CASE REPORT

Case Report: Inflammatory myofibroblastic tumor causes formation of an ileal conglomerate in a patient previously treated for Wilms' tumor [version 1; peer review: awaiting peer review]

Julie Leganger ¹, Rikke Raagaard Soerensen², Jacob Rosenberg¹, Jakob Burcharth³

¹Center for Perioperative Optimization, Department of Surgery D-113, Herlev Hospital, University of Copenhagen, Herlev, Copenhagen, Denmark

²Department of Pathology, Hvidovre Hospital, University of Copenhagen, Hvidovre, Copenhagen, Denmark

³Department of Surgery, Zealand University Hospital, University of Copenhagen, Copenhagen, Denmark

V1 First published: 15 May 2017, 6:677
<https://doi.org/10.12688/f1000research.11373.1>
 Latest published: 25 Sep 2017, 6:677
<https://doi.org/10.12688/f1000research.11373.2>

Abstract

Introduction: Inflammatory myofibroblastic tumors (IMTs) are uncommon mesenchymal lesions classified by WHO as neoplasms of uncertain behavior. Morphologically, IMTs are composed of proliferating spindled myofibroblastic cells accompanied by a marked - usually chronic - inflammatory infiltrate. The etiology is unknown, but several theories have been suggested, including an association with Wilms' tumor. IMTs are rarely diagnosed in adults and have been reported in various organs. IMTs are considered benign but with a potential to recur at their primary site.

Case report: A 44-year-old female experienced intermittent severe abdominal pain, loose stools and a visible abdominal bulge. In early childhood the patient had been treated for a Wilms' tumor. At admission Meckel's diverticulitis was suspected, but during surgery a tumor in the terminal ileum, creating a conglomerate of small intestinal loops, was observed and completely resected. The pathology report characterized the tumor as a histologically benign inflammatory myofibroblastic tumor. Postoperatively, the patient experienced several complications including an anastomotic leakage and subsequent formation of an abscess and transcutaneous fistula.

Discussion: IMTs rarely arise in the small intestine, and to our knowledge the manifestation of a small intestine conglomerate has not been described previously. Making the diagnosis is difficult, and numerous differential diagnoses were possible in this case.

Approximately 8-25% of IMTs in the gastrointestinal tract recur locally. Complete surgical resection is the treatment of choice, and re-excision

Open Peer Review

Approval Status

	1	2
version 2		
(revision)
25 Sep 2017	view	view
version 1		
15 May 2017		

1. **Jiaqi Shi**, University of Michigan, Ann Arbor, USA

2. **Jasmine Kamboj**, Sanford Health, Bemidji, USA

Komal Arora, UHS Hospital, Johnson City, USA

Any reports and responses or comments on the article can be found at the end of the article.

is the preferred therapy for local recurrence. To our knowledge, no guidelines concerning follow-ups are available.

Conclusion: IMTs in the terminal ileum can mimic Meckel's diverticulitis and present with symptoms of obstructive ileus due to the formation of a conglomerate of small intestinal loops.

Furthermore, IMTs should be considered as a diagnostic possibility in patients with a past medical history of Wilms' tumor.

Keywords

Wilms' tumor, inflammatory myofibroblastic tumors, IMTs, neoplasms of uncertain behavior, ileal conglomerate, ileus

Corresponding author: Julie Leganger (julieleganger@gmail.com)

Competing interests: No competing interests were disclosed.

Grant information: The author(s) declared that no grants were involved in supporting this work.

Copyright: © 2017 Leganger J *et al.* This is an open access article distributed under the terms of the [Creative Commons Attribution License](#), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited. Data associated with the article are available under the terms of the [Creative Commons Zero "No rights reserved" data waiver](#) (CC0 1.0 Public domain dedication).

How to cite this article: Leganger J, Raagaard Soerensen R, Rosenberg J and Burcharth J. **Case Report: Inflammatory myofibroblastic tumor causes formation of an ileal conglomerate in a patient previously treated for Wilms' tumor [version 1; peer review: awaiting peer review]** F1000Research 2017, 6:677 <https://doi.org/10.12688/f1000research.11373.1>

First published: 15 May 2017, 6:677 <https://doi.org/10.12688/f1000research.11373.1>

Introduction

Inflammatory myofibroblastic tumors (IMTs) are rare mesenchymal neoplasms composed of proliferating myofibroblastic spindle cells, and an accompanying inflammatory - usually chronic - infiltrate¹. The etiology of IMTs is unknown, but an association with Wilms' tumor, the most common primary renal malignancy in children, has been suggested^{2,3}. Furthermore, theories suggesting a link to infectious agents, tumor associated factors, and cytokines have been proposed⁴. IMTs are most commonly seen in children and young adults¹. IMTs were initially considered a pulmonary tumor², but the lesions have subsequently been reported in various extrapulmonary organs including the mesentery and gastrointestinal tract⁴.

This report presents a case of an IMT in the terminal ileum in a female adult, treated for Wilms' tumor in childhood. The tumor caused a conglomerate of small bowel and mimicked Meckel's diverticulitis, which to our knowledge has not previously been described. We reported this case according to the CARE guidelines⁵.

Case report

Clinical data

A 44-year-old woman was admitted to the hospital due to intermittent severe abdominal pain, during which a visible bulge appeared in the right lower abdominal quadrant. Furthermore, the patient had experienced loose stools over a two year period. The initial blood tests were inconspicuous, with the exception of a slight neutrophilia. The past medical history included treatment for a Wilms' tumor at the age of one with a right-sided nephrectomy and subsequent radiochemotherapy.

Ileus was initially suspected, and an abdominal CT scan was performed, showing a $3 \times 5 \times 4$ cm mass in the small intestine. The imaging report described, that the lesion was composed of a solid and a necrotic portion plus an air-filled space (Figure 1), which lead to the tentative diagnosis of an underlying Meckel's diverticulitis. A diagnostic laparoscopy was initiated, but converted to an open ileocecal resection with a primary anastomosis, as a tumor in the terminal ileum was discovered. There was no sign of intestinal perforation.

Histopathologic findings

Grossly, the specimen consisted of a cecal pole that included the appendix and the distal 40 cm of the terminal ileum bound together in a conglomerate (Figure 2). 10 cm from the proximal resection margin a diverticulum-like pouch arising from the ileal wall was identified. The lesion contained a solid tumor of firm consistency, that measured 3 (depth) $\times$ 5 (width) cm. The cut surface of the tumor elicited heterogeneity with white/grayish areas and focal haemorrhage. The tumor was totally embedded.

Microscopy revealed a mesenchymal tumor infiltrating the ileal wall, the submucosa and adjacent mesentery. No true diverticulum

Figure 1. Abdominal CT performed pre-operatively. Coronal (A) and horizontal (B) planes of the abdominal CT scan performed pre-operatively revealed a mass in relation to the small bowel (indicated by arrows).

was found. The tumor was composed of a dense proliferation of spindle-shaped cells primarily arranged in a fascicular growth pattern, focally also exhibiting a storiform architecture, admixed with a marked chronic inflammatory infiltrate and discrete areas of neutrophilic granulocytes (Figure 3). Immunohistochemically, the tumor cells were positive for actin, vimentin, desmin, and h-caldesmon, focally positive for factor 13A and CD68, and had a negative reaction for ALK1, CD34, CD117, DOG1, S100, AE1/AE3, CK18, CK7, CK19, beta-catenin and MDM2. Ki-67 staining showed a low proliferative rate. Although h-caldesmon reactivity was considered atypical for the tumor, the pathology report characterized the tumor as an IMT based on morphology and additional staining properties.

Figure 2. The gross appearance of the tumor before (A) and after (B) formalin fixation. The tumor (indicated by arrows) infiltrated the wall of the small intestine and the adjacent mesoileum (B), creating a conglomerate of dilated small bowel (A).

Postoperative care and follow-up

Postoperatively, the patient suffered several complications including an intraabdominal abscess caused by a small anastomotic leakage. Subsequent percutaneous drainage of the abscess led to a phlegmone involving the abdominal wall, complicated by the development of a transcutaneous fistula with connection to the peritoneum. Six months after surgery the patient was still having symptoms from the transcutaneous fistula but was otherwise well. She was advised to have a control abdominal magnetic resonance imaging (MRI) after one year.

Discussion

We reported a case of an adult female patient presenting with an IMT in the terminal ileum causing a conglomerate of small

intestinal loops leading to obstructive ileus. A previous study has reported that only 1.2% of IMTs arise from the small intestine³. It has been described that IMTs in the small bowel can cause intestinal obstruction due to intussusception¹. However, in our case a conglomerate of small bowel was the cause of ileus, which to our knowledge has not been presented previously in the literature. In general, patients suffering from IMTs may clinically present with an abdominal mass or non-specific symptoms including abdominal pain⁶. In approximately 15–30% of cases the patients present with a constitutional syndrome of fever, weight loss, malaise and a variety of laboratory abnormalities such as anemia, thrombocytosis, leukocytosis, polyclonal hyperglobulinemia, or elevated erythrocyte sedimentation rate³. In this case the subject only presented with a few of these features.

Figure 3. Histopathology of the tumor. The tumor mass was built from bundles of spindled myofibroblastic cells with a low proliferative rate arranged in a crossing manner, accompanied by a pronounced lymphocytic infiltrate. Focally, infiltration with polynuclear granulocytes was observed (A, shown in the insert). Immunohistochemically, the tumor cells were positive for vimentin (B), actin (C), desmin (D) and h-caldesmon (E).

Macroscopically, IMTs in the gastrointestinal tract are most often characterized as solid, sessile, and solitary lesions, although cases with multiple lesions have been described⁴. In this case the tumor mimicked a diverticulum and based on the clinical presentation, numerous differential diagnoses were possible including Meckel's diverticulitis. The histological differential diagnoses of IMTs depend on the dominant basic histological patterns, which involve the extent of proliferation and sclerosing, and the extent to which the IMT is fibromyxoid/vascular³. It is important to distinguish IMTs from other lesions in the family of inflammatory pseudotumors, as well as from non-neoplastic fibrosclerosing processes and malignant neoplasms with a prominent inflammatory infiltrate, e.g. fibromatosis, sarcoma, gastrointestinal stromal tumor, and mesenteric panniculitis⁶. Immunohistochemically, the spindle cells in consideration in the diagnostic process of IMT are presented in Table 1. The tumor is reactive to antibodies directed against vimentin, smooth muscle actin, and muscle specific actin in the majority of cases. Anaplastic lymphoma kinase expression is detected in approximately 50% of cases^{2,4,6}. The frequency of this finding decreases with age.

A possible association with Wilms' tumor has previously been suggested and different theories have been proposed. One theory is a shared genetic predisposition; another theory is that the treatment of Wilms' tumor involving radiation and chemotherapy may damage the tissue and predispose the patient to development of IMTs. Notable that the latency time in this case is remarkable longer from the other reported cases². The IMTs were at first considered a postinflammatory condition but is now acknowledge as a distinct neoplasm based on clonal rearrangements involving chromosome 2p⁶. The IMTs have been classified by WHO as a neoplasm of uncertain behavior². The tumors are widely acknowledged as benign but with a potential to recur at the primary site. Approximately 8–25% recurs locally^{1,4}. Rare examples are described with tumors undergoing malignant transformation² and/or with metastasis⁴. Several studies have attempted to find predictors of aggressive behavior in IMTs without success⁶. However, it has been suggested that IMTs with a proliferating pattern, a multinodular presentation, or a manifest myofibroblastic or fibroblastic phenotype are more likely to recur⁴. Furthermore, IMTs arising in the gastrointestinal tract are more likely to recur compared with similar tumors arising elsewhere⁷. Anaplastic

Table 1. Clinical and pathological features characteristic for inflammatory myofibroblastic tumors (IMTs). Adapted from 6, to consider in the diagnostic process of IMT.

Features that favour IMT diagnosis	Features that discourage IMT diagnosis
<ul style="list-style-type: none"> • Child or young adult • Mass in lung or soft tissue of abdomen, pelvis, or retroperitoneum • Diffuse inflammatory infiltrate with prominent plasma cells • Mild nuclear atypia including scattered ganglion-like cells • Low mitotic rate without atypical forms • Anaplastic lymphoma kinase positivity by immunohistochemistry or anaplastic lymphoma kinase gene • Wilms' tumor 	<ul style="list-style-type: none"> • Middle-aged or older adult • Mass in skin or subcutis, lymph node, spleen, or bladder • Patchy, predominantly lymphocytic inflammatory infiltrate • Moderate to severe nuclear atypia with hyperchromasia • Atypical mitoses • Necrosis

lymphoma kinase expression is associated with a lower risk of metastasis^{6,7}. Complete surgical resection is the treatment of choice, and re-excision is the preferred therapy for local recurrence⁷. To our knowledge no guideline on IMT follow-up is available.

Conclusion

An IMT in the terminal ileum can mimic Meckel's diverticulitis and the clinical manifestations can include intestinal obstruction due to the formation of a conglomerate of small intestinal loops. Furthermore, IMTs should be considered as a diagnostic possibility in patients with a past medical history of Wilms' tumor².

Consent

Written informed consent for publication of clinical details and clinical images was obtained from the patient.

Author contributions

JR contributed to the conception of the article. JL, RSS, JR, and JB contributed to the design of the work. JL contributed to the acquisition of data and prepared the first draft of the manuscript. RSS provided expertise in pathology and acquisition of clinical photographs. JL, RSS, JR, and JB were involved in the revision of the draft manuscript and have agreed to the final content.

Competing interests

No competing interests were disclosed.

Grant information

The author(s) declared that no grants were involved in supporting this work.

References

1. Ambiru S, Nakamura S, Itabashi T, *et al.*: **Inflammatory myofibroblastic tumor causing ileoileal intussusception: an unusual cause of an unusual neoplasm in an adult, with a clinicopathological review of the literature.** *Clin J Gastroenterol.* 2009; **2**(3): 194–8.
[PubMed Abstract](#) | [Publisher Full Text](#)
2. Ortiz MV, Rossi CT, Hill DA, *et al.*: **Inflammatory myofibroblastic tumor as a second neoplasm after Wilms tumor.** *Pediatr Blood Cancer.* 2015; **62**(6): 1075–7.
[PubMed Abstract](#) | [Publisher Full Text](#)
3. Coffin CM, Humphrey PA, Dehner LP: **Extrapulmonary inflammatory myofibroblastic tumor: a clinical and pathological survey.** *Semin Diagn Pathol.* 1998; **15**(2): 85–101.
[PubMed Abstract](#)
4. Makhlof HR, Sobin LH: **Inflammatory myofibroblastic tumors (inflammatory pseudotumors) of the gastrointestinal tract: how closely are they related to inflammatory fibroid polyps?** *Hum Pathol.* 2002; **33**(3): 307–15.
[PubMed Abstract](#) | [Publisher Full Text](#)
5. Gagnier JJ, Kienle G, Altman DG, *et al.*: **The CARE guidelines: consensus-based clinical case reporting guideline development.** *J Med Case Rep.* 2013; **7**: 223.
[PubMed Abstract](#) | [Publisher Full Text](#) | [Free Full Text](#)
6. Gleason BC, Hornick JL: **Inflammatory myofibroblastic tumours: where are we now?** *J Clin Pathol.* 2008; **61**(4): 428–37.
[PubMed Abstract](#) | [Publisher Full Text](#)
7. Chaudhary P: **Mesenteric inflammatory myofibroblastic tumors.** *Ann Gastroenterol.* 2015; **28**(1): 49–54.
[PubMed Abstract](#) | [Free Full Text](#)

The benefits of publishing with F1000Research:

- Your article is published within days, with no editorial bias
- You can publish traditional articles, null/negative results, case reports, data notes and more
- The peer review process is transparent and collaborative
- Your article is indexed in PubMed after passing peer review
- Dedicated customer support at every stage

For pre-submission enquiries, contact research@f1000.com

F1000Research