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CASE REPORTS

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Intestinal tumor lesions leading to intussusception in children: a report of four cases and literature review

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Abstract

Background: Intussusception is commonly idiopathic in children. Pathologic lead points (PLP) especially intestinal tumors (IT) are extremely rare. The diagnosis of intussusception secondary to pediatric IT is difficult due to the non-specific nature of symptoms. Its management remains a challenge. We report here four pediatric cases of intussusception secondary to IT lesions in order to analyze the clinical and therapeutic characteristics of this unusual condition.

Case presentations: Four children were diagnosed and operated in our center for intussusception secondary to IT during the period from February 2017 to February 2021. Patients' demographics, clinical presentations, investigations, operative data, and postoperative outcome were recorded and analyzed. There were three girls and one boy. Ages ranged between 1 and 8 years (average of 5.5 years). Intermittent abdominal pain with acute exacerbation and vomiting were the main clinical signs. Radiologic investigations showed the intussusception in all cases but the tumor lead point was evident only during the surgical exploration in most cases. All patients underwent surgical treatment with intussusception reduction and mass removal. Pathological examination of these masses revealed: submucosal intestinal lipoma (1 case), hamartomatous polyposis (2 cases), and Burkitt's lymphoma (1 case). For this last case, adjuvant chemotherapy was also needed. The postoperative period was uneventful in all cases.

Conclusion: Pediatric intussusception secondary to IT lesions is an unusual and challenging condition that requires high preoperative diagnostic suspicion, considerate intraoperative judgment, and appropriate postoperative follow-up for successful outcomes.

These tumors should be considered by the pediatric surgeons as possible PLP for recurrent intussusception, especially in older children. Surgeons should be familiar with the various treatment options, because the real cause of the intussusception is often accurately diagnosed by laparotomy.

Keywords: Intestinal tumor lesions, Intussusception, Pathologic lead points, Child, Surgery

Background

Primary IT are rare in children, representing less than 5% of all pediatric neoplasms [1, 2]. Clinical presentations are usually non-specific and variable, leading to diagnosis difficulties. Intestinal obstruction, particularly due to intussusception, has been reported to be a common discovery circumstance for both benign and malignant lesions [3].

We report four new pediatric cases of intussusception secondary to IT lesions, and present a review of the literature data to highlight epidemiologic, clinical, and management particularities of this unusual condition.

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Case presentations

A retrospective review of 4 pediatric cases of intussusception secondary to IT lesions was performed at a single children's hospital (Hedi Chaker Hospital, Sfax, Tunisia). We analyzed several diagnosis and therapeutic characteristics. The most remarkable clinical findings of all patients are summarized in Table 1.

Case 1

An 8-year-old girl was admitted to our department with 2-day history of worsening right-sided lower abdominal and vomiting. Pain had been present for 1 month ago. It was intermittent, and colicky in nature.

Physical examination revealed tenderness at the lower abdominal quadrant without fever nor signs of peritoneal irritation. Abdominal ultrasound (US) showed the typical target sign in the right lower abdomen suggestive of ileocolic intussusception (Fig. 1a). The patient underwent emergent exploratory laparotomy, which revealed a long intussuscepted ileal tract. Manual reduction was performed and an ileo-ileal intussusception was found. The lead point of the intussusception was a 3-cm

Table 1 Characteristics of the four cases of intussusception secondary to IT lesions

Case	1	2	3	4
Age (years)	8	5	8	1
Sex	Female	Male	Female	Female
Clinical presentations	Vomiting + acute abdominal pain + history of intermittent abdominal pain	Acute abdominal pain + history of recurrent abdominal pain with vomiting	Acute abdominal pain + history of intermittent generalized abdominal pain, nausea, and vomiting	Distended abdomen with multiple soft consistency masses
Preoperative imaging	US + CT	US + CT + Barium enema	US	CT
Tumor location/intussusception type	Ileum/ileocolic intussusception	Caecum/ileocolic intussusception	Jejunum/jejuno-jejunal intussusception	Ileum/ileo-ileal intussusception
Management	Resection and end-to-end anastomosis	Enterectomy containing the mass and diverting stoma	Elective removal of intestinal lesion	Resection of the involved segment and end-to-end anastomosis
Histopathological examination	Submucosal intestinal lipoma	Burkitt's lymphoma	Jejunal Peutz-Jeghers polyps	Intestinal hamartomatous polyps

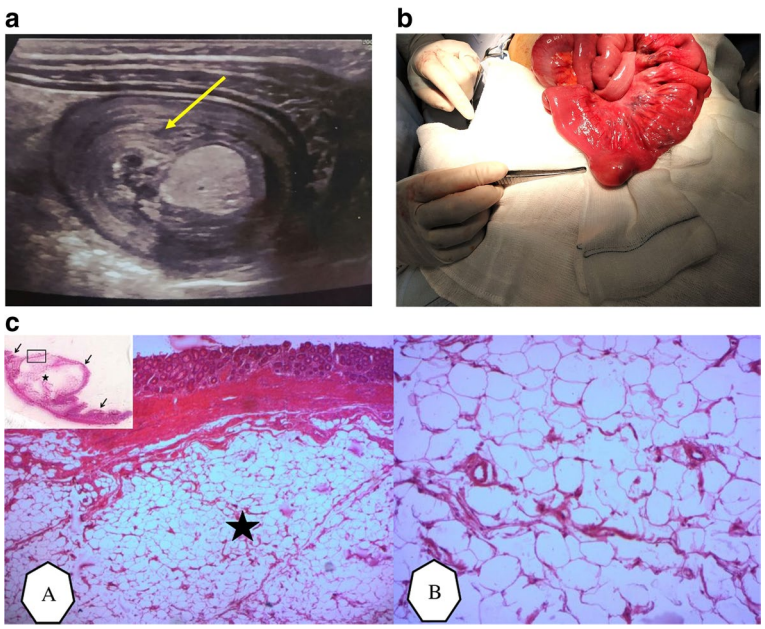

Fig. 1 **a** Sub-hepatic target sign at abdominal ultrasound revealed an intussusception (yellow arrow). **b** Intraoperative view of a submucosal ileal lipoma. **c** Histology image of ileal lipoma. A: Low power examination showed lobules of fat separated by delicate fibrovascular trabeculae (*). (HE × 25). B: A high power examination showed mature fat cells with no malignant features. (HE × 100)

hemispherical mass located 80 cm away from the ileocolic junction and originating from the antimesenteric border of the ileum (Fig. 2a). Resection and end-to-end anastomosis of the ileum segment containing the mass were performed. Histopathological examination confirmed the diagnosis of a submucosal intestinal lipoma (Fig. 1c). The postoperative course was uneventful and the patient was discharged after 1 week.

Case 2

A 5-year-old boy presented to the emergency department with a 2-month history of intermittent generalized abdominal pain and repeated vomiting which had acutely worsened.

On physical examination, he was afebrile with normal vital signs. His abdomen was distended and tender without evident masses or peritoneal signs.

Abdominal US and computed tomography (CT) scan revealed an ileocolic intussusception with a 39.5 × 29.3 mm complex mass in the right lower abdomen that appears to arise from the wall of the caecum (Fig. 2a). The barium enema showed normal small bowel loops, whereas the caecum exhibited a 40.5-mm × 23.1 mm focal filling defect (Fig. 2b). Exploratory laparotomy was performed and confirmed the presence of caeca mass with ileocolic intussusception (Fig. 2c). Enterectomy containing the mass and diverting stoma were performed.

Histopathology showed Burkitt's lymphoma. The child was thereafter referred to the Medical Oncology Department where he underwent adjuvant chemotherapy.

Case 3

An 8-year-old girl was admitted to our hospital with intermittent generalized abdominal pain, nausea and vomiting for the last 3 months, which were suddenly worsened.

Physical examination revealed tenderness in the left lower abdominal quadrant, without fever. Moreover, there were mucocutaneous pigmentations, especially over the lips (Fig. 3a).

Abdominal US showed the typical target sign suggestive of small bowel intussusception. The patient underwent exploratory laparotomy, which revealed a jejuno-jejunal intussusception secondary to multiple jejunal polyps. Reduction of the intussusception and elective removal of intestinal lesion were therefore performed. Pathological examination of the specimens concluded to jejunal Peutz-Jeghers syndrome polyps (Fig. 3b). The postoperative course was uneventful and the patient was discharged after 5 days.

Case 4

A 1-year-old girl, who had fast growing and progressive bilateral lesions on neck and the armpit since the age of 6

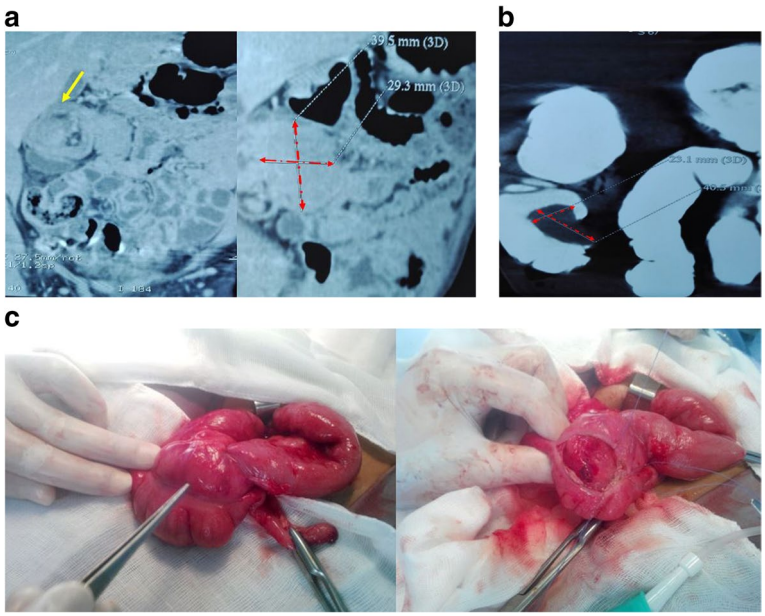

Fig. 2 **a** Abdominal CT scan showing ileocolic intussusception (yellow arrow), with a complex mass (red arrows) in the right lower abdomen that appears to arise from the wall of the caecum. **b** Barium enema showing a focal filling defect at the caecum wall (red arrow). **c** Intraoperative view of caeca mass associating with ileocolic intussusception

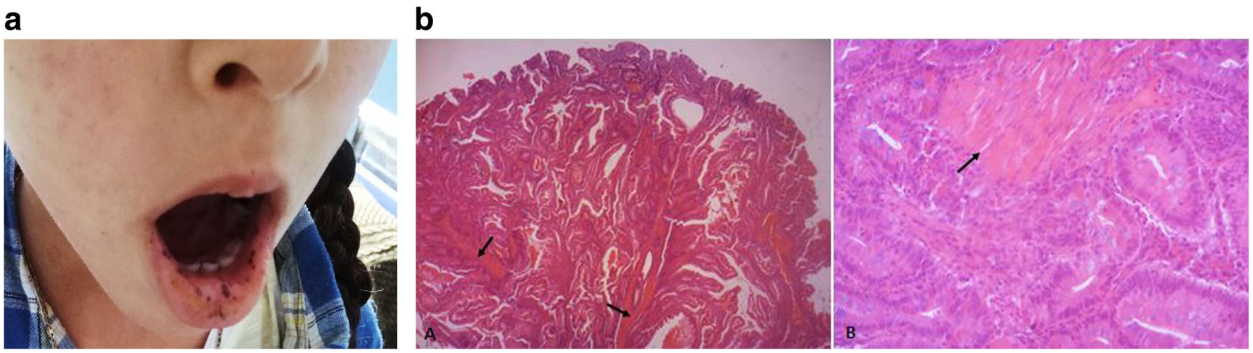

Fig. 3 **a** Dark mucocutaneous pigmentation observed over the lips of the child. **b** Histology image of jejunal Peutz-Jeghers polyp. A: Polyp with hyperplastic foveolar epithelium and inflammatory stroma with some cords of smooth muscle (→) (HE × 25). B: Polyp with inflammatory stroma, smooth muscle fibers (→), and non-dysplastic hyperplastic foveolar epithelium (HE × 200)

months, was admitted to our department for abdominal pain. She had normal psychomotor development.

At physical examination, the child presented macrocephaly, distended abdomen with multiple soft consistency masses, affecting also the neck and thorax (Fig. 4a). Abdominal examination revealed abdominal distention with tenderness all over the abdomen. CT scan revealed extensive lipomatosis in the thorax-neck region and showed a thickening of the bowel wall with intussusception in the left lower quadrant as well as enlarged mesenteric lymph nodes. Exploratory laparotomy was performed and found the presence of 4 ileo-ileal intussusception extending 15 cm (Fig. 4b).

We opted to intussusception reduction, resection of the involved segment, and end-to-end anastomosis (Fig. 4c). Histopathological examination confirmed the diagnosis of intestinal hamartomatous polyps.

One tumor in the left armpit measuring 6 × 8 cm was surgically removed and was pathologically diagnosed as a lipoma. Based on these results, we diagnosed this patient with Bannayan-Riley-Ruvalcaba Syndrome. The postoperative course was also uneventful for this child.

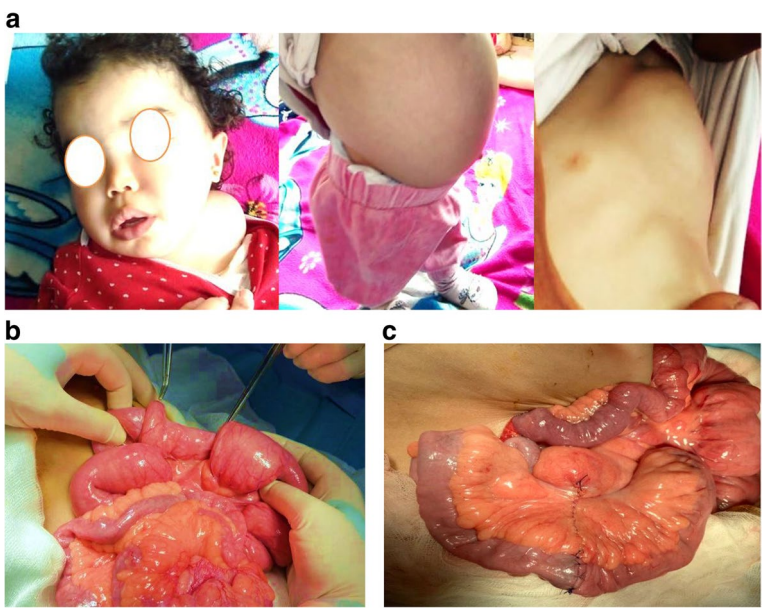

Fig 4 **a** Photo of the child presenting macrocephaly, distended abdomen with multiple masses at the neck and the thorax. **b** Intraoperative view of 2 ileo-ileal intussusceptions. **c** Intraoperative view of intestine after resection of the involved segment and end-to-end anastomosis

Discussion

Intussusception is the most common cause of intestinal obstruction and abdominal emergency in young children. While the majority of cases are idiopathic, underlying PLPs were found in 2.2–15% of childhood intussusception through previous studies [4–6]. They included especially Meckel's diverticulum, intestinal duplication and allergic purpura [5]. In this context, benign and malign intestinal tumors were also noted to be another rare cause of secondary intussusception (SI) in children [5]. Their real prevalence at this age is until unknown, but the paucity of clinical series on the subject reflects the rarity of this entity [1, 2].

Moreover, some particularities were noted, concerning age, clinical presentations and management. So, in a recent study, Zhao et al. [4] have demonstrated that the presence of a PLP is more likely in older children (mean age of 3.8 years), and that intestinal tumors were mostly associated with age more than 2 years (72% of cases). This also was similar to our findings, where the mean age of patients was 5.5 years.

Besides, the classic triad of symptoms (abdominal pain, vomiting and bloody stools) observed in primary intussusceptions are rare in secondary ones [4]. According to the literature data [4, 5], intussusception secondary to PLP, including IT has a wide spectrum of clinical presentations that lack specificity. Intermittent abdominal pain was the main clinical symptom. Other symptoms were described such as nausea, vomiting, bloody stools, abdominal distention, and change in bowel habits. History of recurrent intussusception is another clinical clue for the presence of a PLP [7, 8]. Likewise, three of our patients presented with the history of intermittent abdominal pain which have recently worsened.

On the other hand, the intussusception type, which is based on its anatomic location, varies depending on its idiopathic or secondary character. Then, previous studies [4, 5, 9] showed that complex intussusceptions were the most common type of secondary ones, followed by small intestinal and ileo-colic type. In our study, small bowel intussusception was the most common type.

So, this is because of these atypical clinical presentations and unusual anatomical locations that diagnosis of IT leading to intussusception remains difficult before surgical exploration [4, 5]. Radiologic investigations may help to identify and characterize potential PLP, but their sensibility is very variable [5, 10]. Abdominal US, for example, showed the underlying pathology in 56.8% in the study of Zhang [9] but only in 12% of cases in the study of Zhao [4]. According to Xiao-kun [5] and to our experience, results of abdominal CT were much better than US in looking for IT in the context of recurrent intussusception or atypical symptoms. Enteroscopy could also be useful [4].

Reviewing the literature data, the most common tumoral causes of pediatric SI were benign polyps, Peutz–Jeghers syndrome, hamartoma, and malignant lymphoma [4, 5, 9, 11, 12]. Other rare IT were described such intestinal leiomyoma [9] and lipoma [13, 14]. Comparatively, tumoral lead points in our patients were lipoma, lymphoma, hamartoma, and Peutz–Jeghers polyps.

The management of intussusception secondary to IT in children consists usually on surgical resection, in contrast to primary intussusception where the non-operative approach using pneumatic or hydrostatic reduction is often sufficient [4]. According to our cases and to previous studies [9], the main indications of surgical exploration are the small bowel location of the intussusception, recurrent episodes, or failure of conservative treatment. The preoperative diagnosis of the tumor was infrequent [4, 5, 9].

A reduction at surgery before resection may eliminates unnecessary extensive resection [15]. However, the risk of potential intraluminal seeding or venous tumor dissemination during the manipulation of a malignant lesion should be considered. Then, the main problem remains to distinguish benign from malignant lesions before reduction [16]. Moreover, reduction should not be attempted if there are signs of inflammation or ischemia of the bowel wall [7]. In our series, all intussuscepted tracts were reduced before resection without any incident. In children with multiple small intestinal polyps causing intussusception, such as Peutz-Jeghers syndrome, and as at our case, limited intestinal resections or simple polypectomy through a small enterotomy is preferred [8].

Furthermore, mini-invasive surgical approach could be a safe alternative not only for the reduction of the intussusception, but also to evaluate and treat the PLP [17–19]. Then laparoscopic reduction was successful in 86% in the series of Chan H et al. [16], with identification and excision of PLP in 41.6% of cases. In other studies [20–22], the role of laparoscopic-assisted enteroscopy and colonoscopy was emphasized in intussusceptions secondary to Peutz-Jeghers syndrome. These approaches did not be preconized in our series.

Finally, our patients have been included in specific follow-up programs according to the nature of PLP, which also characterize intussusceptions due to IT as lead point.

Conclusions

Pediatric intussusception secondary to IT lesions is an unusual and challenging condition that requires high preoperative diagnostic suspicion, considerate intraoperative

judgment, and appropriate postoperative follow-up for successful outcomes.

These tumors should be considered by the pediatric surgeons as possible PLP for recurrent intussusception, especially in older children. Surgeons should be familiar with the various treatment options, because the real cause of the intussusception is often accurately diagnosed by laparotomy.

Abbreviations
US: Ultrasound; CT: Computed tomography; IT: Intestinal tumor; PLP: Pathologic lead point.

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Authors' contributions
TC: conception, data analysis, draft the manuscript, revised the manuscript, and approved the submission. RC: data analysis, draft the manuscript, revised the manuscript, and approved the manuscript. MB: revised the manuscript and approved the manuscript. MM: revised the manuscript and approved the manuscript. NB: revised the manuscript and approved the manuscript. RK: revised the manuscript and approved the manuscript. SA: acquisition and analysis, revised the manuscript, and approved the manuscript. HZ: revised the manuscript and approved the manuscript. BTS: revised the manuscript and approved the manuscript. RM: revised the manuscript and approved the manuscript. The authors have read and approved the final manuscript.

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Declarations

Ethics approval and consent to participate
None.

Consent for publication
The parents of the patient have consented to use of clinical photographs for publication and research process.

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

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References

1. Bethel CAI, Bhattacharyya N, Hutchinson C, Ruymann F, Cooney DR. Alimentary tract malignancies in children. J Pediatr Surg. 1997;32:1004–9.
2. Ladd AP, Grosfeld JL. Gastrointestinal tumors in children and adolescents. Semin Pediatr Surg. 2006;15:37–47.
3. Lechner GW, Connolly PJ. Benign neoplasms of the small intestine: with a report of three bleeding benign tumors of the jejunum. JAMA. 1959;169:2003–8.
4. Zhao L, Feng S, Wu P, Lai X-H, Lv C, Chen G. Clinical characteristics and surgical outcome in children with intussusceptions secondary to pathologic lead points: retrospective study in a single institution. Pediatr Surg Int. 2019;35:807–11.
5. Lin X, Xia Q, Huang X, Han Y, He G, Zheng N. Clinical characteristics of intussusception secondary to pathologic lead points in children: a single-center experience with 65 cases. Pediatr Surg Int. 2017;33:793–7.
6. Wang Z, He Q, Zhang H, Zhong W, Xiao W, Lu L, et al. Intussusception patients older than 1 year tend to have early recurrence after pneumatic enema reduction. Pediatr Surg Int. 2015;31:855–8.
7. Guo W, Hu Z, Tan Y, Sheng M, Wang J. Risk factors for recurrent intussusception in children: a retrospective cohort study. BMJ. 2017;7:e018604.
8. Daneman A, Alton DJ, Lobo E, Gravett J, Kim P, Ein SH. Patterns of recurrence of intussusception in children: a 17-year review. Pediatr Radiol. 1998;28:913–9.
9. Zhang Y, Dong Q, Li S-X, Ren W-D, Shi B, Bai Y-Z, et al. Clinical and ultrasonographic features of secondary intussusception in children. Eur Radiol. 2016;26:4329–38.
10. Tan HL, Koh YX, Taufik M, Lye WK, Goh BKP, Ong HS. A clinical scoring system to predict the clinical sequelae of computed tomography diagnosed intussusception. World J Surg. 2018;42:682–7.
11. Scorletti F, Bove K, Brown RL. Small bowel-small bowel intussusception with high grade obstruction due to intramural submucosal ileal hamartoma in a 5-year-old child: a case report. Int J Surg Case Rep. 2019;61:33–7.
12. Saad AAM, Kkalid T, Abbas M, Salih KMA. Rare presentation of chronic ileocecal intussusception secondary to Burkitt's lymphoma in three years Sudanese boy: a case report and literature review. Pan Afr Med J. 2018;31:57–61.
13. Asaumi Y, Miyanaga T, Ishiyama Y, Hattori M, Hashizume Y. Pediatric ileoileal intussusception with a lipoma lead point: a case report. Gastroenterol Rep. 2014;2:70–2.
14. Abdelmohsen SM, Osman MA, Hussien MT. An ileo-ileal intussusception secondary to polypoid lipoma in a child, a case report and review of the literature. Int J Surg Case Rep. 2019;57:88–90.
15. Marinis A, Yiallourou A, Samanides L, Dafnios N, Anastasopoulos G, Vassiliou I, et al. Intussusception of the bowel in adults: a review. World J Gastroenterol. 2009;15:407–11.
16. Bines JE, Ivanoff B, Justice F, Mulholland K. Clinical case definition for the diagnosis of acute intussusception. J Pediatr Gastroenterol Nutr. 2004;39:511–8.
17. Chui CH, Ong LY, Chua JHY, Yap T-L. "Chinese fan spread" distraction technique of laparoscopic reduction of intussusception. JSLS. 2007;11:238–41.
18. Sklar CM, Chan E, Nasr A. Laparoscopic versus open reduction of intussusception in children: a retrospective review and meta-analysis. J Laparoendosc Adv Surg Tech. 2014;24:518–22.
19. Burjonrappa SC. Laparoscopic reduction of intussusception: an Evolving Therapeutic Option. JSLS. 2007;11:235–7.
20. Patel RV, Stanton M, Curry J, Kiely E. Laparoscopic management of Peutz-Jeghers syndrome (PJS) presenting with chronic non-ischaemic jejuno-jejunal intussusception in an adolescent girl. Case Rep Dermatol. 2013;2013:bcr2013200939.
21. Morales Gonzalez AM, Clapp B. Laparoscopic management of small bowel intussusception in a 16-year-old with Peutz-Jeghers syndrome. JSLS. 2008;12:332–4.
22. Zanoni ECA, Averbach M, Borges JLA, Corrêa PAFP, Cutait R. Laparoscopic treatment of intestinal intussusception in the Peutz-Jeghers syndrome: case report and review of the literature. Surg Laparosc Endosc Percutan Tech. 2003;13:280–2.

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