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CASE REPORT

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Vomiting of unclear etiology in an autistic child—multiple possible diagnoses: A case report

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Abstract

Appendicitis is a common complaint in the emergency department (ED) presenting with abdominal pain or vomiting and is often the foremost etiology the provider must rule out using history and physical examination. However, history and physical examination is limited in children and the developmentally delayed who are often non-cooperative. Less commonly, choledochal cysts are found that also require management, or rarer still, multiple possible radiologic or surgical diagnoses. This case report follows a delayed child presenting with vomiting found to have a large type 1 choledochal cyst, cholecystitis, and appendicitis on advanced imaging prompting surgical management of these etiologies. This report reviews the evaluation of children with vomiting and the need for thorough evaluation with advanced imaging when appropriate.

1 | CASE

A 3-year-old male, with a medical history of developmental delay, autism, and obesity, presented to the emergency department with 3 days of non-bloody, non-bilious emesis occurring 5–6 times per day, increased irritability, and decreased appetite. The parent states the patient had increased irritability that was present for most of the day over the last 3 days. The patient had decreased amount of wet diapers on day of admission, and a 3-day history of constipation, both of which he had not previously had. Two days previously, the patient had visited his pediatrician for these symptoms, and was prescribed Pedialyte without improvement. His mother denies hematemesis, fever, lethargy, rash, abdominal distension, cough, and trauma. Patient is up to date on vaccinations.

The patient's vital signs upon presentation revealed a rectal temperature of 99.2°F, a heart rate of 118 bpm, blood pressure of 114/83 mm Hg, a respiratory rate of 24/min, and a weight of 32 kg. Although the exam was limited due to the patient's developmental delay, inability to detail the presence or absence of tenderness due to non-verbal uncooperative status; he demonstrated a non-distended, non-tender abdomen to deep palpation with no guarding or appreciable masses. Bowel sounds were positive and normoactive in all 4 quadrants with no evidence of ascites. His testes were descended and revealed no appreciable tenderness or discoloration. Ear-nose-throat exam revealed no acute abnormalities. Lungs were clear to auscultation, cardiac exam with normal heart sounds. Skin color was normal for his race with no rashes seen.

Pertinent laboratory findings were significant for chloride of 96 mmol/L and carbon dioxide of 18 mmol/L, likely due to dehydration. Sodium and potassium were normal and there was an anion gap of 20. Glucose was 98 mg/dL. His white blood cell count was 16,000/uL with 79% neutrophils. Liver function tests, lipase, and amylase were within normal limits. Attempts to feed the child in the ED resulted in persistent non-bilious vomiting, prompting orders for imaging. CT (computed tomography) was ordered instead of ultrasound to increase diagnostic yield and expedite care. CT of the abdomen and pelvis with intravenous contrast was performed without patient sedation and revealed a large cyst in the porta hepatic region measuring 6.4 × 6.1 cm

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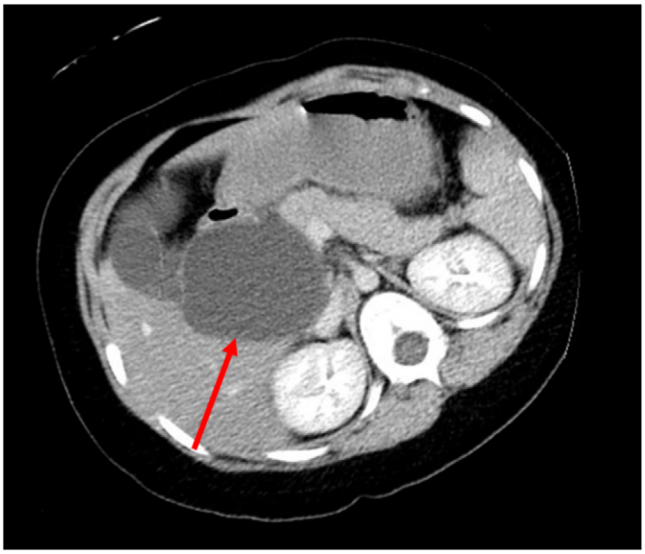

FIGURE 1 CT image of choledochal cyst in transverse view

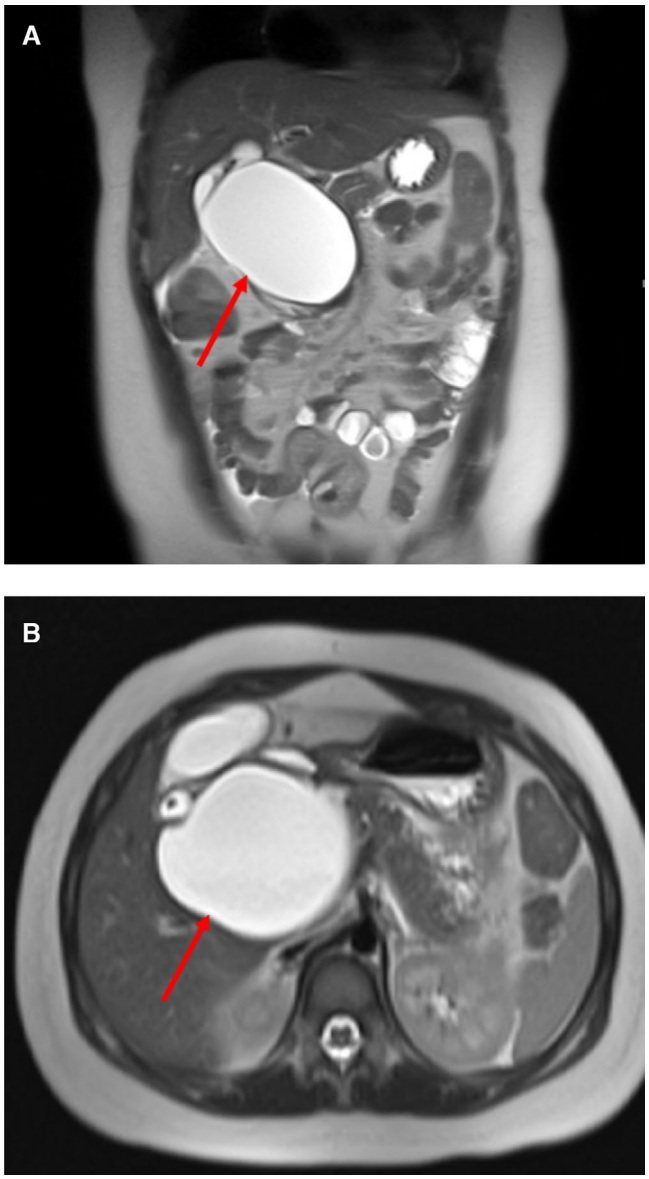

FIGURE 2 (A) MRCP image of choledochal cyst in coronal view. (B) MRCP image of choledochal cyst in transverse view

that was inseparable from the pancreatic head, gallbladder, duodenum, and liver (Figure 1). It also showed a retrocecal appendix measuring 7 mm with a possible appendicolith and trace-free fluid along the right paracolic gutter (Figure 2).

Maintenance fluids, ondansetron, and ceftriaxone were administered to cover for possible appendicitis. The patient was admitted to the pediatric floor and was evaluated by pediatric surgery who recommended gastroenterology consultation. Gastroenterology ordered an ultrasound of the abdomen that confirmed the presence of the cyst, revealed the gallbladder wall was not thickened, there was no pericholecystic fluid, and intrahepatic ducts were not dilated. A magnetic resonance cholangiopancreatography performed on hospital day 2 to further characterize the structure confirmed the presence of a type 1, choledochal cyst with a mass effect on the second portion the duodenum, pancreatic head, portal vein, and inferior vena cava were noted (Figure 3). The patient's clinical status remained unchanged on antibiotics, so the decision was made to operate for removal of the cyst and appendix.

On hospital day 4, the patient underwent a laparotomy for resection of the choledochal cyst, Roux-en-Y hepaticojejunostomy, and appendectomy. The patient tolerated the procedure well, and there were no complications noted in the operative report. A nasogastric tube was placed and maintained on low intermittent suction. Over the next 5 days, the patient's parenteral nutrition was stopped, nasogastric tube removed, and diet gradually advanced. The patient was discharged home on post-op day 8 on cephalexin.

The pathology report of the appendix reported a 7-mm appendix with superficial acute mucosal appendicitis. The appendiceal serosa was pink, had congested vessels, and there was a fecalith in the lumen. The gallbladder showed signs of acute and chronic inflammation without calculi consistent with acute on chronic cholecystitis. Histologic examination of the cyst revealed biliary lined saccular tissue with severe acute inflammation.

2 | DISCUSSION

Autistic children who are non-verbal represent a patient population with diagnostic challenges. Nearly all history and review of symptoms is obtained from their caregivers, and the physical exam can be challenging because the patient is often uncooperative. The location or presence of pain is difficult to determine. The clinician must decide on the reliability of the history, and how the patient looks clinically to decide on how aggressive of a workup to pursue. Advanced imaging techniques such as CT and ultrasound have drastically changed surgical diagnosis in not only this special subset of patients but the population at large.[1] A nonverbal patient who is non-toxic appearing on exam may have their diagnosis delayed as one cannot expose all children with vomiting to ionizing radiation. When a scan is warranted, it may easily

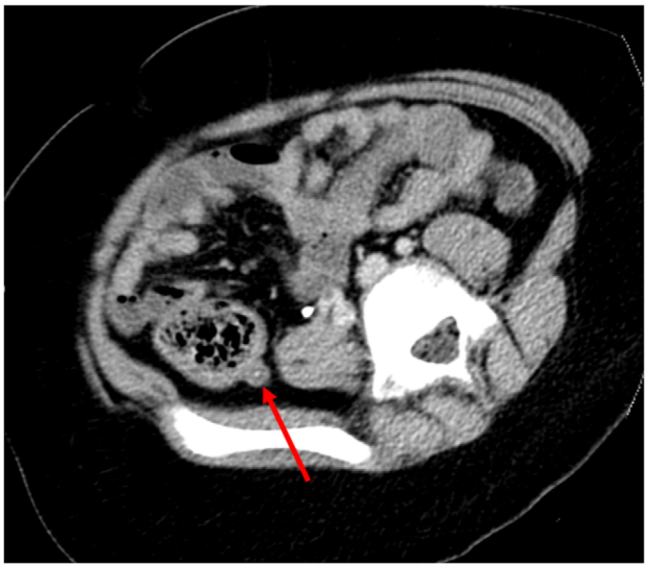

FIGURE 3 CT of abdomen in transverse view demonstrating the retrocecal appendix

pin point a diagnosis; or multiple possible diagnoses that could potentially contribute to the patient's symptoms.[1]

Choledochal or biliary cysts are primarily congenital dilatations of the intrahepatic or extrahepatic biliary ducts. There are multiple types of cysts depending on origin. They remain a part of the differential of abdominal pain, though rare. Typically, they present with right upper quadrant abdominal pain, jaundice, and right upper quadrant mass.[2] Children usually present with jaundice and abdominal mass, although they can present with pancreatitis and variable symptoms. In 1977, the 6 types of biliary cysts, intrahepatic, and extra hepatic cysts were combined into the same classification system.[2] Type 1 cysts have dilatation of the common bile duct and are the most common with 4 different sub types.[2] Type 2 cysts involve the extrahepatic bile duct but are true diverticula. Type 3 cysts involve the intra-duodenal portion of distal common bile duct. Type 4 cysts are characterized by the presence of multiple dilatations. Type 5 cysts are multiple cysts all located in the intrahepatic bile ducts.

The cysts can be present at birth or acquired later in life with an association with many conditions such as biliary atresia, abnormal pancreatobiliary junction, ventricular septal defect, and others.[3] Genetics may also play a role as there seems to be an increased prevalence among certain bloodlines.[3] Often, there may be the presence of chronic and acute inflammation of the cyst and nearby structures on histologic examination, which likely plays a role in some subtypes developing neoplasia.[4]

The average presentation is before 10 years of age and can have the classic triad of abdominal pain, jaundice, and palpable mass in the right upper abdomen.[5] Most commonly only 1 of these of symptoms will be present, or none as this patient did.[5,6] Other presenting symptoms can include vomiting, fever, pruritus, and weight loss with vomiting being the most common symptom.

Liver function studies in patients with biliary cysts are usually normal in the absence of obstruction. Those with type 1 and 4 cysts have an increased risk of cancer with a 10%–30% cancer rate, and therefore usually undergo Roux-en-Y hepaticojejunostomy.[7,8] If a cyst has been identified on computed tomography or ultrasound, the patient should undergo magnetic resonance cholangiopancreatography as it will identify obstruction and other potential complications such as chronic cholecystitis which this patient presented also had.

This patient presented with limited history and exam that easily may have been discharged with a diagnosis of gastroenteritis. If further imaging of the abdomen had not been ordered the cyst, cholecystitis, and appendicitis would not have been found initially. Appendicitis classically presents with periumbilical abdominal pain, radiating to the right lower quadrant within 24–48 hours; followed by fever and vomiting.[7] Many pediatric patients below 6 years of age do not present with a classic adult presentation so there must be a high index of suspicion to rule out or rule in appendicitis. Classically, to evaluate appendicitis, a pediatric patient would undergo ultrasound of the abdomen and then serial abdominal exams or computed tomography. Ultrasound of the abdomen allows for assessment of genitourinary tract, hepatic, gallbladder tree, intestines, and gallbladder pathology without ionizing radiation. Therefore, a workup of abdominal pain with concern for cholecystitis or appendicitis would classically start with ultrasound. A CT scan was performed due to ability to rule out multiple diagnoses such as bowel obstruction, intussusception, and appendicitis, while also expediting care in a busy emergency department at night. The sensitivity of appendicitis with CT in children is 94% to 100% but lower specificity.[9] An abnormal appearing appendix in a child is recommended to undergo appendectomy for resolution of symptoms.[10]

The location of the patient's pain would have been paramount to ascertain which of these diagnoses was truly causing the patient's symptoms. However, given the history and the fact that the cyst needed to be removed for future malignancy risk regardless, the prudent decision by the surgeon and mother was to perform appendectomy, cholecystectomy, and Roux-en-Y. The patient tolerated both of these procedures well and it was completed in a single operation. The operation did not reveal complete obstruction of the duodenum.

3 | CONCLUSION

Developmentally delayed children who are non-verbal represent a population of diagnostic challenge and the emergency physician must be cautious and consider multiple diagnoses, relying heavily on physical examination. The provider must also understand the limitations of the physical examination and use resources available to aid in diagnosis while also attempting to limit ionizing radiation exposure. CT is avoided by many providers in pediatric cases due to the risk of cancer of radiation.[11]

This case highlights the fact that children with intractable vomiting need to be observed and undergo a thorough workup. Appendicitis and other surgical diagnoses should always be a consideration that crosses the physician's mind but ruled out by physical examination and imaging

when warranted. Providers' threshold to pursue advanced imaging may differ and this case shows that there may be more than one etiology contributing to a patient's symptoms.

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