A Very Rare Cause for Intussusception in Children: Inflammatory Fibroid Polip a Case Report and Review of the Literature

Çocuklarda İnvajinasyonun Nadir Bir Sebebi: İnflamatuar Fibroid Polip Olgu Sunumu ve Literatür İncelemesi

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ABSTRACT Intussusception is one of the major causes of intestinal obstruction among infants. The exact etiology cannot be determined usually, but an identifiable cause or a pathological leading point has been reported in a large number of patients. Although majority of these cases consist of juvenile polyps, inflammatory fibroid polyps (IFP) have been reported also among children but only as case reports. IFP is a rare benign lesion of the intestine that may occur throughout the digestive tract. The lesion is usually found in the stomach, and secondly in the ileum. Ileal IFP usually presents as intussusception or obstruction. During the operation of a five-month-old male patient who underwent surgery due to intussusception, a polyp was encountered as a leading point and it was diagnosed as an IFP at the histopathological examination. The lesion described in this report is the first infant reported to date, which is presented with an ileocolic intussusception. The purpose of this report is to emphasize that IFP can be seen among infants too and can cause intussusception at this age.

Key Words: Intussusception; polyps; infant

ÖZET Invajinasyon, bebeklerde barsak tıkanıklıkları nedenleri arasında önemli bir yere sahiptir. Etyolojisi genelde bilinmese de patolojik öncü noktalara bağlı oluşan çok sayıda vaka bildirilmiştir. Bu vakalar arasında polipler önemli yer tutar. Bu poliplerin büyük kısmı juvenil polip yapısında olsa da inflamatuar fibroid polip (EFP) özellikle çocuklarda sadece olgu sunumları şeklinde bildirilmiştir. İnflamatuar fibroid polip (EFP) tüm sindirim sistemi boyunca oluşabilen ve nadir gözlenen bir lezyondur. Bu lezyona genellikle midede, ikinci sıklıkta ileumda rastlanır. Ileal EFP genellikle invajinasyon veya tıkanma ile bulgu verir. Kliniğimizde invajinasyon nedeniyle ameliyat edilen 5 aylık bir erkek olguda öncü nokta olarak bir polipe rastlandı ve polipin histopatolojik incelemesi EFP'yi desteklemekteydi. Bu yazıda bildirilen olgu şu ana kadar EFP'ye bağlı ileokolik invajinasyonla başvuran ilk bebek olma özelliğini taşımaktadır. Bu yazı ile EFP'nin bebeklerde de gözlenebileceğine ve bu yaşta invajinasyona yol açabileceğine dikkat çekmek amaçlanmıştır.

Anahtar Kelimeler: İn tussusepsiyon; polipler; bebek

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Inflammatory Fibroid Polyp (IFP) or Vanek's tumor, which is described by Vanek himself as "eosinophilic inflammatory infiltrate" in 1949 is an uncommon, non-neoplastic proliferating lesion which can develop in various parts of the gastro-intestinal tract but most commonly in the gastric antrum and the ileum.[1] IFP is a protuberant lesion, located near the muscularis mucosae and composed of a proliferation of fusiform cells and

conjunctive fibers surrounding capillaries and a variable inflammatory infiltrate.[2] Most of these lesions are asymptomatic and discovered as incidental findings during endoscopy performed for unrelated reasons. When symptomatic, Vanek's tumours are usually associated with abdominal pain, weight loss, bleeding, dyspeptic symptoms, iron-deficiency anemia and intussusceptions.[3-5] The clinical symptoms may vary according to the location of the lesion. After gastric antrum, the small bowel is the second most common site of origin, where it usually presents as intussusception or obstruction.[6]

We experienced an IFP at the operation of a five-month-old patient, which is not encountered at this age so far.

CASE REPORT

A five-months-old boy was admitted to our hospital with a two-day history of colicky abdominal pain, recurrent vomiting and abdominal distension. There was no history of red jelly stools or melena, nor any history of a previous operation or major illness. On abdominal examination, there was a soft to firm, mobile palpable mass in the right upper quadrant but the abdomen was soft with no sign of peritonitis. Urine examination was clear, but there were remarkably high levels of white blood cells (25.3 K/uL) at full blood count and C-Reactive protein (85 mg/mL). Plain abdominal radiographs revealed multiple loops of dilated small bowel in the left upper quadrant of the abdomen. Ultrasound scan demonstrated a typical "target sign" in the right upper quadrant indicating an intussusception.

A diagnosis of mechanical bowel obstruction was made based on the findings. Because of the long duration of the history and septic apperance of the patient, an exploratory laparotomy was performed instead of pneumatic/hydrostatic reduction. The operation revealed an intussusception, where a long segment of distal ileum was seen to intussuscept into the cecum just proximal to the ileocecal valve and ascending to the splenic flexura. The ileoileal part of the intussusception could not be reduced successfully and an ileal dehiscence occurred. An ileal polyp was discovered at this level by the surgeon (Figure 1). Because of the edematous appearance of the ileocecal region and the terminal ileum an ileostomy was performed, which closed after 1 month successfully.

The resected specimen of jejunum was 13 cm long. Gross pathologic examination revealed a sessile polypoid lesion with a whitish-grey ulcerated surface at the leading point of the intussusceptions (Figure 2). The polyp was 3.0x2.0x1.5 cm in size and occluding the lumen of the jejunum. The polyp and jejunum were infarcted and dilated. The histological study showed an ulcerative inflammation, hemorrhagic necrosis and

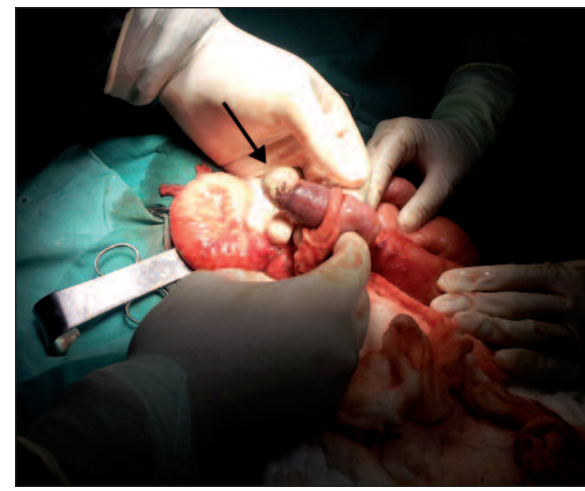

FIGURE 1: An ileal polyp was encountered when ileal dehiscence has occurred. The polyp was arising from the submucosa (arrow) and was acting like a leading point.
(See color figure at http://pediatri.turkiyeklinikleri.com/)

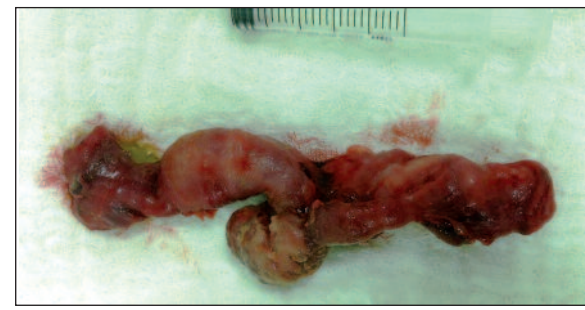

FIGURE 2: Resected specimen of gangrenous jejunojejunal intussusception containing the polyp in the middle.
(See color figure at http://pediatri.turkiyeklinikleri.com/)

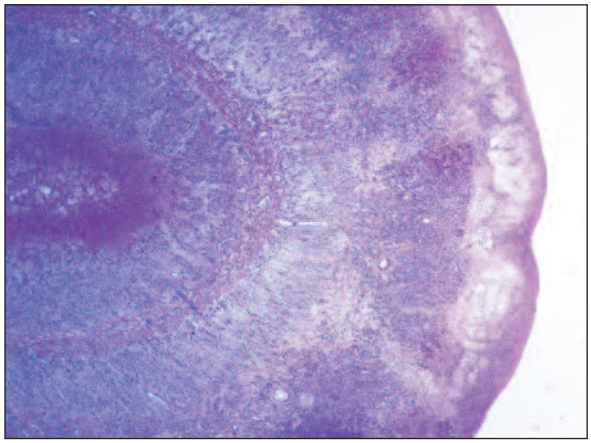

FIGURE 3: The longitudinal section of the polyp. The surface is ulcerated and the amount of the cells is increased. (H&E, x40).
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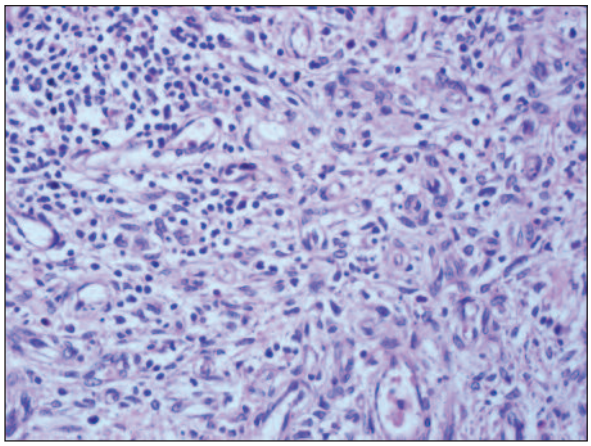

FIGURE 4: In the submucosal area of the polyp, there was intense inflammatory cell population in the fibromyxoid vascularized stroma including spindle-shaped stromal cells and eosinophils around the small blood vessels. (H&E, x200).
(See color figure athttp://pediatri.turkiyeklinikleri.com/)

fibrinous peritoneal response at the ileum. The surface of the lesion was also ulcerated (Figure 3). In the submucosal area of the polyp, there was intense inflammatory cell population in the fibromyxoid vascularized stroma including spindle-shaped stromal cells and eosinophils around the small blood vessels (Figure 4). These findings are considered pathognomonic for the diagnosis of inflammatory fibroid polyp. At immunohistologic staining, the polyp was for CD 34 and vimentin positive and CD 117 and Desmin negative as indicating an IFP.

DISCUSSION

Intussusception is the most common cause of acute intestinal obstruction in infants, occurring when one portion of bowel invaginates into a more distant portion of the bowel. It has a characteristic clinical picture of vomiting, red 'currant jelly' stools, severe colicky abdominal pain and palpable mass. It is an emergent condition where delay in diagnosis is common, which may lead to bowel perforation, obstruction, and necrosis. Most cases in infants and young children are 'idiopathic' in that the etiology of the intussusception is due to hypertrophied lymphoid tissue in the terminal ileum which results in ileocolic intussusception.[7] But it can also result from pathologic lead points, like Meckel's diverticulum, polyps, lymphomas, hematomas of the intestinal wall in Henoch-Schönlein purpura or hemophilia, intestinal duplications, cysts or neurofibromas. In Europe, intussusception has been associated with structural lead points in 1-11% of cases (median, 3%).[8] In a review of 43 cases, Navarro et al. found that 8 (18.6%) of them was due to polyps of different origins and Montgomery et al. found that one of the 63 pediatric patients with intussusceptions was due to inflammatory fibroid polyp.[9,10] Ulukaya-Durakbaşa et al. reported that the histopathology result of one patient among 62 patients was consistent with inflammatory fibroid polyp.[11]

The reported IFP cases in children is limited with case reports. Chongsrisawat et al. have investigated the literature in English language for IFP in children and they found 6 patiens between the ages 3 and 8.[12] Dabral reported a 7-year-old boy who presented with intestinal obstruction due to an IFP.[13]

IFP is a sessile or semi-pedunculated, non-neoplastic, proliferating lesion arising from submucosa, which overlying mucosa is usually ulcerated.[14] At the histological examination IFPs are composed of bland, uniform spindled and/or stellate cells, loose fibromyxoid background with regular vascular pattern and eosinophil rich mixed inflammatory infiltrate. At the immunohistological staining, IFPs are CD34, CD35, vi-

mentin, fascin and cyclin D1 positive and always negative for CD 117, keratin, bcl2 and desmin.[3] With these findings it is possible to differentiate IFP from gastrointestinal (GI) stromal tumors, solitary fibrous tumors, GI schwannomas, GI perineurinomas and gastric plexiform fibromyxomas. In our case, the polyp was for CD 34 and vimentin positive and CD 117 and Desmin negative as indicating an IFP.

The pathogenesis of IFP still remain unknown. It has been hypothesized that several factors like bacterial, chemical, traumatic or metabolic injury could affect the gastrointestinal mucosa and stimulate the formation of the polyps.[15] Several reports showed the association with *Helicobacter pylori (H. pylori)* and it is still unclear whether *H. pylori* was the cause of the ulcer by exposing the stroma to chemical, mechanical and biological irritation, or if the *H. pylori* itself started an immunological reaction which caused the polyp, or if the *H. pylori* only complicated this finding.[16] Buciuto et al. reported one patient with *H. pylori* infection who had also autoimmune diseases (sarcoidosis and rheumatoid arthritis). This report suggests that also immunological factors may be involved at the patogenesis of IFP.[17] Klepinger and Pontius suggested that surgical trauma could be a factor.[18] Ojima et al. reported one patient with IFP which had been caused by repeated colostomy irrigation, and Savargaonkar reported two patients who had a history of intra-abdominal surgical procedures performed several years previously, suggesting that trauma may have initiated the process.[6,19]

This general knowledge for the pathogenesis of the IFP seems not compatible with our case because the patient described in the present report had no history of gastroenteritis, no inflammatory or autoimmune-like symptoms, no associated diseases or previous operation. He did not receive food other than breast milk. The patient was an infant, so we hypothesize that the factors that cause the formation of the polyp may began very early even maybe at the newborn period.

CONCLUSION

Polyps and polypoid lesions of the small intestine present a major challenge to the surgeon especially among children. It is usually difficult to diagnose and it is only apparent when it presents with symptoms like intussusception and bleeding. A high index of suspicion is required for the early diagnosis of these lesions. We reported this case because of the rarity of this disease entity in the field of pediatric surgery.

REFERENCES

1. Vanek J. Gastric submucosal granuloma with eosinophilic infiltration. Am J Pathol 1949;25 (3):397-411.
2. Acero D, Garijo G, Hombrados M, Figa M, Adrados M, Aldeguer X, et al. [Gastrointestinal inflammatory fibroid polyps. Clinical characteristics and follow-up in a series of 26 patients]. Gastroenterol Hepatol 2005;28(4): 215-20.
3. İbiş AC, Albayrak D, Sezer YA, Polat N, Coşkun İ. [Intussusception due to fibroid polyp: case report]. Medical Journal of Bakırköy 2008;4(3):119-21.
4. Vardareli E, Tiftikçi A, Tözün N, Özveri E, Ertem M. [A giant inflammatory fibroid polyp causing anemia: a case report]. ACU Sağlık Bil Derg 2010;1(2):115-7.
5. Paikos D, Moschos J, Tzilves D, Koulaouzidis A, Kouklakis G, Patakiouta F, et al. Inflammatory fibroid polyp or Vanek's tumour. Dig Surg 2007;24(3):231-3.
6. Savargaonkar P, Morgenstern N, Bhuiya T. Inflammatory fibroid polyp of the ileum causing intussusception: report of two cases with emphasis on cytologic diagnosis. Diagn Cytopathol 2003;28(4):217-21.
7. Applegate KE. Intussusception in children: evidence-based diagnosis and treatment. Pediatr Radiol 2009;39(Suppl 2):S140-3.
8. Huppertz HI, Soriano-Gabarró M, Grimprel E, Franco E, Mezner Z, Desselberger U, et al. Intussusception among young children in Europe. Pediatr Infect Dis J 2006;25(1 Suppl): S22-9.
9. Navarro O, Dugougeat F, Kornecki A, Shuckett B, Alton DJ, Daneman A. The impact of imaging in the management of intussusception owing to pathologic lead points in children. A review of 43 cases. Pediatr Radiol 2000; 30(9):594-603.
10. Montgomery EA, Popek EJ. Intussusception, adenovirus, and children: a brief reaffirmation. Hum Pathol 1994;25(2):169-74.
11. Ulukaya-Durakbaşa Ç, Çağlar M, Fettahoğlu S, Zemheri IE, Mutuş HM, Çam S et al. [Clinical experience at pediatric kolorectal polyps]. Göztepe Tıp Dergisi 2012;27(1):1-5.

12. Chongsrisawat V, Yimyeam P, Wisedopas N, Viravaidya D, Poovorawan Y. [Unusual manifestations of gastric inflammatory fibroid polyp in a child]. World J Gastroenterol 2004;10(3):460-2.
13. Dabral C, Singh N, Singh PA, Misra V. [Inflammatory fibroid polyp of small intestine in a child]. Indian J Gastroenterol 2003;22(3): 101.
14. de la Plaza R, Picardo AL, Cuberes R, Jara A, Martínez-Peñalver I, Villanueva MC, et al. Inflammatory fibroid polyps of the large intestine. Dig Dis Sci 1999;44(9):1810-6.
15. Johnstone JM, Morson BC. Inflammatory fibroid polyp of the gastrointestinal tract. Histopathology 1978;2(5):349-61.
16. Shalom A, Wasserman I, Segal M, Orda R. Inflammatory fibroid polyp and Helicobacter pylori. Aetiology or coincidence? Eur J Surg 2000;166(1):54-7.
17. Buciuto R, Kullman E, Boeryd B, Borch K. Helicobacter pylori gastritis associated with a gastric inflammatory fibroid tumour and sarcoidosis. Eur J Surg 1996;162(5):421-4.
18. Klepinger CA, Pontius EE. Inflammatory polyps of the intestinal tract. Am J Clin Pathol 1964;42:371-80.
19. Ojima Y, Okajima M, Asahara T, Arita M, Kobayashi R, Nakahara M, et al. Inflammatory fibroid polyp of the ileum with the appearance of a Borrmann type II lesion, caused by colostomy irrigation: report of a case. Surg Today 1997;27(11):1061-3.