



Reducible appendiceal intussusception: A case report and review the literatures

Abdulrasool Alaei^{1*}

¹Department of Radiology, Faculty of Medicine, Mazandaran University of Medical Sciences, Sari, Iran

ARTICLE INFO

Article type:

Case and Review

Article history:

Received: 19 Sep 2012

Revised: 19 Dec 2012

Accepted: 12Jan 2013

Keywords:

Appendix, Intussusception,
Barium reduction

<http://jpr.mazums.ac.ir>

ABSTRACT

Appendiceal intussusception is a very rare cause of abdominal colic in childhood. This paper reports a reducible appendiceal intussusception in the barium enema in a girl and reviews articles in PubMed.

Introduction

Appendiceal intussusception is a rare condition which has been diagnosed in case reports.^{1,2} The incidence rate of appendiceal intussusception is 0.01% in 11000 human appendix specimens.^{1,2} This condition primarily occurs in children.^{2,3} In most cases, intussusception is limited to appendix but in some conditions cecocolic or ileocolic intussusception may occur.⁴ A variety of underlying anatomical and physiopathological causes as lead point are described.²⁻⁹ So far, several forms of clinical presentations and classifications have been reported.^{2,3} Diagnosis of appendiceal intussusception is by ultrasound,

barium enema, CT scan and colonoscopy^{2, 10-13} and the main routes of treatment are hydrostatic reduction and surgery. A complete review of appendiceal intussusceptions of childrens in pubmed also was performed. The age, sex, presentation, treatment and pathology were reviewed on childrens.¹⁴⁻²⁵

Case presentation

Case report

A four-year old girl referred to our hospital with a history of one week abdominal pain, intermittent nausea, agitation, vomiting and

* Corresponding author: Abdulrasool Alaei, MD., Assistant professor of radiology

Mailing address: Department of Radiology, Bou Ali Sina Hospital, Pasdaran Boulevard, Sari, Iran

TEL: 0098 9111515116

Email: ar_alaei@yahoo.com

diarrhea. The patient had watery voluminous diarrhea without blood and mucous 2-3 times/day. Later, also abdominal distention with no passage of gas and defecation occurred. She was the first child of her family. There was no history of prior diseases, and surgery. The child weighed 12 kg, her heart rate was 100\bpm, and blood pressure was 100/60. Oropharyngeal, cardiovascular and pulmonary examinations were normal. The results of haematological and stool examinations were also normal and fecal occult blood was negative. The patient appeared to be in distress with abdominal tenderness, mild distention and hyperactive bowel sounds. The complete blood count and cell differential were normal. Plain abdominal x-ray showed multiple dilated gas – filled proximal bowel. Ultrasound demonstrated thickened base of appendix with target lesion. Barium enema revealed typical Coil – Spring pattern in Cecal caput with invagination of appendiceal base and filling defect in appendiceal lumen (Figure1).



Figure 1. Barium enema reveals characteristic cecal coil-spring sign with invagination of appendiceal base and filling defects in appendiceal lumen.

Appendiceal intussusception was reduced. The patient was followed and by resolving the symptoms (after four days) she was discharged. She was not improvement of symptoms after one year follow up.

Results

The age, sex, clinical findings, diagnostic imaging, treatment and pathology were evaluated at case reports. The patient age varies from 2month to 15years with average age of 6.9 years (table-1).

Male patients (71.4 %) more affected than female patients (29.6 %)(Table2).

The most clinical findings were abdominal pain (90%) (Table3).

The most of case reports were diagnosed at preoperative imaging procedures (83 %).The most common pathologic finding was inflammation (42%)(Table-4).

Appendectomy was performed in most of the case reports. Surgical Procedures are open (mostly) or laparoscopic .Partial colectomy also wereachieved at patient in appendiceal intussusception without reduction or relapse.

Discussion

Appendiceal intussusception is very uncommon^{1,2} which is seen more in the first two decades of life.²⁶ The first case of appendiceal intussusception was reported in 1858.

Half of pediatric cases occur in children less than 10 years of age and the boy/ girl ratio occurrence is 4-5/1. Intussusception of the appendix probably occurs by the same mechanism and pathogenesis as intussusception elsewhere. Appendiceal intussusceptions mostly simple (primary) form intussusceptions of appendicular mucosa (ie, only spontaneously invagination of appendix into cecum) in some instances might be associated with secondary cecocolic or ileo colic intussusception.^{1, 26, 27} Primary type occure by either partial and complete invagination of the appendix into itself.

Table1: case reports of appendiceal intussusceptions in children

| Author | G | Age | Time of onset | Clinical findings | Diagnostic procedure | Treatment | pathology | year |
|------------|---|--------|---------------|--|--|---|------------------------------------|------|
| Dunavant | M | 8mo | 6d. | Hematochezia Inversion of appendix with anal protrusion | ^a AI | Surgical reduction Appendectomy | - | 1950 |
| Atkinson | F | 2.5y | 1mo. | Cramp/constipation ^b RLQ tenderness | ^c BE: AI without reduction | Appendectomy | Lymphoid Hyperplasia | 1976 |
| " | F | 4.5yr. | 3wk | Cramping /vomiting | ^e US: Soft tissue/mass in RLQ BE: AI without Appendix filling | Right hemicolectomy inverted Appendix | " | 1976 |
| Kleinman | M | 15yr | 4wk | Cramping/ RLQ tenderness | ^d SBT: Filling defect in cecum BE: Reduction with Appendix filling | Ba.reduction | - | 1979 |
| Casteel | F | 14yr | - | Recurrent /abdominal pain/ vomiting | BE: Spiral shell sign (AI) | Appendectomy | - | 1986 |
| Bailey | M | 7yr | - | Recurrent abdominal pain | Colonoscopy Inverted appendix | Polypectomy Appendectomy | Jvvenile cecal polyp | 1987 |
| Ekert. | M | 5yr | - | RLQ pain | US: Target sign in RLQ | Appendectomy Ileocolic resection (After relapse) 2yr. | CF | 1998 |
| Galatioto | F | 27m | - | RLQ pain | US: Target sign in RLQ | Laparoscopic Reduction | - | 1999 |
| Pumberger | M | 4yr | 3wk | Colicky /RLQ mass RLQ Tenderness | US: RLQ multicentric Ring sign ^f WSCM enema reduction (AI) | Appendectomy after recurrence | Chronic inflammation | 2000 |
| Pumberger | | 5yr | 2dy | Pain/ Hematochezia | US: Target sign Wscmeneua No reduction(AI) | Appendectomy | Chronic inflammation | 2000 |
| Gupta | M | 9yr | 1mo | Hematochezia | Colonoscopy /AI | Appendectomy Partial cecectomy | Resolving infectious process | 2000 |
| Attard | M | 5yr | 1wk | Abdominal pain /vomiting Constipation/ RLQ Tenderness | AXR : Fecal impaction BE: AI reduction colonoscopy: inverted appendix | Ileocecal resection | CF | 2000 |
| Koumanidou | M | 6yr | 1wk | RLQ pain /Nausea/ vomiting | US: Donut – Like mass in RLQ BE: Coilspring /nonfilling appendix | Appendectomy | Lymphoid Hyperplasia | 2000 |
| Koumanidou | M | 5yr | 5dy | RLQ pain /Diarrhea / RLQ Tenderness | AXR: Normal US:Donut Like mass in RLQ BE: Coilspring | Appendectomy | Lymphoid Hyperplasia | 2000 |
| Koumanidou | M | 5yr | 7wk | Periumblical pain/ RLQ tenderness | AXR: Normal US:Donut Like mass in RLQ BE: AI | Appendectomy | Lymphoid Hyperplasia | 2000 |

Continued Table1: case reports of appendiceal intussusceptions in children

| Author | G | Age | onset | Clinical findings | Diagnostic procedure | Treatment | pathology | year |
|------------------|---|------|-------|---|---|---|-----------------------------|------|
| Koumanidou | F | 7yr | 3dy | Abdominal pain /Nausea/ Vomiting/ RLQ tenderness | AXR-Bowel obstruction US: Donut Like mass in RLQ Ba – Reduction | Appendectomy | Maltoma | 2005 |
| Karabulut | M | 8yr | 24hr | Abdominal pain /Nausea/ Vomiting/ RLQ tenderness /RLQ tenderness RLQ mass | AXR-Normal US: AI Ba – AI irreducible | Appendectomy | Maltoma | 2005 |
| Ram | M | 6yr | 8wk | Periumblical pain/ Constipation/ vomiting/ Hematochezia /RLQ mass | US: No mass colonoscopy: mass | Diathermy (with/relapse) Right hemicolectomy | CF(inverted appendix) | 2005 |
| Luzier | M | 8yr | 1wk | Periumblical pain/ Constipation/ vomiting/ RLQ mass | US: Donut like mass in RLQ | Laparoscopic Appendectomy Distal cecectomy | - | 2006 |
| Baeza- Herrea | F | 2mo | - | Sever RLQ pain | US: RLQ mass | Appendectomy | Appendix torsion | 2006 |
| Lipskar | M | 6yr | 1dy | Cramp/ RLQ pain/ Tenderness/ Nausea/ Vomiting | Us: AI CT: RLQ mass | Appendectomy partial cecectomy (After recurrence) | Lymphoid Hyperplasia | 2008 |
| Dasilva | M | 2yr | 2dy | Colicky/ RLQ tenderness | US: Onion like in RLQ ^e CDUS: Hyperemia in bowel CT: onion pattern in RLQ | Appendectomy cocostomy | Lymphoid Hyperplasia | 2008 |
| Alehossein | M | 7yr | 2wk | Abdominal pain / vomiting /obstipation/ RLQ tenderness | AXR- normal US – Multi layered sign US- Guided saline enema Reduce BE- Non filling appendix | - | Fecalith appendix | 2009 |
| Wang | M | 10yr | 10dy | Periumblical pain/ Periumblical tenderness | AXR: RLQ mass US: Swollenappendeix/AI | Appendectomy | Burkitt's Lymphoma | 2010 |
| Chang | M | 30mo | 1dy | Fever/ vomiting/ RLQ mass/ RLQ tenderness | Air contrast enema Reduce AXR: Appendicolits US: Target sign/ Appendiculitis | Appendectomy (After recurrence of AI) | Appendicitis perforation | 2011 |
| Echert | M | 12yr | - | RLQ Pain/ Hematochezia | Us: Target sign in RLQ | " | Ulcerative colitis | 2012 |
| Epifanio | M | 6yr | 15hr | Abdomil pain/ vomiting/ RLQ tenderness | US: Target sign in RLQ BE: AI reduction Colonoscopy: polypoid lesion | " | Lymphoid Hyperplasia | 2012 |
| Alaee | F | 4yr | 1wk | Abdominal pain/ Nausea agitation/ vomiting/ Diarrhea | AXR: Bowel distention US: Target lesion BE: coil spring with reduction | Reduction | - | 2013 |

a. AI: Appendiceal intussusception; b. RLQ: Right Lower quadrant; c. BE: Barium enema; d. SBT: Small Bowel Transit; e. US: Ultrasound; f. WSCM: water soluble Contrast Media; g. CDUS: Color Doppler Ultrasound

Table 2. Demographics of Intussusception of appendix

| Sex | N percentage |
|--------|--------------|
| Male | 20 (71.4%) |
| Female | 8 (28.2%) |
| Total | 28(100%) |

Table 3:Clinical findings in Intussusception of appendix

| Clinical findings | N percentage |
|-------------------|--------------|
| Abdominal pain | 20 (90%) |
| Vomiting | 13 (46%) |
| Diarrhea | 12 (42%) |
| Constipation | 6 (21%) |
| RLQ tenderness | 5(17%) |
| Hematochezia | 5 (17%) |
| RLQ mass | 2 (17%) |

Table 4:Pathology of the appendiceal intussusception

| Pathology | N(percentage) |
|--------------------|---------------|
| Inflammation | 12 (42%) |
| Cystic fibrosis | 3 (1%) |
| Lymphoma | 2 (0.7%) |
| Fecal material | 2 (0.7%) |
| Polyp | 1(0.3%) |
| Ulcerative colitis | 1 (0.3%) |
| Torsion | 1 (0.3%) |
| Total | 22 (71%) |

The etiology of appendiceal intussusception is mostly idiopathic, but a series of underlying pathophysiologic and anatomic causes are encountered.the 10% cases of appendiceal intussusceptions is a intraluminal mass have lead points such as fecaliths.Anatomical factors are non-fixed mobile appendix, fetal type of cecum with the appendix originating from its tip, narrow, thin mesoappendix, mobile cecum, wide appendicular lumen and funnel – shaped appendix.^{1,2}Idiopathic Appendiceal

intussusception also occure with no underlying abnormality of the appendix.Pathophysiological factors are follicular lymphoid hyperplasia, helminthiasis, fecaliths, foreign bodies, enteric infection such as adenovirus, polyps, mucocele, postappendiceal stump, endometriosis, ulcerative colitis, crohn's D, cystic fibrosis, peutz – jehers syndrome, melanosis coli, and Tumors (adenomas, cystadenomas, carcinoids, papilloma, adenocarcinomas, maltoma).^{5, 6, 7, 8, 9, 10, 28, 29, 30, 31, 32}

The most common causes of appendiceal intussusceptions in children include fecalith,foreignbody,parasite,lymphoid hypertrophy and polype. Appendiceal intussusception classified in four types is based on the level of appendiceal invagination(Mcswain classification). In type 1, appendiceal tip is the intussusceptum, and the more proximal portion is the intussusciens. In type 2, the base of appendix is the intussusceptum, and the cecum is intussusciens. In type 3, the proximal part of the appendix forms the intussusceptum and the distal portion is the intussusciens and in type 4, there is a complete inversion of the appendix into the cecum.^{11,12,33}Most reports are partial invagination of the appendix in to the cecum. In case with ileocolic intussusceptions that do not reduce in enema must be rule out appendiceal intussusception.

Clinical presentation of appendiceal intussusception include four types asymptomatic, intussusception, recurrent intermittent right lower quadrant pain with vomiting and melena, and acute appendicitis.¹⁻⁴ Diagnosis of appendiceal intussusception is often difficult. Most appendiceal intussusception are founded at the time of operation of the patients suspected to have appendicitis .Ultrasound is diagnostic imaging modality of choice.A few cases in asymptomatic patients have been incidentally diagnosed by barium enema, colonoscopy, CTcolography and endoscopic sonography. In

multiple cases appendiceal intussusceptions was mistaken for cecal polyp in colonoscopy. Sonographic examination demonstrated a lead point within a characteristic multiconcentric ring sign, and longitudinal sonograms showed the inverted appendix protruding into the cecal lumen.^{14,18} CT scan is also a modality for diagnosis of appendiceal intussusceptions^{13,21}. The majority of cases reported in literature were demonstrated in colonoscopic and surgical approaches.³¹ Few cases were managed through laparoscopic surgery^{32,34}. Appendectomy is the treatment of choice unless there is a concern for a neoplasm. Appendiceal intussusception may be reducible in cases without lead point. An intussuscepted appendix may be normal and does not need bowel resection but torsion and perforation of the vermiform appendix can be associated with appendiceal intussusception.^{16,24,35,36} Appendectomy is choice treatment of appendiceal intussusception but it may relapse in spite of previous appendectomy. In these cases partial cecectomy also may be curative. In this case appendiceal intussusception was recognized by barium enema that shows coil-spring sign in cecum with filling defects of appendiceal lumen (Figure). Although surgical removal of appendix is treatment of choice, but in this case hydrostatic reduction of appendiceal intussusception occur.¹⁰ Probably filling defects of appendiceal lumen is due to fecaliths. We recommend diagnostic and therapeutic barium enema in childhood longstanding, persistent abdominal colic, to rule out clinical suspicion of appendiceal intussusception.

Conflict of Interest

None declared.

Funding/Support

None declared.

References

- Honmyo u, Shimada S, Yamamoto S et al. Reversible protruded lesion regarded as idiopathic appendiceal intussusception. *Dig. Endosc.* 2001; 13:90-93.
- Gupta P, Chwals W, Guandalini S. Intussusception of the appendix; another poorly recognized cause of rectal bleeding. *J Ped Gastro enterol Nutr.* 2000; 30:320-3.
- Karabulut R, Sonmez K, Turkyimaz Z, Yilmaz Y, Akyurek N, Basaklar A, Kale N. Mucose – associated lymphoid tissue lymphoma in the appendix, a lead point for intussusception. *J of ped surgery.* 2005; 40:872-874.
- Atkinson Go, Gay BB JR, Naffis D. Intussusception of the appendix in children. *Am J Roentgenol.* 1976; 126:1164-8.
- McIntosh JC, Mroczek EC, Balwinc, Mestre J. Intussusception of the appendix in a patient with cystic fibrosis. *Jped Gastroenterol Nutr.* 1990; 11: 542-4.
- Yoshikawa A, Kuramoto S, Mimura T. A Peutz-Jeghers syndrome manifesting complete intussusception of the appendix and associated with a focal cancer of the duodenum and a cystadenocarcinoma of the pancreas: Report of a case. *Dis Colon Rectum* 1995; 41:517-21.
- Porter HJ, Padfield LJ, Peresl c, et al. Adenovirus and intranuclear inclusion in appendices in intussusception. *J Clin Pathol.* 1993; 46: -8.
- Yunise Y, Hashida Y. Electron microscopic demonstration of adenovirus in appendix vermiformis in a case of ileocecal intussusception. *Ped.* 1973; 51:566-70.
- Solomon DJ, Freson M, Price SK. Complete appendicular inversion: the inside-out appendix: An unusual presentation of Crohn's disease: A case report and review of the literature. *J Belg Redio.* 1991; 74:115-6.
- Jevon GP, Daya D, Qizilbash AH. Intussusception of the appendix: A report of 4 cases and review of the literature. *Arch Pathol Lab Med.* 1992; 116:960-4.
- Casteels M, Eggermont E, Kerremans R, Ponnelle E et al. Intussusception of the vermiform appendix: a preoperative diagnosis in an adolescent girl. *J ped Gastroenterol Nutr.* 1986; s:156-62.
- Kleinman PK. Intussusception of the appendix: Hydrostatic reduction. *AJR.* 1980, 134(6): 1268-70.
- Swanger R, Davis S, McBride W, et al. Multimodality imaging of an appendiceal intussusception. *Pediatr Radiol.* 2007 Sep; 37(9):929-32.
- Silva MRC, Pinus J, Ganc AJ, Falleiros G, Garcia RG, Ganc R, et al. Intussuscepção do apêndice cecal: Intussusception of cecal appendix. *Einstein (São Paulo).* 2008; 6(2):206-8.
- Ekert P, Mougnot JF, de Lagausie P, Gerardin M, Le Bourgeois M, Munck A, et al. Iterative intestinal intussusception and appendiceal mucocele in an infant with mucoviscidosis. *Arch Pediatr.* 1998 Apr; 5(4):400-3.

16. Baeza-Herrera C, García-Cabello LM, León-Cruz A, Martínez-Rivera Mde L. Torsion of the vermiform appendix associated with intussusception. *Cir Cir*. 2006 Sep-Oct;74(5):369-71.
17. Wang SM, Huang FC, Wu CH, Ko SF, Lee SY, Hsiao CC. Ileocecal Burkitt's Lymphoma Presenting as Ileocolic Intussusception With Appendiceal Invagination and Acute Appendicitis. *J Formos Med Assoc*. 2010 Jun;109(6):476-9.
18. Koumanidou C, Vakaki M, Theofanopoulou M, Nikas J, Pitsoulakis G, Kakavakis K. Appendiceal and appendiceal-ileocolic intussusception: sonographic and radiographic evaluation. *Pediatr Radiol*. 2001 Mar;31(3):180-3.
19. Attard TM, Askin FB, Cuffari C. *J Pediatr Gastroenterol Nutr*. Appendiceal inversion as a lead point for ileocolic intussusception in a child with cystic fibrosis. 2000 Sep;31(3):300-2.
20. Bailey DJ, Courington KR, Andres JM, Bagwell CE, Hitchcock CL. Cecal polyp and appendiceal intussusception in a child with recurrent abdominal pain: diagnosis by colonoscopy. *J Pediatr Gastroenterol Nutr*. 1987 Sep-Oct;6(5):818-20.
21. Luzier J, Verhey P, Dobos N. Preoperative CT diagnosis of appendiceal intussusception. *Am J Roentgenol* 2006; 87:325–326.
22. Ram AD, Peckham C, Akobeng AK et al (2005) Inverted appendix mistaken for a polyp during colonoscopy and leading to intussusception. *J Cyst Fibros* 4:203–204
23. Galatioto C, Angrisano C, Blois M, Goletti O, Bucciatti P, Lorenzetti L, Massimetti M, Palla G, Seccia M, Cavina E. Laparoscopic treatment of appendiceal intussusceptions. *Surg Laparosc Endosc Percutan Tech*. 1999 Oct;9(5):362-4.
24. Chang TP, Russell SA. Perforated appendicitis and appendicolith in a child presenting as intussusception: a case report. *Pediatr Emerg Care*. 2011 Jul;27(7):635-8.
25. Epifanio M, Lima MA, Spolidoro JV, Eloi J, Baldissera M, Lorenzi D. Appendiceal intussusceptions. *J Pediatr Gastroenterol Nutr*. 2012 Dec 18.
26. Pumberger W, Hörmann M, Pomberger GH, Hallwirth U. Sonographic diagnosis of intussusception of the appendix vermiformis. *J Clin Ultrasound*. 2000 Nov-Dec; 28(9):492-6.
27. Arenal Vera JJ, Blanco Alvarez JI, Del Villar Negro A, Marcos Rodríguez JL, Lorenzo Fernández Y, Tinoco Carrasco C. Appendicular invagination. *Rev Esp Enferm Dig*. 1993 Aug; 84(2):116-8.
28. Eckert K, Radeloff E, Liedgens P. Intussusception of the appendix, A rare cause of acute abdominal pain in childhood. *chirurg*. 2012 feb; 83(2):172_5
29. M. Ashrafi, V. Joshi, M. Zammit, and K. Telford. Intussusception of the appendix secondary to mucinous cystadenoma: A rare cause of abdominal pain. *Int J Surg Case Rep*. 2011; 2(2): 26-27.
30. Chaar CI, Wexelman B, Zuckerman K, Longo W. Intussusception of the appendix comprehensive review of the literature. *Am J Surg*. 2009 Jul;198(1):122-8.
31. Salehzadeh A, Scala A, Simson JN. Appendiceal intussusception mistaken for a polyp at colonoscopy: case report and review of literature. *Ann R Coll Surg Engl*. 2010 Sep; 92(6).
32. Akbayir N, Yildirim S, Sökmen HM, Kiliç G, Erdem L, Alkim C. Intussusception of vermiform appendix with microscopic melanosis coli: a case report. *J Gastroenterol*. 2006 Sep; 17(3):233-5.
33. Alehossein M, Alizadeh H, Nahvi H, Ghasemi Esfe AR, Vaziri Bozorg SM. Preoperative sonographic diagnosis of appendiceal intussusception: a case report. *J Clin Ultrasound*. 2009 Jul-Aug;37(6):363-5.
34. Schmidt J.S., Maier D., Raj P., Remine S.G. Laparoscopic Management of Appendiceal Intussusception Associated with Villous Adenocarcinoma. *Journal of Laparoendoscopic Surgery*. October 1994, 4(5): 369-373.
35. Flint R and Wright T. Intussusception of a normal appendix: how to avoid a right hemicolectomy. *NZ Med Journal*. 2003 Apr 17; 116(1172):u403.
36. Lipskar A, Telem D, Masseaux J, Midulla P, Dolgin S. Failure of appendectomy to resolve appendiceal intussusception. *J Pediatr Surg*. 2008 Aug; 43(8):1554-6.