware of this entity and its varying presentation so as to manage it properly.

REFERENCES

1. Pena A. Anorectal malformations. Semin Pediatr Surg. 1995; 4(1): 35–47.
2. Uphadaya P. Rectal atresia. In Newborn Surgery, Puri P (ed), Oxford, UK. 1996: 395–398.
3. Bentley JF, Smith JR. Developmental posterior enteric remnants and spinal malformations: the split notochord syndrome. Arch Dis Child. 1960; 35(2): 76-86.
4. Stringer MD, Spitz L, Abel R, Kiely E, Drake DP, Agrawal M, et al. Management of alimentary tract duplications in children. Br J Surg. 1995; 82(1): 74-8.
5. Rajah S, Ramanujam TM, Anas SR, Jayaram G, Baskaran P, Ganesan J, et al . Duplication of rectum: Report of four cases and review of literature. Pediatr Surg Int. 1998; 13(5-6): 373-6.
6. Mehta MH, Mehta SG, Trivedi PS, Patel RV. Pararectal duplication. Indian J Med Sci. 1992; 46(3): 83-5.
7. Amjadi K, Poenaru D, Soboleski D, Hurlbut D, Kamal I. Anterior rectal duplication: A diagnostic challenge. J Pediatr Surg. 2000; 35(4): 613-4.
8. Delarue A, Garcia Meric P, Martin C, Piguet C, Andre N, Galli G, et al. Antenatal rupture of diverticular rectal duplication with neonatal perineal fistualization. Pediatr Surg Int. 1998; 13(4): 288-9.
9. Park WH, Chai SO, Park KK. Cystic rectal duplication: A rare cause of neonatal bladder outlet obstruction and hydronephrosis. Pediatr Surg Int. 2001; 17(2-3): 221-3.
10. Mboyo A, Monek O, Massicot R, Martin L, Destuynder O, Lemouel A, et al . Cystic rectal duplication: A rare cause of neonatal intestinal obstruction. Pediatr Surg Int. 1997; 12(5-6): 452-4.
11. Bremer JL. Diverticula and Duplications of the Intestinal Tract. Arch Pathol Lab Med. 1944; 38: 132-140.
12. Ravitch MM. Hind gut duplication: Doubling of colon and genital urinary tracts. Ann Surg. 1953; 137(5): 588-601.
13. Yousefzadeh DK, Bieckers GH, Jackson JH Jr, Benton C. Tubular Colonic Duplications: Review of 1876-1981 Literature. Pediatr Radiol. 1983; 13(2): 65-71.

Funding: None; Conflict of Interest: None Stated

How to cite this article: Patil PS, Kothari P, Gupta R, Gupta P, Kekre G, Dixit KV, Kamble R. An unusual presentation of rectal atresia with rectal duplication cyst in a neonate: A case report. Indian J Case Reports. 2015; 1(2): 47-49.