Cytomegalovirus enterocolitis presenting as abdominal compartment syndrome in a premature neonate

Steven L. Lee, Hege Johnsen, Harry Applebaum

Los Angeles, California, USA

Case report

Background: **Cytomegalovirus (CMV) enterocolitis is an uncommon intestinal disorder of newborns that is often initially misdiagnosed as necrotizing enterocolitis.**

Methods: **We treated a premature twin boy with CMV enterocolitis who presented with abdominal compartment syndrome requiring urgent decompression. All patients with neonatal CMV enterocolitis reported were reviewed.**

Results: **Nine previously reported patients with neonatal CMV enterocolitis presented with abdominal distention and signs of sepsis. At the time of surgery, either perforation or stricture was identified. The current report is the first to present with clinical signs of abdominal compartment syndrome.**

Conclusion: **CMV is a rare cause of neonatal enterocolitis. Surgical intervention is required for bowel perforation, stricture, or abdominal compartment syndrome.**

World J Pediatr 2012;8(1):80-82

Key words: **abdominal compartment syndrome;**
cytomegalovirus;
enterocolitis;
necrotizing enterocolitis

Introduction

Congenital cytomegalovirus (CMV) can cause perinatal disease. Approximately 1%-2.5% of all newborns are positive for CMV, but only 5%-10% have clinical symptoms.[1,2] CMV enterocolitis is seen commonly in the immunocompromised patients, but rarely in newborns, and it is often mistaken for necrotizing enterocolitis (NEC). We present a neonate with CMV enterocolitis progressing to abdominal compartment syndrome and reviewed all published reports on CMV enterocolitis.

Case report

The male baby was one of the twins. He was born at $27^{4/7}$ weeks of gestation to a 28-year-old female, gravida 1, para 0. The boy had a birth weight of 1130 g and measured 36.5 cm in length. Apgar scores were 7 and 8 at 1 and 5 minutes, respectively. The baby was intubated shortly after birth due to respiratory distress and suspected sepsis, and was extubated 24 days later.

He was started on total parental nutrition on day 1 after birth. On day 2 he was also started on enteral feedings with breast milk. The total parental nutrition was tapered and discontinued on day 15 of life. He was on full feedings by day 18 of life. The patient developed abdominal distention and bloody stool on day 34 of life. An abdominal radiograph did not show free air or pneumatosis intestinalis. He was intubated for apneic episodes; enteral feeding was held and antibiotics were started. The symptoms resolved and he was extubated and started on enteral feedings again, which he tolerated well. He did well and was getting ready for discharge home when he developed nasal congestion on day 74 of life. Nasal swabs were negative for respiratory syncytial virus. He then developed a maculopapular rash with petechia over the face and trunk. Over the following days the platelets dropped to 24 000/μL and he was transfused with platelets. Four days after the symptoms started he developed severe abdominal distension, emesis and respiratory distress which required intubation. An abdominal radiograph showed dilated bowel loops, but no free air or pneumatosis intestinalis. Air was also noted in the scrotum. Examination of the groin revealed an inguinal hernia that was easily reduced at bedside. The abdominal distention was not improved after reduction of the inguinal hernia. Bloody stool was also noted. A repeat abdominal radiograph showed worsening of the bowel dilatation. The baby was then transferred to our NICU on day 78 of life with suspicion of NEC.

Author Affiliations: Division of Pediatric Surgery, Harbor-UCLA, Torrance, California, USA (Lee S); Department of General Surgery (Johnsen H) and Division of Pediatric Surgery (Applebaum H), Kaiser Permanente, Los Angeles Medical Center, Los Angeles, California, USA

Corresponding Author: Steven L. Lee, MD, Division of Pediatric Surgery, Harbor-UCLA Medical Center, Torrance, CA 90509, USA (Tel: (310) 222-2706; Fax: (310) 782-1562; Email: slleemd@yahoo.com)

doi: 10.1007/s12519-011-0307-3

By this time the rash had disappeared. The baby was still thrombocytopenic with a platelet count of 47 000/μL, despite platelet transfusion prior to transfer. He had a white blood cell count of 15 600/mm^3 and C-reactive protein level of 158 mg/L. An abdominal X-ray suggested possible pneumatosis intestinalis in the right upper quadrant, but no free air (Fig. 1). The abdominal distension dramatically worsened. The patient had no urine output, required dopamine and epinephrine to maintain his blood pressure, and high frequency ventilation with high pressure in order to ventilate. The patient was promptly brought to the operating room for an urgent exploratory laparotomy due to a clinical abdominal compartment syndrome.

A transverse, supraumbilical incision was made over the abdomen. Upon opening the abdomen, the patient's ventilatory and hemodynamic status significantly improved. The bowel was found viable with no signs of NEC. Two areas of circumferential color changes were found in the distal ileum. Bowel resection was not indicated. Mesenteric adenopathy was not found and no malrotation was noted. Because of the dilated bowel, the abdomen could not be closed primarily. A 6.5 cm silo was therefore sutured to the fascia for temporary abdominal wall closure. The patient was transferred back to the NICU intubated and in fair condition. Over the next couple of days the silo was reduced at bedside. He was quickly weaned off the epinephrine, and dopamine drips and switched back to conventional ventilation with minimal settings.

Seven days later, the patient was brought back to the operating room for a wash-out and abdominal closure. The silo was removed and the bowel was explored. Two strictures were found in the distal ileum. At this point it was unclear what caused the strictures. A 10 cm segment, including the two strictures, was resected. The proximal end was brought up as an end ileostomy and the distal end was closed. An open internal ring was identified in the right groin and closed with a purse-string suture. The fascia was closed with a biologic-mesh because of tension with attempted primary closure.

Five days after the operation, pathological report was positive for CMV, confirmed by immunohistochemistry (Fig. 2). The patient was started on ganciclovir immediately. Additional work up included urine sample that was postive for CMV and tracheal secretions negative for CMV. An ophthalmology examination was also negative for CMV.

The baby was extubated on post-operative day 13 from the original operation, and enteral feedings were started on post-operative day 14. The patient was discharged with improved condition. The ileostomy was closed two months later.

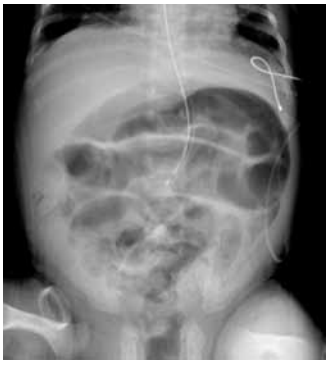

Fig. 1. Abdominal radiograph demonstrating dilated loops of the bowel with possible pneumatosis intestinalis in the right upper quadrant.

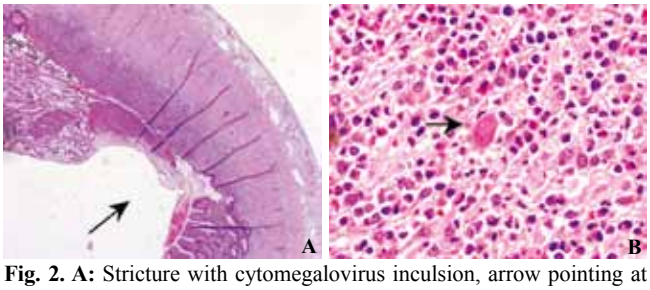

Fig. 2. A: Stricture with cytomegalovirus inculsion, arrow pointing at ulcer; **B:** Close up of cytomegalovirus inclusion (arrow).

Discussion

The patient developed clinical signs of abdominal compartment syndrome that required urgent decompression. In retrospect, the diagnosis of CMV should have been suspected with the patient's rash and thrombocytopenia one week prior to presenting at our NICU. Interestingly, the twin sister had developed NEC on day 17 of life and a 15 cm segment of necrotic terminal ileum was resected. Review of the pathology specimen did not show evidence of CMV enterocolitis. Unfortunately, CMV testing was not done routinely at the institution where the twins were born, so it was unknown if the baby boy had congenital CMV or contracted the virus from breast milk feeding. Neither the breast milk nor the mother was tested for CMV before giving birth. CMV infection is very common and the mother was most likely a carrier of the virus.

Congenital CMV can cause perinatal diseases. The postnatal transmission of CMV can also occur via mother's breast milk. Full-term babies are protected by maternal antibodies, but preterm infants born before the transfer of the majority of the protective immunoglobulins, are more susceptible to develop symptomatic CMV infection.[3] The majority of immunoglobulins are transferred after week 28 of gestation and the baby presented in this paper was

Table. Summary of reported cases of neonatal cytomegalovirus enterocolitis

Ref	GA (wk)	Weigtht (g)	Clinical presentation	Operative findings
1	30	Unknown	Abdominal ditension, emesis	Inflamed ileocecal area, perforated appendix fistulizing to cecum
2	33	2200	Emesis, abd distension, mild respiratory distress	5 cm ischemic stricture in the descending colon
3	29	1490	Abd distension, emesis, gray color of skin	Strictures in ascending and transverse colon
4	23	580	Loose stools, met acidosis, abd distension, emesis	Stricture 20 cm proximal to ileocecal valve
5	37	2490	Abd distension, emesis, mimicking Hirschsprung's disease	Stricture at the sigmoid colon
6	Term	3500	Fever, abd distension, hepatosplenomegaly, diarrhea	Ileal perforation 12 cm proximal to ileocecal valve
7	34	2695	Abd distension, barium enema showing multiple colonic strictures	Colonic strictures with severe colitis and mucosal ulceration
8	36	Unknown	Abd distension	Ileal ulceration
9	27	490	Abd distension, mottled pale skin, tachycardia	Colonic strictures
Present case	27	1130	Abd distention progressing to ACS	Strictures in terminal ileum

GA: gestational age; Ref: reference; abd: abdominal; ACS: abdominal compartment syndrome.

born prematurely at week $27^{4/7}$.[3] CMV enterocolitis is seen commonly in the immunocompromised adult and pediatric population, but there are only a few of case reports of CMV enterocolitis in the newborns who are not immunosuppressed. Most of these babies presented with symptoms similar to NEC, including abdominal distention, tenderness, and bloody stools (Table).[1-9] The present case is the first reported with abdominal compartment syndrome associated with CMV enterocolitis. Eight out of the ten patients who required surgical intervention were for strictures including 5 in the colon and 3 in the ileum; the other 2 patients were operated on for intestinal perforation. The diagnosis of CMV enterocolitis in this study was not made until the specimen was examined microscopically. CMV enterocolitis typically presents with abdominal distention and bloody stools. Strictures are commonly found at the time of surgery. The pathology reports show these strictures to be due to ulcers containing CMV inclusion bodies.[4] When CMV enterocolitis is discovered, further investigation should be performed to rule out other CMV infection. In our patient, CMV was also identified in the urine. Typically, gancylcovir is administered for systemic or disseminated disease. Use of gancyclovir in patients with CMV enterocolitis is controversial and not well described in the literature.

In conclusion, CMV enterocolitis is rare in newborns, and most commonly seen in premature infants. Most of the patients present with a clinically picture similar to NEC. CMV enterocolitis differs from NEC in that it presents with ulceration progressing to stricture, rather than gangrene and perforation. Our patient presented with abdominal compartment syndrome, which is a previously unrecognized presentation of CMV enterocolitis. Most patients can recover from the CMV enterocolitis without significant morbidity.

Funding: None.
Ethical approval: Not needed.
Competing interest: No benefits in any form have been received or will be received from any commercial party related directly or indirectly to the subject of this article.
Contributors: Lee S proposed the study and wrote the first draft. Johnsen H analyzed the data. All authors contributed to the design and interpretation of the study and to further drafts. Lee S is the guarantor.

References

1 Terry NE, Fowler CL. Cytomegalovirus enterocolitis complicated by perforated appendicitis in a premature infant. J Pediatr Surg 2006;41:1476-1478.
2 Reyes C, Pereira S, Warden MJ, Sills J. Cytomegalovirus enteritis in a premature infant. J Pediatr Surg 1997;32:1545-1547.
3 Gessler P, Bischoff GA, Wiegand D, Essers B, Bossart W. Cytomegalovirus-associated necrotizing enterocolitis in a preterm twin after breastfeeding. J Perinatol 2004;24:124-126.
4 Srinivasjois RM, Kava MP, Thomas A, Rao SC. Cytomegalovirus-associated ileal stricture in a preterm neonate. J Paediatr Child Health 2008;44:80-82.
5 Ekema G, Pedersini P, Milianti S, Ubertazzi M, Minoli D, Manciana A. Colonic stricture mimicking Hirschsprung's disease: a localized cytomegalovirus infection. J Pediatr Surg 2006;41:850-852.
6 Huang YC, Lin TY, Huang CS, Hseun C. Ileal perforation caused by congenital or perinatal cytomegalovirus infection. J Pediatr 1996;129:931-934.
7 Stiskal J, Jacquette M, Kaplan G, Ritterman R, Stavis R, Palder S. Congenital cytomegalovirus infection with gastrointestinal involvement. J Pediatr 1997;131(1 Pt 1):168.
8 Hakim A, Mimouni F, Payandeh F, Morawski J. Immunochemical staining in congenital cytomegalovirus-induced ileal ulceration. J Pediatr 1997;131(1 Pt 1):168-170.
9 Shetty A, Barnes R, Lazda E, Doherty C, Maxwell N. Cytomegalovirus: a cause of colonic stricture in a premature infant. J Infect 2007;54:e37-e99.

Received December 4, 2009
Accepted after revision June 1, 2010