Case Report

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Idiopathic gastric perforation in an asplenic infant

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ABSTRACT

Introduction: The cause of idiopathic gastric perforations in neonates remains unknown. Perforations of the abdominal oesophagus, stomach and duodenum in infants and children are the rarest type of intestinal perforations. There are 21 reported cases of an idiopathic gastric rupture in non-neonates. **Case Report:** A 6-week-old boy presented with nausea, vomiting and decreased oral intake. Physical examination revealed a firm, distended abdomen tender. Abdominal lateral decubitus radiograph revealed massive free air. Patient was found to have an idiopathic gastric perforation of the greater curvature with incidental findings of asplenia. **Conclusion:** Paediatric abdominal foregut perforations have diverse aetiologies. The foregut is the least likely portion of the gastrointestinal tract to perforate in children. The aetiology is unknown but there are many postulated theories. We present a case of an idiopathic gastric rupture involving the greater curvature of 6-week-old boy with incidental findings of being asplenic.

Key words: Asplenia syndrome, idiopathic gastric perforation, foregut perforation

INTRODUCTION

Idiopathic gastric perforation is a rare, serious event in neonates associated with poor prognosis. Its aetiology is controversial with many postulated theories. Neonatal gastric perforation was thought to occur spontaneously without any associated gastrointestinal conditions and was originally thought to be caused by a congenital absence of gastric musculature.[1] Prematurity and NSAIDS has been shown to be a predisposing risk factor. However, it is rarely seen in infants. There are 21 reported cases of an idiopathic gastric rupture in non-neonates.[2-5] We present a case of an idiopathic gastric rupture involving the greater curvature of 6-week-old boy with incidental findings of asplenia.

CASE REPORT

A 6-week-old boy presented with 1-day history of vomiting and decreased oral intake. Family noticed an episode of non-projectile, non-bilious emesis after a feeding. He was offered but refused further feedings. The following day, patient continued to refuse feedings and his family decided to take him to the emergency department. He was born full term through spontaneous vaginal delivery without any complications. There were no abnormalities with prenatal ultrasounds. Patient neither required admission to the neonatal ICU nor received any nasogastric feedings. Family history was significant for trisomy 18 in an older sibling. Physical examination revealed a firm, distended abdomen tender to palpitation. Abdominal ultrasound showed no pyloric stenosis, whereas a lateral decubitus abdominal radiograph revealed massive-free air [Figure 1]. Patient

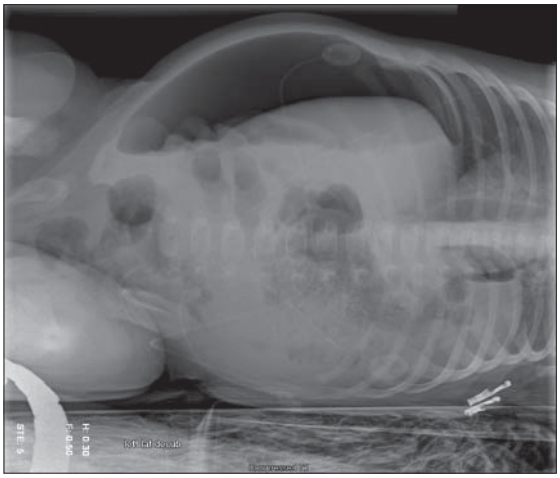


Figure 1: Lateral decubitus abdominal x-ray showing massive free air

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was taken emergently for an exploratory laparotomy. The abdomen was explored through a right upper quadrant transverse incision extending across the midline. Exploration revealed necrosis involving the greater curvature of the stomach. The stomach was found not to be volvulised. Duodenal rotation was normal. The small bowel was eviscerated and found to be free of any signs of perforation or obstruction. The caecum and appendix were identified in the right lower quadrant. The left upper quadrant and tail of the pancreas were inspected and no spleen was identified. The omentum and greater curvature were biopsied and sent off as a specimen. No signs of infection were appreciated and no cultures were obtained. The greater curvature of stomach was imbricated with interrupted 3-0 Vicryl sutures. Two anterior gastropexy sutures were placed to keep the stomach from twisting, although no signs of a gastric volvulus were appreciated. He was taken back for a re-exploration the following day for his acidosis. The stomach was found to be viable and pink. No areas of necrosis were found. Patient was transferred to the intensive care unit. Postoperatively, patient had a complicated course. He developed a wound infection with *Pseudomonas* on post-operative day # 8 and placed on appropriate antibiotics. Patient was extubated on post-operative day # 10. An upper GI gastrografin study showed no overt extraluminal contrast on post-operative day # 14 and his diet was advanced without difficulty. Pathology report showed necrotic smooth muscle and fibrofatty connective tissue consistent with history of gastric perforation. There were no signs of *Helicobacter pylori* infection on pathology. Patient was placed on oral antibiotics upon discharge to prevent overwhelming post-splenectomy infection (OPSI). Family was educated on possible symptoms of OPSI with instructions to seek medical attention if should ever occur.

DISCUSSION

The cause of idiopathic gastric perforation in neonates remains unknown, with many theories advocating explaining its aetiology. Risk factors predisposing neonates to idiopathic perforations include prematurity, NEC and NSAIDS usage.[6] Perforations of the abdominal oesophagus, stomach and duodenum in infants and children are the rarest type of intestinal perforations.[1] Spontaneous gastric perforation was considered a congenital anomaly by Herbut in 1943.[7] He explained these findings as congenital agenesis of the musculature on the greater curvature, such that mucosa is covered by serosa without smooth muscle; ulcerations easily occur, with no muscle to protect the mucosa.[1] Shaw *et al*. in 1965 experimentally disproved this theory in dogs by tying off the oesophageal and duodenal ends of the stomach and insufflating with air until the organ perforated. He found that all perforations occurred on the greater curvature, and when examined histologically, all specimens showed lack of musculature near the perforation.[8] Holgerson in 1981 repeated Shaw's experiment on 10 post-mortem newborn infants' stomachs and found the identical gross findings.[9] Currently, there are 21 reported cases of idiopathic gastric perforations in non-neonates in the literature. Review revealed that there was a higher incidence in females with the most common site of perforation along the greater curvature of the posterior gastric wall.[3]

The aetiology of idiopathic gastric perforation is still controversial. The most recent findings suggest that lack of interstitial cells of Cajal (ICC) may be responsible.[10] The ICC expresses tyrosine kinase receptor by the name of C-kit which functions as gastrointestinal pacemaker cells.[11] Stem cell factor, a ligand of C-kit, is important for immune system homeostasis in the gastrointestinal tract. Lack of C-kit could impair the immune response and contribute to bacterial overgrowth and invasion of the gastric wall.[12] Thus, perforation may result from a combination of hypomotility causing gastric distention and bacterial invasion of the weakened gastric wall.[11] A popular theory is the closed loop phenomenon. An over-distended stomach causes an acute decrease in the angle of His creating a one-way valve by occluding the gastro-oesophageal junction with the fundus and right crus.[3]

Our patient did not have any of the risk factors for necrotising enterocolitis. Pneumatosis intestinalis was not seen on abdominal radiographs and no evidence of obstruction was seen at laparotomy. Based on the incidental findings of asplenia, a cardiac workup was performed. Asplenia syndrome, also called Ivemark syndrome, heterotaxy syndrome, cardiosplenic syndrome, right isomerism or bilateral right-sidedness, is associated with a centrally located liver, absence of spleen, two morphologic right lungs and congenital heart disease.[13] No abnormalities were found on echocardiogram or chest X-ray. It is unknown whether the incidental finding of asplenia lead to or could have attributed to an idiopathic gastric perforation. Follow-up visit revealed a well-healed incision and no complications.

CONCLUSION

Paediatric abdominal foregut perforations have diverse aetiologies. Half occur in neonates but seem evenly

distributed between premature and term babies. The foregut is the least likely portion of the gastrointestinal tract to perforate in children. The aetiology is unknown but there are many postulated theories. This is a unique case of an idiopathic gastric perforation in a 6-week-old asplenic boy.

REFERENCES

1. Leone RJ Jr, Krasna IH. 'Spontaneous' neonatal gastric perforation: Is it really spontaneous? J Pediatr Surg 2000;35:1066-9.
2. Abadir J, Emil S, Nguyen N. Abdominal foregut perforations in children: A 10-year experience. J Pediatr Surg 2005;40:1903-7.
3. Shimizu T, Horiuchi T, Hirooka S, Yonekura T, Tamai H. Idiopathic gastric rupture in a 3-month-old girl. Acta Paediatr 2003;92:628-30.
4. Libeer F, Vanhamel N, Huyghe M, Verlinden E. Spontaneous gastric rupture in non-neonatal children: A case report. Acta Chir Belg 2007;107:560-3.
5. Guven A, Demirbag S, Atabek C, Ozturk H. Spontaneous gastric perforation in a child with Burkitt lymphoma. J Pediatr Hematol Oncol 2007;29:862-4.
6. Kawase Y, Ishii T, Arai H, Uga N. Gastrointestinal perforation in very low-birthweight infants. Pediatr Int 2006;48:599-603.
7. Herbut PA. Congenital defect in the musculature of the stomach with rupture in a newborn infant. Arch Pathol 1943;36:191-4.
8. Shaw A, Blanc WA, Santulli TV, Kaiser G. Spontaneous rupture of the stomach in the newborn: A clinical and experimental study. Surgery 1965;58:561-71.
9. Holgerson LO. The etiology of spontaneous gastric perforation of the newborn: A reevaluation. J Pediatr Surg 1981;16(4 Suppl 1):608-13.
10. Kara CS, Ilçe Z, Celayir S, Sarimurat N, Erdogan E, Yeker D. Neonatal gastric perforation: Review of 23 years experience. Surg Today 2004;34:243-5.
11. Ohshiro K, Yamataka A, Kobayashi H, Hirai S, Miyahara K, Sueyoshi N, *et al*. Idiopathic gastric perforation in neonates and abnormal distribution of intestinal pacemaker cells. J Pediatr Surg 2000;35:673-6.
12. Yamataka A, Yamataka T, Kobayashi H, Sueyoshi N, Miyano T. Lack of C-KIT + mast cells and the development of idiopathic gastric perforation in neonates. J Pediatr Surg 1999;34:34-8.
13. Schinasi DA, Ellison AM. Spontaneous gastric perforation in a child with heterotaxy syndrome. Pediatr Emerg Care 2010;26:925-7.

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