Adenomyoma of the small intestine: a rare pathological lead point for intussusception in an infant

You-Jung Bak, Udo Rolle, Stefan Gfroerer and Henning C Fiegel*

Abstract

Introduction: Intussusception is a typical abdominal emergency in early childhood.

Case description: We report a case of an infant in the typically affected age group with intussusception triggered by a rare benign intramural intestinal adenomyoma as a pathological lead point. The infant had the typical symptoms of a recurrent idiopathic ileocolic intussusception.

Discussion and evaluation: Idiopathic intussusception is frequent in the infant age group. Contrary to that, reports on pathological lead points for intussusceptions are sparse in the toddler age.

Conclusions: That case illustrates that even in intussusceptions in the typically affected age group, it is important to be aware of pathological lead points, especially if the intussusceptions are recurrent.

Keywords: Intussusception; Intestinal adenomyoma; Pathological lead point

Background

Intussusception occurs when a proximal part of the bowel invaginates into a more distal part, typically within the ileocecal region, which occurs commonly in infants and children between 3 months and 4 years of age. Typical symptoms in these patients include a triad of acute abdominal pain, vomiting and bloody stools; however, regularly, patients present with variable, non-specific symptoms. Ultrasonography is the established standard for diagnosis of intussusception and has a high sensitivity and specificity (Lehnert et al. 2009). Idiopathic intussusception occurs due to swollen mesenteric lymph nodes in patients in the typically affected age group that have been affected by viral infection or non-specific immunologic factors. If recurrent intussusception or intussusception occur in older children, the presence of a pathological lead point must be considered. Herein, we report and discuss the case of an infant in the typically affected age group with an ileocolic intussusception triggered by an adenomyoma of the distal ileum wall, a rare benign intramural intestinal tumor, acting as pathological lead point.

Case description

A previously healthy 11-month-old girl was admitted to our department with a 2-day history of colicky abdominal pain, vomiting and bloody stools; however, regularly, patients present with variable, non-specific symptoms. Ultrasonography is the established standard for diagnosis of intussusception and has a high sensitivity and specificity (Lehnert et al. 2009). Idiopathic intussusception occurs due to swollen mesenteric lymph nodes in patients in the typically affected age group that have been affected by viral infection or non-specific immunologic factors. If recurrent intussusception or intussusception occur in older children, the presence of a pathological lead point must be considered. Herein, we report and discuss the case of an infant in the typically affected age group with an ileocolic intussusception triggered by an adenomyoma of the distal ileum wall, a rare benign intramural intestinal tumor, acting as pathological lead point.

© 2014 Bak et al; licensee Springer. This is an Open Access article distributed under the terms of the Creative Commons Attribution License (http://creativecommons.org/licenses/by/4.0), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly credited.
emergency surgery was indicated. During laparotomy, an ileoileocolic intussusception was identified and reduced (Figure 3). After reduction, a palpable intraluminal mass presented as possible lead point of the intussusception approximately 10 cm from the ileocecal valve (Figure 4). Segmental resection of the ileum and reanastomosis were performed. The further recovery period was uneventful, and the infant was discharged 6 days after the operation.

Pathological findings were as follows. The mass was a 1×1×1 cm polypoid lesion covered with hemorrhagic and partly necrotic mucosa. Microscopically, the tumor was located in the submucosa and composed of glandular structures lined by mucin-secreting columnar epithelium and smooth muscle bundles (Figure 5). These findings were compatible with the diagnosis of adenomyoma of the ileum. Elsewhere, the ileum showed severe mucosal ulceration and necrosis in addition to subtotal perforating enteritis with hemorrhagic infarction, all of which were consistent with changes resulting from the intussusception.

Discussion and evaluation
Intussusception is a common cause of bowel obstruction in infants and toddlers, with the greatest incidence in infants aged 3–9 months (Lehnert et al. 2009, Gfroer et al. 2009). There is a seasonal incidence, with peaks in spring and autumn resembling the most typical periods of seasonal gastroenteritis and respiratory tract infections. Most infants do not have a specific lead point. Hypertrophied Peyer’s patches and reactive lymph node hyperplasia, which result from prior viral infection, can serve as a lead point for idiopathic intussusception. Specific lead points (e.g., Meckel diverticulum, intestinal polyps, lymphomas, and intestinal duplication) are more
commonly found in older children and adults. Ultrasonography is the preferred diagnostic tool in intussusception and has a sensitivity of 98-100% and a specificity of 88-100% (Lehnert et al. 2009, Gfrorer et al. 2009). Hydrostatic reduction under ultrasound control and contrast enema are established therapies for the treatment of intussusception, with a success rate of 70-90% (Lehnert et al. 2009, Gfrorer et al. 2009). Immediate surgery is indicated in patients who have peritonitis, sepsis, evidence of perforation, unsuccessful non-operative repositioning or a clear finding of pathological lead points. In cases occurring in individuals not in the typical age group or in cases of recurrent intussusceptions, a pathological lead point must be excluded.

Adenomyoma of the gastrointestinal tract is a rare benign lesion localized at the stomach, small intestine and biliary ducts (Zhu et al. 2010). Adenomyoma of the stomach is usually asymptomatic. Its occurrence in the small intestine of children is extremely rare. However, in the small intestine, intussusception is its most common complication, which has been reported in 13 cases so far (Table 1). The reported cases had significantly varied ages, with a range from 2 days to 82 years. In our case, the infant was of the typical age and had the symptoms most commonly associated with idiopathic ileocolic intussusception, but the intussusception was nonetheless due to a pathological finding.

**Conclusions**

Adenomyoma of the small bowel is a rare cause of intussusception in all age groups. The here presented case shows, that even in patients where intussusceptions occur in the typically affected age group, it is important to be aware of pathological lead points, especially in recurrent intussusceptions.

**Table 1** Previous reported cases of adenomyoma in intussusception

| No. | First author     | Year | Age  | Surgical diagnosis | Histopathology                     |
|-----|------------------|------|------|--------------------|------------------------------------|
| 1   | Schwartz et al.  | 1958 | 8 months | intussusception   | myoepithelial hamartoms            |
| 2   | Gal et al.       | 1986 | 82 years | intussusception   | adenomatous hamartoma              |
| 3   | Kim et al.       | 1990 | 7 years | intussusception    | adenomyoma                         |
| 4   | Gal et al.       | 1991 | 9 months | intussusception    | adenomyoma                         |
| 5   | Lamki et al.     | 1993 | 1 year | intussusception    | adenomyoma                         |
| 6   | Serour et al.    | 1994 | 3 years | intussusception    | adenomyoma                         |
| 7   | Chan et al.      | 1994 | 5 months | intussusception    | adenomyoma                         |
| 8   | Gonzalvez et al. | 1995 | 2 years | intussusception    | adenomyoma                         |
| 9   | Yamagami et al.  | 1997 | 4 months | intussusception    | adenomyoma                         |
| 10  | Lee et al.       | 2001 | 18 years | intussusception    | adenomyoma                         |
| 11  | Park et al.      | 2003 | 7 months | intussusception    | adenomyoma                         |
| 12  | Mouravas et al.  | 2003 | 18 months | intussusception   | adenomyoma                         |
| 13  | Takeda et al.    | 2011 | 68 years | intussusception    | adenomyoma                         |
|     | here described case | 2013 | 11 months | intussusception    | adenomyoma                         |
Consent
Written informed consent was obtained from the parents for the publication of this report and any accompanying images.

Competing interests
The authors declare that they have no competing interests.

Authors’ contributions
YJB collected the data and drafted the manuscript. HCF performed the literature study and assisted in drafting the manuscript. SG and UR were involved in the case and in the critical revision of the drafted manuscript. All authors read and approved the final manuscript.

Acknowledgments
We wish to acknowledge ML Hansmann (Institute of Pathology, Goethe-University Frankfurt/M.) for providing the histological images.

Received: 24 July 2014 Accepted: 8 October 2014
Published: 18 October 2014

References
Chan YF, Roche D (1994) Adenomyoma of the small intestine in children. J Pediatr Surg 29(12):1611–1612
Gal R, Kolkow Z, Nobel M (1986) Adenomyomatosis hamartoma of the small intestine: a rare cause of intussusception in an adult. Am J Gastroenterol 12:1209–1211
Gal R, Rath-Wolfson GM, Kessler E (1991) Adenomyoma of the small intestine. Histopathology 18:369–371
Giofer F, Fiegel H, Rolle U (2009) Invagination. Monatschr Kinderheilkd 157:917–924
Gonzalez J, Marco A, Andujar M, Iniguez L (1995) Myoepithelial hamartoma of the ileum: a rare cause of intestinal intussusception in children. Eur J Ped Surg 5:303–304
Kim CJ, Choe GY, Chi JG (1990) Foregut choristoma of the ileum (adenomyoma) – a case report. Ped Pathol 10:799–805
Lamki N, Woo CL, Watson AB Jr, Kim HS (1993) Adenomyomatosis hamartoma causing ileoileal intussusception in a young child. Clin Imaging 17:183–185
Lee JS, Kim HS, Jung JJ, Kim YB (2001) Adenomyoma of the small intestines in an adult: a rare cause of intussusception. J Gastroenterol 37(7):556–559
Lehnert T, Sorge I, Till H, Rolle U (2009) Intussusception in children – clinical presentation, diagnosis and management. Int J Colorectal Dis 24:1187–1192
Mouravas V, Koutoumiou G, Patoulas J, Kostopoulos I, Kottakidou R, Kallergis K, Kepertis C, Liolios N (2003) Adenomyoma of the small intestine in children: a rare cause of intussusception: a case report. Turk J Pediatr 45:345–347
Park HS, Lee SG, Lee JM, Kang MJ, Lee DG, Chung MJ (2003) Adenomyoma of small intestine: report of two cases and review of the literature. Pathol Int 53:111–114
Schwartz SI, Radwini HM (1958) Myoepithelial hamartoma of the ileum causing intussusception. AMA Arch Surg 77:102–104
Sorour F, Gorenstein A, Lipnitzky V, Zaidel L (1994) Adenomyoma of the small bowel: a rare cause of intussusception in childhood. J Pediatr Gastroenterol Nutr 18(2):247–249
Takeda M, Shoji T, Yamazaki M, Higashi Y, Maruo H (2011) Adenomyoma of the jejunum leading to intussusception. Case Rep Gastroenterol 5(3):602–609
Yamagami T, Tokiwa K, Iwai N (1997) Myoepithelial hamartoma of the ileum causing intussusception in an infant. Pediatr Surg Int 12:206–207
Zhu HN, Yu JP, Luo J, Jiang YH, Li QJ, Sun WY (2010) Gastric adenomyoma presenting as melena; a case report and literature review. World J Gastroenterol 16(15):1934–1936

doi:10.1186/2193-1801-3-616
Cite this article as: Bak et al.: Adenomyoma of the small intestine a rare pathological lead point for intussusception in an infant. SpringerPlus 2014 3:616.

Submit your manuscript to a SpringerOpen journal and benefit from:
- Convenient online submission
- Rigorous peer review
- Immediate publication on acceptance
- Open access: articles freely available online
- High visibility within the field
- Retaining the copyright to your article

Submit your next manuscript at ➔ springeropen.com