UDK 616.13/.14-002-053.2
COBISS.SR-ID 159495689

ISSN 2466-2992 (Online) (2024) br. 2 p. 35-42

IgA VASCULITIS AND ABDOMINAL PAIN – CASE REPORT AND REVIEW

IgA VASKULITIS I ABDOMINALNI BOL – PRIKAZ SLUČAJA I PREGLED LITERATURE

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Summary: INTRODUCTION Acute abdominal pain is one of the most common complaints in the pediatric population. There are various causes of abdominal pain. IgA vasculitis is characterized by purpura, abdominal pain, arthritis or arthralgias, and kidney problems. Atypical forms of this disease have been proven difficult to diagnose and treat. Here we present a case of atypical IgA vasculitis in a child who first presented with an episode of hematochezia. CASE REPORT: A 6-year-old boy presented with abdominal pain, vomiting, diarrhea and hematochezia, pale and dry skin, and dry mucus membranes. He was adynamic on admission but vital signs were normal. Initially, the patient was treated at his hometown hospital due to dehydration which was thought to be caused by viral gastroenteritis. Due to the persistence of symptoms and worsening of abdominal pain, he was then transferred to the Pediatric Surgery Clinic. Just after purpuric skin changes were noted, a pediatrician was called for a consultation, and IgA vasculitis was diagnosed. CONCLUSION Abdominal pain in children, although mostly caused by common pathology and conditions, can present a significant diagnostic problem, especially in pediatric patients presenting with atypical forms of IgA vasculitis. Physicians must be aware of rare causes of abdominal pain and include them in differential diagnosis.
***Key words*:** ultrasound; purpura; gastrointestinal symptoms; diagnostic dilemma.

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INTRODUCTION

Acute abdominal pain is one of the most common complaints in pediatric population. The key to distinguishing underlying causes is a detailed history, full clinical examination, and carefully selected investigations. Common causes of abdominal pain (viral gastroenteritis, constipation, etc.) are usually self-limiting [1]. Surgical causes of abdominal pain, such as acute appendicitis, acute intestinal obstruction, acute cholecystitis, appendicular abscess, volvulus, etc. are usually not self-limiting and must be included in differential diagnosis [2, 3]. IgA vasculitis (formerly Henoch-Schönlein purpura) is the most common form of systemic vasculitis in children. It is a self-limiting disease that presents as a tetrad of clinical manifestations: palpable purpura, arthritis/arthralgia, abdominal pain, and kidney disease. Abdominal pain in IgA vasculitis (IgAV) is usually acute, diffuse, and may be colicky. It is caused by submucosal hemorrhage and edema [4]. Common, rare, and life-threatening diseases in children presenting with abdominal pain along with ultrasound presentation and management are shown in Table 1. Here we present an atypical case of IgAV in a male child who first presented with acute abdominal pain and gastrointestinal symptomatology.

CASE REPORT

A 6-year-old male presented with abdominal pain, vomiting, diarrhea and hematochezia, pale and dry skin, and dry mucus membranes. He was adynamic on admission. Vital signs were normal: HR 132/min, BP 90/60 mmHg, RR 24/min, SaO2 98%. Height and weight were at the 52nd percentile. Symptoms began 7 days before admission with diarrhea, vomiting, and abdominal pain. He was treated at a local hospital with intravenous rehydration due to suspicious viral gastroenteritis. However, due to the persistence of symptoms, and especially the worsening of abdominal pain along with the development of hematochezia, the patient was transferred to Pediatric Surgical Clinic, Clinical Center Niš, for further evaluation. Due to suspicion of Meckel diverticulum, a radionuclide scan was done, but the results came back inconclusive. On a 2nd hospital day, the child developed periocular edema, and right knee swelling, followed by limited and painful joint movement. On the 3rd hospital day, an urticarial diffuse skin rash occurred. After pediatric consultation, a diagnosis of IgA vasculitis was suspected and the patient was transferred to the nephrology department of the Pediatric Clinic. On admission, the patient was hypertensive (TA 135/96 mmHg). The initial laboratory has shown leukocytosis, thrombocytosis, proteinuria, hypoalbuminemia, and IgA immunoglobulin elevation. The skin rash prograded to skin lesions noted in Figure 1. Treatment was induced with methylprednisolone, enalapril, and a short course of diuretics. The patient was discharged ten days later with tapering doses of corticosteroids and enalapril. A nephrologist did the follow-up, and the patient had fully recovered with no relapses.

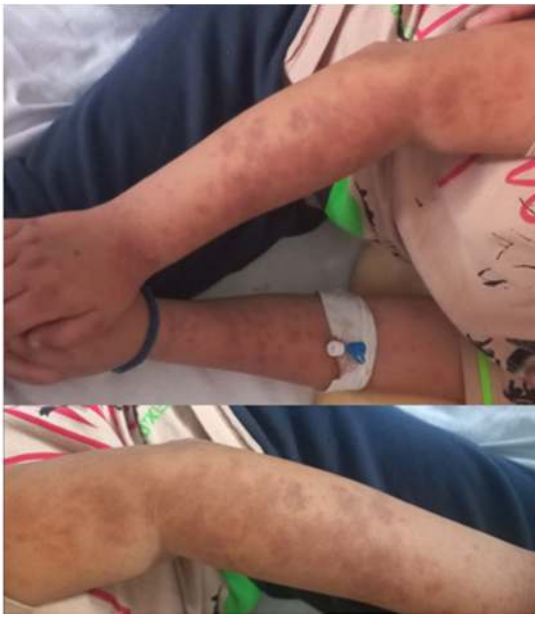

Figure 1. Skin changes in upper extremities of our patient

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DISCUSSION

IgA vasculitis is triggered by an abnormal immunologic response possibly linked to HLA-B35 and HLA-DRB1*01 [5]. Gastrointestinal symptoms may precede skin changes. This can present a diagnostic dilemma, due to many conditions that could mimic IgAV (table 1). A wide range of diagnostic procedures and thinking outside the box need to be used to differentiate this condition from other diseases.

IgAV therefore may be considered as a diagnosis of exclusion. In acute settings (child presenting with hematochezia), an emergency physician's way of thinking (thinking of rare, dangerous causes, and excluding them toward more common, benign conditions) has to be implemented. We have tried to simplify and illustrate the pathway for diagnosing abdominal pain in Figure 2.

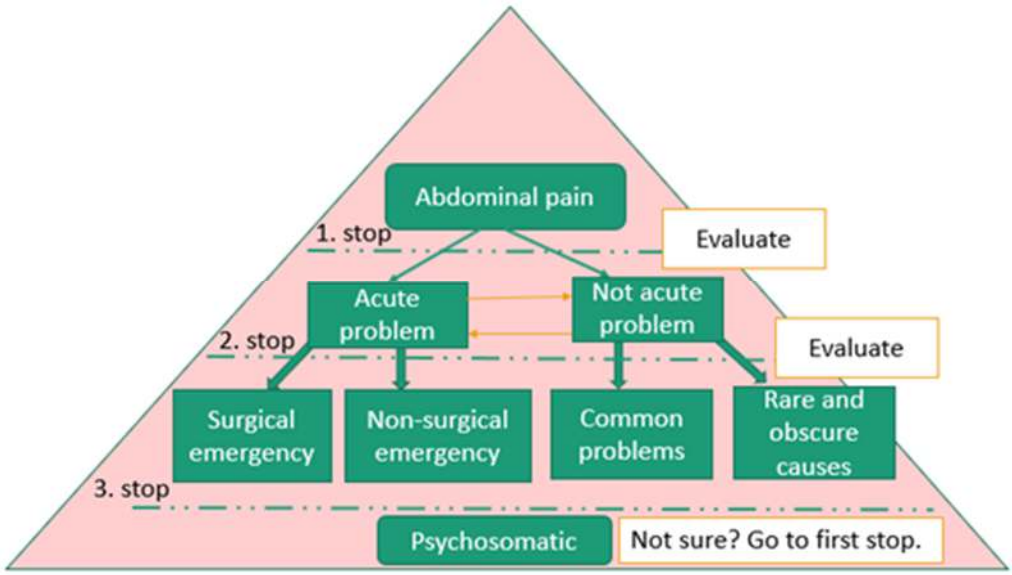

Figure 2. Pyramid of abdominal pain.

Viral gastroenteritis is a common complaint at primary care and presents as diarrhea, abdominal pain, and vomiting [6]. Acute appendicitis presents with positive Rovsing (severe RLQ pain with palpation of LLQ), and psoas sign (because the cecum acts as a cushion that blocks the examiner's hand). These patients need to be thoroughly clinically examined. Ultrasound is a valuable method in the diagnosis of appendicitis [7]. Contrast CT of the abdomen with fine-tuning to lessen radiation dosage and MRI (although not widely available) can also be used [8]. Ultrasound may also be useful in excluding causes such as testicular and ovarian torsion that present as sudden severe pain and swelling in the scrotum, abdominal pain, nausea and vomiting, and severe pelvic pain with nausea and vomiting respectively. Testicular torsion can be accessed with cremasteric reflex and TWIST score where a cutoff of 5 out of 7 has a positive predictive value and specificity of 100% [9]. In our case, unilateral scrotal pain, erythema, and swelling mimicked testicular torsion. Ultrasound in IgAV shows testis with normal morphology, epididymal enlargement, scrotal skin thickening, and hydrocele [10]. Hematemesis and hematochezia may be a sign

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of coagulopathy disorders. Sometimes, hematemesis in IgAV can precede other symptoms and may appear dramatic and require gastroenterologist consult [11]. Physicians may also think of renal colic. Point of care ultrasound (PoCUS) may provide early detection of lithiasis in renal colic and faster clinical decision-making with possible surgical referral and lower radiology (CT) referral [12].

Duodenal hematoma is a rare condition in children, that can present with acute abdominal pain. It may occur after a diagnostic procedure, but also after blunt abdominal trauma, coagulopathy disorders, and leukemia [13]. Another condition that should be considered is gastric ulcer with or without perforation [14]. Gastric ulcers in children may occur with H. pylori infection, but detecting H. Pylori alone without peptic ulcer disease (PUD) in children shouldn't be a reason for antibiotic treatment. In addition, there may even be a not-yet well-understood role of H. pylori's beneficial effect on children's immune system development [15]. Malrotation and volvulus are described in infants, and therefore early diagnosed. Rarely they occur in older children and as such may clinically look like pancreatitis (author's personal experience), gastroenteritis, or diabetic ketoacidosis. Developing ultrasound diagnostic algorithms in order to speed up diagnosis and treatment is very important [16, 17]. In females of reproductive age, ruling out ectopic pregnancy must be considered in any case where a definitive cause of acute abdominal pain is still unknown. Severe lower abdominal pain, accompanied by elevated hCG and ultrasound may point to diagnosis [18]. In patients with IgAV, there may be a risk for intussusception. Children with age at onset of IgAV below 6 years, children not receiving glucocorticoid therapy within 72 h of onset of gastrointestinal (GI) symptoms such as hematochezia, and patients with increased D-dimer levels are at risk for intussusception [19].

Acute non-calculous cholecystitis may co-occur with IgAV and other immune-related disorders (Systemic lupus erythematosus, Kawasaki disease, Juvenile dermatomyositis), but also occurs in critically ill patients, post-surgically, and may be triggered by viral (Epstein-Barr virus) and other infectious pathogens [20]. Acute and chronic pancreatitis are rare in children. They may occur in congenital (pancreas divisum, pancreaticobiliary maljunction), genetic predisposition (PRSS1, SPINK1 mutations, CFTR in cystic fibrosis, etc.), autoimmunity, infections, and toxic-metabolic risk factors (alcohol, chronic renal failure, hypercalcemia, hyperlipidemia) [21].

Paroxysmal nocturnal hemoglobinuria (PNH) is a rare life-threatening disease mostly caused by phosphatidylinositol glycan anchor biosynthesis class A (PIGA) somatic mutation leading to GPI-anchor deficiency for the decay-accelerating factor (DAF/CD55) and membrane inhibitor of reactive lysis (MIRL/CD59). With the lack of GPI-anchor proteins erythrocytes are prone to lysis by membrane attack complex [22]. These patients may suffer from hemolytic anemia and vomiting, but may also present with abdominal pain due to mesenteric microthrombosis, and rare complication of bilateral renal vein thrombosis [23]. Familial Mediterranean fever (FMF) is caused by missense and nonsense mutations in the MEFV gene, which codes for a protein called pyrin, involved in the function of inflammasome multiprotein complexes. FMF can manifest with acute abdominal pain during acute attacks, and signs of peritonitis. Patients may often undergo unnecessary procedures and even repeated surgical interventions such as laparotomy. FMF can also present with scrotal attacks and erysipelas-like erythema. Additionally, joint attacks may occur. All of this can resemble atypical IgA vasculitis (IgAV). Yalcinkaya–Ozen criteria are used in children with suspected FMF [24].

Table 1. Some of the causes of acute abdominal pain and related diagnostic with clinical cues.

Disease	Clinical features	Ultrasound	Management	Ref.
Appendicitis	Right lower quadrant pain, guarding, migration of periumbilical pain to the right lower quadrant	>0.6 cm, fat standing (increased echogenicity of periappendiceal fat), hyperemia, appendicolith (sometimes normal), echogenic free fluid suspicious for rupture	Surgical or conservative	2, 7
Duodenal hematoma	Epigastric pain, vomiting	Uniform echogenic mass along the duodenal convexity, intestinal wall thickening, obstruction of the duodenal lumen	Primarly non-surgical	13
Gastric ulcer	Burning epigastric pain triggered by meal	Gastric antrum or duodenal bulb show marked, diffuse, and circumferential wall thickening, “HH sign“	Primarily non-surgical	14
Gastroenteritis	Abdominal cramping	Not significant	Primary non-surgical, symptomatic, IV fluids	6
IgA vasculitis	Diffuse (colicky) abdominal pain	Echogenic kidneys, target or doughnut sign (intussusception), ddx scrotal edema	Primarily non-surgical	4
Intussusception	Vomiting, hematochezia, abdominal pain	Concentric ring sign	Surgical	19
Ovarian/ Testicular torsion	Abdominal pain, nausea, vomiting	Twisting of spermatic chord/alterated blood flow, increase in size/ovarian edema/variable echogenicity	Primarily surgical	10
Pancreatitis	Severe, dull pain in the upper left abdominal quadrant or central epigastric pain	Increased pancreatic volume with a marked decrease in echogenicity displacement of the transverse colon and/or stomach	Conservative	21
Renal colic	Sudden flank pain that extends anteriorly and inferiorly towards the groin	Echogenic foci, acoustic shadowing, twinkle artifact on Doppler, color comet-tail artifact	Primarily non-surgical. Analgesia, fluids	12, 1
Volvulus	Bilious vomiting in neonate, abdominal distension and pain	Clockwise whirlpool sign, abnormal superior mesenteric vessels, abnormal bowel, free intra-abdominal fluid	Surgical	16

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Acute hepatic porphyrias (AHP) are rare, life-threatening genetic disorders that are caused by enzyme deficiencies in the heme biosynthetic pathway. AHP may present in a child with fever, crampy abdominal pain, darkening of the urine, mental confusion, and cutaneous manifestations on sun-exposed skin. In cases of recurrent and unexplained episodes of abdominal pain in a child, a urinary analysis for δ-aminolaevulinic acid (ALA), porphobilinogen (PBG), and porphyrins may prove diagnostic [25]. Mutations of the SERPING1 gene are thought to give rise to hereditary angioedema (HA), which can lead to recurrent abdominal pain in a high number of cases. Symptoms may overlap with other rare diseases, but cutaneous swelling failing to respond to usual treatment may provoke consideration of HA [26]. Paroxysmal attacks of dull, colicky and/or diffuse abdominal pain that last 17 hours on average can be due to a rare clinical entity of abdominal migraine which can be diagnosed using Rome IV criteria. Various pathophysiology models are proposed to explain this disorder in children. They include theories about visceral hypersensitivity, gut-brain axis, and psychological stressors [27]. Abdominal pain can pose a significant diagnostic challenge, particularly in pediatric patients with atypical forms of IgA vasculitis. Physicians should be mindful of rare causes of abdominal pain and include them in differential diagnosis.

CONCLUSION.

Abdominal pain in children, although mostly caused by common pathology and conditions, can present a significant diagnostic problem, especially in pediatric patients presenting with atypical forms of IgA vasculitis. Physicians must be aware of rare causes of abdominal pain and include them in differential diagnosis.

Conflict of interest:
The author declares no conflict of interest

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IgA VASKULITIS I ABDOMINALNI BOL – PRIKAZ SLUČAJA I PREGLED LITERATURE

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Sažetak: UVOD.Akutni abdominalni bol je jedan od najčešćih razloga posete pedijatrijskoj ambulanti. Etiologija abdominalnog bola je široka. IgA vaskultis karakterišu purpura, abdominalni bol, artritis, artralgija, kao i bubrežna patologija. Atipični slučajevi, naročito gde gastrointestinalni problemi prethode pojavi ospe po koži, mogu predstavljati dijagnostički i terapijski problem. Prikazujemo atipični slučaj „IgA" vaskulitisa kod pedijatrijskog pacijenta gde je bolest počela sa hematohezijom.
PRIKAZ BOLESNIKA.Šestogodišnji dečak primljen je zbog abdominalnog bola, povraćanja, prolivastih stolica, hematohezije, blede i suve kože i suvih sluznica, sa znacima dehidracije. Bio je adinamičan, ali vitalni znaci su bili uredni. Inicijalno je tretiran u matičnoj ustanovi zbog dehidracije pod sumnjom na virusni gastroenteritis. Zbog održavanja simptoma i pogoršanja abdominalnog bola preveden je na Kliniku za dečiju hirurgiju. Po pojavi kožnih promena i nakon konsultacije pedijatra preveden je na Kliniku za pedijatriju gde je postavljena dijagnoza „IgA" vaskulitisa. ZAKLJUČAK.Možemo zaključiti da abdominalni bol kod dece može predstavljati dijagnostičku dilemu, naročito kod pacijenata sa atipičnim „IgA" vaskulitisom. Lekari moraju biti svesni i povremeno uzeti u obzir i retka stanja i oboljenja u diferencijalnoj dijagnozi abdominalnog bola.
Ključne reči ultrazvuk; purpura; gastrointestinalni simptomi; dijagnostička dilema

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