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Case report

Intra-abdominal multicentric inflammatory myofibroblastic tumors mimicking ruptured appendicitis

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ABSTRACT

Inflammatory myofibroblastic tumors (IMTs) are a rare soft tissue neoplasm, usually seen in children and adolescents, which are predominantly found in the pulmonary region. The extrapulmonary multicentric lesions are exceedingly rare. We herein report the case of a 19-year-old female who developed acute bowel obstruction which caused by multicentric IMTs. We described her clinical presentations, operative finding, and pathological finding.

1. Introduction

Inflammatory myofibroblastic tumors (IMTs) are a rare soft tissue neoplasm, usually seen in children and adolescents. Even though they are predominantly found in the pulmonary region [1,2], extrapulmonary occurrences have been reported, and the most common extrapulmonary site is the abdominopelvic region. Multicentric lesions have also been reported, which are associated with a high local recurrence rate and metastasis [3]. Although IMTs are characterized by diverse clinical presentation and intermediate biological potential, it is rare to find mesenchymal tumors that contain myofibroblastic spindle cells along with lymphocytes, plasma cells, and eosinophils. Importantly, such variation in clinical presentation and nonspecificity of imaging results render distinguishing between IMT and other mesenteric mass and their preoperative diagnosis challenging. Complete surgical resection is the most effective treatment [2], but given the difficulties in establishing the correct diagnosis, surgical plans may require improvisations. Here we report a rare case of multicentric IMT that mimicked ruptured appendicitis and describe the surgical procedure employed, intra-operative and pathological findings observed, and prognosis according to the 2020 Surgical CAse REport (SCARE) guidelines [4].

2. Case presentation

A 19-year-old woman without systemic disease presented to the emergent department with a 4-day history of persistent epigastric cramping pain, followed by vomiting, constipation, and fever. Initial physical examination showed mild abdominal distention with local tenderness over the epigastric area. As abdominal radiography revealed a dilated small bowel with air-fluid levels, a small bowel obstruction was suspected. However, abdominal computed tomography (CT) with contrast revealed a dilated small bowel with a transitional zone and loculated fluid accumulation around the tip of the appendix and paracolic region (Fig. 1). Therefore, ruptured appendicitis with adhesion band-induced bowel obstruction was suspected and an exploratory laparotomy was scheduled.

Intraoperatively, we found multiple fibrotic bands leading out from the mesentery of the terminal ileum, with one of them causing bowel obstruction as it had adhered to the mesentery of the ileum (Fig. 2). Further, palpable mesenteric lymphadenopathy and multiple nodules (>10) located over the peritoneum, omentum, liver surface, and mesentery were noted, along with clear ascites over the pelvic cavity. A right hemicolectomy with multicentric tumor excision was performed.

Histological evaluation of the tumor in the terminal iliac mesentery, fibrotic bands, and omental tumor enabled the identification of IMTs. Microscopically, they were composed of short spindle cells in interlacing bundles with lymphoplasmacytic infiltration that partially contained myxoid stroma (Fig. 3). Further, they were positive for actin but negative for anaplastic lymphoma kinase 1 (ALK-1) staining. The other peritoneal nodules, mesenteric nodules, and the submucosal tumor of

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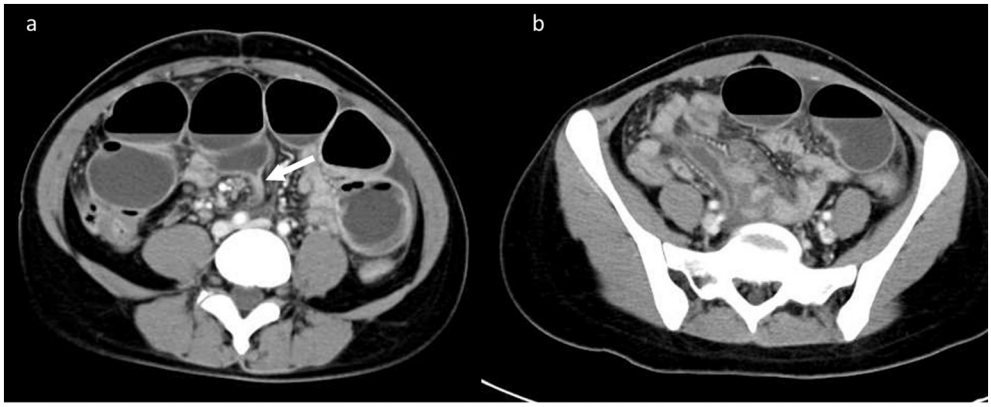

Fig. 1. (a) Abdominal CT showed small bowel obstruction with an adhesion band (white arrow). (b) Loculated fluid accumulation around the appendix with perifocal infiltration, which rupture appendicitis was suspected.

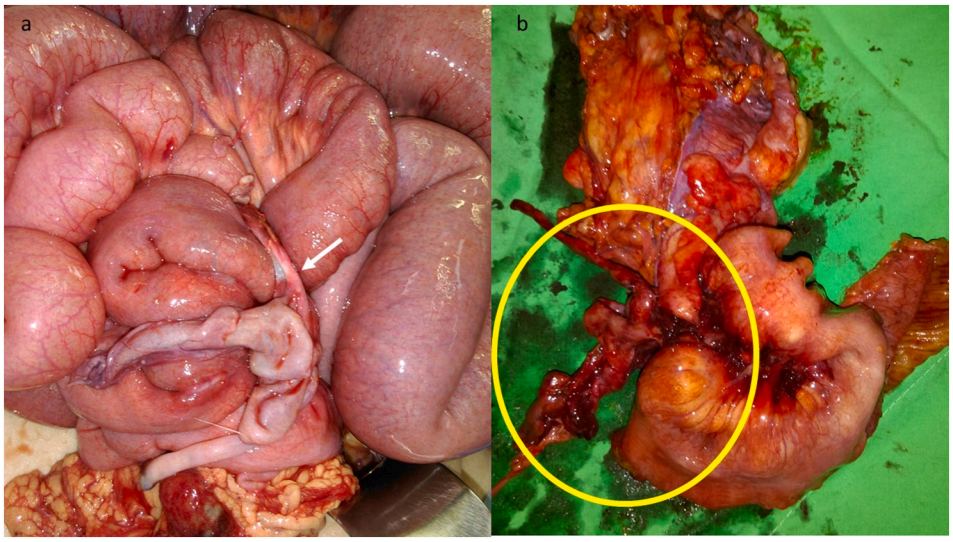

Fig. 2. (a) One of the fibrotic bands (white arrow) causing small bowel obstructive ileus which compatible with CT image Fig. 1a. (b) The terminal ileum mesenteric tumor and the suspicious involved appendix with irregular surface (yellow circle). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

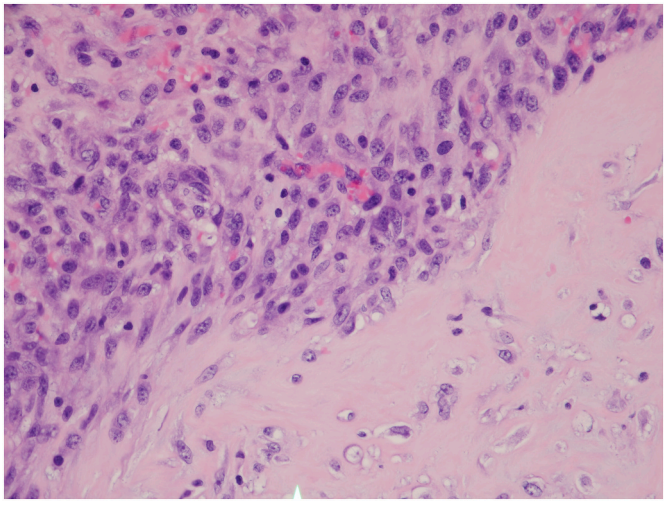

Fig. 3. H&E stains revealed spindles cells in both compact and loose myxoid stroma (400×).

ileum showed features of calcifying fibrous tumors (CFTs), such as paucicellular fibroblastic proliferation and presence of lymphoid follicles and psammomatous calcification. The ascending colon, appendix, and regional lymph nodes were tumor free, and the pathological diagnosis was multicentric intra-abdominal IMT with multiple CFT.

Her postoperative course was unremarkable, and she regained digestive function smoothly. Given the diagnosis of IMT, a chest CT scan was acquired but no obvious pulmonary lesions were found. She was discharged on postoperative day 12 without any complications, and regular chest–abdomen CT scans acquired during a follow-up period of 24 months showed no evidence of recurrence or metastasis.

3. Discussion

IMTs are rare, distinctive lesions with an unclear etiology that were first described by Dr. H. Brunn in 1939 [5]. Grossly, IMTs may be firm, fleshy, or gelatinous, with a white or tan cut surface [2]. They typically occur in children and young adults and predominantly affect the lungs as only 5% of IMTs occur in extrapulmonary locations. Notably, such extrapulmonary lesions may occur in the head/neck, respiratory tract, intra-abdomen, extremities, and uterus [2]. The clinical presentation of IMTs is related to the location of the lesion and its affected organs

because, for e.g., while patients with intra-abdominal tumors present with intermittent pain, abdominal distension, weight loss, malaise, anorexia, and vomiting [6]. In our patient, IMTs had affected the terminal ileum and caused an obstruction that mimicked ruptured appendicitis, which necessitated additional caution and improvisation during surgery due to the presence of unusual intra-operative findings. It is important to differential diagnosis between IMT and Gastrointestinal stromal tumor (GIST). During pre-operative CT scan, IMT usually presented as well-circumscribed masses with variable enhancement patterns and extend to the adjacent bowel; but GIST usually presented as a single solid tumor [2]. The clinical presentations were also different; GIST usually presented as gastrointestinal tract bleeding [7,8]. The pathological findings were also different. IMT characterized by a variably cellular spindle cell in a myxoid to collagenous stroma with a prominent inflammatory infiltrate plasma cells and lymphocytes; but GISTs were composed of spindle cells with palely eosinophilic fibrillary cytoplasm, ovoid nuclei, and syncytial cell borders [9,10].

Although CFTs are mainly seen in the gastrointestinal tract, they are also found in several organ systems. Even though most CFT lesions are solitary, multiple CFTs have also been reported, as seen in our patient. All CFTs were removed during laparoscopy, even though they were relatively benign. The coexistence of IMTs and CFTs was first reported in 1999, and thus, CFTs were hypothesized to be the sclerosing end stage of IMTs [11]. Additionally, Tomassen et al have reported that these two tumor types share a DNA methylation profile [12], thereby supporting the theory that CFTs may be the end stage of IMTs. However, others consider them to be distinct entities based on histological findings, for e.g., while IMTs are positive for anaplastic lymphoma kinase, CFTs are negative. Further, IMTs stain positive for muscle-specific actin, smooth muscle actin, desmin, and vimentin, whereas CFTs are negative for all these molecules.

IMTs are classified as an intermediate neoplasm in the World Health Organization classification of soft tissue tumors due to their tendency to recur. In contrast, although CFTs are generally benign, case descriptions of familial occurrence have been reported [13]. The current treatment of choice for IMTs and CFTs is complete surgical resection [6,14], and alternative treatment strategies, such as chemotherapy, radiotherapy, or immunomodulation drugs remain controversial as they have shown inconsistent results [6]. Further, IMT recurrence is determined by primary lesion site and ranges from 2% for lung lesions to 25%–37% for extrapulmonary lesions [1,15] with low incidence (5%) of distant metastases, with common sites being the lungs and brain. As major risk factors for recurrence are incomplete surgical resection, ALK negativity, and *p*53 positivity [9], long-term clinical and radiological follow-up is recommended.

4. Conclusion

Available literature shows that the coexistence of intra-abdominal multicentric IMTs and CFTs is an exceedingly rare occurrence. Clinical presentation depends on tumor location, and preoperative diagnosis is challenging. The most effective treatment is complete surgical excision. In our patient, while we suspected IMT based on intra-operative findings, radical complete excision was strongly recommended, along with long-term post-surgical follow-up due to the potential for recurrence.

Declaration of competing interest

The authors declare that they have no competing interests.

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Ethical approval

This study was approved by the ethics committee of National Cheng Kung University Hospital, College of Medicine, National Cheng Kung University, Tainan, Taiwan. Institutional Review Board Statement: This study was approved by the institutional review board of NCKUH (A-EC-110-015).

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

FF Yang and TH Li contributed equally to the study design, collection of data, and the writing of the manuscript. RH Chan contributed to data review and revision of manuscript. CT Lee contributed to collection of data and pathology review.

Research registration

Not applicable – single case report.

Guarantor

RH Chan.

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