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Case report

Co-existence of acute appendicitis and inflammatory myofibroblastic tumor of the small intestine: A case report☆

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HIGHLIGHTS

- Besides its rarity, coexistence of inflammatory myofibroblastic tumor with acute appendicitis is another rare event.
- Immunohistochemistry plays a crucial role including positivity for actin, vimentin, CD68 and anaplastic lymphoma kinase.
- Complete surgical excision followed by serial imaging techniques should be regarded as the mainstay of the treatment.

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ABSTRACT

Introduction: Inflammatory myofibroblastic tumor as a rare neoplastic lesion is seen most commonly in the pulmonary system. Beside the presence of limited number of inflammatory myofibroblastic tumors of the gastrointestinal tract in the literature, co-existence with acute appendicitis has not been reported before.

Presentation of case: A 27-year-old woman admitted to emergency department with acute abdominal pain at the right lower quadrant. The initial diagnosis was as acute appendicitis. Intraoperatively, a mass with a diameter of almost 5 cm originated from the distal ileal segments neighboring the appendix was seen. The patient was managed by segmental resection of the small intestine including the mass with appendectomy. Histologically, there were bundles of spindle cells accompanied by lymphoplasmocytic infiltration. The immunohistochemical studies showed that tumor cells were positive for smooth muscle actin, vimentin, perinuclear activity for anaplastic lymphoma kinase and CD 68. The final pathologic diagnosis was inflammatory myofibroblastic tumor.

Discussion: Concomitant resection of tumoral lesions detected in the neighbor intestinal segments during appendectomy should be considered to diagnose and treat.

For the diagnosis of inflammatory myofibroblastic tumor, immunohistochemistry pattern including positivity for actin, vimentin, CD 68 and anaplastic lymphoma kinase plays a crucial role. Therefore, detailed immunohistochemistry analysis should be performed in suspicious cases.

Conclusion: Coexistence of inflammatory myofibroblastic tumor located in the gastrointestinal system with acute appendicitis is a rare event. Complete surgical excision should be regarded as the mainstay of the treatment. Long-term follow up with serial imaging techniques is recommended.

1. Introduction

Inflammatory myofibroblastic tumor (IMT) is a rare neoplastic lesion with a tendency toward local aggressive behavior and recurrence [1]. Although several names such as plasma cell granuloma, inflammatory pseudotumor, and myxoid hamartoma have been used previously, the lesion has been first described by

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Bahadori and Liebow in 1973 as IMT [2]. It is a reactive or inflammatory process in the pulmonary system mostly occurring in children and young adults, however, latest research shows that many cases have been reported in adult patients with unusual extra-pulmonary sites [2,3]. Prognosis of IMT shows variations with regard to the localization of the tumor like a benign clinical course in soft tissues or an aggressive behavior with intra-abdominal and retroperitoneal diseases [3,4].

There is limited number of IMT cases found in the gastrointestinal tract [1,5,6]. However, co-existence of IMT with acute appendicitis has not been reported before. The extent of surgery in such cases has not been studied because of the rarity of these cases.

In this paper, it was aimed to report an ileal IMT in a young female patient with acute appendicitis.

2. Case presentation

A 27-year-old woman presented with acute abdominal pain at the right lower quadrant to the emergency department. Medical, family, and psychosocial history were all normal. On admission, physical examination revealed abdominal tenderness and rigidity around the periumbilical region and the right lower quadrant. Laboratory tests showed normal leukocyte count and hemoglobin level of 9.3 g/dL. All other biochemical values were within the normal range. Abdominal ultrasonography revealed acute appendicitis and peri-appendiceal inflammation appearing as a mass. With a presumptive diagnosis of acute appendicitis with plastron formation, laparotomy was performed. Intraoperatively, acute appendicitis was detected. In addition, a mass with a diameter of almost 5.5 cm originated from the distal ileal segments in the vicinity of the inflamed appendix vermiformis was seen. Therefore, appendectomy and segmental resection of the small intestine including the mass were performed (Fig. 1). The patient was discharged at the fifth postoperative day with normal recovery. On the 6th postoperative month, she had no complaint with a normal abdominal computed tomography.

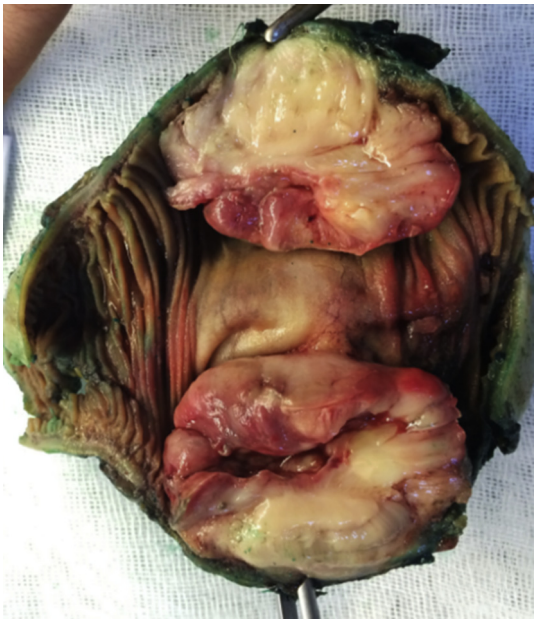

Fig. 1. The macroscopic appearance of inflammatory myofibroblastic tumor with a diameter of almost 5.5 cm originated from the distal ileal segments neighboring the appendix.

3. Histopathology

The specimen was collected from the appendix with a length of 7 cm and the ileum with a length of 12 cm after fixation in 10% neutral buffered formalin. Histopathology of the appendix was concordant with acute appendicitis as infiltration of the muscularis propria by polymorphonuclear leukocytes. A serial section of the ileum revealed a polypoid mass with a diameter of 5.5 cm was detected in the ileum. The tumor was covered with normal mucosa and it was located from the subepithelial to subserosal planes, mostly intraluminal. Cut surface section of the lesion showed diffuse yellowish-tan appearance with well demarcated borders. Perforation, necrosis, hemorrhage, calcification, and ulceration were not identified macroscopically (Fig. 1).

Histologically, the tumor appeared to have bundles of spindle cells accompanied by lymphoplasmocytic infiltration including leukocytes, lymphocytes and eosinophils (Figs. 2 and 3). The immunohistochemical studies showed that tumor cells were positive for smooth muscle actin, vimentin, perinuclear activity for anaplastic lymphoma kinase (ALK) (Fig. 4) and CD 68. Additionally, negative results were detected for antibodies to p53, CD117, S100, CD34, CD 31, CD30, HBME-1, pansitokeratin, caldesmin and desmin. Ki-67 was positive in 15% of the cells. The final pathologic diagnosis was IMT.

The written consent was taken from the patient.

4. Discussion

Histologically, IMTs are defined by the World Health Organization as lesions composed of a myofibroblastic spindle cell population accompanied by an inflammatory infiltration of plasma cells, lymphocytes, and eosinophils. Presence of such inflammatory cells shows chronic inflammatory state underlying this pathology [7]. In this setting, IMT occurs in various age groups and is slightly more common in women than in men [3,6]. Although the lung is the most common site of the involvement, IMT has also been reported in other organs including the stomach, small bowel mesentery, omentum and retroperitoneum as well as the kidneys, renal pelvis, the liver, the spleen and the esophagus [3,4,7–9]. In Malkhlouf's study, the stomach was found to be the most common extra-pulmonary site observed in 34% of the total 38 cases [10]. Extra pulmonary site lesions usually present with non-specific signs and

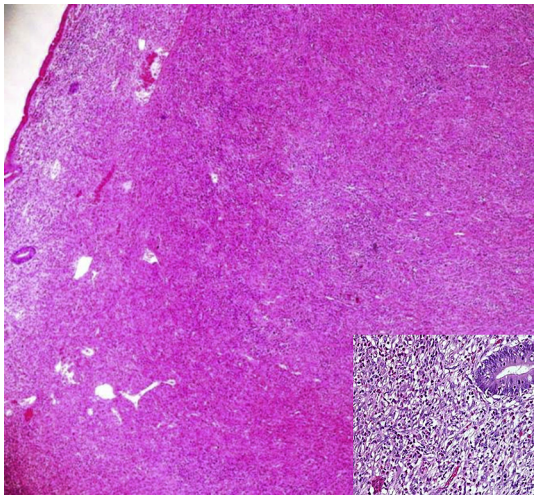

Fig. 2. Low power view demonstrating fasicular patternized areas and chronic inflammatory cells on submucosal lesion Inflammatory Myofibroblastic Tumor (H&E ×100).

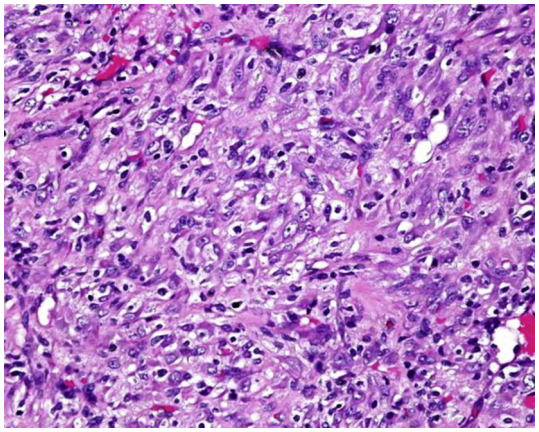

Fig. 3. Inflammatory Myofibroblastic Tumor: High-power magnification demonstrating the presence of tumor consist spindle cells shaped of myofibroblasts, mature plasma cells, eosinophils and lymphocytes (H&E ×200).

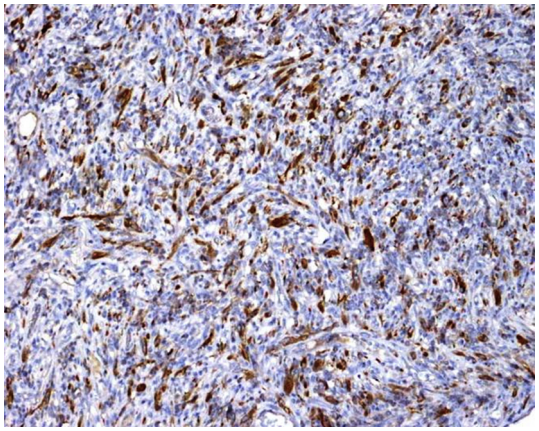

Fig. 4. Immunohistochemical staining for ALK positive (×200).

symptoms including abdominal pain, gastric and intestinal mass with occasional obstruction and growth retardation in children [11]. It has been thought that incidental finding of IMT with acute appendicitis was the first case in the English literature. Although intestinal obstruction caused by IMT has been reported in a pediatric patient, acute presentation either primary pathology or an incidental finding can be regarded as a rare event [12]. Therefore, concomitant resection of tumoral lesions found in the neighbor intestinal segments should be considered to diagnose and treat such rare entities even during appendectomy as in the present case.

The treatment of IMT is believed to be complete surgical excision. It has also been thought that long-term follow up with serial imaging techniques are required [13,14]. However, the choice of the imaging test and the interval has not been determined yet. As a marker for proliferative index, Ki-67 value may be helpful to determine the time interval for follow up. Therefore, an individualized follow up should be planned for each patient. Computed tomography at the postoperative 6th month was evaluated as normal for the patient. Although higher Ki-76 value may be regarded as an ominous sign for the patient presented in the paper, symptom based follow-up can be regarded as a more appropriate approach for young female patients. The choice of imaging either by ultrasonography or computed tomography should be determined according to the symptomatology of the patients.

Radiotherapy and chemotherapy using cisplatin, doxorubicin and methotrexate have also been tried as an adjunct to surgery with no evidence of substantial benefit for the patients [15,16]. Although it was reported some degree of regression with use of steroid therapy or non-steroidal anti-inflammatory medications, the benefit of such medical treatment is still controversial [17]. Based on the results of these adjuvant therapies, surgery should be regarded as the mainstay of the treatment of IMT.

For the exact and differential diagnosis of IMT, immunohistochemistry pattern plays a crucial role. Tumor cells were shown to be positive for actin, vimentin, CD68 and ALK, and they did not express CD 117 and CD 34. Additionally, the cells were focally positive for SMA without desmin expression. Although microscopic similarity between IMT and gastrointestinal stromal tumor has been reported, it is thought that immunohistochemical features of IMT can be helpful for such differentiation as in the case. Although it is normally expressed in neural tissues, ALK has been recently detected in the spindle cells of some IMT's with a range from 33% to 67% [10]. Therefore, ALK positivity was regarded as an important diagnostic criterion for IMT in the current paper.

Lack of preoperative ultrasonographic findings and intraoperative image showing the co-existence of acute appendicitis and IMT were regarded as the main limitations of this study.

5. Conclusion

IMT is a rare neoplastic lesion occurring in pulmonary and extrapulmonary sites, mostly occurring in children and young adults. For the diagnosis of IMT, immunohistochemistry pattern plays a crucial role including positivity for actin, vimentin, CD68 and ALK. Complete surgical excision should be regarded as the mainstay of the treatment of IMT. Long-term follow up with serial imaging techniques may be recommended for possible local aggressive behavior and recurrence.

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