



Case report

Small intestinal myxoma presented with bowel obstruction, a rare case report

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ABSTRACT

Introduction: Myxomas are rare benign mesenchymal neoplasms and mostly occur in cardiac atrium and with lower prevalence, appear in sinonasal tract, gnathic bone, skin and joints. Benign primary tumors of the small intestine are quite unusual accounting about 3% of all the gastrointestinal tract neoplasms.

Presentation of case: In this report, we present a rare case of small intestinal myxoma in a 43-year-old man complicated by ileoileal intussusception causing bowel obstruction. The patient underwent segmental resection of small bowel and the histopathologic evaluation confirmed the diagnosis of small intestinal myxoma.

Discussion: Among benign primary neoplasms of small bowel, Solitary intestinal myxomas are extremely rare mostly locate in submucosal layer of gastric, small bowel and rarely in cecum. To our knowledge only 9 cases of small bowel myxoma is reported in the literature so far.

Conclusion: This case report aware surgeons and pathologists about rare occurrence of intestinal myxomas as a differential diagnosis of intussusception.

1. Introduction

Myxomas are rare benign mesenchymal neoplasms and mostly occur in cardiac atrium and with lower prevalence, appear in sinonasal tract, gnathic bone, skin and joints [1]. Benign primary tumors of the small intestine are quite unusual accounting about 3% of all the gastrointestinal tract neoplasms [2]. Among these benign neoplasms, Solitary intestinal myxomas are extremely rare mostly locate in submucosal layer of gastric, small bowel and rarely in cecum [3]. These tumors cause disabling pain, vomiting, intestinal bleeding and obstruction and patients refer with difficulties in passing stool [4]. To our knowledge only 9 cases of small bowel myxoma is reported in the literature so far. In this report, we present the tenth case of small intestinal myxoma in a 43-year-old man complicated by ileoileal intussusception causing bowel obstruction.

2. Case presentation

A 43-year-old man referred to our center with severe abdominal pain and vomiting. The patient started complaining of intense and colicky pain in the periumbilical area for one week before consultation. The pain was intermittent, without any irradiation and no remitting or

exacerbating factor. Two days after the onset of the pain, constipation had started and a complete stoppage of stool passage occurred with vomiting. No previous history of any medical condition was reported. Drug and family history was unremarkable. Upon arrival, the patient was anxious, and a physical exam revealed a conserved general state, stable vitals, a periumbilical tenderness, with no abdominal distention and no signs of dehydration. A plain abdominal radiograph revealed distended small bowel gathering in the left upper quadrant with air fluid levels (Fig. 1). An abdominal ultrasound showed significantly dilated, fluid-filled and hyperperistaltic bowel loops. CT Scan imaging was performed and eccentric wall thickening and obstruction in the proximal ileum was revealed (Fig. 1). Complete blood count showed moderate leukocytosis and liver function test revealed hyperbilirubinemia. Laparotomy surgery was done and found a distal small bowel obstruction complicated by ileoileal intussusception with a large encapsulated polypoid yellowish mass measuring 4 cm without any adhesion and there was no lymph node. Segmental resection of 16 cm of small bowel was done and intestinal end-to-end anastomosis was also performed. The specimen was sent to pathology laboratory for histologic examination. In gross examination of 16 cm intestinal specimen a yellow polypoid mass protruded within the bowel's lumen measuring 4x3x2 cm was identified in central part of specimen 6 cm from closest resected

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margin. Histopathologic evaluation showed neoplastic myxoid lesion in submucosa made of stellate and spindle shaped cells with no sign of atypia or mitotic activity, which were surrounded by abundant, myxomatous stroma. There were also scattered eosinophils and lymphoplasmacytes within the tissue (Fig. 2). Adjacent sections of the small intestine and the mesentery indicated normal structures. Immunohistochemical (IHC) studies showed that neoplastic cells were negative for S100, CD117, Cytokeratin and ALK markers, but they were strongly positive for Vimentin which proved their mesenchymal origin and SMA. Endothelial marker CD31 stained the tumor's vessels. Ki-67 was also low. A final diagnosis of benign intestinal myxoma with clear resected margins was made upon all the previously mentioned histologic and IHC findings. Postoperatively, the patient had a stable and uneventful clinical course. She was discharged 8 days after admission with normal gastrointestinal function. One month later, echocardiographic evaluation was conducted which revealed normal heart activity without any evidence of cardiac myxoma. Reevaluation at six months and 1 year follow up with abdominal and thoracic C.T. scanning did not show any metastatic lesions, enlarged lymph-nodes, recurrent lesions or intracardiac masses.

3. Discussion

Small bowel obstruction is a major cause of mortality and morbidity, accounting for 15% of all emergency admissions for abdominal pain. Bowel obstruction is rarely caused by intussusception (about 1% of all bowel obstructions). Intussusception in adults occurs due to a pathological condition as a trigger point. Benign tumors of the small bowel account for a small percentage of all gastrointestinal tumors. These tumors can cause intestinal obstruction, hemorrhage, tumor rupture, or malignant degeneration. Myxoma is originated from mesenchymal tissue and is characterized by the loose textured slimy tissue of stellate cells, reticulin fibers, and mucoid substance. It is mainly found in the skin, soft tissue, and heart [5]. Myxomas are rarely reported in the small intestine. To the best of our knowledge, this is the tenth case of intestinal myxoma reported in the literature.

All of the reported cases in the literature including our case are summarized in Table 1. The median (range) age of the patients was 44.6 (20–68) years. The female to male ratio was 7:3 (2.3). Nine out of ten patients (90%) had ileal myxoma, while one (10%) had a jejunal myxoma. All patients presented as mechanical intestinal obstruction sign & symptoms. Two patients had ileocecal intussusception; both were males. All cases of ileal myxoma presented with intussusception, while the jejunal one had small bowel obstruction without intussusception. All the ten cases needed surgical bowel resection with primary anastomosis, all survived and had a favorable outcome. There was one report of synchronous intestinal and cardiac myxomas in these reported cases [6].

Myxomas should be included in the differential diagnosis of intestinal tumors in adults presenting with intussusception. Patients presented with intestinal myxoma should undergo echocardiography, CT, or MRI scanning of the heart to evaluate the presence of cardiac myxoma. Carney complex is an inherited, autosomal dominant disorder presented by multiple tumors, including atrial and extracardiac myxomas, schwannomas, endocrine tumors, and various pigmentation abnormalities [1]. True myxomas are benign and tend to infiltrate surrounding tissues, and do not metastasize. Therefore, treatment of intestinal myxomas should comprise of simple, wide resection of the intestinal segment with primary anastomosis and close follow-up to detect recurrence of the tumor [5].

Small intestine myxoma is an extremely rare disease but it should be included in the differential diagnosis of small bowel obstruction mainly in the presence of intussusception in young patients and surgeons and pathologists should be aware of occurrence of this rare entity in clinical practice.

The work has been reported in line with the SCARE 2020 criteria [13].

Sources of funding

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Ethical approval

This case report is exempt from ethical approval in our institution.

Consent

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

Author contribution

Maryam Maghbool, MD: study concept and design, data interpretation, manuscript writing.

Babak Samizadeh, MD: data interpretation, manuscript writing.

Research registration

This does not apply as it is a case report of a patient who has given written consent and has been de-identified. It is therefore not prospective research involving human participants.

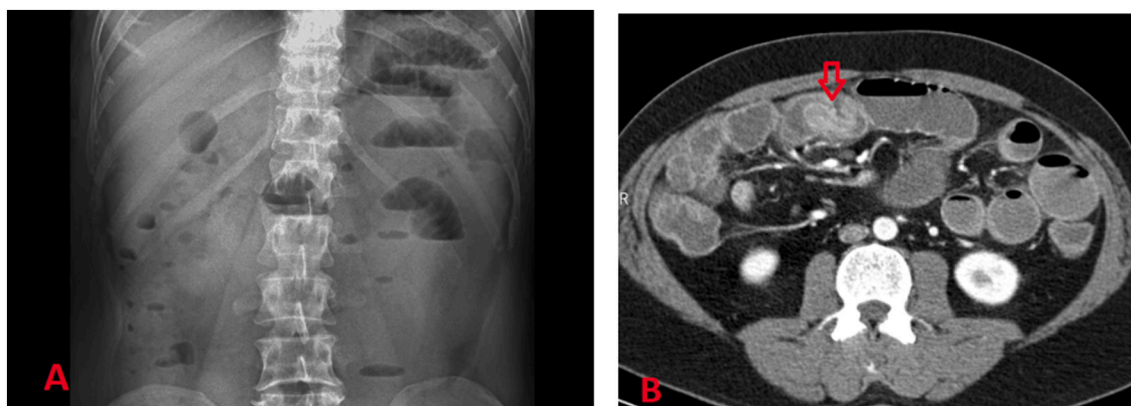


Fig. 1. A, plain abdominal X-ray showing multiple air fluid level; B, abdominopelvic CT scan showed eccentric wall thickening and obstruction in the proximal ileum (red arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

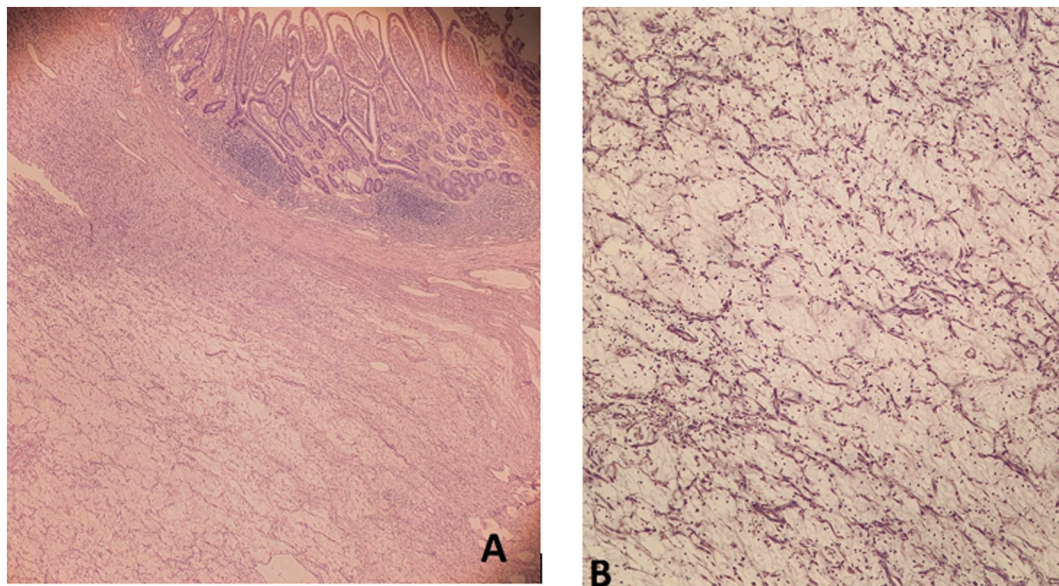


Fig. 2. A, Small intestine mucosa with submucosal myxoid lesion (H&E, $\times 40$); B, stellate and spindle shaped cells with no sign of atypia or mitotic activity surrounded by abundant, myxomatous stroma with scattered eosinophils and lymphoplasmacytes within the tissue (H&E, $\times 200$).

Table 1
Summary of intestinal myxoma reported cases in the literature.

Case	Reference	Age (yrs.)	Sex	Clinical presentation	Site	Surgical intervention
1	Bourgett et al. [7]	20	Female	Abdominal pain, constipation	Ileum	Segmental resection
2	Brachetto et al. [8]	54	Male	Abdominal pain	Ileum	Segmental resection
3	Larimore et al. [9]	48	Female	Abdominal pain, nausea, vomiting	Ileum	Segmental resection
4	Sullivan et al. [10]	44	Female	Abdominal pain, distention, nausea	Jejunum	Segmental resection
5	Stout et al. [11]	68	Female	Abdominal pain, distention, nausea	Ileum	Segmental resection
6	Weinberg et al. [12]	40	Female	Abdominal pain, nausea, vomiting	Ileum	Segmental resection
7	Wang et al. [6]	47	Female	Abdominal pain, constipation	Ileum	Segmental resection
8	Varsamis et al. [1]	44	Female	Abdominal pain, distention, nausea	Ileum	Segmental resection
9	Mousa et al. [5]	38	Male	Abdominal pain, distention, nausea, bloody diarrhea	Ileum	Right hemicolectomy
10	Current case	43	Male	Abdominal pain, nausea, vomiting, constipation	Ileum	Segmental resection

Guarantor

Dr. Maryam Maghbool.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Declaration of competing interest

None.

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