

CAS CLINIQUE/CASE REPORT

JEJUNOJEJUNAL INTUSSUSCEPTION CAUSED BY AN INFLAMMATORY FIBROID POLYP Case Report and Review of the Literature

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El Hajj II, Sharara AI. Jejunojejunal intussusception caused by an inflammatory fibroid polyp. Case report and review of the literature. *J Med Liban* 2007 ; 55 (2) : 108-111.

ABSTRACT : Inflammatory fibroid polyps (IFPs) are rare benign submucosal growth of the gastrointestinal tract. The exact pathogenesis is still not well known. Clinical symptoms are variable. Physical exam and laboratory tests are helpful in establishing the diagnosis ; however, microscopic examination of the resected lesions is required to confirm the diagnosis of IFPs. Surgical resection of the lesion remains the primary therapy.

Jejunojejunal intussusception secondary to IFPs in particular has only rarely been reported. We have reviewed all documented cases and added a new one to the literature. In this paper, we examine pertinent pathologic, epidemiologic, clinical, diagnostic, and therapeutic characteristics of this clinical entity.

INTRODUCTION

Inflammatory fibroid polyps (IFPs) are rare benign tumor-like lesions that originate in the submucosa of the gastrointestinal (GI) tract. IFPs may occur anywhere in the alimentary tract, but are seen mainly in the antral portion of the stomach, less frequently in the ileum, and only occasionally in the colon, jejunum, duodenum, and esophagus [1-2]. Jejunojejunal intussusception secondary to IFPs is uncommon and has only been reported in few cases.

CASE REPORT

A 52-year-old woman presented to the hospital with 10 days history of intermittent abdominal pain, nausea, and three episodes of vomiting. No fever, no chills reported. No weight change. Normal bowel movements, no fresh blood per rectum, no hematemesis reported. Past medical history is positive for osteopenia, hypertension, and dyslipidemia. Past surgical history is positive for hysterectomy and unilateral oophorectomy. Physical exam

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El Hajj II, Sharara AI. Invagination jéjunojéjunale secondaire à un polype fibroïde inflammatoire. Cas clinique et revue de la littérature. *J Med Liban* 2007 ; 55 (2) : 108-111.

RÉSUMÉ : Les polypes fibroïdes inflammatoires sont des élévations bénignes de la sous-muqueuse du système digestif. La pathogénie exacte n'est pas bien connue. Les symptômes cliniques sont variables. L'examen physique et les tests de laboratoire aident à établir le diagnostic, mais l'examen microscopique de la lésion est indispensable pour confirmer le diagnostic. La résection chirurgicale de la lésion est le traitement radical.

L'invagination jéjunojéjunale secondaire au polype fibroïde inflammatoire est rarement rapportée. Nous avons revu tous les cas documentés et nous avons ajouté un nouveau cas à la littérature. Les caractéristiques pertinentes de l'anatomie pathologique, de l'épidémiologie, de la présentation clinique, et des modalités diagnostiques et thérapeutiques de cette entité clinique sont examinées dans cet article.

revealed tenderness of the mid and left abdomen and hyperactive bowel sounds. CBC, BUN, creatinine, electrolytes, and liver function tests were within normal limits. Chest X-ray was normal. X-ray of the abdomen showed disturbed gas distribution and distended small bowels. Enhanced CT scan of abdomen and pelvis showed jejunojejunal intussusception with an intraluminal well-defined soft tissue mass (Figure 1). The patient underwent laparoscopic enterectomy and enterostomy. Grossly, the mass showed a soft polypoid lesion, measuring 3.5 x 2.5 x 2.5 cm, with a yellow cut surface and focal areas of hemorrhage located at the tip (Figure 2). Microscopic examination showed a capillary network with stellate stromal cells and mixed inflammatory cell infiltrates (prominent eosinophils with neutrophils, lymphocytes and plasma cells) in a myxomatous stroma (Figure 3). Pathologic findings are consistent with an inflammatory fibroid polyp. The postoperative course was uneventful.

DISCUSSION

We searched the medical literature using the National Library of Medicine PubMed system and OLDMEDLINE from pre-1966 citations, from January 1945 through June 2006. Case reports of jejunojejunal intussusception were identified by querying the key words



FIGURE 1
Enhanced CT scan of abdomen and pelvis showing jejunal intussusception with an intraluminal well-defined soft tissue mass.

“inflammatory fibroid polyp” and “jejunojejunal intussusception.” Sixteen cases of jejunojejunal intussusception secondary to IFPs reported from 1981 to 2006, were identified (Table I) [2-15]. We herein describe a new case of IFP of the jejunum causing jejunojejunal intussusception.

IFPs have been first described by Vanek [16] in 1949, as “*gastric submucosal granuloma with eosinophilia*,” but similar lesions have been described in the small bowel and colon. Various names have been suggested, including eosinophilic granuloma, submucosal fibroma, hemangiopericytoma, inflammatory pseudotumor, and fibroma. The term IFP, first proposed by Helwig and Ranier [17] for the gastric polyps, has gained acceptance for similar lesions throughout the GI tract.

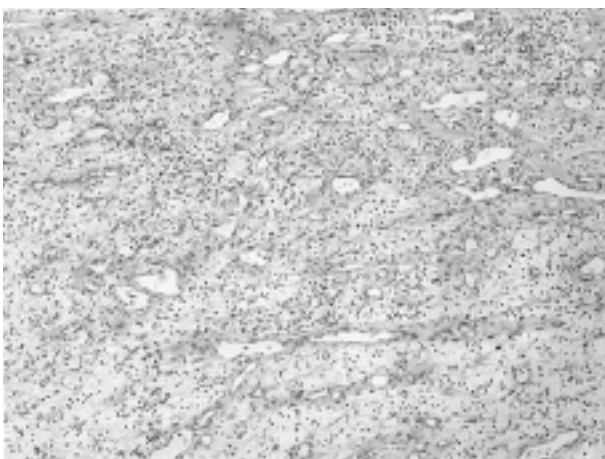


FIGURE 3
Microscopic examination showing a capillary network with stellate stromal cells and mixed inflammatory cell infiltrates (prominent eosinophils with neutrophils, lymphocytes and plasma cells) in a myxomatous stroma.

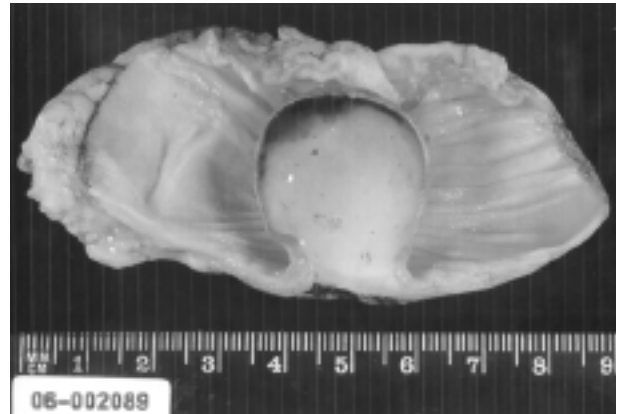


FIGURE 2
Gross examination showing a soft polypoid lesion, measuring 3.5 x 2.5 x 2.5 cm, with a yellow cut surface and focal areas of hemorrhage located at the tip.

IFPs are non neoplastic cellular proliferations originating primarily from the submucosa of the GI tract. They are composed of fibroblasts, blood vessels, and inflammatory cells (particularly eosinophils) within an edematous and collagenous stroma. IFPs appear as sessile or polypoid, usually solitary, circumscribed, and round to ovoid nodules, and occasionally as nodular thickening of the bowel wall [1-2]. The microscopic features are similar to those in our patient.

The pathogenesis of fibroid polyps is unknown ; a reactive process is favored. Johnstone and Morson postulated that the condition is a result of an uncontrolled proliferation of the mesenchymal tissue of the submucosa of the gut [1]. Helwig and Ranier [17] suggested that the striking features of IFPs are their polypoid nature and the characteristic arrangement of fibrosis and vascular elements. The degree of eosinophilic infiltration is variable and of doubtful significance.

IFPs may occur anywhere in the alimentary tract, but are seen mainly in the antral portion of the stomach, less frequently in the ileum (usually in the distal half), and only occasionally in the colon, jejunum, duodenum, and esophagus [2, 18]. In a review of 76 cases of IFPs, Johnstone and Morson [1] found a relative incidence of 75% in the stomach, 18% in the small intestine, 7% in the colon, and 1% in the esophagus.

Intussusception is a frequently and mostly benign condition in childhood. Conversely, it is a rare condition during adulthood and generally associated with an underlying malignancy [19]. IFPs affect both sexes and all age groups, with a peak incidence in the fifth and sixth decades, and a slight male predominance [1]. In a review of 64 cases of IFPs, Shimmer and Helwig [2] found that 61% of patients were males, and 39% were females. The patients ranged in age at diagnosis from 3 to 80 years, with a mean of 48.8 years. In the particular case of jejunojejunal intussusception secondary to IFP, we found a slight female predominance. The patients ranged

in age from 17 to 66 years, with a mean age of diagnosis of 49.1 years (Table I).

Clinical symptoms of IFPs are variable depending on the location and size of the lesion. Abdominal pain is the main symptom in patients whose lesions are in the stomach. Obstruction or intussusception is the most frequent initial symptom when located in the small intestine. Other GI symptoms such as vomiting, diarrhea, bloody stools, tenesmus, and alteration in bowel habits are infrequent [1-2, 12].

Physical exam, radiological studies, and laboratory tests are helpful in establishing the diagnosis ; however microscopic examination of the resected lesion is required. Radiographically, the plain abdominal film is usually the initial imaging study. The findings include small bowel distension with air-fluids levels and a small amount of gas in the colon. Ultrasound has a 100% sensitivity and 89% specificity rate in depicting intussusception. Classical ultrasound imaging features of intussusception included doughnut, target and bull's eye signs [20]. CT scan of abdomen and pelvis with oral and IV contrast is the most accurate diagnostic tool for evaluation of intussusception. There are three classic CT patterns pathognomonic for intestinal intussusception : 1) target lesion-intraluminal soft tissue mass with eccentric fat density due to invaginated mesentery ; 2) reniform pattern ; 3) and finally sausage pattern [21]. Magnetic resonance imaging (MRI) can contribute to the radiologic diagnosis of intussusception by demonstrating the "bowel-within-bowel" or "coiled-spring" appearance [22]. Retrograde double balloon enteroscopy was reported to be useful for the preoperative diagnosis of IFP causing small bowel intussusception in one case [13].

The primary treatment of IFPs is exploratory laparotomy. The lesion seems to have no malignant potential, so local excision of the polyp is curative and recurrence of the polyps has been reported only once [1].

In summary, IFPs should be included in the differential diagnosis of medium-sized, single, mural, or intramural lesions of the GI tract, as well as in the differential diagnosis of intussusception or intestinal obstruction in elderly patients.

REFERENCES

1. Johnstone JM, Morson BC. Inflammatory fibroid polyp of the gastrointestinal tract. *Histopathology* 1978 ; 2 : 349-61.
2. Shimer GR, Helwig EB. Inflammatory fibroid polyps of the intestine. *Am J Clin Pathol* 1984 ; 81 : 708-14.
3. Williams RM. An ultrastructural study of a jejunal inflammatory fibroid polyp. *Histopathology* 1981 ; 5 : 193-203.
4. Winkler H, Zelikovski A, Gutman H et al. Inflammatory fibroid polyp of the jejunum causing intussusception. *Am J Gastroenterol* 1986 ; 81 : 598-601.
5. Kim JS, Kwon SY, Byun KS et al. Jejunal inflammatory fibroid polyp presenting as intussusception – a case report with review of the literature. *Korean J Intern Med* 1994 ; 9 : 51-4.
6. Ling CC, Hsu TC, Shih SC et al. Inflammatory fibroid polyp of the jejunum causing intussusception : a case report. *Chinese Med J* 1994 ; 53 : 127-30.
7. Bandyopadhyay PK, Ishaq N, Malik AK, Mahroos S. Inflammatory fibroid polyp of proximal ileum causing recurrent intussusception. *Br J Clin Pract* 1997 ; 51 : 125-6.

TABLE I
DOCUMENTED CASES OF JEJUNOJEJUNAL INTUSSUSCEPTION SECONDARY TO INFLAMMATORY FIBROID POLYPS SINCE 1945

| AUTHOR | Number of cases | Year | Age | Sex | Symptoms | Duration of symptoms |
|----------------------------|-----------------|------|-----|-----|----------------------------------|----------------------|
| Williams [3] | 1 | 1981 | NS | NS | Abd. pain | NS |
| Shimer et al. [2] | 3 | 1984 | NS | NS | Abd. pain | 2 weeks |
| Winkler et al. [4] | 1 | 1986 | 61 | M | Abd. pain, nausea, diarrhea | 1 week |
| Kim et al. [5] | 1 | 1994 | 52 | F | Abd. pain, vomiting | 10 days |
| Ling et al. [6] | 1 | 1994 | 56 | F | Abd. pain, nausea, diarrhea | 8 weeks |
| Bandyopadhyay et al. [7] | 1 | 1997 | 64 | F | Abd. pain, vomiting | 3 weeks |
| Shih et al. [8] | 1 | 1997 | 66 | M | Abd. pain, vomiting, weight loss | 4 days |
| Kuestermann et al. [9] | 1 | 1999 | 34 | F | Abd. pain, nausea, vomiting | 5 weeks |
| Sah et al. [10] | 1 | 2002 | 45 | M | Abd. pain | 2 weeks |
| Topaloglu et al. [11] | 1 | 2003 | 56 | M | Abd. pain | NS |
| Bays et al. [12] | 1 | 2004 | 17 | M | Abd. pain | 8 weeks |
| Miyata et al. [13] | 1 | 2004 | 64 | F | Abd. Pain, constipation/diarrhea | 6 weeks |
| Spengler & Jeschke [14] | 1 | 2004 | 55 | F | Abd. pain, vomiting | 3 weeks |
| Vijayaraghavan et al. [15] | 1 | 2004 | 20 | F | Abd. pain | 3 weeks |
| Present case | 1 | 2006 | 52 | F | Abd. pain, nausea, vomiting | 10 days |

Abd : abdominal NS : not stated

8. Shih LN, Chang SL, Chuang SM, Kuo CF. Inflammatory fibroid polyp of the jejunum causing intussusception. *Am J Gastroenterol* 1997 ; 92 : 162-4.
9. Kuestermann SA, Saleeb SF, Teplick SK. General case of the day. Jejunal intussusception caused by an inflammatory fibroid polyp (IFP). *Radiographics* 1999 ; 19 : 539-41.
10. Sah SP, Agrawal CS, Rani S. Inflammatory fibroid polyp of the jejunum presenting as intussusception. *Indian J Pathol Microbiol* 2002 ; 45 : 119-21.
11. Topaloglu S, Ozel H, Saygun O et al. Jejunal intussusception caused by an inflammatory fibroid polyp. *Hepato-gastroenterology* 2003 ; 50 (Suppl 2) : ccliv-cclv.
12. Bays D, Anagnostopoulos GK, Katsaounos E et al. Inflammatory fibroid polyp of the small intestine causing intussusception : a report of two cases. *Dig Dis Sci* 2004 ; 49 : 1677-80.
13. Miyata T, Yamamoto H, Kita H et al. A case of inflammatory fibroid polyp causing small-bowel intussusception in which retrograde double-balloon enteroscopy was useful for the preoperative diagnosis. *Endoscopy* 2004 ; 36 : 344-7.
14. Spengler J, Jeschke A. Jejunal intussusception of an inflammatory fibrinoid polyp. *Ultraschall Med* 2004 ; 25 : 450-2.
15. Vijayaraghavan R, Sujatha Y, Santosh KV, Belagavi CS. Inflammatory fibroid polyp of jejunum causing jejuno-jejunal intussusception. *Indian J Gastroenterol* 2004 ; 23 : 190-2.
16. Vanek J. Gastric submucosal granuloma with eosinophilic infiltration. *Am J Pathol* 1949 ; 25 : 397-411.
17. Helwig EB, Ranier A. Inflammatory fibroid polyps of the stomach. *Surg Gynecol Obstet* 1953 ; 96 : 335-67.
18. Kim YI, Kim WH. Inflammatory fibroid polyps of gastrointestinal tract. Evolution of histologic patterns. *Am J Clin Pathol* 1988 ; 89 : 721-7.
19. Bayle S, Rossi P, Bagneres D et al. Ileum inflammatory fibroid polyp revealed by intussusception. About one familial case. *Rev Med Interne* 2005 ; 26 : 233-7.
20. Dicle O, Erbay G, Haciyanli M, Bora S. Inflammatory fibroid polyp presenting with intestinal invagination : sonographic and correlative imaging findings. *J Clin Ultrasound* 1999 ; 27 : 89-91.
21. Gayer G, Apter S, Hofmann C et al. Intussusception in adults : CT diagnosis. *Clin Radiol* 1998 ; 53 : 53-7.
22. Marcos HB, Semelca RC, Worawattanakui S. Adult intussusception : demonstration by current MR techniques. *Magn Reson Imaging*. 1997 ; 15 : 1095-8.

انغلاف صائمي صائمي بسبب التهاب سليلة ليفية سيرة حالة ومراجعة المنشورات

موجز: التهاب السليلات الليفية تمثل ارتفاعات سليمة تحت مخاطية الجهاز الهضمي. المرضية الحقيقية ليست معروفة والاعراض السريرية متعددة. المعاينة واختبارات المختبر تساعد على وضع التشخيص ولكن الفحص المجهرى للآفة ضروري لتأكيد المعالجة الجذرية تتم باستئصال الآفة. الانغلاف الثانوي للصائم بسبب التهاب السليلة الليفية نادرا ما يذكر في المنشورات التي راجعناها راضفنا اليها حالة مريضنا. وقد بحثنا الخصائص الموافقة للتشريح المرضي وللويائيات وللمشهد السريري ولطرق التشخيص والمعالجة لهذه الحالة السريرية.