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Case report

Forshal type IE appendiceal intussusception: A case report

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ABSTRACT

Introduction: Appendiceal intussusception is a rare condition. Clinical features are not specific for it. Patients may present with abdominal pain and vomiting. These symptoms represent a variety of abdominal pathology. Preoperative diagnosis is difficult because of the non-specific clinical features. We present a case report of a child who initially presented with ileocolic intussusception.

Case presentation: This is a case report of a 5-years-old boy with abdominal pain and vomiting. He had an ileocolic intussusception 2 days back, and was successfully managed by hydrostatic reduction and discharged. On ultrasonography, an intussusception was identified in the ileocaecal region. Hydrostatic reduction failed this time and laparotomy was performed. On laparotomy, there was complete intussusception of the appendix with normal ileocaecal junction. Appendectomy was performed. Post-operative period was uneventful.

Discussion: Appendiceal intussusceptions are mostly diagnosed intra-operatively. The clinical features may mimic various other acute and chronic abdominal conditions. Type IE appendiceal intussusception, as described by Forshal, is a rare condition. Appendectomy with a rim of the caecum is the procedure of choice.

Conclusion: Though ileocaecal intussusceptions are common in children, appendiceal intussusceptions are rare and are usually diagnosed during the operative procedure. Radiologists and pediatric surgeons should be aware of this rare entity. Appendectomy is the treatment of choice in most of the appendiceal intussusceptions.

1. Introduction

Appendiceal intussusception is a rare condition and its incidence is approximately 0.01% [1]. This is mostly idiopathic; however, there are certain anatomical or pathological predisposing factors responsible for appendiceal intussusceptions [2]. Anatomical factors include freely mobile appendix, narrow thin mesoappendix, poorly fixed high caecum, and hyperperistalsis. Pathological conditions include appendicular inflammation, calcified faecolith, various benign and malignant conditions, and foreign bodies [2]. Ileocolic intussusception in children is a common condition where the appendix enters into the caecum. This has a definite symptoms set of intermittent abdominal pain, vomiting, diarrhea, fever, and rectal bleeding. True appendiceal intussusception with eversion of the appendix is extremely rare. Appendiceal intussusception has no definitive symptoms that differentiate it from the common ileocolic intussusception. Clinical manifestations range from asymptomatic patients to patients with an acute abdomen that mimics acute appendicitis [3]. Preoperative diagnosis of appendiceal intussusception is difficult because of non-specific symptoms and the rarity of the entity. Diagnosis is usually made intra-operatively [1]. Recently, preoperative diagnoses have been reported by imaging and colonoscopy [3].

We report a case of a 5-year-old boy who presented to the emergency department of our institute with intermittent abdominal pain and was diagnosed as an appendiceal intussusception intraoperatively. The case has been reported in line with SCARE 2020 criteria [4].

2. Case presentation

Our case report presents a case of a 5-year-old male patient from the Terai region of central Nepal. He presented to the emergency department of our institution, with intermittent abdominal pain for 6 h. It was associated with two episodes of vomiting. There was no fever, constipation, diarrhea, bleeding per rectum, and urinary complaints. He had

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an ileocolic intussusception 2 days back and was discharged after successful hydrostatic reduction, in the same hospital. He had red currant jelly like stool in the previous episode.

On physical examination, he was ill-looking and irritable, and the signs like pallor, icterus, cyanosis, dehydration, rashes, petechiae, and purpura were absent. On examination of the abdomen, on palpation, there was mild tenderness at the center and right lower abdomen. Abdominal ultrasonography revealed a concentric doughnut-shaped hypo and hyperechoic ring of size 2.5·2.5 cm in the sub-hepatic region, suggestive of an intussusception.

After resuscitation, he was subjected to ultrasound-guided saline hydrostatic reduction. Hydrostatic reduction was performed under the standard protocol with 3 ft height of saline, for 3 attempts, each of 3 min. The procedure failed to reduce the intussusception and he was planned for laparotomy.

The surgical procedure was performed by a team of pediatric surgery unit of our center. A right subcostal transverse incision was given and the abdomen was opened. The intraoperative finding was an intussusception involving the entire length of the appendix. The appendix was found everted and invaginating into the caecum as shown in Fig. 1.

Reduction was tried manually by retrograde milking of the intussusceptum but it could not be achieved. Hence approximately 1 cm of longitudinal incision was given in the caecum along the tenia coli. The appendix was found swollen, everted, in a sleeve pattern, with mucosal aspect evident as shown in Fig. 2. It was difficult to reduce the intussusception, so appendectomy was performed from inside the caecum. Caecotomy incision and the defect after appendectomy were repaired in two layers and the abdomen was closed.

The resected appendix specimen was sent for histopathologic examination. On gross examination, a dark brown mucosal surface was observed. On microscopic examination, it revealed mucosal ulceration, submucosal edema, and congested blood vessels, indicating inflammation and ischemic changes (Fig. 3).

He was kept nil by mouth for 24 h and started feeding gradually. Post-operative period was uneventful. He was discharged on the 4th post-operative day. He had no issues after 2 weeks of follow-up. The patient's party is satisfied with the treatment they received.

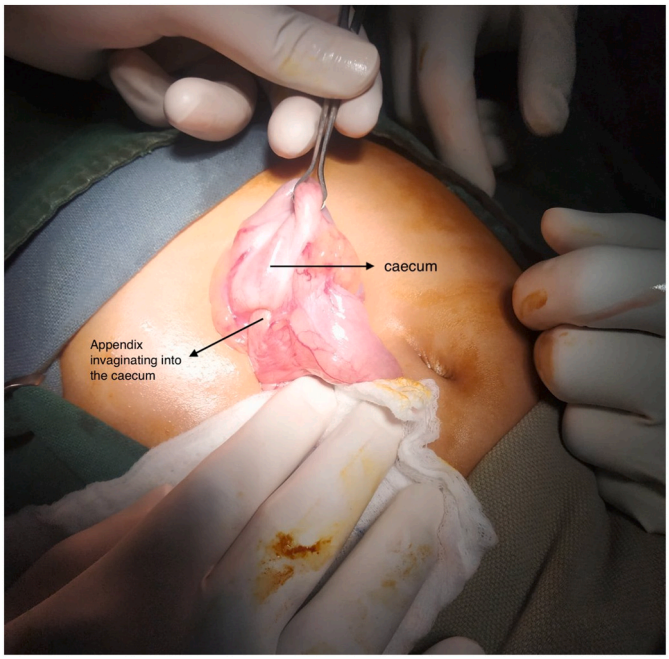

Fig. 1. Invagination of the appendix into the caecum.

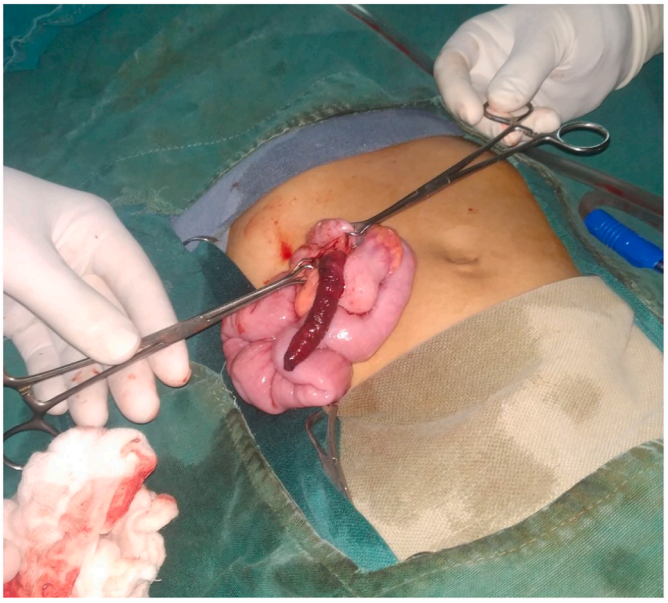

Fig. 2. Inner view of the intussusceptum (appendix) after the caecotomy.

3. Discussion

Ileocolic intussusception in children is a common condition whereas appendiceal intussusception is rare. Most of them are idiopathic in origin and less than 10% of the cases have an identifiable pathological lead point [5]. Invasion of the appendix into the caecum can lead to further invagination of ileum into the caecum. True appendiceal intussusception, where invagination of appendix into the appendix is rare. In a landmark study by Collins, 71,000 appendiceal specimens were examined over 40 years and found an incidence of appendiceal intussusception as just 0.01% [1].

Preoperative diagnosis is challenging and is rarely achieved. The condition itself is rare and clinical features are not specific for the condition [6]. Clinical presentation of appendiceal intussusception may be grouped in 4 different ways. The first group of patients may have features of acute appendicitis. The second variety of patients has features similar to the common ileocolic intussusception. These include intermittent pain, vomiting, diarrhea, and rectal bleeding. Our patient had abdominal pain, vomiting, and a previous episode of rectal bleeding and may be considered as this group. The third group has recurrent features of abdominal pain, vomiting, and bleeding per rectum. These features are explained by the recurrent pattern of intussusception and self-reduction. The fourth group of patients are asymptomatic and discovered incidentally [3].

Our patient had a typical feature of childhood intussusception and was managed by ultrasound-guided hydrostatic reduction successfully. He had a recurrence after 2 days. We performed laparotomy after the failure of hydrostatic reduction in the second presentation. Appendiceal intussusception was diagnosed only after laparotomy.

Appendiceal intussusception was found more in adults than in children. Jevon noted partial or complete intussusception of appendix in adult females. All of them had either villous adenoma or endometriosis [7]. Chaar also found it more common in adults. All had some appendicular pathology [8].

Preoperative diagnosis of appendiceal intussusception can be made by contrast-enhanced CT scan of the abdomen [9]. Experienced ultrasonologist may identify intussusception confined to the caecum [10]. There are few reports of diagnosis of recurrent or partial appendiceal intussusception by colonoscopy [3]. On the other hand, it may be detected on diagnostic colonoscopy for recurrent pain abdomen or bleeding rectum [11].

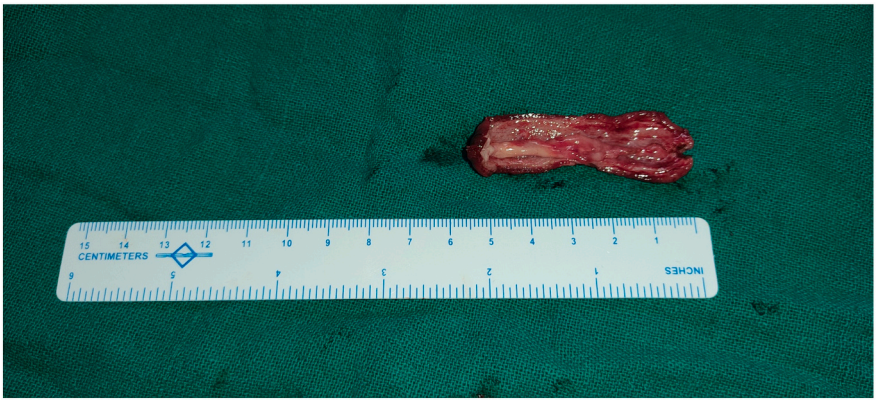

Fig. 3. Post-appendectomy specimen after cutting longitudinally, showing 5.7 cm long appendix.

In view of the high volume of ileocolic intussusception in our center and a clear ultrasonic finding of intussusception in subhepatic region, we did not perform an additional investigation to confirm the diagnosis. We performed hydrostatic reduction twice alleviating the symptoms, the maneuver proved to be temporary and ultimately unsuccessful. This phenomenon was consistent with other reports [12].

Intussusception of the appendix was first classified into 6 types by Moschcowitz (1910) [13], which was later expanded by McSwain (1941) [13,14]. Later, Forshal (1953) provided a comprehensive classification system [13].

This is classified as type 1 for primary intussusception of the appendix only. Type 2 describes compound intussusception of the appendix with caecocolic intussusception. Type 3 describes compound intussusception with ileocolic variety. Type 4 is the intussusception of the appendix without turning inside out. Type 5 is the involvement of the appendix in any intussusception whereas type 6 is the intussusception of stump appendix. Type 1 is further classified as below -

IA: Invagination of appendix tip into proximal the appendix
IB: Intussusception of the distal appendix into the proximal appendix
IC: Intussusception starting along the length of the appendix
ID: Retrograde intussusception of the appendix
IE: Complete invagination of the appendix

In our patient, the appendix was completely everted and invaginated into the caecum consistent with type IE as described by Forshal. Complete inversion of the appendix is again a rare type of appendiceal intussusception.

Type IE is the result of the progression of types IA, IB, and IC [13]. It can involve the entire colon and may protrude from the anus [15]. Although primary appendiceal intussusception itself acts as a lead point in secondary cecocolic or ileocolic intussusception. That was not the case in our patient, possibly due to early intervention.

When the appendix is totally invaginated, manual reduction is rarely possible [12] as evidenced in this case. Although there are no guidelines in the management due to its rarity, appendectomy remains the procedure of choice for all types of appendiceal intussusception. We performed appendectomy through caecotomy and from inside the caecum. Removal of a rim of caecum around the appendix is advised to prevent recurrent stump intussusception by some authors [12,16]. Although the appendix was found to be inflamed in our case, it is yet unclear whether the inflamed appendix acts as a lead point or the appendiceal intussusception leads to acute appendicitis [17].

4. Conclusion

Appendiceal intussusception is a rare condition in children. Preoperative diagnosis is difficult and it can be easily missed and/or misdiagnosed as other entities. Forshal type IE, the complete invagination of the appendix inside the caecum, is an even rarer form that can lead to secondary intussusceptions. Appendectomy remains the procedure of choice.

Ethical approval

Case reports are exempt from ethical approval in our institution, Tribhuvan University Institute of Medicine, Maharajgunj.

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Author contribution

All the authors contributed equally for the preparation of this case report.

Naveen C. Bhatta (NB), Dinesh Prasad Koirala (DK), Geha Raj Dahal (GD) = study concept and surgical therapy for the patient.

Kshitiz Acharya (KA), Aramva Bikram Adhikari (AA), Karishma Kathayat(KK) = Data collection, obtaining consent from patient's party, review of previous literatures and preparation of the manuscript.

NB and Karishma Kathayat(KK) = Editing and writing of the manuscript.

DK and GD = Senior author and manuscript reviewer.

All the authors individually did the final proof-reading of the manuscript before submission.

Guarantor

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Research registration

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Consent

Written informed consent was obtained from the patient's father for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this

journal on request.

Declaration of competing interest

There are no conflicts of interest.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.ijscr.2021.106151.

References

[1] D.C. Collins, 71,000 human appendix specimens. A final report, summarizing forty years' study, Dec, Am. J. Proctol. 14 (1963) 265–281.

[2] S.B. Joshi, E. H, V. Kinhal, S.K. Kola, S. V. K, Intussusception in children with a pathological appendix acting as a "lead point" - a series of 3 cases, J. Clin. Diagn, Res 9 (7) (2015), PD03–PD04, https://doi.org/10.7860/JCDR/2015/9325.6214. Jul.

[3] Aug H. Tavakkoli, S.M. Sadrkabir, P. Mahzouni, Colonoscopic diagnosis of appendiceal intussusception in a patient with intermittent abdominal pain: a case report, World. J. Gastroenterol. 13 (31) (2007) 4274–4277, https://doi.org/10.3748/wjg.v13.i31.4274.

[4] for the SCARE Group, R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, The SCARE 2020 guideline: updating consensus Surgical CAse REport (SCARE) guidelines, Int. J. Surg. 84 (2020) 226–230, https://doi.org/10.1016/j.ijsu.2020.10.034.

[5] N.T. Ong, S.W. Beasley, The leadpoint in intussusception, J. Pediatr. Surg. 25 (6) (1990) 640–643, https://doi.org/10.1016/0022-3468(90) 90353-b. Jun.

[6] E. Dainius, L. Pankratjevaite, S. Bradulskis, D. Venskutonis, Intussusception of the appendix and ileum endometriosis: a case report, Visc Med. 32 (2016 Jun) 211–213, https://doi.org/10.1159/000445394. Epub 2016 Jun 6. PMID: 27493950; PMCID: PMC4945780.

[7] G.P. Jevon, D. Daya, A.H. Qizilbash, Intussusception of the appendix. A report of four cases and review of the literature, Sep, Arch. Pathol. Lab. Med. 116 (9) (1992) 960–964. PMID: 1524463.

[8] C.I. Chaar, B. Wexelman, K. Zuckerman, W. Longo, Intussusception of the appendix: comprehensive review of the literature, Am. J. Surg. 198 (1) (2009) 122–128, https://doi.org/10.1016/j.amjsurg.2008.08.023. Jul.

[9] J. Miller, S. Shukla, B. Baigorri, H. Tejero, CT diagnosis of appendiceal intussusception in a middle-aged female, J. Radiol. Case. Rep. (6) (2020) 8–14, https://doi.org/10.3941/jrcr.v14i6.3809. Jun.

[10] X.X. Duan, Y. Peng, L. Yang, W.J. Chen, X.L. Tang, Preoperative sonographic diagnosis of McSwain type V appendiceal intussusception: a case report, Medicine 99 (49) (2020), e23452, https://doi.org/10.1097/MD.0000000000023452.

[11] A. Salehzadeh, A. Scala, J.N. Simson, Appendiceal intussusception mistaken for a polyp at colonoscopy: case report and review of literature, Ann. R. Coll. Surg. Engl. 92 (6) (2010), W46-8, https://doi.org/10.1308/147870810X12699662981591. Sep.

[12] I. Samuk, A. Nica, Y. Lakovski, E. Freud, Appendiceal intussusception: a diagnostic challenge, Available from: Eur. J. Pediatr. Surg. 28 (1) (2018 Feb 1) 30–33 http://www.thieme-connect.de/DOI/DOI?10.1055/s-0037-1604400.

[13] I. Forshal, Intussusception of the vermiform appendix with a report of seven cases in children, Br. J. Surg. 40 (162) (1953) 305–312, https://doi.org/10.1002/bjs.18004016203. Jan.

[14] PMID: 30112244; PMCID: PMC6077606 J. Haug, A.S. Katkar, J. Covelli, Appendiceal intussusception mimicking cecal mass and fecal matter: a report of two rare cases, Case Rep. Radiol. 16 (2018) (2018 Jul) 4809650, https://doi.org/10.1155/2018/4809650.

[15] D. Dunavant, H. Wilson, Intussusception of the appendix, with complete inversion of the appendix and protrusion from the anus, Ann. Surg 135 (2) (1952) 287–288, https://doi.org/10.1097/00000658-195202000-00017.

[16] Aug A. Lipskar, D. Telem, J. Masseaux, P. Midulla, S. Dolgin, Failure of appendectomy to resolve appendiceal intussusception, J. Pediatr. Surg. 43 (8) (2008) 1554–1556, https://doi.org/10.1016/j.jpedsurg.2008.03.038.

[17] D.H. Phan, M.K. Hong, M.J. Morgan, Appendiceal intussusception causing appendicitis ANZ, J. Surg 88 (2018) E851–E852, https://doi.org/10.1111/ans.13931.