

Shahirah Adilla MS¹, Sakinah AF¹, Muhammad CY¹, Rahilah AR¹, Faizatul AD¹, S Zaraqah²

¹Paediatric Surgery Unit, Department of General Surgery, Hospital Sultanah Nur Zahirah, Terengganu, Malaysia

²Anatomic Pathology Unit, Department of Pathology, Hospital Sultanah Nur Zahirah, Terengganu, Malaysia

Introduction

Appendicitis is an uncommon entity among neonates and rarely included in the differential diagnosis of acute abdomen.(1) It carries a high morbidity and mortality especially among premature infants. Challenges to diagnose are due to the nonspecific symptoms and clinical signs which need a high index of suspicion.

Case Summary

A premature baby born at 32 weeks, weighed 835g, admitted for respiratory distress syndrome and neonatal sepsis. Patient tolerated feeding well and had normal bowel output.

At day 28 of life he presented with abdominal distension and blood in stool. He was septic which required intubation. Plain abdominal radiograph illustrated dilatation of small bowels with pneumatosis intestinalis and portal venous gas (Figure 1a). Exploratory laparotomy performed, appendix was perforated with slough tissue covering surrounding caecum and appendix (Figure 1b). The rest of the bowels were normal. Appendicectomy and peritoneal lavage done. Histopathology examination (HPE) for appendicectomy showed *mucosal ulceration and moderate transmural infiltration by neutrophils, areas of disruptions are seen in muscularis propria, ganglion cells seen within inter-myenteric nerve plexus and no hypertrophic nerve bundle seen.* (Figure 2a and 2b)

Three weeks later, he developed feeding intolerance and abdominal distension. Lower gastrointestinal fluoroscopy revealed stricture at the descending colon, hence, laparotomy, adhesiolysis and colostomy done. Findings were interloop bowel adhesions and narrowing at mid-descending colon corresponded to the contrast study. HPE of resected bowel reported as *congested colonic mucosa, ganglionic cells seen with no hypertrophic nerve bundles.* Rectal biopsy taken at term came back as normal ganglionic sample.

Discussion

Low incidence of neonatal appendicitis compared to older age group may be attributed to certain factors namely the funnel shape of the appendix organ, neonatal diet predominantly milk, recumbent posture of neonates, and infrequent incidence of gastrointestinal infection among neonatal population.(2,3,4)

Most of the cases presented with nonspecific clinical presentation and features causing delay in diagnosis establishment.(1,5,6) Symptoms and signs such as generalised abdominal distension, vomiting, feeding intolerance, pyrexia and abdominal tenderness are all vague clinical manifestations in which later, postoperatively diagnosed as neonatal appendicitis.(2,5,6)

Blood investigations in neonatal appendicitis are non specific such as increased in leucocyte count and c-reactive protein. (7) About 52% of patient showed pneumoperitoneum.(1,5) Ultrasound abdomen may only show secondary features such as right iliac fossa free fluid or intra-abdominal collection.(1,4) CT scan may be more specific modality but come with the risk of exposing neonates or infants to high dose of radiation.(4)

It is proposed that in the management of neonatal appendicitis is early diagnosis and surgical intervention. Reason being, high chances for appendicitis to get early perforation and causing high morbidity and mortality as previously stated. The nature of neonatal viscera in which the appendix is fragile and indistensible cecum leads to early perforation plus inability of the underdeveloped omentum to contain the perforation with the immature immune system further contributing to the high fatality index. (8,9)

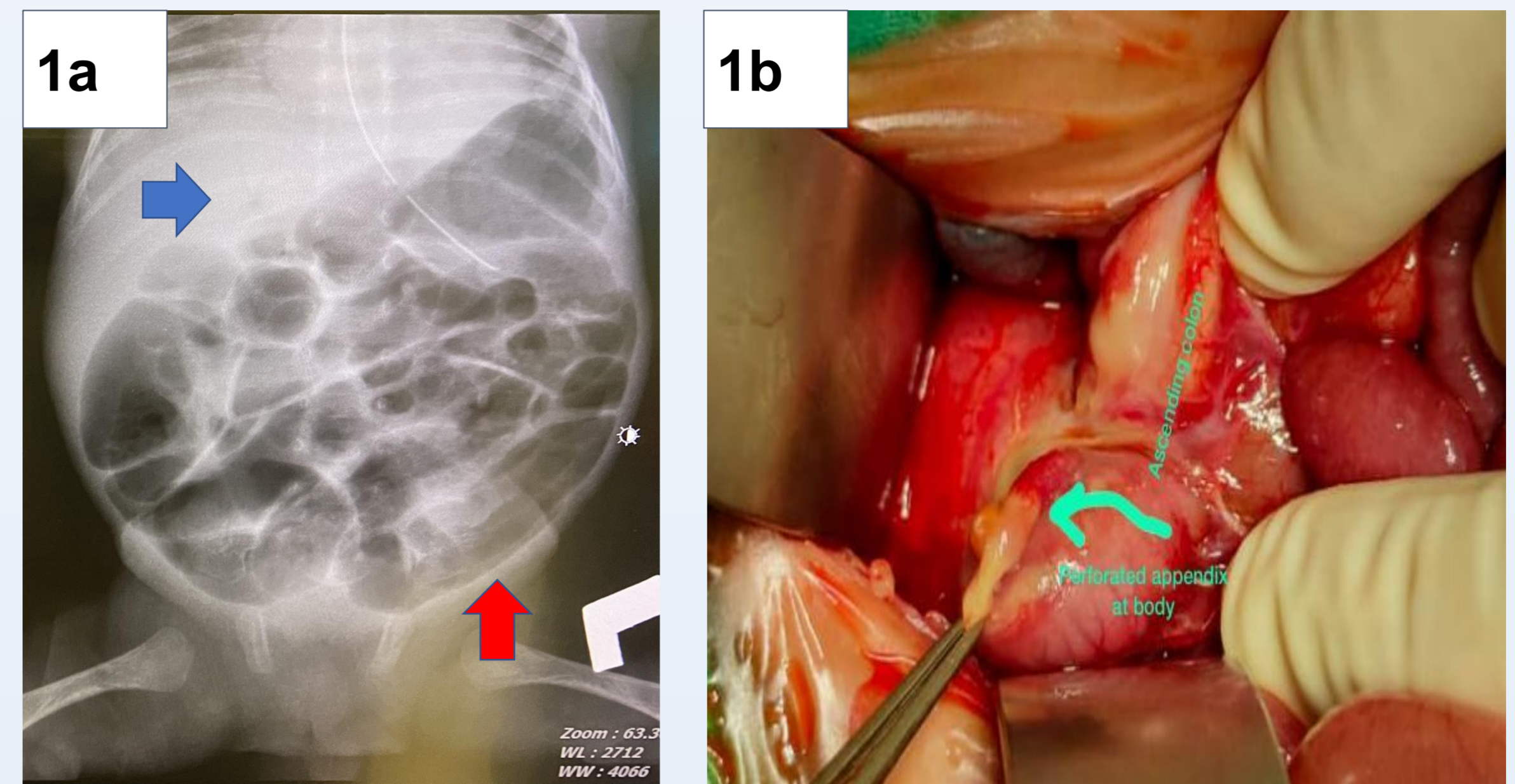


Figure 1a Abdominal x-ray showed dilatation of small bowels with pneumatosis intestinalis (red arrow) and portal venous gas (blue arrow)

Figure 1b Intraoperative findings showed perforation at the body of appendix with slough tissue covering the cecum and appendix

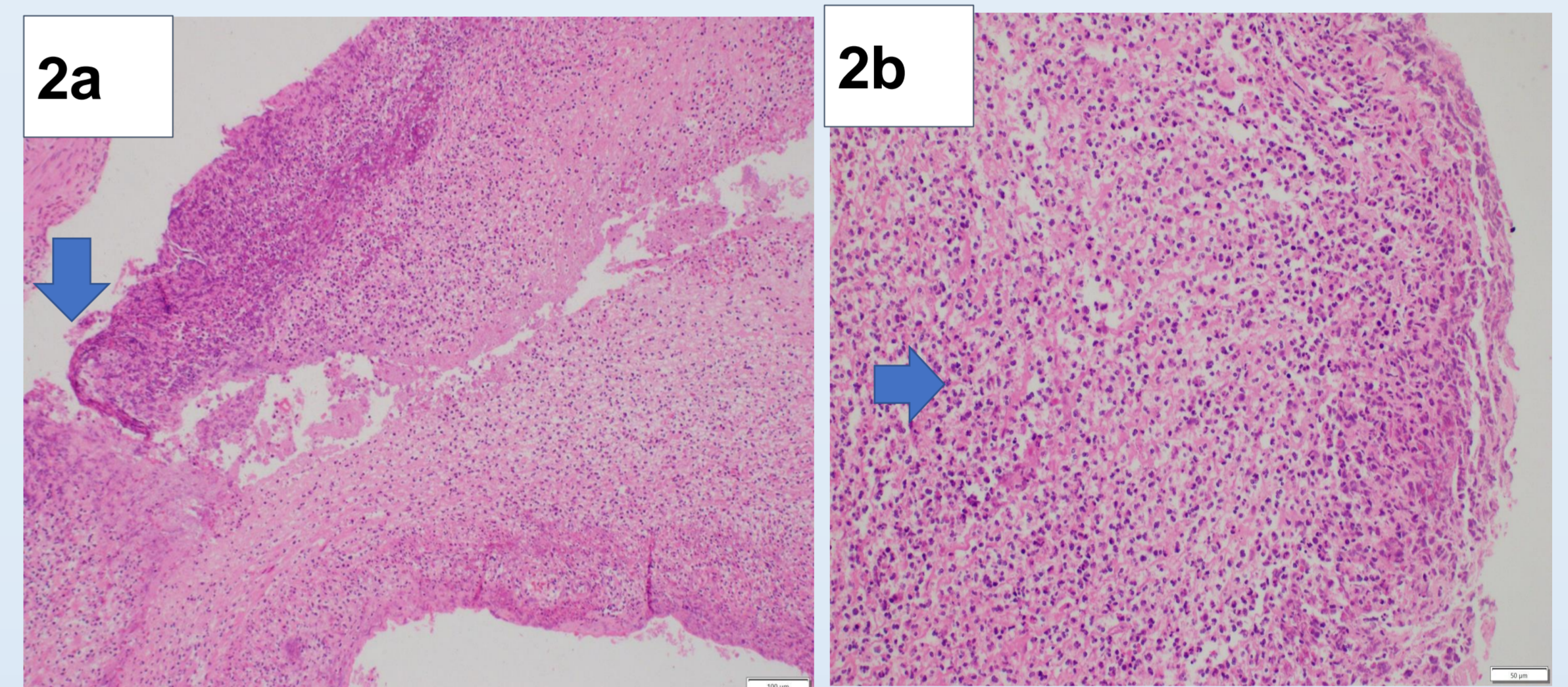


Figure 2a H&E, higher magnification : Muscularis propria disruption (arrow) leading to perforation.

Figure 2b H&E, highest magnification : Transmural neutrophilic infiltration (arrow) leading to muscularis layer necrosis.

Martin and Perrin et al once postulated the pathogenesis of neonatal appendicitis may be caused by luminal obstruction secondary to underlying Hirschsprung disease and intraoperatively presented as perforated appendicitis.(10) To exclude the possibility, we proceeded with suction rectal biopsy and similar to other publications, our case showed no evidence of such.

Conclusion

To conclude, neonatal appendicitis is not frequently seen and with relation to its vague clinical presentation, making it hard to establish the diagnosis early. Despite the unfamiliarity, we need to keep in mind the diagnosis of appendicitis in neonates with acute abdomen as delay in recognising this condition may lead to late intervention with less favourable outcomes in this age group.

References

1. Raveenthiran V. Neonatal appendicitis (part 1): a review of 52 cases with abdominal manifestation. *J Neonatal Surg.* 2015;4(1):4.
2. Karaman A, Çavuşoğlu YH, Karaman İ, Çakmak Ö. Seven cases of neonatal appendicitis with a review of the English language literature of the last century. *Pediatr Surg Int.* 2003;19(11):707–9.
3. Jancelewicz T, Kim G, Miniati D. Neonatal appendicitis: a new look at an old zebra. *J Pediatr Surg.* 2008;43(10):e1–5.
4. Schwartz, K., Gilad, E., Sigalet, D., Yu, W. and Wong, A., 2011. Neonatal acute appendicitis: a proposed algorithm for timely diagnosis. *Journal of Pediatric Surgery*, 46(11), pp.2060-2064.
5. Huang X, Huang X, Mbugi J, Wei L, Ziyu H, Ya H, et al. An experience with 31 cases of neonatal appendicitis: a retrospective study. *J Neonatal Surg.* 2019;8(3):21
6. Stifle D, Stallmach T, Sachet P. Acute appendicitis in neonates: Complication or morgues suit generis? *Pediatr Surg Int.* 1998;14:122–3
7. Okumuş, M. and Zübarioğlu, A., 2020. Neonatal and infantile appendicitis still confuses minds: report of two cases. *Annals of Pediatric Surgery*, 16(1).
8. Ruff ME, Southgate WM, Wood BP. Neonatal appendicitis with perforation. *Am J Dis Child.* 1991;145:111–2. [PubMed: 1985420]
9. Kwong MS, Dinner M. Neonatal appendiceal perforation and Hirschsprung's disease. *J Pediatr.* 1980;96:917–8. [PubMed: 7365602]
10. Martin LW, Perrin EV. Neonatal perforation of the appendix in association with Hirschsprung's disease. *Ann Surg.* 1967; 166:799-802.