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Iron Deficiency Anemia Masking Adenomyoma of the Jejunum

Intestinal adenomyomas, also known as myoepithelialhamartomas, are benign non-neoplastic tumor-like lesions characterized by glandular structures lined by cuboidal or tall columnar epithelium and surrounded by smooth muscle bundles. Reported cases of adenomyoma of the gastrointestinal tract have been largely confined to the stomach and duodenum, with lesions distal to the duodenum occurring less frequently. Here we report a case of adenomyoma of the jejunum that presented with anemia, along with a review of the literature.

Case Report

A 62 year old woman with history of heartburn and cholecystectomy presented for bronchial thermoplasty for treatment of asthma. She was incidentally found to have iron deficiency anemia. Anemia was evaluated with upper and lower endoscopy, and findings were negative for suspicious pathology or source of bleeding. Capsule video endoscopy revealed a pedunculated 10 mm polyp in the distal jejunum/proximal ileum. Endoscopic removal of the polyp was completed by double balloon enteroscopy. Grossly, the lesion was polypoid shaped measuring 0.9x0.8x0.6 cm **(Figure 1)**. Microscopically, it was a well-circumscribed submucosal lesion composed of variably sized glandular structures surrounded by smooth muscle bundles. The glandular structures showed pyloric morphology and were lined by cuboidal or columnar epithelium with interspersed goblet cells and occasional Paneth cells **(Figure 2)**. Immune-stains were performed with appropriate controls. The lesional epithelial component was strongly and diffusely positive for CK7 and CK19 and weakly positive for CDX2, but negative for CK20 **(Figure 3)**. The smooth muscle component was positive for desmin and alpha-SMA. Ki-67/MIB-1, highlighting rare lymphocytes in the lesion and basal cells of the small intestinal mucosal epithelium **(Figure 4)**. All pathologic features supported a final diagnosis of adenomyoma. The patient's postoperative course was complicated by transient abdominal pain and bloating after the procedure. Abdominal X-ray was negative for free intra-abdominal air or perforation. Her pain resolved overnight and she was discharged the following morning. Upon follow-up one year later, she continues to do well, and the anemia has resolved.

Discussion

Myoepithelialhamartoma of the gastrointestinal tract was first described by Clarkein 1940.[5] While the incidence of small intestine adenomyoma is relatively rare, the actual incidence is unclear because very few cases

have been reported. Review of the literature reveals 34 reported cases in the small intestine,[3,5-13,15-21,23-34] including the duodenum, ampulla of Vater, and ileum, with 11 well-documented cases reported in the jejunum **(Table 1)**.[3,5,9-10,16-17, 20-21,29,31,34] Of these cases, the patients were between the ages of three to eighty one years old, with six women and five men. While they are typically asymptomatic, symptoms of intestinal adenomyoma vary depending on the size, location of the lesion, and patient age. In the periampullary region, adenomyoma typically presents with biliary obstruction or abdominal pain, with reported cases of acute pancreatitis.[14,22] Lesions in the ileum and jejunum have presented with intussusception and intestinal obstruction,[29] but many reported cases have been incidentally detected. In the small intestine, intussusception is its most common complication, reported in 19 cases.[1-3,6-8,11-13,15-20,24-25,27,32-33] Anemia has been reported in 11.7% of the cases described (4 cases).

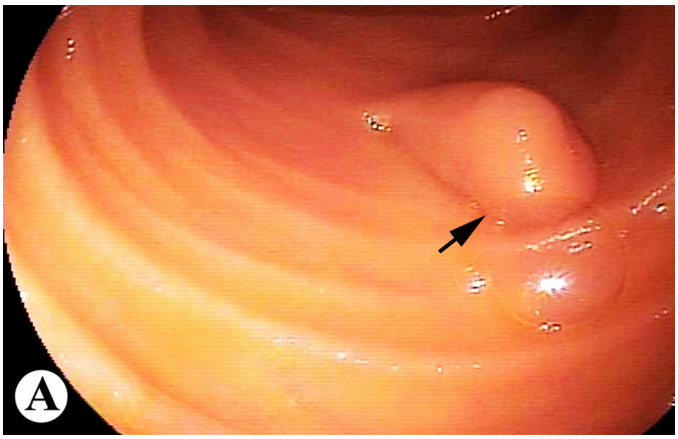

Figure 1: Polypoid shaped mass (0.9 x 0.8 x 0.6 cm) identified and removed by double balloon enteroscopy.

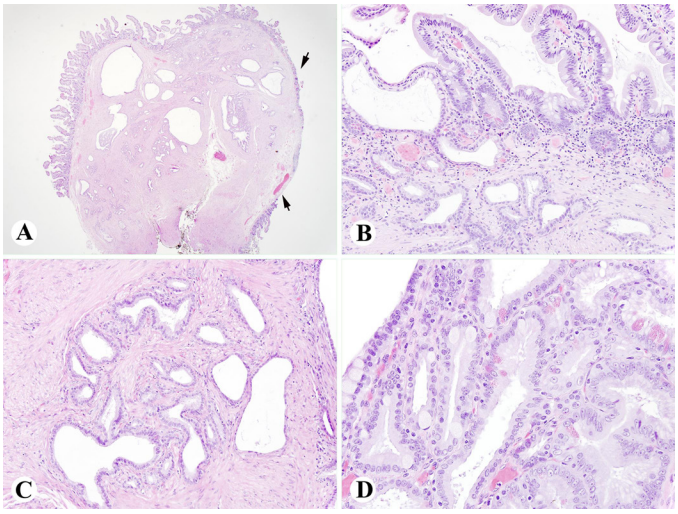

Figure 2: Histology of the jejunaladenomyoma. (A-D): On H&E stain, the submucosal mass showing dilatation of glandular architecture with focal mucosal erosion (A, arrows, 20x), extension into the lamina propria (B, 200x), interlacing with smooth muscle (C, 200x) and cytological bland foveolar gland metaplasia with focal intestinal metaplasia (D, 400x).

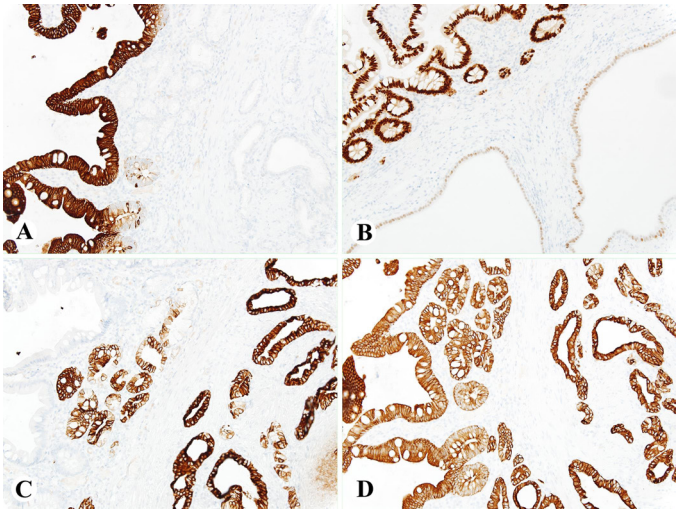

Figure 3: Immunostains of the jejunaladenomyoma showing (A): CK20 is positive in the normal surface epithelium of the jejunum, negative in the adenomyoma; (B): CDX2 is strongly positive in the normal jejunal mucosa and weekly positive in the adenomyoma; (C): CK7 is negative in the normal surface epithelium of the jejunum, positive in the adenomyoma; and (D): CK19 is positive in both normal jejunal mucosa and adenomyoma. (A-D, 200x).

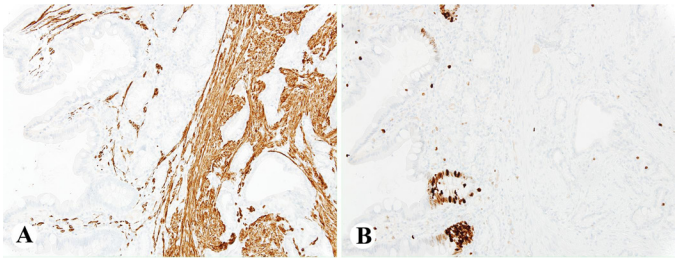

Figure 4(A): Desminimmunostain highlighting the interlacing smooth muscle; (B): Ki-67 immunostain showing a low proliferation index (<2%) of the adenomyoma (A-B, 200x).

Table 1: Adenomyoma of the Jejunum.

Author	Age (years)	Sex	Associated Symptoms	Size of lesion	Presence of ectopic pancreatic tissue	Tumor Markers
Chan et al, 1994	3	Male	Abdominal pain, distension, vomiting	8 mm	No	NA
Clarke et al, 1940	64	Male		1x1x1 cm		
Gourtsoyiannis et al, 1993	51	Female	Anemia	NA	NA	NA
Hizawa et al, 1996	23	Female	Gardner's syndrome	NA	NA	NA
Lee et al, 2002	18	Male	Intussusception; abdominal pain and vomiting	4.5x3x2.5 cm	No	NA
Lo Bello Gemma et al, 2003	13	Female	Intussusception, volvulus			
Park et al, 2003	63	Male	Constipation	1.3x0.8x08 cm	No	NA
Qing et al, 2009	61	Female	Abdominal pain, weight loss, diarrhea	1.3x1.1x1.1 cm	No	AE1/AE3 (+) CK-7 (+) CK-20 (-) CDX-2 (-) CA19-9 (+) in large glands
Tomibayashi et al, 2011	81	Female	Intestinal obstruction, anemia	2.0x1.5 cm	Yes	CK-7 (+) CK-20 (-) Some expression of CA19-9
Van Helden et al, 1998	65	Male	Anemia, melena	5-15 mm	NA	NA
Yu et al, 2008	74	Female	Anemia, melena	1.5 cm	No	NA
Current Case	62	Female	Anemia	0.9x0.8x0.6 cm	No	CK7 (+) CK19 (+) CK20 (-) CDX2 weak expression

Most cases were surgically removed (26 cases), which may be attributed to a high number of laparotomies due to the adenomymoma presenting with intussusception, and a small percentage were removed endoscopically (3 cases). Microscopically, intestinaladenomyomas are submucosal tumors characterized by glandular structures lined by cuboidal or tall columnar epithelium and surrounded by bundles of smooth muscle.[21] The differential diagnosis of this pathological presentation includes: enteritis cysticaprofunda, hamartomatous polyps in Peutz-Jeghers syndrome, adenocarcinoma, and pneumatosiscystoides. The pathogenesis of adenomyoma remains unclear, but the most widely accepted hypothesis is that it represents either a form of myoepithelialhamartoma or pancreatic heterotopia.[26] The term 'hamartoma' refers to a focal but excessive overgrowth of cells indigenous to the particular site, while the term 'heterotopia' refers to microscopically normal cells that are present in an abnormal location. Clarke suggested that the classification of 'adenomyoma' be used only for lesions with exocrine-type ducts without

ectopic pancreatic acini or islets surrounded by smooth muscle, and that the term 'pancreatic heterotopia' should refer to lesions with ectopic pancreatic acini or islets.[5] In our case, we performed immunohistochemical testing in an attempt to increase our understanding of the origins of the adenomyoma. We detected the expression of CK7 and CK19, weak expression of CDX2, and the absence of CK20 expression. In general, CK7 is distributed in the pancreatic duct epithelium but is essentially absent in GI epithelium. CK20 is distributed in the GI epithelium but is absent in pancreatic duct epithelium.[4] Thus, theimmunohistochemical features of the lesion are similar to that of the pancreatic duct epithelium and appear to support the heterotopic pancreas hypothesis. However, most reported cases of adenomyoma, including our present case, do not contain pancreatic tissue. In fact, the glandular structures showed pyloric morphology and were lined by cuboidal or columnar epithelium with interspersed goblet cells and occasional Paneth cells, suggesting a pyloric origin. Qing et al.(2009) and Tomibayashi et al. (2011) also reported cases of adenomyoma of the jejunum with goblet cells. Goblet cells were also found in a case of adenomyoma in a Meckel's diverticulum.[33]

Conclusion

Adenomyoma of the small intestine is an extremely rare tumor largely unknown to most clinicians. While typically asymptomatic, symptoms of intestinal adenomyoma vary depending on size, location, and patient age. In our patient this lesion was likely the source of chronic GI bleeding manifesting as anemia. The prognosis of adenomyoma is excellent, with no evidence of recurrence or metastatic transformation in reported cases. Treatment of asymptomatic or bleeding adenomyomas is simple resection. Nevertheless, the pathogenesis of small bowel adenomyoma is not well understood due to its rarity, making further studies necessary.

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Dengue Transmission from Donor to Recipient After Living Donor Liver Transplant

Organ transplantation has been associated with a small but worrying risk of transmission of disease from donor to recipient. Initially reported for viral and bacterial infections following renal transplants[1], similar reports are now mushrooming world over following liver transplantation.[2] Whereas the common culprits, namely HIV, HBV, HCV, etc have been covered by screening