Non-NEC and Pneumoperitoneum

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Background

Pneumatosis intestinalis (PI) is a radiographic finding that is uncommon. There are various etiologies of this finding with the most life-threatening being bowel necrosis. Immediate surgical intervention is indicated in the setting of bowel necrosis; however for the less threatening etiologies, a conservative approach may suffice. The clinical presentation of PI can be vague in symptomatology, especially in the pediatric population. This presents a particularly difficult challenge to clinicians, when determining the correct management.

Objective

A 28-day-old premature infant with a history of gastroschisis presented to the emergency department with vomiting and hematochezia. An abdominal radiograph and computed tomography scan demonstrated widespread PI and pneumatosis coli with massive pneumoperitoneum. At exploration, however, She underwent emergent laparotomy, but was not found to have any she had no evidence of perforation or necrotizing enterocolitis. She has recovered fully.

Methods

We present a case of a premature infant who suffered from pneumoperitoneum and pneumatosis intestinalis. The course of management is described in detail to allow our findings to be shared with providers facing similar clinical dilemmas.

Introduction

Pneumatosis intestinalis (PI) is a condition in which air is found within the bowel wall. Classification of the major pathogenic mechanisms causally involved in intramural gas formation include bowel necrosis, mucosal disruption, increased mucosal permeability, and pulmonary disease.[1] Of these possible etiologies, bowel necrosis is the most life-threatening cause of intramural gas formation. In such cases, portal venous gas embolization may be present.[1] While pneumatosis and pneumoperitoneum can often be identified on plain radiography, cross-sectional imaging with computed tomography (CT) should be obtained for further investigation and to identify an etiology, if possible. The presence of free intraperitoneal gas on a routine radiograph usually indicates bowel perforation. Plain film radiography is sensitive in only 50–70% of cases, and the site of perforation is almost never elucidated.[4,5]

Causes of pneumatosis PI in an infant patient are mainly related to prematurity. NEC is the most common and frequently dangerous gastrointestinal emergency in premature infants in the neonatal intensive care unit (NICU).[2] Ninety percent of infants who develop NEC are born premature, however, near-term and full-term infants can also develop the disease.[3] The definite diagnosis of NEC is made from either surgical or postmortem intestinal specimens that demonstrate the histological findings of inflammation, infarction, and necrosis. However, a pathologic diagnosis is not always possible. At times, radiographic findings may be equivocal and treatment decisions should be based upon clinical suspicion and findings.[6] Some key risk factors have been consistently identified as important prerequisites for initiation of intestinal injury leading to NEC. These include prematurity, breast feeding, abnormal microbial intestinal colonization, gastroschisis, and ischemia. [7,8,9]

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Images

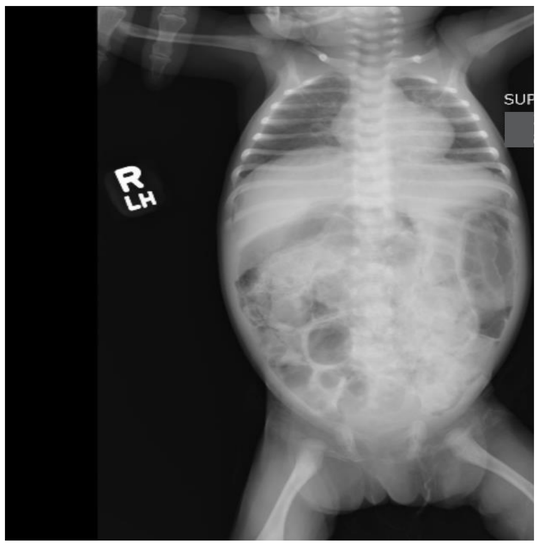

Figure 1. Abdominal X-Ray showing air within the bowel wall

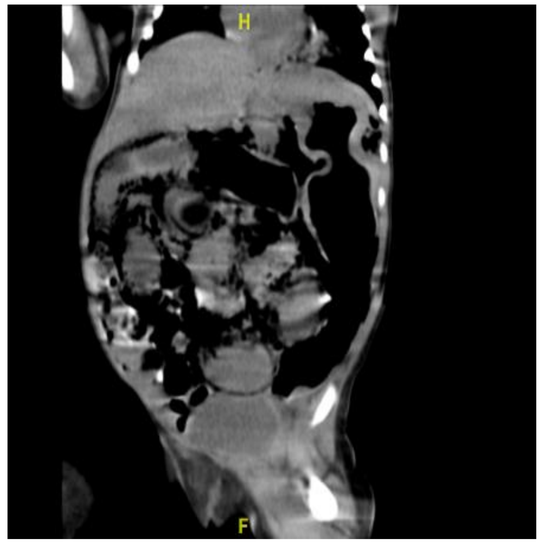

Figure 2. Computerized tomography of the abdomen showing free intraperitoneal air

Discussion

PI in the newborn is a distinctive radiographic finding that frequently portends a calamitous clinical course. Associated findings of advanced disease include portal venous gas and thumbprint sign, both of which were absent in this case. Ginglen et. al. describe the typical findings in NEC mentioned in their summary.[10] Pneumoperitoneum with PI, while not pathognomonic of perforated NEC, strongly suggests the diagnosis. The initial management of PI is commonly conservative with bowel rest, bowel decompression, and antibiotics. The definite diagnosis of NEC is made from either surgical or postmortem intestinal specimens that demonstrate the histological findings of inflammation, infarction, and necrosis. However, a pathologic diagnosis is not always possible. At times, radiographic findings may be equivocal and treatment decisions should be based upon clinical suspicion and findings.[6] Exploratory laparotomy is reserved for patients with persistent bowel obstruction and evidence of perforated bowel. In this case, it was the finding of massive pneumoperitoneum in the context of PI that prompted emergent exploration. Even though our patient looked remarkably well, we felt obligated to explore her – particularly given her NEC risk factors of prematurity and gastroschisis. Prompt exploration has been shown to minimize morbidity and mortality in necrotizing enterocolitis. In a study performed by Grosfeld et. al., it was found that early intervention was associated with a decreased mortality rate (36%).[11] Most surgeons agree that pneumoperitoneum is an absolute indication for operation.[12-15] However, in this case, the findings of pneumoperitoneum with PI were not due to perforation or even to NEC. We theorize that the large-volume free air in the abdomen may have coalesced from the rupture of innumerable tiny subserosal gas pockets which were identified at laparotomy. Other than this widespread pneumatosis, the bowel was negative for any ischemic changes. Furthermore, we hypothesize that the ultimate cause of this widespread PI was food protein-induced enterocolitis syndrome (FPIES) due to a cow's milk supplement added to her mother's breast milk. FPIES is caused most commonly by cow's milk and soy proteins. FPIES rarely occurs in exclusively breastfed infants, but Monti et. al. similarly describe an infant with FPIES caused by cow's milk proteins passed through the breast milk after accidental maternal ingestion.[3] FPIES is underrecognized; children are often mismanaged as having acute viral gastrointestinal illness, sepsis, or surgical disease, delaying diagnosis of FPIES for many months. [3] The fact that our patient recovered uneventfully on – and continues to thrive on – HAAIF supports, but does not prove, our hypothesis.

Conclusion

We fully recognize that virtually all neonates with PI and pneumoperitoneum will continue to require emergent abdominal exploration for what is statistically very likely to be perforated NEC. However, we also believe that this case should serve as a reminder to clinicians who care for newborns that they should consider non-surgical conditions such as FPIES in the differential diagnosis of any patient who presents with PI – even with massive pneumoperitoneum.

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