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Non-Surgical Management of Colo-Colonic Intussusception in Patients with Hereditary Angioedema

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Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
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Conflict of interest: None declared

Patient: Female, 17
Final Diagnosis: Colo-colonic intussusception
Symptoms: Abdominal pain
Medication: —
Clinical Procedure: —
Specialty: Radiology

Objective: Unusual clinical course

Background: Hereditary angioedema (HAE) is an autosomal disease caused either by deficiency or presence of a non-functioning C1 inhibitor. The lack or non-functionality of said inhibitors leads to activation of an inflammatory cascade, which result in cutaneous and mucosal edema. Most patients with HAE present with either cutaneous, laryngeal/pharyngeal, or gastrointestinal exacerbations. An uncommon gastrointestinal manifestation of HAE is an intussusception, which in most cases require invasive/surgical management.

Case Report: A 17-year-old Hispanic female patient with past medical history of HAE, presented with a 4-day history of episodic abdominal pain, worsening during the last 2 days with associated nausea, vomiting, and bright red blood per rectum. The abdominal ultrasound performed at our institution showed an elongated region of hypoechoic and hyperechoic concentric rings, raising suspicion of an intussusception. The patient was treated conservatively, with 30 mg of ecallantide and a unit of fresh frozen plasma (FFP). Follow-up abdominopelvic computed tomography scan was performed approximately 20 hours after the administration of fresh frozen plasma revealing complete interval resolution of the colo-colonic intussusception. Subsequently, the patient was kept under hospital care for the next 4 days with adequate progression of diet and without recurrence of intussusception.

Conclusions: To the best of our knowledge, most cases of patient with HAE presenting with intussusception have been treated with invasive/surgical procedures. In our case, conservative management has proven successful to reduce edema with subsequent non-surgical reduction of the intussusception. By directly targeting the pathophysiologic aspects of HAE, an unnecessary invasive procedure, as well as its potential complications, were avoided.

MeSH Keywords: Angioedemas, Hereditary • Enema • Intussusception • Kallikreins • Plasma

Full-text PDF: <https://www.amjcaserep.com/abstract/index/idArt/910223>



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Background

Hereditary angioedema (HAE) is a rare autosomal dominant disease, caused by a non-functioning C1 inhibitor or by C1 inhibitor deficiency [1]. The lack or non-functionality of C1 inhibitors leads to production and release of vasodilators, which in turn leads to cutaneous and mucosal edema. An uncommon gastrointestinal manifestation of HAE is an intussusception. It is caused by telescoping of bowel loops within the lumen of another segment of bowel [1]. In most reported cases, invasive/surgical management is needed for reduction of the intussusception. In the case presented, a non-surgical approach directly targeting the pathophysiology of HAE proved to decrease the edema, which resulted in reduction of the intussusception.

Case Report

A 17-year-old Hispanic female patient, with past medical history of HAE diagnosed at approximately 2 years of age, presented with a 4-day history of abdominal pain. The patient stated pain began as a constant periumbilical pressure, a 5 out of 10 on the pain scale. Approximately 4 hours later, the pain worsened, increasing to a 7 out of 10 on the pain scale. The patient's mother initially took the patient to an outside institution where the patient was administered 30 mg of ecallantide subcutaneously, as previously used to treat patient's symptoms of HAE. After symptoms improved, the patient was discharged home.

Two days later, the patient was taken to same outside institution due to recurring abdominal pain. The patient referred abdominal pain became intermittent with episodes lasting approximately 2 to 5 minutes, accompanied by nausea and vomiting. She then experienced several loose bowel movements with bright red blood per rectum. Pain began to irradiate to her upper abdomen, predominantly to the left upper quadrant. An abdominal ultrasound, as well as an abdominopelvic computed tomography (CT) scan were performed.

The latter showed evidence to suggest a colo-colonic intussusception at the level of the distal splenic flexure and proximal descending colon, measuring up to 18 cm in length, with associated moderate amount of free fluid. There was no free air identified, to suggest bowel perforation. The patient was then transferred to our institution for further management.

The patient denied fever, weight loss, skin rash, headaches, dizziness, palpitations, respiratory difficulty, loss of appetite, or hematuria. The patient reported a recent upper respiratory tract infection with clear nasal secretions and odynophagia. Her last exacerbation of known HAE was approximately 2 months ago, when she was treated with ecallantide for symptom relief. The patient's family history revealed her father as well as paternal grandfather both suffer from HAE. Physical examination showed soft and depressible abdomen, non-tender to palpation, and with increased bowel sounds. No masses or organomegaly were detected.

Laboratory workup performed in our institution revealed normal white blood cell and platelet count. Hemoglobin and hematocrit levels were within normal limits. Additionally, the patient's complete metabolic and coagulation panels were unremarkable.

An abdominal ultrasound was performed in order to confirm the suspected intussusception. An elongated region of hypoechoic and hyperechoic concentric rings, measuring at least 12 cm long by 4.4 cm AP by 5.4 cm transverse, was appreciated (Figure 1). Sonographic findings were highly suggestive of a colo-colonic intussusception. Additionally, intra-abdominal and pelvic free fluid were observed.

A unit of fresh frozen plasma (FFP) was administered, and the patient was treated conservatively. A follow-up abdominopelvic CT scan was performed at approximately 7 hours after performing the ultrasound. The CT scan showed interval improvement of the previously identified colo-colonic intussusception, which had reduced to approximately 8 cm in length (Figure 2). Oral contrast was seen passing through the



Figure 1. Gray scale abdominal ultrasound in a patient with colo-colonic intussusception. Transverse view (A) and 2 longitudinal views (B, C) show an elongated region of hypoechoic and hyperechoic concentric rings, measuring at least 12 cm long by 4.4 cm AP by 5.4 cm transverse, concerning for colo-colonic intussusception.

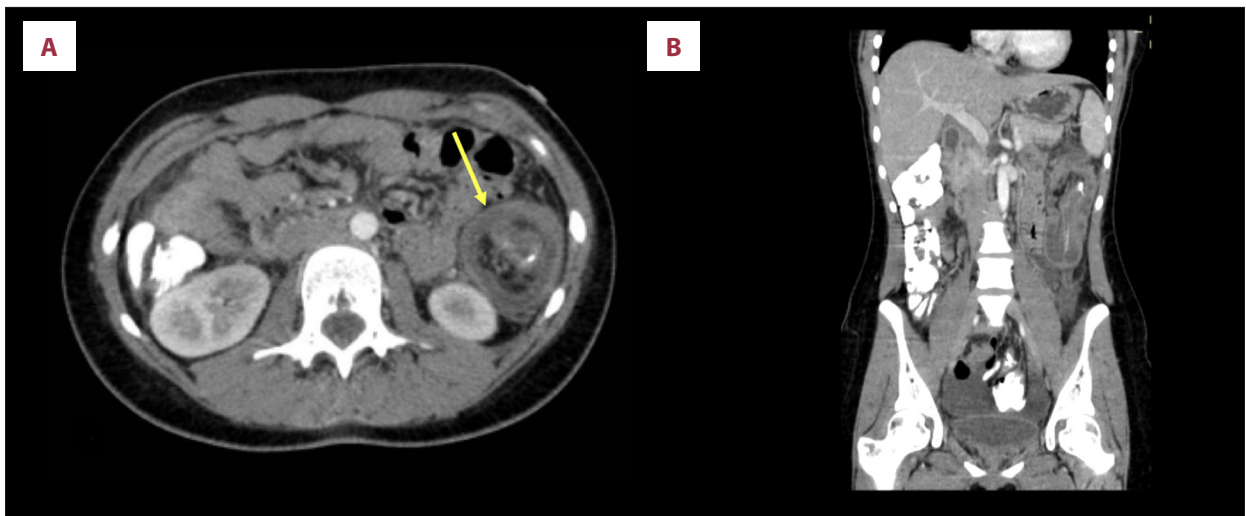


Figure 2. Contrast-enhanced abdominopelvic computed tomography (CT) in patient in colo-colonic intussusception. Axial (A) and coronal (B) reconstructions of a follow-up abdominopelvic CT scan performed approximately 7 hours after the abdominal ultrasound (shown in Figure 1) show the previously identified colo-colonic intussusception (arrow), now measuring approximately 8 cm long. Oral contrast seen passing through intussusception into the distal colon and rectum. Severe mucosal edema noted, more prominent along the distal aspect of the intussusception. There was no CT evidence to suggest bowel ischemia, bowel perforation or obstruction.

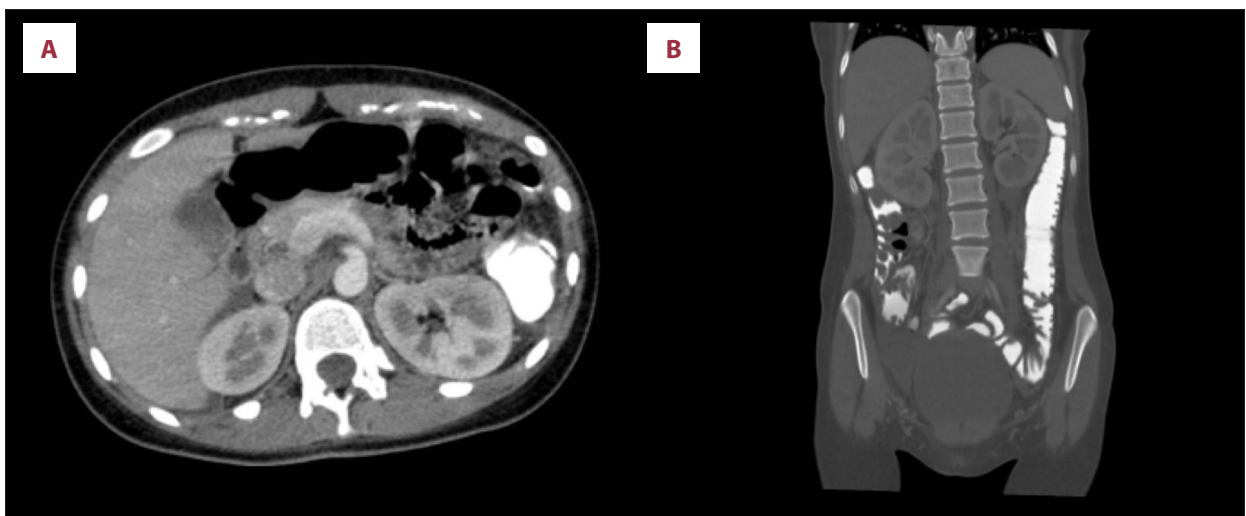


Figure 3. Follow-up contrast-enhanced abdominopelvic computed tomography (CT) scan. Axial (A) and coronal (B) reconstructions of a follow-up abdominopelvic CT scan performed approximately 13 hours after prior abdominopelvic CT scan (shown in Figure 2), show interval resolution of the colo-colonic intussusception with residual bowel wall edema.

intussusception into the distal colon and rectum. Severe mucosal edema was also noted, more prominent along the distal aspect of the intussusception. There was no CT evidence to suggest bowel ischemia, bowel perforation, or obstruction identified at the time. Close follow-up with abdominopelvic CT scan was recommended.

Interval resolution of the intussusception was observed in a second follow-up abdominopelvic CT scan performed approximately 13 hours after first study. It showed residual bowel

wall edema of a long segment of small bowel, involving the distal ileum and jejunum (Figure 3). Additionally, a segment of large bowel showed haustral thickening and associated fat stranding. The patient was kept under hospital care for the next 4 days with adequate progression of diet and without recurrence of intussusception.

Discussion

Hereditary angioedema (HAE) is a rare autosomal dominant disease, with an estimated prevalence of 1 patient for every 50 000 individuals, of which 40% experience their first angioedema attack before the age of 5 years [1]. HAE can be caused by either a deficiency of C1 inhibitor (known as HAE type I) or a presence of a non-functioning C1 inhibitor (known as HAE type II) [2]. Through molecular pathways (beyond the scope of this report) the lack or non-functionality of C1 inhibitor leads to production and release of vasodilators, which in turn lead to cutaneous and mucosal edema.

Most patients with HAE present either cutaneous, laryngeal/pharyngeal, or gastrointestinal exacerbations [1], some of which may be severe and life-threatening. An uncommon gastrointestinal manifestation of this disease is an intussusception. Intussusception is caused by telescoping of bowel loops within the lumen of another segment of bowel. The most common type is ileo-colonic intussusception, where the ileum prolapses into the lumen of the adjacent colon [3]. It is usually seen in pediatric patients and presents with colicky abdominal pain and currant jelly stools [3]. The radiographic study of choice for diagnosis of intussusception is an abdominal ultrasound [3], which shows hypoechoic and hyperechoic concentric rings corresponding to alternating layers of bowel wall with mesenteric fat [4]; known as the “target sign”. Once diagnosed, reduction with air or contrast enema is the gold standard for management of the intussusception. However, in patients with HAE, an invasive procedure such as air or contrast enema, or even a surgical intervention, may worsen the patient’s condition [5].

To the best of our knowledge, only a few cases of non-surgical management of intussusception in patients with HAE have been reported. In the case presented, conservative management has proven to reduce edema with subsequent non-invasive

reduction of the intussusception. FFP and ecallantide were both administered, leading to resolution of symptoms and reduction of intussusception in less than 24 hours. Ecallantide, a recombinant plasma kallikrein inhibitor, is considered part of the first-line therapy for acute attacks of HAE. It is usually administered as 3 separate subcutaneous injections of 10 mg/mL each [6]. Acting as a kallikrein inhibitor, it blocks the activation of bradykinin, which leads to a decrease in the degree of vasodilatation and vascular permeability. On the other hand, FFP is considered a second-line agent for treatment of acute angioedema attacks. Two units of FFP are administered initially, and this dose can be repeated every 2 to 4 hours until improvement of symptoms [7]. FFP contains C1 esterase inhibitor, which inhibits the kallikrein-kinin pathway and ultimately blocks the activation of bradykinin. This treatment combination leads to a decrease in edema with conservative reduction of the patient’s colo-colonic intussusception by directly targeting the pathophysiologic aspects of HAE.

Conclusions

HAE, due either to the lack or non-functionality of C1 inhibitors, leads to cutaneous and mucosal edema. An uncommon gastrointestinal manifestation of HAE is an intussusception. In the case presented, conservative management has proven to reduce edema with subsequent non-surgical reduction of the intussusception. By directly targeting the pathophysiologic aspects of HAE, an unnecessary invasive procedure, as well as its potential complications, were avoided.

Conflict of interest

None.

References:

1. Cicardi M, Zuraw B: Hereditary angioedema: Epidemiology, clinical manifestations, exacerbating factors, and prognosis. UpToDate 2016 Oct. Available from: URL: https://www.uptodate.com/contents/hereditary-angioedema-epidemiology-clinical-manifestations-exacerbating-factors-and-prognosis?source=search_result&search=hereditary%20angioedema&selectedTitle=3-150
2. Patel N, Suarez L, Kapur S, Bielory L: Hereditary angioedema and gastrointestinal complications: an extensive review of the literature. *Case Reports Immunol*, 2015; 2015: 925861
3. Donnelly LF: Gastrointestinal. In: *Pediatric imaging the fundamentals*. 1st ed. Pennsylvania: Elsevier, 2009; 86–124
4. Hertzberg BS, Middleton WD: General abdomen. In: *Ultrasound: The requisites*. 3rd ed. Pennsylvania: Elsevier, 2016; 204–28
5. Craig T, Pursun EA, Bork K et al: WAO guideline for the management of hereditary angioedema. *World Allergy Organ J*, 2012; 5: 182–99
6. Ecallantide: drug information. UpToDate. Available from: URL: https://www.uptodate.com/contents/ecallantide-drug-information?source=see_link
7. Cicardi M, Zuraw B: Hereditary angioedema: treatment of acute attacks. UpToDate 2016. Available from: URL: https://www.uptodate.com/contents/hereditary-angioedema-treatment-of-acute-attacks?search=hereditary%20angioedema%20&source=search_result&selectedTitle=2-56&usage_type=default&display_rank=2#H18