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Infantile Adenomyomatosis of the Gallbladder in a 3-Month-Old

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Abstract: Adenomyomatosis of the gallbladder is an acquired condition of the gallbladder with epithelial, mucosal, and muscular hypertrophy. The result is usually gallbladder wall thickening with associated diverticula known as Rokitansky-Aschoff's sinuses. These mucosal invaginations of the gallbladder wall may extend beyond the muscular layer. The condition is typically asymptomatic and is predominantly diagnosed in adults between 50 and 60 years of age, usually with concomitant cholelithiasis, motility disorders, or chronic inflammation. Few cases within the literature have been described in the pediatric population and even fewer within this subset have been diagnosed in infants. We describe a case of a 3-month-old male with failure to thrive, persistent nonbilious, nonbloody emesis, and elevated transaminases with ultrasound evidence of gallbladder adenomyomatosis. The patient was managed with outpatient laboratory monitoring and follow-up imaging.

Key Words: infantile adenomyomatosis, adenomyomatosis, gallbladder, pediatric

INTRODUCTION

Adenomyomatosis of the gallbladder is an acquired condition defined by mucosal hypertrophy with invaginations into a thickened muscularis, which are termed Rokitansky-Aschoff sinuses (1). Adenomyomatosis is typically diagnosed in patients 50 to 60 years of age, is asymptomatic, and is reported in 5% of cholecystectomy specimens within the adult population (1). Rarely, adenomyomatosis has been described in the pediatric population; however, even fewer cases have been described in infants (2–5). We present a case of a 3-month-old male with failure to thrive and persistent emesis found to have gallbladder adenomyomatosis.

CASE REPORT

A 3-month-old male, born full term via spontaneous vaginal delivery, presented to the emergency department with chronic nonbilious, nonbloody emesis and 3 oz of weight loss. The infant was undergoing evaluation for failure to thrive as an outpatient with chronic emesis beginning shortly after birth. There was no family history of gastrointestinal disorders and the infant was exclusively breastfed. Neonatal screening was unremarkable. Initially, the patient was diagnosed with gastroesophageal reflux and was started on lansoprazole as an outpatient, which only decreased the frequency of the vomiting.

On evaluation in the emergency department, vital signs were normal and physical exam was unremarkable. The patient's weight was 4.26 kg, which was less than first percentile. Laboratory testing was notable for thrombocytosis (672 × 10^3/UL; reference range 150–350 × 10^3/UL), elevated aspartate transaminase (99 IU/L; reference range 20–67 IU/L), and elevated alanine transaminase (110 IU/L; reference range 5–33 IU/L). Stool guaiac was negative. Pyloric ultrasound demonstrated a normal pylorus but extensive echogenic foci within the gallbladder wall. A dedicated gallbladder ultrasound confirmed multiple echogenic foci within the gallbladder wall suggestive of diffuse adenomyomatosis of the gallbladder (Fig. 1). Subsequently, a KUB and an upper GI series were normal. The patient was managed with follow-up laboratory analysis in 2 weeks and recommended repeat ultrasound in 4 to 6 months. On follow-up laboratory analysis, the patient's aspartate transaminase normalized and the alanine transaminase decreased to 45 UI/L. Follow-up abdominal ultrasound 4 months later demonstrated resolution of the imaging findings of adenomyomatosis. The patient was seen in clinic 5 months after presentation with persistent intermittent emesis and improving weight, within the 25 to 50th percentile for length.

DISCUSSION

Gallbladder adenomyomatosis can involve varying amounts of the gallbladder wall and may be segmental in approximately >60% of cases, fundal in 30% of cases, or diffuse in <5% of cases (6). Motility disorders, chronic inflammation, and cholelithiasis are suspected to be predisposing factors for the development of adenomyomatosis, but the exact pathogenesis remains uncertain (6). While adenomyomatosis is overall regarded as a benign condition, there have been prior studies reporting its malignant potential, particularly with the segmental type (7,8). Adenomyomatosis is predominantly diagnosed in adults, however, has been rarely reported in the pediatric population with only a few reported cases in infants (1–5). The clinical presentation of pediatric adenomyomatosis is variable with many cases found incidentally. The principal symptom in most cases was recurrent abdominal pain with few cases also reporting emesis (1).

Adult symptomatic adenomyomatosis is typically managed with cholecystectomy after exclusion of other causes of pain. In asymptomatic adult patients, guidelines have stated that cholecystectomy is not indicated for incidentally discovered adenomyomatosis (9). However, differences in management have been proposed for different asymptomatic subtypes. For asymptomatic fundal adenomyomatosis, cholecystectomy is not currently recommended (6). With previously reported associations of the segmental type of adenomyomatosis with adenocarcinoma, surgical management is recommended on a case-by-case basis for patients with this type, balancing patient risk factors and the morbidity of surgery (6).

Guidelines for the management of pediatric adenomyomatosis are not established given the rarity of the condition in this patient population. Many pediatric cases in which the initial presentation was abdominal pain were managed with cholecystectomy. If cholecystectomy is pursued, preoperative evaluation with magnetic resonance cholangiopancreatography (MRCP) may be helpful to characterize the extrahepatic biliary anatomy to avoid bile leak from resection of an

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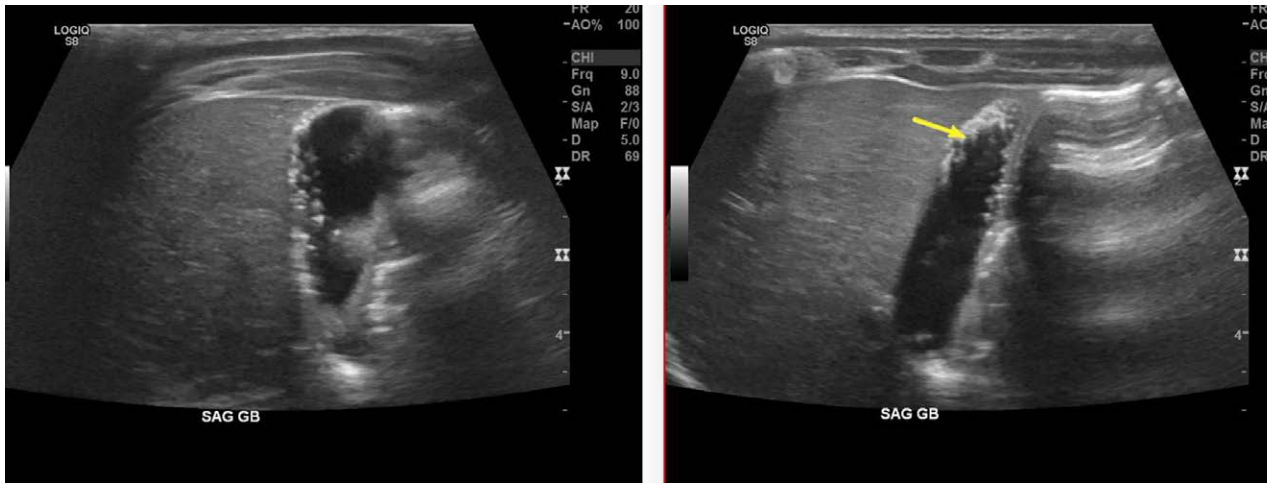

FIGURE 1. Sagittal ultrasound images of the gallbladder demonstrate diffuse echogenic foci throughout the gallbladder wall with comet tail artifact (arrow).

aberrant duct. In the cases of infantile adenomyomatosis, all patients in the literature were managed with follow up ultrasound in 3 to 8 months, with resolution of the imaging findings of adenomyomatosis in three of four patients after several months (2–4). Findings were unchanged in one patient followed with ultrasound after 7 months and follow-up ultrasound evaluation was planned for every 3 months to coincide with the patient's tumor surveillance for Beckwidth-Weidemann syndrome (5). Our patient was initially managed conservatively with follow-up laboratory analysis and recommended imaging in 4 to 6 months as the patient's failure to thrive could not be definitively attributed to adenomyomatosis. Consistent with the majority of reported infantile adenomyomatosis cases, follow-up abdominal ultrasound demonstrated resolution of the imaging features.

Adenomyomatosis can be identified sonographically by the presence of Rokitansky-Aschoff sinuses, which appear as well-defined anechoic regions within the gallbladder wall if they are bile-filled or echogenic foci if they are sludge or gallstone-filled as well as gallbladder wall thickening (1). Comet tail artifact may be seen due to reverberation between the sinuses. Differential considerations based on the imaging features include gallbladder carcinoma (although rare in the pediatric population), adherent cholelithiasis, and cholesterol polyps. Although using ionizing radiation, computed tomography may also suggest the diagnosis of adenomyomatosis if there is gallbladder wall thickening and a pseudocystic wall appearance with or without intramural calcifications, although this can mimic neoplastic findings (6). MRI with MRCP is superior in differentiating adenomyomatosis from cancer, but often requires sedation in the pediatric population. Adenomyomatosis shows the "pearl necklace" sign, seen as rounded foci of high signal intensity matching that of bile on fluid sensitive sequences within a thickened gallbladder wall which is 92% specific (10). Ultrasound is the first-line imaging study for diagnosis with a sensitivity of 65%; however, MRCP is the gold standard (6).

CONCLUSIONS

Pediatric, particularly infantile, adenomyomatosis of the gallbladder represents a rare presentation of a common pathology within the adult population and may present clinically with abdominal pain and emesis or be incidental. Ultrasound is an efficacious way to diagnose the condition.

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