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Case Report

Acalculous cholecystitis in the setting of immunoglobulin a vasculitis: a case report with literature review

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ABSTRACT

Immunoglobulin A vasculitis (IgAV), also known as Henoch-Schönlein Purpura, is the most common childhood vasculitis which presents with a tetrad of symptoms: palpable purpura, abdominal pain, arthralgias, and renal involvement. Gastrointestinal involvement has been reported with findings including abdominal pain, pancreatitis, intussusception, ischemia, and gallbladder involvement. Our patient had the classical presentation of IgAV with additional gallbladder thickening found on imaging. He was managed conservatively for his symptoms and gallbladder thickening. This case adds to the six prior cases that we found in our literature search reported of gallbladder thickening alongside IgAV. However, this was the only case that was managed conservatively resulting in a complete resolution of symptoms.

Keywords: IgAV vasculitis, Henoch-Schönlein Purpura, Acalculous cholecystitis

INTRODUCTION

Immunoglobulin A vasculitis (IgAV), formerly and more commonly known as Henoch-Schönlein Purpura (HSP), is the most common childhood vasculitis with an incidence of approximately 6 to 20 annual cases per 100,000 children.[2,3] IgAV clinically presents with the tetrad of palpable purpura, abdominal pain, arthralgias, and kidney involvement.[1] Although gastrointestinal involvement with IgAV is most commonly limited to abdominal pain, additional findings may include pancreatitis, intussusception, bowel ischemia, and gallbladder involvement.[4-13] Here, we report a unique case of an acute onset of IgAV with simultaneous acalculous cholecystitis.

CASE REPORT

A 4-year-old male with no significant past medical history presented to the emergency department of a tertiary referral hospital in late August 2022 after referral from his primary care provider. The patient presented with a recent history of an upper respiratory infection and a 1-week history of abdominal pain, recurrent non-bilious non-bloody vomiting, edema of the extremities, eyes, and occiput, arthralgias, and a purple, raised rash on his anterior legs bilaterally. The patient was initially seen by his primary care provider after the onset of the rash where he was given a presumed diagnosis of impetigo and prescribed a trial of cephalexin. Upon return to his primary care provider, urinalysis revealed microscopic proteinuria and hematuria, and the patient was sent to the emergency department.

In the emergency department, a complete blood count (CBC), complete metabolic panel (CMP), urinalysis, and abdominal ultrasound (US) were obtained. CBC was significant for a white blood cell count of 15,200/mm^3 and a neutrophil count of 9,970/mm^3. CMP was significant for a BUN of 15 mg/dL, an anion gap of 19 mmol/L, and an albumin of 3.9 g/dL. Urinalysis was significant for 30 mg/dL of protein, >80 mg/dL of

ketones, and 11-15 red blood cells per high power field. Given the provider's suspicion for IgAV, an abdominal ultrasound US was obtained to evaluate for intussusception. No evidence of intussusception was seen on US, but the gallbladder was incidentally found to be acalculous, hyperemic, and edematous with a thickened wall. The patient was given a fluid bolus, maintenance fluids, and pain management. He was admitted to the inpatient pediatrics service and both nephrology and pediatric surgery were consulted at this time.

The following morning, a C-reactive protein (CRP) level was obtained and found to be elevated at 14 mg/L. C3 complement, C4 complement, and anti-nuclear antibody levels were obtained and found to be within normal limits at 137 mg/dL, 23 mg/dL, and <1:80, respectively. The nephrology consultation recommended against steroids given the patient's clinical improvement overnight and his urine protein-creatinine ratio of 0.4. The pediatric surgery consultation did not recommend surgical intervention given the lack of postprandial pain or focal right-upper quadrant tenderness. Given the patient's clinical improvement regarding pain and emesis and his low degree of renal involvement, the patient was discharged after one day of care.

Four days after the patient's initial presentation, the patient returned to the emergency department with a new petechial rash of the buttocks, abdominal pain, scrotal swelling, and left hemiscrotal discoloration. At this time, CBC, CMP, urinalysis, abdominal US, and scrotal US were obtained. CBC revealed a decreasing white blood cell count from discharge. CMP and CRP were within normal limits. Urinalysis revealed no proteinuria and 4-10 red blood cells per high power field. Abdominal US revealed no intussusception, and scrotal US revealed edema without torsion or epididymitis. The patient was started on a prednisone taper and discharged for follow-up in 2 weeks.

DISCUSSION

IgAV is an immune-mediated leukocytoclastic small vessel vasculitis characterized by IgA1 immune complex deposition and complement activation.[14] It is a common vasculitis with the majority of cases occurring among children.[15,16] IgAV typically presents with the tetrad of palpable purpura, arthralgias, abdominal pain, and renal involvement. A clinical diagnosis of IgAV requires palpable purpura without thrombocytopenia or coagulopathy and with either two or three of the additional features listed in the tetrad.[17]

Of the four common symptoms, abdominal pain is the third most common and presents in approximately 50 to 75% of patients with IgAV.[18] In approximately 25% of patients, abdominal pain presents prior to the skin manifestations of IgAV.[19] Although the abdominal presentation is typically limited to colicky abdominal pain, intussusception occurs in approximately 0.6% to 2.3% of patients diagnosed with IgAV.[20,21] When the clinical suspicion for IgAV is high and accompanied by abdominal pain, an abdominal US can be obtained to evaluate for peritoneal fluid, bowel wall thickening, intussusception, and other etiologies. In our case, the patient presented with all four symptoms associated with IgAV and subsequently underwent an abdominal US to evaluate for possible intussusception. However, findings were more consistent with acalculous cholecystitis.

Acalculous cholecystitis (AC) is an acute inflammatory condition of the gallbladder resulting in gallbladder stasis and ischemia. This condition is most commonly seen in critically ill patients due to gallbladder hypomotility and ischemia.[22] In a pediatric setting, AC is often associated with viral infections, immune-mediated disorders, and critical illnesses.[23] Although surgery is a common therapeutic approach to AC in adults, surgery can theoretically be avoided in pediatric patients with different etiologies if the underlying cause for gallbladder inflammation can be addressed.[24] In the setting of acute immune-mediated AC such as IgAV-associated AC, supportive care and steroid use will likely address the underlying vasculitis and prevent the need for surgical interventions. In our patient, supportive care was sufficient for symptomatic resolution, and surgical interventions and invasive imaging were avoided.

Of the previously reported cases of IgAV-associated AC, three of the patients underwent surgical intervention and three did not (Table 1).[9-13] The three patients that underwent surgical interventions did not initially present with palpable purpura and, therefore, received a post-operative diagnosis of IgAV once the purpura appeared. Conversely, all three patients that initially presented with the IgAV tetrad were treated with supportive care and, in some cases, steroids. However, the patients that did not undergo surgery all underwent some form of invasive imaging such as endoscopic retrograde cholangio-pancreatography or esophagogastro-duodenoscopy. Therefore, in non-critically ill pediatric patients that present with findings consistent with AC such as in our case, clinicians should be cognizant of possible immune-mediated and infectious etiologies and should consider alternative therapies when appropriate.

Table 1: Literature review for case reports of IgAV with associated acalculous cholecystitis.

Study	Year	Age (Years)	Surgery	Description
Kumon et al[9]	1988	32	Yes	Patient presented with palpable purpura and myalgias. On day 5 of admission, patient developed abdominal pain, nausea, and vomiting. Patient underwent exploratory laparotomy which revealed an edematous gallbladder. Re-operation on day 7

Continued.

Study	Year	Age (Years)	Surgery	Description
				revealed small-bowel necrosis. Gallbladder histopathology revealed fibrinoid necrosis and small-vessel vasculitis.
Amemoto et al[10]	1994	8, 7	No, no	Case 1: Patient admitted with purpura, abdominal pain, and myalgias. Patient was treated with total parenteral nutrition for 5 days, and on day 5, abdominal US revealed gallbladder wall thickening. Patient underwent ERCP during stay and subsequently exhibited spontaneous recovery of gallbladder inflammation.
				Case 2: Patient was admitted with purpura, myalgias, and abdominal pain. Abdominal US revealed gallbladder wall thickening. Patient was started on steroid therapy, improved, and was discharged. She had recurrence of IgAV and cholecystitis 5 months later. During the course of her admissions, patient underwent 3 EGDs.
Hoffmann et al[13]	2004	53	Yes	Patient was admitted with abdominal pain. Abdominal US revealed thickened gallbladder without stones. Laparoscopic cholecystectomy revealed fibrinoid necrosis with transmural inflammation of intramural arteries. On day 4 post-op, patient developed palpable purpura and arthralgias. Patient was treated with methylprednisolone and pantoprazole.
Bacelli et al[11]	2013	9	No	Patient presented with IgAV tetrad and underwent abdominal US which revealed gallbladder wall thickening. MRCP was performed which revealed gallbladder fundus thickening. Patient was treated with IV meropenem and methylprednisolone.
Özkaya et al[12]	2016	7	Yes	Patient presented with abdominal pain, scleral icterus, and fever. Abdominal US revealed a thickened gallbladder wall with peri-cholecystic fluid. Ileo-colic intussusception was also seen. Exploratory laparotomy was performed with cholecystectomy. 2 weeks later, patient was re-admitted with arthralgias and palpable purpura. Patient underwent appendectomy for appendicitis 11 months after discharge.

There are several limitations to our report. Primarily, given the lack of surgical intervention in our case report, there is no histopathological information to definitively diagnose our patient with IgAV. However, given that the patient exhibited the characteristic history and tetrad associated with IgAV, a clinical diagnosis was appropriate. Additionally, given the patient's short inpatient duration, skin biopsies for immunofluorescence were not obtained.

CONCLUSION

IgAV is an immune-mediated small vessel vasculitis commonly seen in children. Abdominal pain is frequently seen with this condition, and pediatric patients presenting with the tetrad of symptoms associated with IgAV often undergo abdominal US to rule out intussusception. Here, we report a unique case of a patient with IgAV with simultaneous acalculous cholecystitis that was treated with supportive care without surgery or invasive imaging.

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