Original Article

An Audit of Non-Idiopathic Intussusception in Children

Wajeeh Uddin, Nandlal Kella, Javed Ahmed, Talat Mehmood, Noshad Ahmed Shaikh

ABSTRACT

OBJECTIVES: To outline the causes of non-idiopathic intussusception in children and to document the clinical characteristics of the different etiologies.

METHODOLOGY: It is a retrospective review of 19 cases of proved secondary intussusception, over a span of 5 years i.e. from January 2004 to December 2008. Data was collected from private as well as from government hospitals in different centers. Patient's age ranges from three months to 12 years. Demographic data, clinical presentations, surgical management and follow up were recorded on pre designed proforma. Baseline blood investigations and radiological tests were noted. Surgery was performed in all patients due to delayed presentation and non availability of image intensifier.

RESULTS: Among 19 patients, 13 (68.42 %) were boys and 6 (31.57%) were girls with male to female ratio of 2.16:1. The mean age was 2.15 years. Meckel's diverticulum was the most common cause and found in 8 (42.10%) children. Four (21.05%) had lymphoma, 2 (10.52%) had haemangiomas and 2 (10.52%) with polyps of the colon. Round worms, duplication of gut and Henoch-Schönlein's purpura were the other causes of non-idiopathic intussusceptions.

CONCLUSION: Intussusception caused by an underlying disease or secondary to some pathological lead point is not uncommon and it is important to be vigilant for pathological lead points in children of any age.

KEY WORDS: Intussusception, Non-idiopathic, Pathological lead point, Meckel's diverticulum, lymphoma.

INTRODUCTION

Etiology of intussusceptions is uncertain and more than 90% cases are considered idiopathic. Only 6-8% has pathological lead points[1], common lead points are Meckel's diverticulum, polyps, lymphoma, and worms. Classical presentation of intussusceptions is pain, vomiting and palpable mass. Except for the age of presentation, there are no distinguishing parameters regarding clinical presentation of idiopathic and non-idiopathic intussusceptions. Occasionally, systemic manifestations of an underlying disease indicate the specific cause of an intussusception (e.g. perioral pigmentation in Peutz-Jeghers syndrome). The incidence of lead point is higher in adult age as compared to paediatric age group.[2]

After the age of one year, the proportion of intussusceptions due to a pathological lesion at the lead point increases. There is an identifiable lesion in the majority of children over 5 years of age. The objective of this study is to outline the causes of non-idiopathic intussusception in children and to outline some of the clinical characteristics of the different etiologies in our set-up.

PATIENTS AND METHODS

This study is based on a retrospective analysis of 19 cases of intussusceptions, secondary to lead point, out of 150 children which were managed for intussusceptions. It was over a span of five years i.e. from January 2004 to December 2008 with age ranges from 3-months to 12-years. The record were collected from a private as well as government hospitals. Demographic data, clinical presentations, investigations, types of intussusception, lead points, surgical procedures and complications were recorded on a predesigned proforma. Investigations recorded were complete blood count, serum electrolytes, urea, X-ray and ultrasound abdomen. All patients under went laparotomy due to late presentation and non availability of image intensifier.

Surgical options opted were manual reduction followed by excision of lead point and primary closure, resection of gangrenous or irreducible obstructed segment and primary anastomosis or stoma formation. Resected tissues were sent for histopathology.

RESULTS

There were 13 (68.42%) boys and 6 girls (31.57%), with male-female ratio of 2.16:1. The mean age at diagnosis was 2.15 years with a range of 3-months to 12-years. Out of the 19 patients, 10 (52.63%) were below one year and amongst these, 80% were between six months and one year of age. The mean duration of symptoms was 3.5 days, ranging from one day to seven days.

Pain was most common symptoms in 94.7% patients. An abdominal lump was palpable in 9(47.36%) pa-

tients and a mass was felt rectally in 3(15.78%) while other signs and symptoms were shown in **Table I.** Ten (52.63%) patients had classical signs and symptoms of intussusception, while remaining were explored with features of intestinal obstruction and found intussusception incidentally. X-ray abdomen was suggestive of intestinal obstruction in all cases, while ultrasound detected intussusception in 9 (47.3%) patients. Types of intussusception found in this study were Ileocolic in 9 (43.63%), Ileoileal 8 (42.10%) and Colocolic 2 (10.52%) patients. Amongst the 19 patients, manual reduction was possible in 6 (31.57%), including five cases of Meckel's diverticulum and one case of haemangioma, in all other cases resection of lesions and primary closure was made. Resection of segment of intestine and end to end anastomosis was made in 12 (63.17%) patients, of which five cases were for gangrene of intestine and seven cases were secondary to failure of manual reduction due to lead points. Stoma was made in one case due to compromised vascularity. All the resected specimens were sent for histopathology. In this series three patients were re-operated due to post operative adhesions. Lead points/pathological causes leading to non-idiopathic intussusception in this series are depicted in **Table II.**

TABLE I: CLINICAL PRESENTATIONS OF INTUSSUSCEPTION

Signs and Symptoms	Number (%)
Pain	18 (94.7%)
Vomiting	16 (84.2%)
Bleeding per rectum	15 (78.9%)
Distension of abdomen	15 (78.9%)
Mass per abdomen	9 (47.3%)
Fever	8 (42.1%)
Diarrhea	5 (26.31%)
Mass per rectum	3 (15.7%)

DISCUSSION

The most frequent cause of intestinal obstruction in pediatric age group is intussusception. It usually occurs from six months to three years of age. It is most common in infants and 80% of cases are reported in children less than two years. Its occurrence less than three months and above six years of age is rare.[1] Intussusception mostly occurs in otherwise healthy and well-nourished children and has male predominance, with a male-female ratio of approximately 3:2.

Approximately 90 percent cases of intussusception in children are considered to be idiopathic[1] while 6-8%

TABLE II: NON-IDIOPATHIC CAUSES OF INTUSSUSCEPTION

Causes	Number (%)
Meckels diverticulum	8 (42.10%)
Lymphoma	4 (21.05%)
Intestinal Polyps	2 (10.52%)
Haemangioma	2 (10.52%)
Duplication of intestine	1 (5.26%)
Henoch schonlein perpura	1 (5.26%)
Worms infestation	1 (5.26%)

patients have an underlying lesion acting as lead point for the intussusception. Lead points are responsible for a greater proportion of cases of intussusception in children less than three months or more than five years[1, 3].

Conditions that may act as a lead point include; Meckel's diverticulum[4], polyps[5], small bowel lymphoma[6], duplication cysts[7], vascular malformations[8], inverted appendiceal stumps [9], Ascaris lumbricoides [10], Henoch-Schönlein purpura[11], cystic fibrosis[12], and intestinal lymphangioma.

At the time of presentation, the classically described triad; as described by Bode,[13] of pain, a palpable sausage-shaped abdominal mass, and currant-jelly stool was seen in cases of this series, 61% while in our study pain, palpable abdominal mass and bleeding per rectum was found in 94.7%, 47.3% and 78.7% respectively.

Diagnosis of intussusception is based on history and physical examination while X-ray abdomen may be helpful. Ultrasonography is the best tool for diagnosis, while CT scan is mostly done in doubtful cases of intussusception. In our study ultrasound helped in 47.3% cases for diagnosis of intussusception.

Once the diagnosis of intussusception has been made surgical or non surgical methods should be applied. Non surgical reduction is gold standard in idiopathic intussusception while in non-idiopathic cases its results are not promising hence laparotomy is best option[14,15], which we applied in all our cases.

Types of intussusception in our study are ileocolic 43.63%.ileoileal 42.10% and colocolic 10.52% .While Mansoor[16] described types as ileocolic in 33%, ileoileal in 56% and colocolic in 11%, which matches our results.

Meckel's diverticulum occurs in 2% of the population and may present at any age[17] .In a large series of Meckel's diverticulum it was found that 71.3% was symptomatic. Common presentation was intestinal

obstruction, bleeding per rectum, diverticulitis and umbilical pathology. Among intestinal obstruction intussusception and volvulous were major pathologies[4]. Common ectopic tissues found were gastric (88%), pancreatic (7%), and gastric with pancreatic (3%). In our study population common cause of non-idiopathic intussusceptions was Meckel's diverticulum (42.10%) which is almost similar to other study[16] .

Lymphomas are rarely revealed by acute intestinal intussusception in children .In our study population we have 21% cases of lymphoma causing intussusception. Gupta et al[6] described in his study that 17.5% patients of primary abdominal Burkit's lymphoma presented with non-idiopathic intussusception. Intussusception as a presenting feature of Burkitt's lymphoma may be associated with early stage disease, which is curable with less intensive therapy. Although the role of surgery in intra abdominal Burkitt's lymphoma remains controversial and different opinions are present in the literature, but in acute abdominal diseases like intestinal obstruction, intussusception, intestinal perforation, and acute appendicitis surgery is the only option.

Inflammatory fibroid polyp (IFP) is an entity of old age, commonly found in stomach and distal ileum and rarely encountered in pediatric age group[18]. Its presence in colon is a rare finding. In our study we have two cases of solitary inflammatory polyps in colon causing colocolic intussusceptions. Hence the inflammatory solitary polyps, which are benign in nature, should be considered in differentials of lead points in pediatric intussusceptions as well.

Gastrointestinal haemangiomas make up 0.05% of all intestinal neoplasms. They may be solitary or multiple Either localized or diffuse. They may present as pain, bleeding, volvuluos, perforation and rarely intussusception[19] as in our two cases.

Duplication cyst of the alimentary tract is well known as one of the rare causes of non-idiopathic intussusception[20]. We have in present study, one 9 months old male with ileocolic intussusception caused by the duplication cyst in the terminal ileum.

Henoch-Schönlein purpura (HSP) is an IgA mediated auto immune hypersensitivity vasculitis in children. Its incidence is 20 cases per 100000 populations. Surgical complication reported are intestinal perforation, gastro intestinal bleeding, necrosis and intussusception[21] being the most common problem in HSP which occurs in 2-3% patients of HSP. A small bowel wall hematoma acts as the lead point[22]. Ultrasound is a best tool for diagnosis, Emergency laparotomy is generally considered the appropriate course of action for small bowel intussusception associated with Henoch-Schönlein's purpura (HSP) [22]. In our study a 5 year old girl presented with pain in abdomen and bleeding per rectum. Clinical examination favored the intussusception and diagnosis was confirmed on ultrasonography. Henoch-Schönlein's purpura was diagnosed by the appearance of a purpuric rash without thrombocytopenia. Emergency laparotomy was performed and an ileoileal intussusception was found with a suspected less vascular part of gut, therefore resection and anastomosis was made.

CONCLUSION

Although the majority has idiopathic aetiology, intussusception caused by an underlying disease or secondary to some pathological lead point is not uncommon. Surgical treatment is indicated as a primary intervention for patients with suspected intussusception who are acutely ill or have evidence of perforation or where the radiographic facilities and expertise to perform nonoperative reduction are not readily available or unsuccessful, and for evaluation or resection of a pathological lead point. Nonetheless, it is important to be vigilant for pathological lead points in children of any age.

REFERENCES

1. Blakelock RT, Beasley SW .The clinical implications of non-idiopathic intussusception. Pediatr Surg Int. 1998;14(3):63-7
2. Ein SH, Daneman A. Intussusception. In: O'Neill JA Jr, Rowe MI, Grosfeld JL, (eds). Pediatric Surgery. 6th edition. Chicago: Year Book; 2006. p.1313-7.
3. Navarro O, Daneman A. Intussusception. Part 3:Diagnosis and management of those with an identifiable or predisposing cause and those that reduce spontaneously. Pediatr Radiol 2004;34 (4):305-12
4. St-Vil, D, Brandt ML, Panic S. Bensaussant AL, Blanchard H. Meckel's diverticulum in children:a 20-year review. J Pediatr Surg 1991;26(11):1289-92
5. Hwang CS, Chu CC, Chen KC, Chen A. Duodenojejunal intussusception secondary to hamartomatous polyps of duodenum surrounding the ampulla of Vater. J Pediatr Surg 2001;36(7): 1073-5
6. Gupta H, Davidoff AM, Pui CH, Sochat SJ, Sandlund JL. Clinical implications and surgical management of intussusception in pediatric patients with Burkitt lymphoma. J Pediatr Surg 2007;42 (6):998-1001
7. Chen Y, Ni YH, Chen CC. Neonatal intussusception due to a caecal duplication cyst. J Formos Med Assoc 2000;99(4):352-3
8. Morgan DR, Mylankal K, Barghouti N, Dixon MF. Small bowel haemangioma with local lymph node

involvement presenting as intussusception. J Clin Pathol 2000;53(7):552-3
9. Hamada Y, Fukunaga S, Takada K, Sato M, Hioki K. Postoperative intussusception after incidental appendicectomy. Pediatr Surg Int 2002;18(4):284-6
10. Yetim I, Ozkan OV, Semerci E, Abanoz R. Rare cause of intestinal obstruction;Ascaris lumbricoides infestation two case reports. Cases J 2009;17:2:7970.
11. Choong CK, Beasley SW. Intra-abdominal manifestations of Henoch-Schonlein purpura. J Paediatr Child Health 1998;34(5):405-9
12. Holmes M, Murphy V, Taylor M, Denham B. Intussusception in cystic fibrosis. Arch Dis Child 1991;66(6):726-7
13. Bode CO. Presentation and management outcome of childhood intussusception in Lagos. A prospective study. Afr J Paeds Surg 2008;5(1):24-8
14. Ikeda T, Koshinaga T, Inoue M, Goto H, Sugitou K, Hagiwara N. Intussusception in children of school age. Pediatr Int 2007 :94(1):58-63
15. Kella N, Rathi PK, Soomro B, Qureshi MA. Childhood intussusception:Delayed presentation and surgical management. JLUMHS 2009;8(1):46-9
16. Mansoor H, Yousuf UF. Intussusception: What leads in children beyond infancy. Ann King Edward Med Coll 1998;4(4):31-2
17. Amin MU, Siddique MK, Mahmood R. Inverted Meckles diverticulum causing intussusception in adults. J Coll Physcian Surg Pak 2008;18(9):574-5
18. Dabral C, Singh N, Singh PA, Misra V. Inflammatory fibroid polyp in a child. Indian Gastroenterol 2003;22(3):101-3
19. Kella N, Rathi PK, Rathi S, Qureshi MA. Small bowel haemangioma:A rare cause of intestinal obstuction in infant. Pak J Med Sci 2008;24 (4):629-31
20. Milbandt K, Sigalet D. Intussusception associated with Meckles diverticulum and duplication cyst. J Pediatr Surg 2008;43(12):21-3
21. Cull DL, Rosario V, Lally KP, Partner I, Mahour GH.Surgical implications of Henoch-Schönlein's purpura. J Pediatr Surg 1990;25(7):741-3
22. Sonmez K, Turkyilmaz Z, Demirogullari B, Karabulut R, Aral YZ, Sönmez K, et al. Conservative treatment for small intestinal intussusception associated with Henoch-Schönlein's purpura. Surg

AUTHOR AFFILIATION:

Dr. Wajeeh Uddin
Consultant Paediatric Surgeon
Sindh Govt. Children Hospital
Karachi, Sindh-Pakistan.

Dr. Nandlal Kella *(Corresponding Author)*
Assistant Professor, Department of Paediatric Surgery
Liaqat University of Medical and Health Sciences
Jamshoro, Sindh-Pakistan.
Email: nckella@yahoo.com

Dr. Javed Ahmed
Assistant Professor, Surgical Unit – VIII
Daw University of Health Sciences
Karachi, Sindh-Karachi.

Dr. Talat Mehmood
Professor, Paediatric Surgery
Dow University of Health Science
Karachi.

Dr. Noshad Ahmed Shaikh
Professor of Surgery and Vice Chancellor
LUMHS, Jamshoro, Sindh-Pakistan.