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Myxoma of the small intestine complicated by ileo-ileal intussusception: Report of an extremely rare case

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ABSTRACT

INTRODUCTION: Myxomas of the small intestine are extremely rare types of primary bowel neoplasms. Their presence can trigger intestinal intussusception in the adults. We present the eighth case of intestinal myxoma reported in the English literature.
PRESENTATION OF CASE: Our patient is a 44-year-old Caucasian female who presented with clinical and imaging findings of intestinal intussusception. Laparotomy revealed ileo-ileal intussusception caused by an intramural mass of the middle-ileum which was resected. Histological and immunohistochemical studies pointed to the diagnosis of benign intestinal myxoma, while imaging studies of the heart excluded a synchronous cardiac myxoma. Twenty months after surgery she remains disease-free.
DISCUSSION: The myxoma is a benign, true neoplasm which resembles primitive mesenchyme. It occurs predominantly in the heart and is also found in several soft tissues and bones. Myxomas seem to grow at different rates of speed, they infiltrate adjacent structures and they do not metastasize, apart from cardiac variants. Intestinal myxomas share some clinical characteristics which are emphasized.
CONCLUSION: Myxomas of the small intestine should be included in the differential diagnosis of ileal tumors in middle-aged women manifesting as intestinal intussusception. Treatment should include wide resection of the affected intestinal segment with primary anastomosis. A close follow-up control of the patients along with cardiac imaging evaluation is recommended postoperatively, in order to detect and treat any possible recurrence of the tumor or a synchronous cardiac myxoma.

1. Introduction

Benign primary tumors of the small intestine are quite unusual, comprising approximately 3% of all the gastrointestinal tract neoplasms.[1,2] They can either remain asymptomatic or manifest as complications, including intestinal bleeding, obstruction or perforation.[3,4] Intussusception is an uncommon form of bowel obstruction, especially in adults. Adult to child intussusception ratio is about 1/20. Moreover, adult intussusception is secondary in 90% of cases, deriving from a pathologic lesion that acts as a "lead point". Such lesions could be adhesions, polyps, Meckel's or colonic diverticula, benign or malignant intestinal or appendiceal tumors.[5–7]

Myxomas of the small intestine are extremely rare. Stout reported five cases of intestinal myxomas in a review of 1948, only one of which was encountered by him.[8] In 1956 Weinberg published a case of intussusception due to seven segmental myxomas of the small intestine.[9] These six cases were also included in the review of Wilson et al. in 1974 referring to benign small bowel tumors.[3] Finally, Wang and Sharkey reported in 2003 the case of a woman with the simultaneous presence of intestinal and left atrial myxoma.[10]

We present the case of a patient we treated who displayed a myxoma of the small intestine with ileo-ileal intussusception, focusing on the diagnostic approach and management of this rare condition. To the best of our knowledge, this is the eighth case of benign small bowel myxoma reported in the English literature.

2. Presentation of case

A 44-year-old Caucasian female patient referred to our emergency room with the "typical–atypical" clinical findings of intestinal intussusception. She complained of paroxysmal, crampy pain of the lower-right abdomen that had started 48 h ago, flatulence, nausea and multiple episodes of emesis. She had not passed

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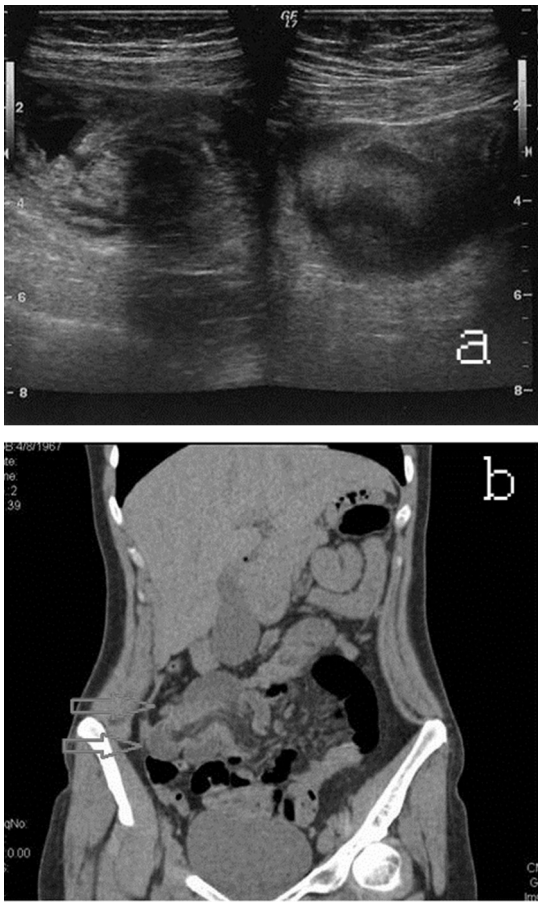

Fig. 1. (a) Abdominal ultrasonography showing a "target-shaped" mass. (b) Abdominal C.T. scanning manifesting ileo-ileal intussusception (arrows).

any stool or air for the last 48 h. Physical examination revealed a soft abdomen with mild distension and no signs of rebound tenderness. Nevertheless, a mass could be palpated on the lower-right abdominal quadrant and metallic intestinal sounds were present in auscultation of this region. Rectal examination was negative for blood.

Her medical history included goiter with normal thyroid function. Laboratory values and temperature were within normal limits, apart from leukocytosis with a "left shift" (W.B.C. = 15,100/mm^3, neut. = 80.9%, lymp. = 13.7%). Abdominal X-ray demonstrated "air-fluid" levels of the small intestine indicative of mechanical bowel obstruction and was followed by ultrasonographic examination of the abdomen which showed a typical "target-shaped" mass adjacent to the caecum (Fig. 1a). Finally, abdominal C.T. scanning pointed to the diagnosis of ileo-ileal intussusception with dilated intestinal loops (Fig. 1b).

Emergency laparotomy through a lower-right paramedian incision was performed soon after admission. Intraoperatively, antegrade small bowel intussusception extending for about 5 cm was found and reduced carefully with firm traction of the involved bowel's wall (Fig. 2a). An intramural, smooth mass of the ileum's antimesenteric wall, lying at about 50 cm from the ileocaecal valve was identified (Fig. 2b). A thorough investigation of the peritoneal cavity followed, which revealed no other pathological lesions or signs of metastatic disease. We resected a 12 cm segment of middle-ileum containing the mass and performed a side-to-side isoperistaltic anastomosis. Free intraperitoneal liquid was also collected for microbiological culture and cytological examination, which were both negative for infection or malignancy.

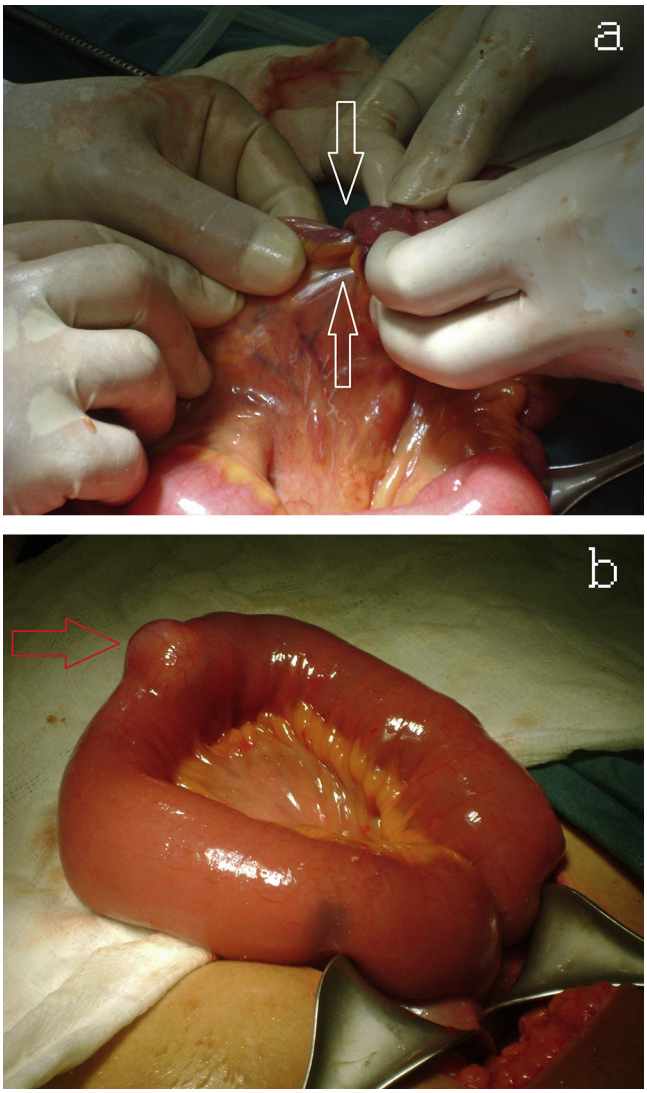

Fig. 2. Intraoperative photos. (a) Antegrade ileo-ileal intussusception (arrows). (b) Intramural, smooth tumor in the antimesenteric wall of ileum, identified after reseting of the intussusception (arrow).

Gross pathologic examination of our 12 cm intestinal specimen indicated an intramural tumor made of elastic, gelatinous, whitish tissue, which measured 2.5 cm in diameter and protruded firmly within the bowel's lumen (Fig. 3a). It was located 3.5 cm from the closest resection margin. Histological examination of multiple cut surfaces showed a neoplastic myxoid lesion which sprang from the submucosa (Fig. 3b). Superficially, the tumor was extending at sites up to the mucosa, causing ulceration. At its deepest margin it was infiltrating focally the muscular layer and the intestinal serosa. Adjacent sections of the small intestine and the mesentery indicated normal structures and no vascular neoplastic emboli.

Microscopically, the tumor was made of stellate and spindle-shaped cells with no elements of atypia or mitotic activity, which were surrounded by an abundant, Alcian Blue positively stained myxomatous stroma (Fig. 3c). There were also scattered eosinophils and lymphoplasmatocytes circumferentially. Immunohistochemical studies showed that neoplastic cells were negative for S100, NSE, GFAP, Synaptophisin, CD117, Actin, Desmin and BCL-2 markers, but they were strongly positive for Vimentin which proved their mesenchymal origin (Fig. 3d). Endothelial marker CD34 stained the tumor's vessels. A final diagnosis of benign intestinal myxoma with

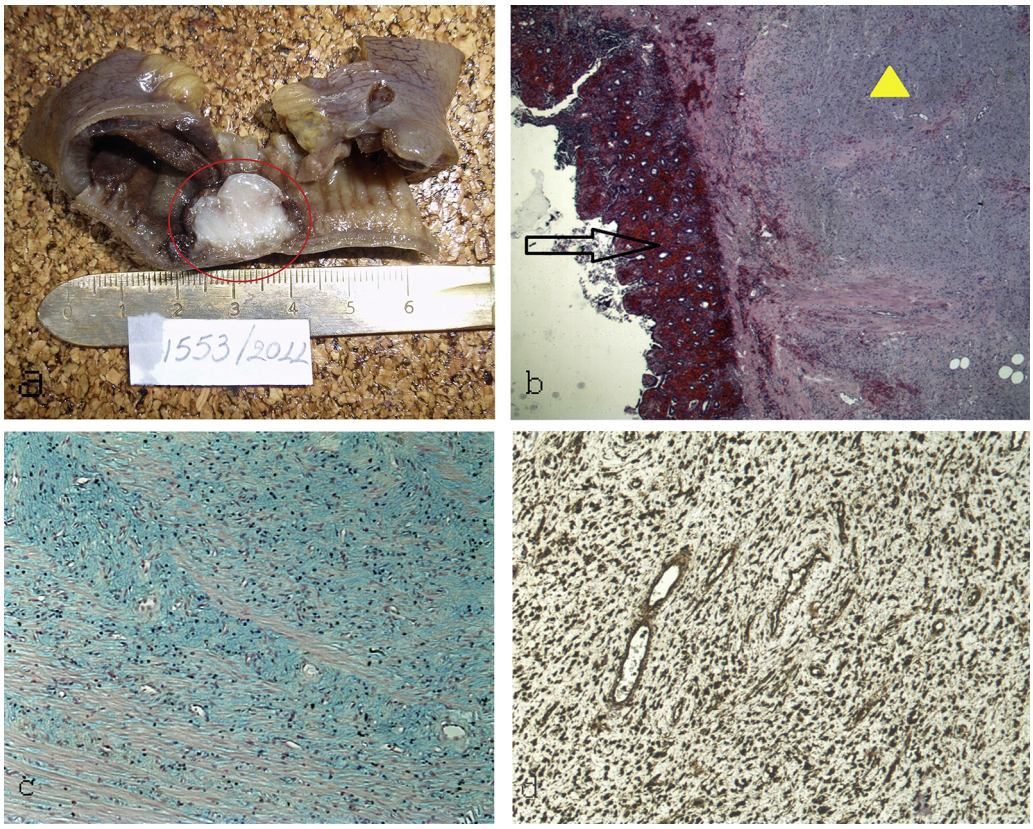

Fig. 3. (a) Gross image of our intestinal specimen containing the myxoma (circle). (b) Submucosal tumor (triangle) adjacent to normal intestinal mucosa (arrow) (H&E 40×). (c) Abundant Alcian Blue positively stained myxoid stroma of the tumor and scattered neoplastic cells (Alcian Blue 400×). (d) Stellate tumor's cells and vessels stained positively for Vimentin immunohistochemical marker (Vimentin 400×).

clear resection margins was established upon all the previously mentioned findings.

Postoperatively, the patient had a stable and uneventful clinical course. She was discharged seven days after admission with normal gastrointestinal function. One month later, ultrasonographic cardiac evaluation was conducted to her which revealed normal heart activity. Reevaluation at six months and 1 year postoperatively with abdominal and thoracic C.T. scanning did not show any metastatic lesions, enlarged lymph-nodes or intracardiac masses (Fig. 4). The patient remains with no signs of recurrence and normal levels of CEA (0.487 ng/ml) and CA 19.9 (2.4 U/ml) twenty months after surgery.

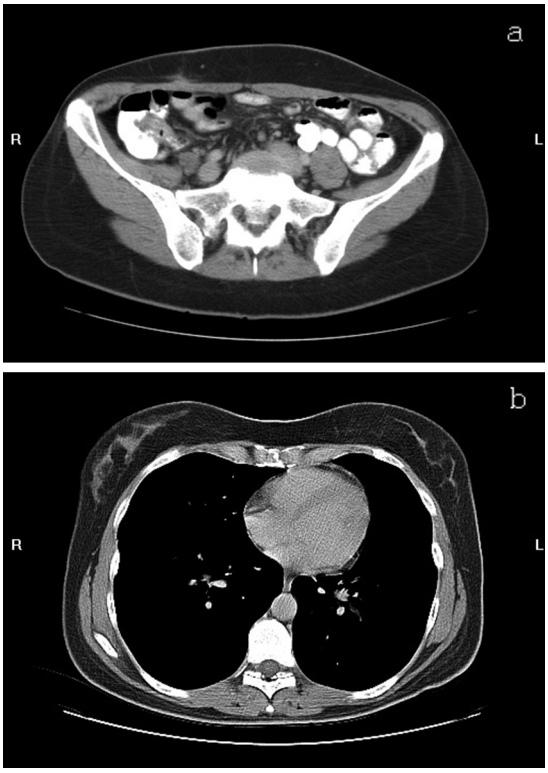

Fig. 4. Abdominal and thoracic C.T. scanning six months after surgery. No signs of intraperitoneal recurrence of the myxoma (a) or intracardiac myxoid masses (b).

3. Discussion

The myxoma is a true neoplasm of mesenchymal origin which is invariably benign in its cellular conduct and is made of a tissue resembling none found in the normal adult organism. It consists of stellate cells set in a myxoid stroma with fine reticulin fibers, which is probably made of hyaluronic acid and shows the staining characteristics of mesenchymal mucus rather than mucus of epithelial origin. Therefore, it reproduces almost accurately the histological appearance of primitive mesenchyme.[8–12]

The term "myxoma" was introduced by Virchow in 1863 for a tumor that was similar in histology with the umbilical cord.[13] The main source of true primary myxomas may be found in the early undifferentiated connective tissue of the embryo which has a mucous quality and is widely distributed in the umbilical cord and subcutaneous areas. Remaining islands of this embryonic tissue may give rise to primary myxomas, whereas secondary myxomas may develop by degeneration of mesoblastic tumors like fibromas,

lipomas, and chondromas.[11] Recently, it has been proposed that myxomas have heterogeneous origin and are not single entities.[14]

The myxoma is a tumor that occurs predominantly in the heart.[15] Less frequently it is found in the skin (fingers), subcutaneous and aponeurotic tissues, in bones (jaws) and the lower genitor-urinary tract (bladder).[8–10] A familial syndrome of multiple synchronous myxomas (Carney complex) has also been described.[16]

Myxomas seem to grow at different rates of speed. There are long periods of inactivity, followed by shorter periods of rapid enlargement. They also tend to push and progressively infiltrate adjacent structures, a fact that should be considered in treatment strategy. Cases of recurrence after close excision margins have been reported, followed by multiple re-operations and even fatal outcomes if vital structures were involved. However, metastases have never been reported, apart from cardiac myxomas. These tumors were most probably myxoid variants of cardiac sarcomas.[8–10] There has also been published a case of cardiac myxosarcoma with a previously detected jejunal metastasis causing intussusception.[17]

Myxomas of the small intestine are scarce entities and only few published cases can be found. However almost all of them, including ours, bear some similarities which should be emphasized. Intestinal myxomas mainly occurred in middle-aged women and were predominantly located in the ileum in seven out of eight cases (87.5%). Moreover, they originated from the submucosa and tended to protrude into the intestinal lumen as pedunculated masses. Finally, they acted as lead points for intestinal intussusception, which was a common manifestation among all patients.[3,8–10]

4. Conclusion

We presented the rare case of a primary intestinal myxoma complicated by ileo-ileal intussusception. Myxomas of the small intestine should be included in the differential diagnosis of masses causing intussusception, especially in cases of middle-aged women and ileal localization of the intussusception. Since true myxomas are benign, tend to infiltrate surrounding tissues and do not give metastases, treatment should include simple, wide resection of the involved intestinal segment with primary anastomosis and close follow-up control of the patient, in order to detect any possible recurrence of the tumor. Additionally, all patients with intestinal myxomas should undergo echocardiography, C.T. or M.R.I. scanning of the heart so as to exclude or treat the synchronous presence of a cardiac myxoma.[10,15]

Conflict of interest statement

None.

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None.

Ethical approval

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 contributions

Theodoros Tavlaridis, Nikolaos Varsamis and Eftychios Lostoridis performed the operation and collected the data. Eirini Tziastoudi conducted the pathology report and provided the histopathologic images of our specimen. Nikolaos Salveridis participated in the postoperative evaluation of the patient. Chrysanthi Chatzipourgani performed, interpreted and provided the diagnostic imaging studies. Constantinos Pouggouras, Athanasios Pakataridis and Constantinos Christodoulidis contributed in data analysis.

Nikolaos Varsamis wrote the manuscript.

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