J Pediatr Rev. 2013;1(1)70-76

JPR
Journal of Pediatrics Review
Mazandaran University of Medical Sciences

Reducible appendiceal intussusception: A case report and review the literatures

Abdulrasool Alaee[1*]

[1]Department 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.[1,2,3]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 Alaee, MD., Assistant professor of radiology*

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

TEL: 0098 9111515116

Email: ar_alaee@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).

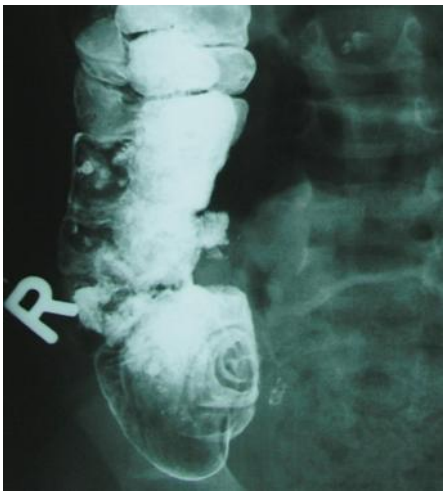

Figure 1. Barium enema reveals characteristic cecal coil-spring sign with invagination of appendiceal base and filling defects in appendiceal lumen.

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 intussusception 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/ Hematochczia	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 temderness	AXR-Bowel obstruction US: Donut Like mass in RLQ Ba – Reduction	Appendectomy	Maltoma	2005
Karabulut	M	8yr	24hr	Abdominal pain /Nausea/ Vomiting/ RLQ temderness /RLQ tenderness RLQ mass	AXR-Normal US: AI Ba – AI irreducable	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 [g]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: Swellenappedeix/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)	Appendictis 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%)
Hematochczia	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 – jeghers 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 intussuscipiens. In type 2, the base of appendix is the intussusceptum, and the cecum is intussuscipiens. In type 3, the proximal part of the appendix forms the intussusceptum and the distal portion is the intussuscipiens 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 laparascopical 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.

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