

## CASE REPORT

### Synchronous adenomyomas of the ileum in an adult-an exceptional cause of intussusception

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## Introduction

Adenomyoma of the gastrointestinal (GI) tract is an infrequent benign lesion. The pathogenesis of adenomyoma remains unclear. Endoscopic and radiologic evaluations are by no means pathognomonic of this entity [1–3]. Most cases of jejunal and ileal adenomyoma have been reported as single case reports.

Herein, we report an adult case of two synchronous adenomyomas of the ileum leading to intussusceptions as a rare condition and uncommon complication. To the best of our knowledge, this is the first case of synchronous adenomyomas of the ileum.

## Case Report

A 29-year-old male, without medical history, was hospitalized for an occlusive syndrome with nausea vomiting, and abdominal cramps, which had lasted for 3 days. The patient's physical status on admission was as follows: body temperature 36.7°C, blood pressure 120/60 mm Hg, and pulse rate 85 beats/min. Physical examination indicated distension of the abdomen, without tenderness.

Contrast-enhanced computed tomography (CT) (Fig. 1), showed wall thickening, a dilatation of the

#### Key Clinical Message

In this article, we report a case of two synchronous ileal adenomyomas leading to intussusception. This rare occurrence has never been reported in the literature. Our case is noteworthy, because the lesion is rare and should be considered in the differential diagnosis of intussusception in adults.

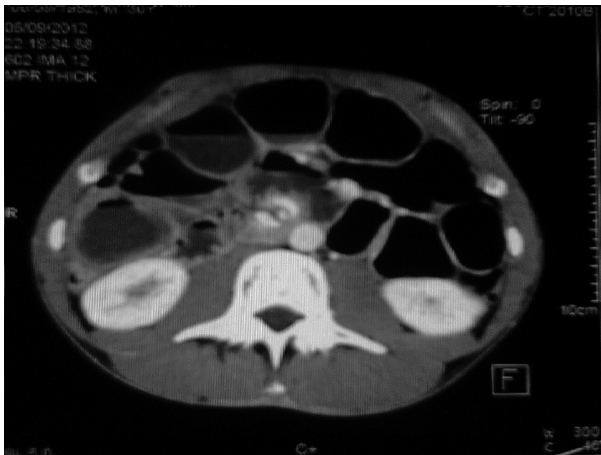
#### Keywords

Adenomyoma, adult, ileum, intussusception.

small bowel and invagination of the small bowel into itself upstream a characteristic sausage sign.

He was diagnosed with small bowel intussusception of the ileum and immediately admitted to our hospital. Open laparotomy was performed and normograde intussusception was observed in the ileum. In addition, two lesions were found in the small intestinal lumen 80 cm proximal to the ileocecal valve. Those lesions have acted as a lead point and hence caused the intussusception. The patient's postoperative course was uneventful.

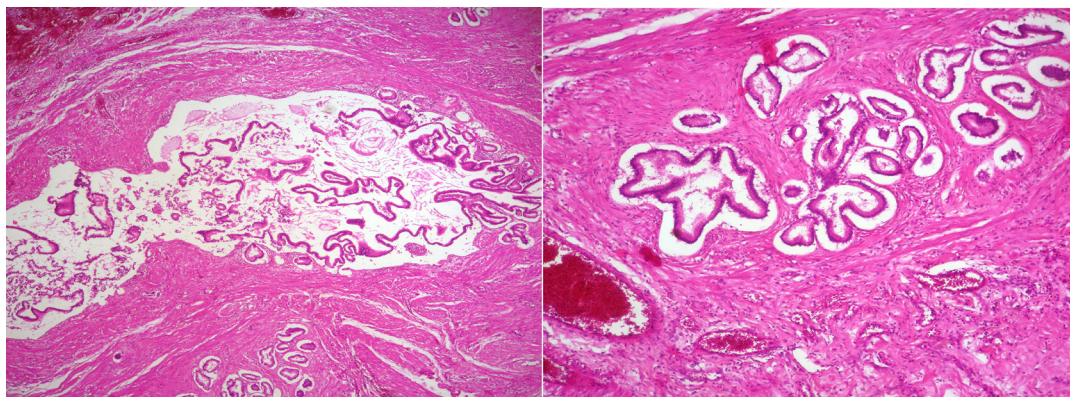
The macroscopic findings of two lesions were consistent with a submucosal tumors, the lesions were sessile and solid, measured, respectively, 2.3 × 2 × 0.8 cm and 1 × 0.5 × 0.5 cm (Fig. 2). The distance between the first and second lesions was 2.5 cm. Microscopically, the two lesions were located in the small intestine submucosa, and were composed of hyperplastic glandular structures of variable sizes and in varying morphologic appearance as well as smooth muscular bundles surrounding the glandular elements. The glandular structures were lined by cytologically benign cuboidal and tall columnar epithelium with basally located nuclei. The large glands were cystically dilated. No ectopic pancreatic acini or islets were found in the lesion (Fig. 3). Immunohistochemical examination was also performed. In the duct epithelial cells, cytokeratin 7 (CK 7)



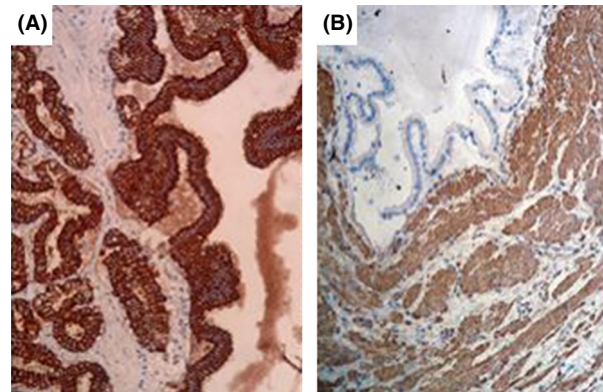
**Figure 1.** CT images showing a dilatation of the small bowel.



**Figure 2.** The macroscopic findings: The resected ileum, which contained 2 lesions covered with normal mucosa.



**Figure 3.** Features of adenomyoma of the ileum: composed of proliferative glandular structures of variable size and irregular muscular bundles surrounding the glandular elements.



**Figure 4.** immunohistochemical profile of adenomyoma of the ileum. (A) CK 7 positivity, (B)  $\alpha$ -smooth muscle actin positivity.

was strongly expressed, whereas CK 20 was not, and the smooth muscle cells surrounding the glandular elements were positive for  $\alpha$ -smooth muscle actin (Fig. 4). The diagnosis of adenomyomas of small bowel was made.

## Discussion

Adenomyoma of the gastrointestinal tract was first described in the stomach by Magnus-Alsleben in 1903 [4]. The actual incidence remains unclear, because very few cases have been reported since then. Most common sites involved in the gastrointestinal tract are the gallbladder and the biliary tree [3]. Adenomyoma of the small intestine is exceedingly rare.

To date, only 26 well-documented cases of adenomyoma involving the small intestine have been reported in the literature (Table 1). The patient age ranges from 2 days to 82 years (mean 25 years), including 15 pediatric patients and 11 adult patients. This distribution was clearly revealed to be bimodal and consisted of two

**Table 1.** Review of the literature: Clinicopathological findings of adenomyoma of the small intestine [6, 7].

First author	Age/Sex	Symptoms	Size (cm)	Location
Clarke (1940)	64 y/M	Incidental	1	Jejunum
Schwartz (1958)	8 m/M	Intussusception	2	Ileum
Rosenmann (1980)	2 d/F	Intestinal obstruction	NA	Ileum
Gal (1986)	82 y/F	Intussusception	2	Ileum
Kim (1990)	7 y/M	Intussusception	4	Ileum
Gal (1991)	9 m/M	Intussusception	1.2	Ileum
	79 y/M	Incidental	0.6	Ileum
Lamki (1993)	1 y/M	Intussusception	1.5	Ileum
Chan (1994)	5 m/F	Intussusception	0.8	Ileum
	3 y/M	Incidental	0.8	Ileum
Serour (1994)	3 y/M	Intussusception	2	Ileum
Gonzalvez (1995)	2 y/M	Intussusception	2	Ileum
Tanaka (1996)	24 y/M	Melena	1.2	Ileum
Hizawa (1996)	23 y/F	Incidental	NA	Jejunum
Yamagami (1997)	4 m/M	Intussusception	NA	Ileum
Yao (2000)	1 y/M	Intussusception	NA	Meckel diverticulum
Ueyama (2001)	52 y/F	Incidental	NA	Ileum
Lee (2002)	18 y/M	Intussusception	4.5	Jejunum
Mouravas (2003)	1 y/M	Intussusception	1.5	Ileum
Park (2003)	7 m/M	Intussusception	1.2	Ileum
	63 y/M	Incidental	1.3	Jejunum
Takahashi (2006)	75 y/M	Incidental	0.9	Ileum
Yu (2008)	74 y/F	Tarry stool	1.5	Jejunum
Qing (2009)	61 y/F	Incidental	1.5	Jejunum
Tomibayashi (2011)	81 y/F	Intestinal obstruction	2	Jejunum
Takeda (2011)	68 y/M	Intussusception	NA	Ileum

y, years; m, months; d, days; NA, not available.

peaks: <30 years old and >50 years old. The male-to-female ratio is approximately 2:1. The lesion occurs 2 to 3 times more frequently in the ileum than in the jejunum. One lesion was found in a Meckel diverticulum [5].

Symptoms of adenomyoma of the GI tract depend on the location of the lesion and patient age. Jejunal and ileal adenomyoma of pediatric patients usually present with intussusception (11 cases), but one reported case presented with intestinal obstruction [8]. In adult patients, intussusception is an infrequent complication; most reported adult cases are incidental findings although these lesions may occasionally cause gastrointestinal bleeding or abdominal pain [3].

Clinically, our case presented with intussusceptions caused by adenomyomas of the ileum. To our knowledge, our case is the first report of two synchronous adenomyomas of small intestine, and the third report of adenomyomas causing intestinal intussusceptions in an adult [6, 7].

Grossly, adenomyoma of the GI tract is an intramural nodule covered by mucosa and it protrudes into the lumen. The diameter of adenomyoma range from 0.6 cm to 4.5 cm. Histologically, adenomyoma of the small intestine mainly occupies the submucosa, and often extends into the muscularis propria [9]. The lesion consists of glandular structures of various sizes and interlacing smooth muscle bundles surrounding the glandular elements. Cystically dilated glands are usually observed. The glandular structures are lined by cuboidal to tall columnar epithelium with basally oriented nuclei. Goblet cells and Paneth cells are occasionally observed. Those glands are surrounded by interlacing smooth muscle bundles. Myofibroblasts and fibroblasts may also proliferate [10, 11]. Both the epithelial and smooth muscle cells lack nuclear atypia. Pancreatic acini and islet tissue are not present [12]. Pathological diagnosis by biopsy specimen is usually difficult, partly because the lesion mainly occupies the submucosa. Immunohistochemically, the glandular element of adenomyoma of the small intestine is positive for cytokeratine (CK) 7 and CA19-9, negative for CK 20 and mucin antigen 2 (MUC 2) [3, 13], whereas normal intestinal epithelial cells around the lesion are negative for CK 7 and positive for CK 20. The glandular epithelial cells of the lesion do not express CDX-2, a marker of intestinal mucosal epithelium [3]. The smooth muscle cells surrounding the glandular elements are positive for  $\alpha$ -smooth muscle actin and desmin [12].

Differential diagnosis include pneumatosis cystoides intestinalis, adenocarcinoma, and hamartomatous polyp in Peutz-Jeghers syndrome. Cysts of pneumatosis cystoides intestinalis contain gas and are lined by multinucleated giant cells. The characteristics of adenomyoma that differentiate it from adenocarcinoma include the absence of cellular atypia and the presence of smooth muscle bundles surrounding the glands and cysts. In Peutz-Jeghers syndrome, the essential feature is branching cores of muscular fibers derived from the muscularis mucosae and covered by normal mucosa, whereas adenomyoma is located in submucosa and/or muscularis propria [9].

The optimal management of adult intussusception remains controversial. Most of the debate focuses on the issue of primary en bloc resection versus initial reduction, followed by a more limited resection [14]. Proponents of primary resection cite the high incidence of underlying malignancy, which mandates en bloc resection. Also the reduction in an intussusception includes the increased risk of anastomotic complications (the bowel wall may be weakened during manipulation) and the potential for bowel perforation [15].

In summary, adenomyoma of the small intestine is a rare, benign condition that might occur at any age. The prognosis for its treatment is very good.

## Consent

Written informed consent was obtained from the patient for publication of this Case Report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

## Conflict of Interest

None declared.

## References

1. Bedirli, A., T. E. Patiroglu, E. M. Sozuer, and O. Sakrak. 2002. Periampullary adenomyoma: report of two cases. *Surg. Today* 32:1016–1018.
2. Kwon, T. H., H. Park do, K. Y. Shim, H. D. Cho, J. H. Park, S. H. Lee, et al. 2007. Ampullary adenomyoma presenting as acute recurrent pancreatitis. *World J. Gastroenterol.* 13:2892–2894.
3. Qing, X., B. A. Petrie, V. Buslon, and S. French. 2009. Adenomyoma of the jejunum. *Exp. Mol. Pathol.* 86:127–130.
4. Takahashi, Y., and T. Fukusato. 2011. Adenomyoma of the small intestine. *World J. Gastrointest. Pathophysiol.* 2:88–92.
5. Higashi, M., M. Goto, M. Saitou, T. Shimizu, K. Rousseau, S. K. Batra, et al. 2010. Immunohistochemical study of mucin expression in periampullary adenomyoma. *J. Hepatobiliary Pancreat. Sci.* 17:275–283.
6. Tomibayashi, A., S. Sasaki, H. Nakayama, S. Yoneyama, T. Ishii, and T. Watanabe. 2011. Adenomyoma of the small intestine in an adult: report of a case. *Surg. Today* 41:1101–1105.
7. Takeda, M., T. Shoji, M. Yamazaki, Y. Higashi, and H. Maruo. 2011. Adenomyoma of the ileum leading to intussusception. *Case Rep. Gastroenterol.* 5:602–609.
8. Van Helden, S. H., G. Jutten, H. Van Hoey, and A. M. Dierick. 1998. Jejunal hamartoma as a rare cause of gastrointestinal haemorrhage. *Histopathology* 32:574–575.
9. Yao, J. L., H. Zhou, K. Roche, B. S. Bangaru, H. Ginsburg, and M. A. Greco. 2000. Adenomyoma arising in a meckel diverticulum: case report and review of the literature. *Pediatr. Dev. Pathol.* 3:497–500.
10. Rosenmann, E., C. Maayan, and O. Lernau. 1980. Leiomyomatous hamartosis with congenital jejunoileal atresia. *Isr. J. Med. Sci.* 16:775–779.
11. Handra-Luca, A., B. Terris, A. Couvelard, H. Bonte, and J. F. Flejou. 2003. Adenomyoma and adenomyomatous hyperplasia of the Vaterian system: clinical, pathological, and new immunohistochemical features of 13 cases. *Mod. Pathol.* 16:530–536.
12. Mouravas, V., G. Koutsoumis, J. Patoulias, I. Kostopoulos, R. Kottakidou, K. Kallergis, et al. 2003. Adenomyoma of the small intestine in children: a rare cause of intussusception: a case report. *Turk. J. Pediatr.* 45:345–347.
13. Takahashi, Y., J. Fukushima, T. Fukusato, and S. Mori. 2006. Adenomyoma with goblet and Paneth cells of the ileum. *Pathol. Res. Pract.* 202:549–553.
14. Yakan, S., C. Caliskan, O. Makay, A. G. Denecli, and M. A. Korkut. 2009. Intussusception in adults: clinical characteristics, diagnosis and operative strategies. *World J. Gastroenterol.* 15:1985–1989.
15. Yamada, H., T. Morita, M. Fujita, Y. Miyasaka, N. Senmaru, and T. Oshikiri. 2007. Adult intussusception due to enteric neoplasms. *Dig. Dis. Sci.* 52:764–766.