

Pediatric emphysematous cystitis: a report and review of a rare diagnosis in children

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An 11-year-old female with spastic quadriplegia was seen in the emergency room with abdominal pain, vomiting and anorexia. Labs revealed possible pancreatitis and signs of a urinary tract infection. A CT scan was performed to assess her abdominal pain and demonstrated circumferential air

within the bladder wall. Following cultures being drawn, she was started on broad spectrum antibiotics. Her urine eventually grew Klebsiella Pneumoniae. Follow up imaging 2 weeks later demonstrated resolution of the air. Emphysematous cystitis is an exceedingly rare condition in the pediatric population, with this report representing the second case within the literature.

Key Words: emphysematous cystitis, UTI, urinary tract infection, bladder

Introduction

Emphysematous cystitis (EC) represents a condition that many urologists are familiar with, but that few have treated. Classically seen in elderly diabetic females, EC has been reported in the literature roughly 150 times since 1961 when the term "cystitis emphysematosa" was introduced by Bailey.¹

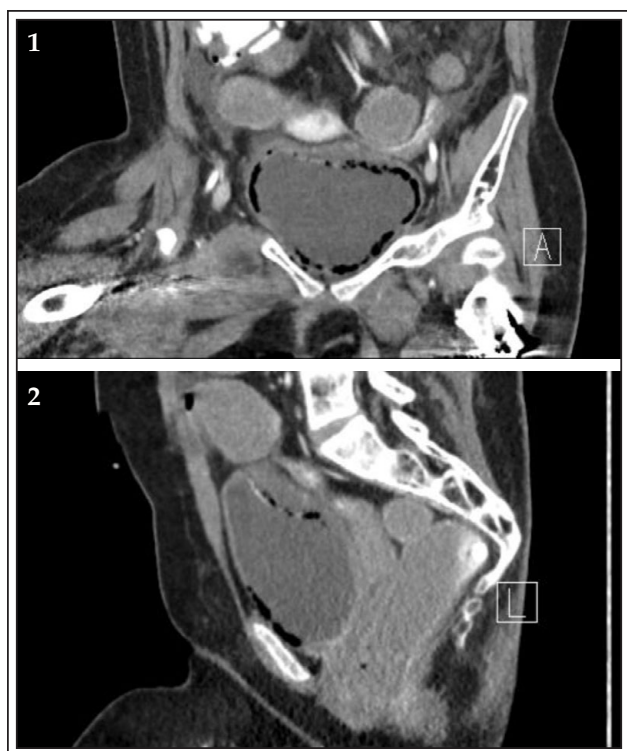
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While EC is an uncommon diagnosis in adults, it is exceedingly rare in the pediatric population, with a single case report published in an infant with bilateral ureteropelvic junction obstruction.² Here we report a case of EC in an 11-year-old female with no history of recurrent urinary tract infections or immunosuppression.

Case report

An 11-year-old female with a history of spastic quadriplegic cerebral palsy, encephalopathy, and Lennox-Gastaut syndrome presented to our emergency room with vomiting, abdominal pain, decreased urine



Figures 1 and 2. Coronal and sagittal CT images showing circumferential air in the bladder wall.

output and constipation for 5 days. Labs were drawn and demonstrated an elevated amylase, lipase, a blood glucose of 305 and white blood cell count (WBC) of 29.7. Blood and urine cultures were drawn and a CT scan performed to assess her abdomen. The CT showed acute severe necrotizing pancreatitis with a secondary finding of circumferential air within the bladder wall, Figures 1 and 2. Her upper tracts were normal however. A urinalysis was performed and showed high levels of glucose in addition to being concerning for a urinary tract infection (UTI). Urology was consulted and after identifying that she was hemodynamically stable, recommended immediate bladder decompression and initiation of broad spectrum antibiotics, including fungal coverage. In discussion with her mother, she endorsed a remote history of a urinary tract infection as an infant, but no recent or recurrent UTIs. After 24 hours of antibiotics, the patient's WBC count decreased to 11.8 and continued to decrease over the next several days with no further fevers. Her urine results returned 72 hours later, demonstrating *Klebsiella Pneumoniae*.

Over the next week she was maintained on IV antibiotics with continued hemodynamic stability, no change in her examination and clinical improvement

and on hospital day 7, a pelvic CT was performed to reassess her bladder. This demonstrated near resolution of all bladder wall air with continued normal upper tracts. She was maintained on 14 days of antibiotics. An additional CT scan was performed 2 weeks later on a readmission for pancreatitis and showed resolution of EC. At follow up, 6 weeks after this admission, urodynamics were performed given her history of cerebral palsy and this unusual urinary infection. This study revealed normal bladder compliance, no detrusor over-activity and a normal bladder capacity with normal bladder emptying.

Discussion

A diagnosis of EC is made when gas is seen within the lumen of the bladder and/or within the bladder wall. Air within the bladder is a familiar, but uncommon finding on imaging studies with potential causes ranging from the benign, including recent instrumentation or catheterization, to more insidious, including fistulae or infection with a gas forming organism. When air is seen within the bladder wall, it is most often the sequela of a complicated UTI and requires prompt attention. Patients should have prompt decompression of the bladder and other infected areas of the urinary tract, with extreme cases potentially requiring surgical intervention as up to 10% of patients may require surgical debridement. Delayed treatment can be life threatening with one study reporting a mortality rate of 7% for patients with EC and 14% for patients with EC and an emphysematous condition elsewhere in the urinary tract.³

Classic urinary symptoms (frequency, urgency, dysuria) are present in just over half (53%) of patients, while abdominal tenderness/pain is the most common presenting symptom in up to 80%.⁴ As a result, like in this case, a diagnosis of EC is often delayed or incidentally found. Given the abstruseness of the diagnosis, imaging is generally helpful (and technically necessary) in confirming the diagnosis. Plain abdominal x-ray is highly sensitive (97.4%), but has a low specificity due to potential confounding from rectal gas, radiation penetration issues, gas containing abscess, etc. Additional studies have shown the rate of EC diagnosis from plain x-ray to be as low as 13%.⁵ As such, CT is recommended to confirm the diagnosis as it is highly sensitive and specific. Bladder ultrasound is generally not recommended as studies have found it to be much less sensitive.⁴ Over the past decade, the number of EC cases reported in the literature has increased, almost certainly owing to an increase in the use of diagnostic imaging. However, the condition is rarely reported in children.

The vast majority of patients with EC are reported to have diabetes and not infrequently, patients have been noted to have frequent UTIs, urinary retention or neurogenic bladder – all conditions that increase the likelihood of infection.⁶ Although our patient does not have diabetes, perhaps her concurrent episode of pancreatitis may have been a contributing factor causing an elevated serum glucose and glucosuria. *Escherichia Coli* is the causal organism in greater than 50% of cases, with other enteric bacteria including *Klebsiella* and *Enterobacter* species, accounting for the majority of additional cases.³ Interestingly, many causal agents including *Candida* sp, *Pseudomonas*, *Proteus*, *Aspergillus*, *Staphylococcus*, *Streptococcus* and *Clostridium* species, including *Clostridium Difficile* (7) have been described, highlighting the need for broad spectrum coverage if EC is suspected.

Given the infrequency with which EC is seen in children, we followed a protocol as to what would normally be carried out for an adult with the condition. Given her hemodynamic stability, lack of air within the upper tracts and clinical improvement over time, we were able to manage this patient conservatively. In this child, given her comorbidities, we did perform urodynamics, but this may not be necessary in all children with EC. Instead, we would recommend a high suspicion for underlying bladder and bowel dysfunction, given its known close association with UTI development.

EC is an exceedingly rare entity in children with our report representing the second case within the literature. Practitioner should however have a high degree of suspicion in a patient with glucosuria or diabetes. If EC is suspected, CT imaging should be obtained to make the diagnosis and evaluate the upper tracts. Provided that the patient is hemodynamically stable, conservative therapy may be followed with prompt decompression of the bladder, urine culture, initiation of broad spectrum antibiotics, including fungal coverage, and investigation of the underlying cause. □

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