Benign pneumatosis in children

Laura Z. Fenton
Carlo Buonomo

Abstract Background. In pediatrics, pneumatosis intestinalis (PI) is usually due to necrotizing enterocolitis in premature newborns. Beyond infancy, PI is uncommon. “Benign pneumatosis” is PI in patients with few or no symptoms that resolves with conservative management.

Objective. Our goal was to better characterize benign PI in children. Our investigation focused on identifying underlying risk factors, symptoms at time of diagnosis, management and outcome.

Materials and methods. Available medical records and radiographs of children with pneumatosis intestinalis from 1990 to 1998 were reviewed for underlying conditions, symptoms at time of radiographs, management and outcome.

Results. Thirty-seven children (mean age 4 years) were included. Thirty-two children had identifiable risk factors. Twenty-five children were immunocompromised by their underlying conditions or therapeutic regimen. Thirty-five children were managed conservatively with resolution of PI. Two patients, however, required surgery and one patient died.

Conclusion. Benign pneumatosis does occur in children. The majority have underlying risk factors, most commonly related to immunosuppression. Clinical deterioration is the most useful indicator for surgical intervention. In most patients PI resolves with conservative management.

Introduction

Pneumatosis intestinalis (PI) is defined as the presence of gas within the bowel wall regardless of cause. In pediatric radiology, PI is usually seen in premature neonates with necrotizing enterocolitis. These neonates are usually quite ill and surgical intervention is frequently necessary to prevent extensive necrosis of the bowel, sepsis, and death. PI is an unusual finding in children beyond the 1st year of life, but may be seen rarely with midgut volvulus, intussusception, and intestinal ischemia of other cause. There is, however, a population of older infants and children with PI who have a more benign presentation and clinical course. Several underlying conditions have been associated with pneumatosis in children, including immunosuppression (steroids, chemotherapy, acquired immunodeficiency syndrome), mucosal disruption (trauma, indwelling catheters, bowel obstruction), obstructive pulmonary conditions (asthma, cystic fibrosis, barotrauma) [1], short-gut syndrome, congenital heart disease, and collagen vascular disease [2]. Our investigation included identifying underlying conditions that may represent risk factors for the development of pneumatosis. For the purpose of this paper, “benign pneumatosis” is defined as radiographic pneumatosis in an older infant or child with few or minimal symptoms, who usually have an underlying condition that has been associated with PI. This group of children with “benign pneumatosis” have not been well characterized and data on clinical manifestations, management and outcome are limited. Our goal was to define the characteristics of benign pneumatosis better so that
when encountering a child with pneumatosis, imaging findings can be appropriately correlated with the clinical signs and symptoms to aid in management decisions.

**Patients and methods**

A retrospective review of imaging studies and medical records was performed for all older infants and children with diagnosis of pneumatosis intestinalis at Children’s Hospital, Boston, over a 9-year period (1990–1998). To exclude infants with necrotizing enterocolitis, only those older than 6 months were included. The underlying condition(s), signs and symptoms at presentation, medications, radiographic findings, management, and outcome were analyzed.

**Results**

Thirty-seven infants and children with PI were identified, ranging in age from 8 months to 17 years (mean 4 years). There were 20 boys and 17 girls. The diagnosis of PI was established by abdominal radiographs in all children. Three children also had abdominal CT scans. Thirty-two of 37 patients had underlying conditions that correspond to established risk factors for the development of PI, which included leukemia (9), bone-marrow transplant (7), solid-organ transplant (5), short-gut syndrome (4), AIDS (3), chronic pulmonary disease (3), malignancy (3), congenital heart disease (1), and chronic immunosuppression (steroids) (19). Note that 15 of these patients had two risk factors: steroid administration in addition to leukemia (7), chronic pulmonary disease (2), solid organ transplant (5), and bone-marrow transplant (1). Five children had three risk factors: bone-marrow transplant for leukemia and taking steroids at the time of PI. Only five children had no identifiable risk factor; their diagnoses are as follows: end-stage renal disease, end-stage liver disease, pyruvate dehydrogenase deficiency, mental retardation/cerebral palsy and Goldenhar’s syndrome. Twenty-five of 37 children were immunocompromised by their underlying conditions: leukemia (9), AIDS (3), agammaglobulinemia (2), and/or their treatment regimen: bone-marrow transplant (7), solid-organ transplant (5), and steroids (19). Thirteen of the 37 children had and were being fed by gastrostomy tube at the time of PI (Tables 1, 2).

Of note, 4 of the 37 children (Nos. 7, 16, 19, 35) had more than one episode of PI during the 9-year study period (each was only included once in the data set; the most severe episode of PI was used for analysis). All of these children were immunocompromised (liver transplant, lung transplant, ALL/bone-marrow transplant, autoimmune hemolytic anemia) and were taking steroids.

The majority (28 of 37 children) had clinically mild gastrointestinal symptoms at the time of PI. Symptoms included diarrhea (17), abdominal pain (13), fever (12), vomiting (9), and abdominal distention (5). Multiple symptoms were present in 18 children. Nine children had no gastrointestinal symptoms: six of these nine children were taking steroids at the time of PI. Rotavirus infection has been implicated in some children with PI [6]; however, only six in our series were tested, with one positive (no. 24).

Abdominal radiographs demonstrated PI of the colon in 36 children (right colon 11, left colon 7, and diffuse 18), and small bowel and colon in one child (Figs. 1, 2). A subserosal (bubbly) pattern was seen in 16, submucosal (linear) pattern in 6, and a mixed pattern in 15. Pneumoperitoneum was identified in 4 children (nos. 25, 28, 29, and 33), portal venous air in 2 (nos. 7, 31) and both in 1 (no. 16) (Figs. 3, 4). No pneumoretroperitoneum, pneumomediastinum, or pneumothorax was identified. Three children had CT scans (nos. 25, 28, and 29), which confirmed colonic PI in all, pneumoperitoneum.