

## Pneumatosis intestinalis due to rotavirus infection in a child with Prader-Willi syndrome

Uluç Yiş, Yasemin Topçu, Erhan Bayram, Pakize Karakaya, Handan Çakmakçı, Semra Hız-Kurul

Division of Pediatric Neurology, Department of Pediatrics and Department of Radiology, Dokuz Eylül University Faculty of Medicine, İzmir, Turkey. E-mail: dr.erhanbayram@yahoo.com

Pneumatosis intestinalis (PI) is a radiographic finding that is defined as the presence of gas within the bowel wall regardless of the cause. PI is less common and has a good prognosis after the first year of life<sup>1</sup>. A large variety of underlying diseases are associated with PI in children. Among the triggering conditions, gastrointestinal infections are common. Rotavirus infection as a cause of PI has been reported rarely<sup>2-4</sup>. Prader-Willi syndrome (PWS) is a rare genetic disorder characterized by low muscle tone, short stature, cognitive disabilities, incomplete sexual development, increased appetite, and obesity. Patients with PWS are prone to sudden death, but the pathophysiologic mechanisms underlying the sudden death are not completely understood<sup>5</sup>. We report an eight-month-old girl with PWS whose clinical condition deteriorated rapidly and who developed PI during the course of rotavirus gastroenteritis.

An eight-month-old girl was hospitalized for temperature dysregulation, feeding difficulty

and hypotonia. She was diagnosed as PWS by fluorescent in situ hybridization test (Fig. 1). During her hospitalization in November 2011, she developed vomiting and watery diarrhea occurring more than 10 times daily. There were also cases of rotavirus infection hospitalized during this period. On the first day of illness, in addition to frequent vomiting and diarrhea, she was also febrile (38.9°C) and lethargic. Intravenous hydration was started for poor oral uptake and stool examination revealed a few leukocytes. The stool was also positive for rotavirus antigen, which was tested on the first day of her illness. Rotavirus was detected by ELISA test (SD BIOLINE Rota/Adeno Rapid, Standard Diagnostics, Inc., Korea), which has 100% sensitivity and 99.7% specificity. On the third day of illness, the frequency of vomiting and diarrhea decreased, but the clinical condition of the patient deteriorated with signs of shock. She had hypotension,



Fig. 1. Hypotonia, frog-like position and obesity of the patient.

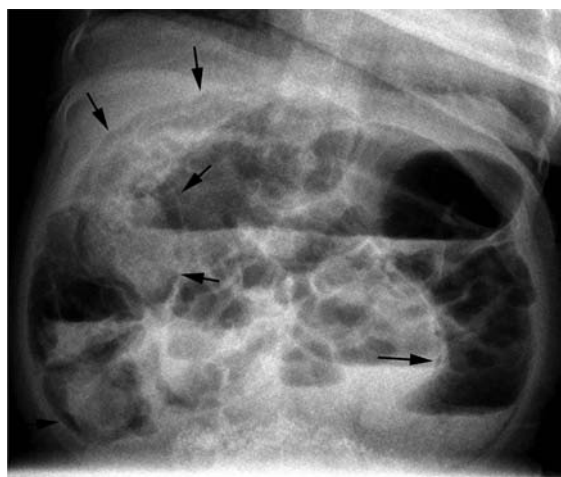
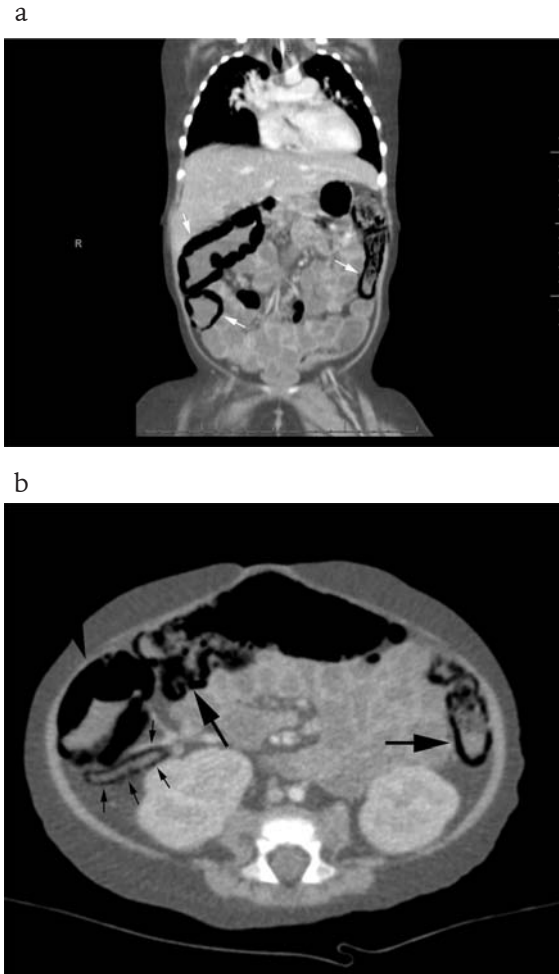


Fig. 2. Upper abdominal radiograph in erect position shows pneumatosis intestinalis (arrows) and gas distention of the bowel with air-fluid levels.



**Fig. 3.** Abdominal CT, coronal reformatted image (a) shows the gas was mostly within the colonic segments of the cecum and the ascending, transverse and descending colon. Axial CT scan (b) shows colonic pneumatosis intestinalis (arrows) with normal small intestine walls; the gas within the appendix walls (small arrows) is seen as well.

tachycardia, prolonged capillary filling time, and diffuse abdominal tenderness. The laboratory examinations on the third day of illness showed mild leukocytosis ( $16,000/\text{mm}^3$ , absolute neutrophil count 70%) with normal basic metabolic panel, liver function tests and blood gases analysis. Blood, urine and stool cultures were taken while aggressive fluid and antibiotic (ceftriaxone and metronidazole) were administered on the third day of illness. Abdominal radiography in the erect position revealed air distention of the bowel with diffuse gas within the intestine walls and air-fluid levels of the intestine (Fig. 2). Computed tomography (CT) revealed the gas was mostly

within the colonic segments of the cecum and the ascending, transverse and descending colon. The small intestine wall appeared normal on CT images (Fig. 3a, b). No abdominal free air was detected on CT as a sign of intestinal perforation. Cultures of blood, urine and stool, which were taken on the third day of illness, revealed no growth of any pathogen, and stool was also negative for ova and parasites. After one week of clinical deterioration, the patient recovered and the diarrhea resolved. A rotavirus stool antigen was negative.

Pneumatosis intestinalis (PI) in children is rare. It occurs frequently in premature infants with necrotizing enterocolitis. The incidence of the disease is not known, but 37 episodes of PI were identified from 1992 to 1999 in 32 patients aged 30 days and older in a tertiary children's hospital located in Denver, Colorado, United States<sup>6</sup>. Mechanical, bacterial, viral, and pulmonary causes and increased mucosal permeability have been proposed for the pathogenesis of PI. It rarely occurs in healthy children, and most of the patients have an underlying disease like organ and bone marrow transplant, decompensated congenital heart disease, motility disorders, gastroschisis, and short bowel syndrome<sup>6</sup>. The most common preceding factors are noninfectious colitis, acute enteric infection or toxin, bowel ischemia, and gastrointestinal dysmotility<sup>6</sup>. *Clostridium difficile*, cryptosporidium, cytomegalovirus, adenovirus, and norovirus infections are associated with PI, but rotavirus infection is reported rarely<sup>2-4,7</sup>. PI due to rotavirus infection may occur in healthy and immunocompromised children<sup>2-4</sup>. Patients with PWS are prone to sudden death. Respiratory tract infections, gastroenteritis, sudden cardiac death, hypoventilation, growth hormone treatment, and acute gastric dilatation are associated with sudden death<sup>5</sup>. There are two reports of children aged two and three years with PWS who died shortly after the start of gastrointestinal symptoms and high fever. Some children and adults with PWS with or without obesity have also been reported to die or become seriously ill from acute gastric dilatation<sup>5</sup>. The mechanism underlying the acute gastric dilatation is not clear and not related to overfeeding. It is hypothesized that an abnormal gastric homeostasis may be a

component of PWS. We suggest that abnormal gastrointestinal homeostasis may also lead to PI during the course of rotavirus gastroenteritis. The prognosis of PI is generally good with medical management, but about 20% of patients may be lost or require surgery<sup>6</sup>. Patients with organ transplant, decompensated cardiac disease, low serum bicarbonate concentration, and portal venous gas have a poorer prognosis<sup>6</sup>. Our patient was hospitalized early, when she developed the signs of PI and shock, and she recovered quickly with appropriate medical treatment. In conclusion, gastrointestinal symptoms in children with PWS during enteritis should be taken seriously, as early diagnosis may be life-saving.

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