Fetal extraperitoneal rectal perforation: a case report

Saurabh Garge and Anil Chacko

Fetal extraperitoneal rectal perforation is a very rare condition, but prompt diagnosis and appropriate treatment produce overall good outcome. Its etiology and pathophysiology are poorly understood. Only 16 cases have been reported worldwide. We report another case of fetal extraperitoneal rectal perforation managed by us. *Ann Pediatr Surg* 13:104–106 © 2017 Annals of Pediatric Surgery.

Annals of Pediatric Surgery 2017, **13**:104–106

Keywords: extraperitoneal, fetal, rectal perforation

Department of Pediatric Surgery, Choithram Hospital & Research Centre, Indore, Madhya Pradesh, India

Correspondence to Saurabh Garge, MS, MCh, Consultant, Department of Pediatric Surgery, Choithram Hospital & Research Centre, Indore 452010, Madhya Pradesh, India
Tel: +91 963 095 6311; fax: +91 731 420 6707;
e-mail: saurabhgarge8@gmail.com

Received 11 February 2016 **accepted** 22 June 2016

Introduction

Fetal extraperitoneal rectal perforation (FERP) is very rare, but it presents with a classical clinical appearance, with all babies having an immediately recognizable abnormality at birth [1]. It typically presents in neonates with a gas-filled or meconium-filled perineal or buttock swelling, leading to a risk of skin rupture, intraperitoneal perforation, and systemic sepsis [2]. Early diagnosis and referral is the only variable on which the prognosis and survival depends [1–9]. Only 16 cases have been reported of this rare entity previously [1–9]. If treated timely, this disease is associated with good prognosis and normal function.

Case report

A 2.1-kg full-term boy born by Cesarean section was admitted to the neonatal intensive care unit with gradual progressive abdominal distension along with increasing scrotal swelling since birth. He had not passed meconium since birth. He did not have any history of rectal washes or rectal instrumentation. On examination, he had very feeble pulses with delayed capillary refill time and sclerema over both lower limbs. Both buttocks had fluctuant swellings with skin discoloration (Fig. 1). Systemic examination showed mild abdominal distension with absent bowel sounds. On local examination, right scrotal swelling was present, testis was not palpable separately, and fluctuation was present but the skin over the scrotum was normal (Fig. 1). The left testis was normal. Aspirate from the scrotum revealed meconium, vernix, and lanugo. Anal opening was normal. Fecal staining was seen on inserting a 10 Fr infant feeding tube per rectum. Nasogastric aspirate was bilious in nature. Erect abdominal radiograph did not show any gas under the diaphragm; however, gas was seen lining the scrotal walls (Fig. 2). Abdominal ultrasound was normal. Scrotal ultrasonography revealed heterogeneous mixed echogenic collection in both tunica and extending to both inguinal canals bilaterally (Fig. 3). Perineal ultrasonography was also suggestive of the presence of echogenic contents. Exploratory laparotomy revealed a posterolateral extraperitoneal rectal perforation below peritoneal reflection. A diverting divided sigmoid colostomy along with drainage of bilateral gluteal abscess was done. The patient tolerated the procedure well and was kept on broad spectrum antibiotics. Colostomy was functional on postoperative day 2, and the neonate was started on feeds. The feeds were increased gradually and the patient was discharged on the seventh postoperative day. The patient awaits a distal loopogram, based on which further course will be decided.

Discussion

Previously reported cases have demonstrated the whole spectrum of FERP [1–9]. The extravasated meconium and air can go in any direction. If it goes anteriorly it can present as rectourethral fistula [10]. If it goes posteriorly, it may present in the form of a neuroenteric fistula or sinus [11]. If it goes in the infralevator direction, it presents in the form of scrotal and buttock swelling and perineal sepsis [1–9]. If it goes superiorly, it can present in the form of neonatal peritonitis [1].

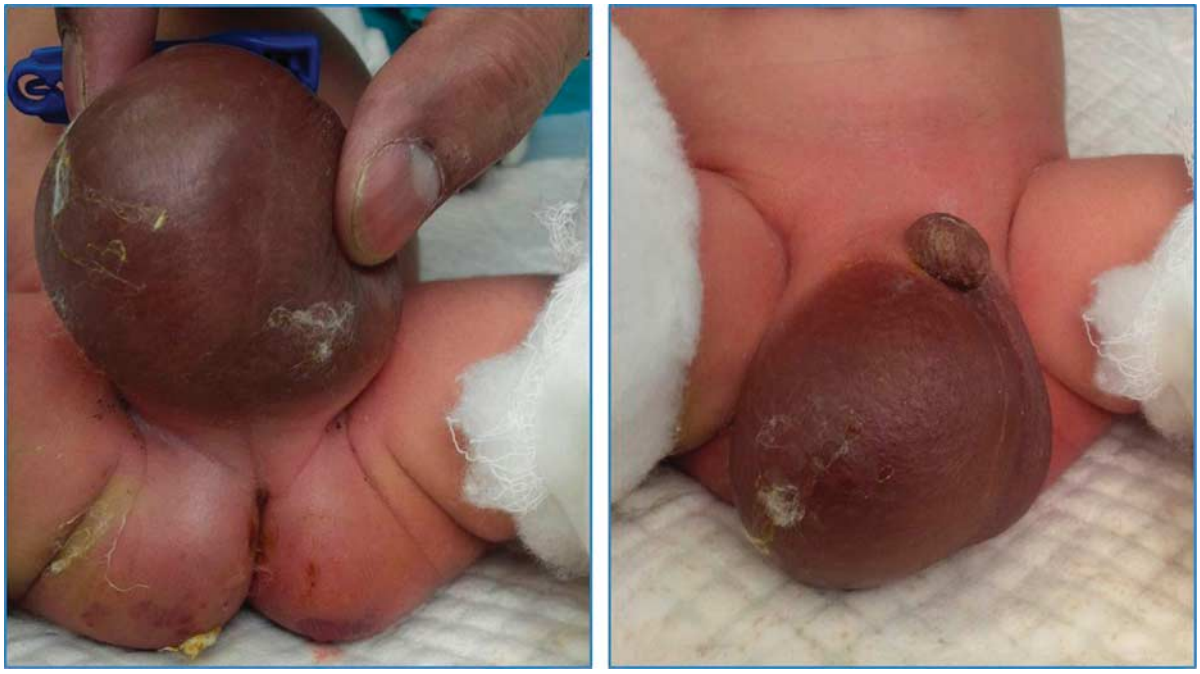
The presentation depends upon the direction of the spread of meconium [1–9]. Most commonly, it resents as a buttock, perineal, or scrotal mass with or without fistulization [1–9]. The buttock mass can be left sided, right sided, or midline posteriorly [1,3,8]. In the series of Pitcher *et al.* [1], two cases of FERP with swelling in the posterior midline were referred as meningomyelocoele and sacrococcygeal teratoma. It was also observed that intraperitoneal extension was more frequently associated with severe sepsis and difficult postoperative course [1]. The swelling can be soft cystic to firm induration [1,3]. It can resent as frank fistulization, skin discoloration, or aplasia cutis [1–9].

Several theories are advocated for explaining FERP embryologically [1,2,9]. One theory states that the perforation may occur because of a late ischemic event [1]. This is based on the fact that the perforation site in many cases was situated at the region of the watershed line between the supply of middle and inferior rectal arteries. This theory is refuted by the fact that there is a good transmural blood supply of the rectum [1,9].

Another theory states that FERP may be caused by forceful peristalsis against a distal obstruction [1,9]. The presence of huge postnatal swellings filled with meconium and air were thought to be in agreement with this

1687-4137 © 2017 Annals of Pediatric Surgery DOI: 10.1097/01.XPS.0000489150.40422.c6

Fig. 1



Scrotal and buttock swelling skin discoloration.

hypothesis. However, distal obstruction could not be demonstrated in any of the previous studies [1–9]. Furthermore, there was an FERP demonstrated in a child with duodenal atresia, which precludes above theories [9]. In case of proximal atresia, it is unlikely that intestinal transit would have generated enough pressure to cause rectal perforation.

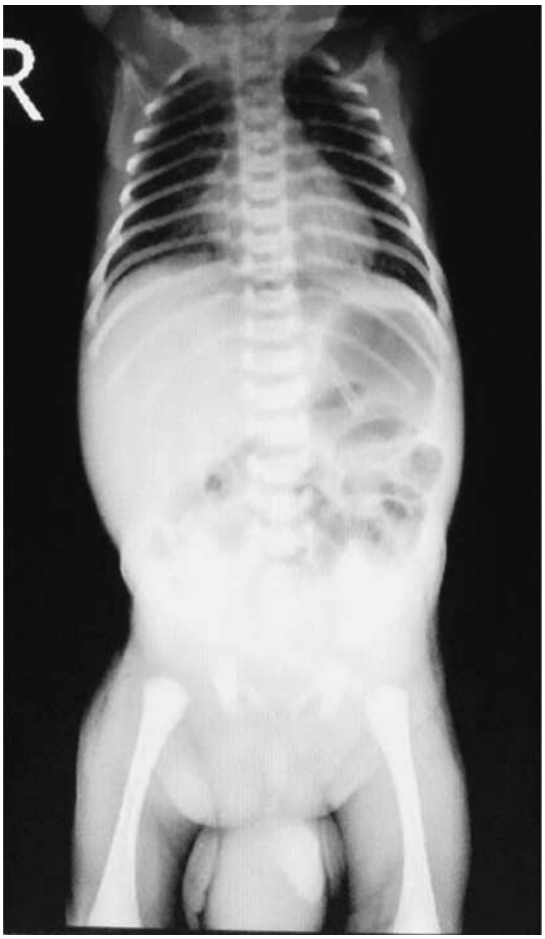
Another theory, proposed by Davies *et al.* [2], involves herniation of the supra-levator rectum via a pelvic floor defect. This may lead to localized ischemic insult and perforation, similar to that seen in Richters hernia [1,2,9]. Buttock swelling from a perineal hernia through a levator ani defect has been reported previously [12]. There have also been similar presentations due to rupture of rectal diverticular duplications [6].

Apart from the embryological causes, rectal perforation has been also reported because of rectal thermometers or probes and also because of amniocentesis [13].

Many previous cases are an evidence that the preceding event is a late ischemic one [1–9]. In most of the cases, perineal swelling was apparent within few hours of birth [3]. In a case of Pitcher *et al.* [1], the patient even had aplasia cutis, suggesting skin perforation in the buttock region antenatally.

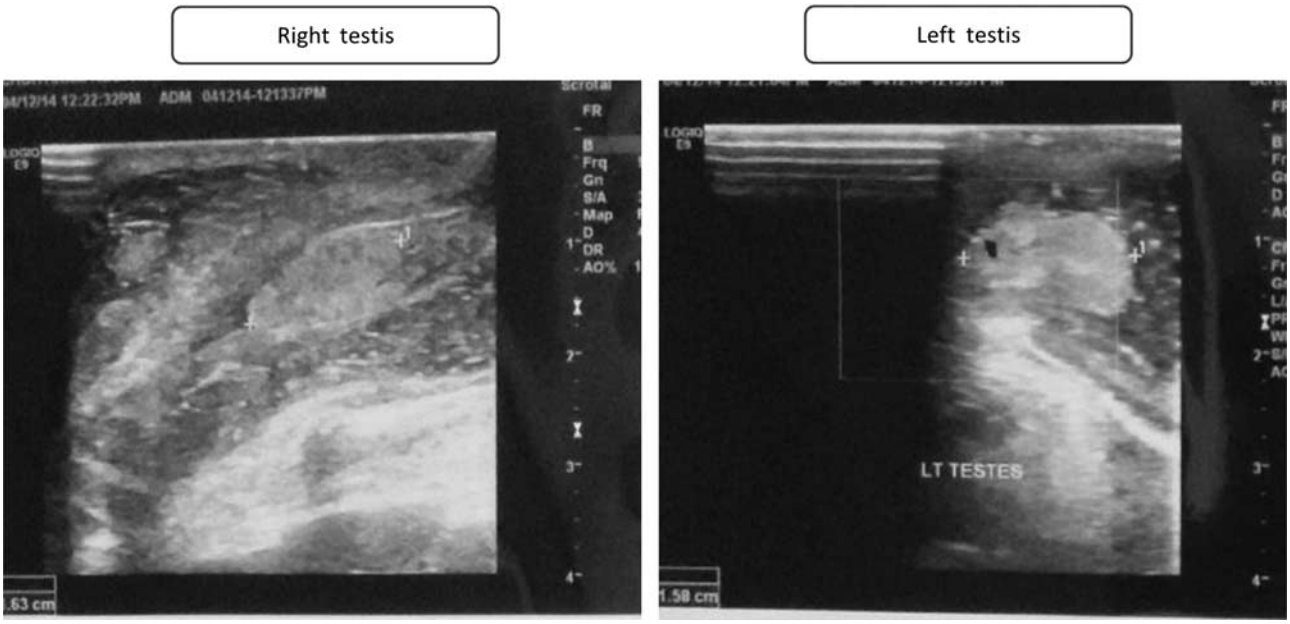
Investigations recommended depend upon the presentation [1–9]. Most of the clinicians will be encountering this rare entity for the first time. Thus, diagnosing this entity purely on clinical grounds may be difficult [3]. Initial diagnostic imaging modalities recommended are an ultrasound of the perineum, pelvis, and abdomen, and a contrast study of the rectum [3]. The typical findings on ultrasound are of a cavity containing echogenic material

Fig. 2



Erect radiograph showing gas in the scrotum.

Fig. 3



Ultrasound images showing echogenic contents in both tunica (right>left).

with anterior displacement of the rectum and compression of the bladder [1,3]. Aspiration of the material from perineal swelling may show the presence of lanugo, verni, and meconium. The contrast enema may or may not show a perforation depending on whether the perforation has had time to heal over by the time of birth [1–9]. It also depends upon the size of the rectal perforation. In cases with neuroenteric fistulization, MRI may be required [11].

Treatment usually involves initial resuscitation and higher antibiotics [1–9]. Because of the rarity, late diagnosis and late referrals are common. The patients are usually in sepsis. After proper resuscitation, exploratory laparotomy with fecal diversion in the form of a sigmoid colostomy and perineal drainage is recommended [1–9]. In case of peritoneal extension, proper peritoneal toilet is advocated. In cases of neuroenteric fistulae, a laminectomy and debridement are recommended [9,11]. The perineal sepsis usually resolves promptly without apparent damage to the anal canal or sphincteric mechanism [1–9].

A follow-up distal loopogram to rule out distal obstruction and fistulization is recommended. If normal, closure of the fecal diversion is done [1–9]. Most of the cases reported previously had no problems related to distal obstruction and continence. The survival for this entity is good with achievement of normal function if the diagnosis is made timely.

Conclusion

Antenatal perforation of the rectum is a rare entity, and most clinicians encountering this condition will be doing so for the first time. The timing of presentation is usually within hours of birth. Given that delay in diagnosis is associated with increased morbidity such as generalized peritonitis, it is essential that diagnosis is immediate to expedite transfer to a surgical center.

Conflicts of interest

There are no conflicts of interest.

References

1 Pitcher GJ, Davies MR, Bowley DM, Numanoglu A, Rode H. Fetal extraperitoneal rectal perforation: a rare neonatal emergency. *J Pediatr Surg* 2009; **44**:1405–1409.
2 Davies MR, Cywes S, Rode H. Prenatal perforation of the extraperitoneal part of the rectum, associated with a developmental defect of the pelvic floor. *Z Kinderchir* 1984; **39**:271–273.
3 Sundararajan L, Patel D, Jawaheer G. Antenatal rectal perforation presenting in the neonate. *Pediatr Surg Int* 2008; **24**:601–603.
4 Mitsudo SM, Boley SJ, Rosenzweig MJ, Campbell DE. Extraperitoneal pelvic meconium extravasation in a newborn infant. *J Pediatr* 1983; **103**:598–600.
5 Dickinson SJ, Moumgis B. Prenatal volvulus with pseudocyst perforating the perineum. *J Pediatr Surg* 1966; **1**:288–291.
6 Delarue A, Garcia-Meric P, Martin C, Piguet C, André N, Galli G, Guys JM, *et al*. Antenatal rupture of a diverticular rectal duplication with neonatal perineal fistulisation. *Pediatr Surg Int* 1998; **13**:288–289.
7 West DK, Touloukian RJ. Meconium pseudocyst presenting as a buttock mass. *J Pediatr Surg* 1988; **23**:864–865.
8 Lockwood C, Ghidini A, Romero R, Hobbins JC. Fetal bowel perforation simulating sacrococcygeal teratoma. *J Ultrasound Med* 1988; **7**:227–229.
9 Tan YW, Motiwale S. Fetal extraperitoneal rectal perforation presenting after duodenal atresia repair. *J Pediatr Surg* 2010; **45**:2447–2449.
10 Waldbaum RS, Glendinning AF. Tubular duplication of the rectum with a rectourethral fistula. *J Urol* 1975; **113**:876–879.
11 Darwish B, Stanley TV, Wickremesekera A, Bowkett B, Leadbitter M. Presacral neuroenteric fistula in a newborn presenting with an epidural abscess: case report and review of the literature. *Pediatrics* 2004; **114**:527–531.
12 Doig CM, Nixon HH. Pelvic hernias in children. *J Pediatr Surg* 1972; **7**:44–47.
13 Casaccia G, Giorlandino C, Catalano OA, Bagolan P. Prenatal rectal perforation: an unsuspected cause of isolated ascites. *J Perinatol* 2006; **26**:717–719.