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Case Report
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Inflammatory Myofibroblastic Tumor of the Abdominal Wall in an Adult Male: A Case Report and Literature Review

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Abstract

Background: Inflammatory Myofibroblastic Tumor (IMT) of the abdominal wall is an exceedingly rare entity and to our best knowledge, there are only 4 cases are reported in the English literature (excluding of this case). The clinical symptoms, treatment and prognosis of this tumor are still uncertain.

Case Summary: A 49-year-old Chinese man was admitted to our hospital with approximately 45 mm palpable mass located in the abdominal wall, without any other systemic symptoms. Computed Tomography (CT) demonstrated a 41 mm × 23 mm abdominal wall mass that was well-limited, heterogenous enhancement in portal venous image. The lesion was resected and histopathologic examination revealed that the tumor was predominantly comprised of spindle cells arranged in myxoid stroma with an inflammatory infiltrate of lymphocytes, plasma cells and histiocytes. An immunohistochemical analysis demonstrated positive staining for vimentin, CD34, D2-40 and negative staining for Smooth Muscle Actin (SMA), desmin, S100, CD21, CD23, CD1α, HMB45, Melan-A, SOX10, Myogenin, STAT6, CD117. Hence, we had come to a diagnosis of IMT. The patient recovered well without any complications, and we had been followed up for the last 27 months without clinical or radiographic evidence of recurrence or metastasis.

Conclusion: Since abdominal wall IMT is uncommon lesion and lacks characteristic clinical manifestation with uncertain biological behavior, we have great challenge to have a clear statement on preoperational diagnosis. Surgical resection remains the best option for diagnosis and treatment.

Keywords: Inflammatory myofibroblastic tumor; Abdominal wall; Case report; Anaplastic lymphoma kinase; Diagnosis; Literature review

Core Tip

Inflammatory Myofibroblastic Tumor (IMT) is an exceedingly rare entity and lacks characteristic clinical manifestation with uncertain biological behavior; we have great challenge to have a clear statement on preoperational diagnosis. Surgical resection remains the best option for diagnosis and treatment.

Introduction

Inflammatory Myofibroblastic Tumor (IMT) is a distinctive type of fibroblastic/myofibroblastic tumors composed of differentiated myofibroblastic spindle cells with an inflammatory infiltrate of lymphocytes, plasma cells and eosinophils [1]. Approximately 150 to 200 cases are newly diagnosed with IMT annually in the America [2]. IMT can occur at any age with a predilection for children, adolescents and young adults [3]. The tumor primarily affects visceral organs and soft tissue. The most common sites of involvement are the lung, mesentery and omentum [1]. To the best of our knowledge, IMT originating from the abdominal wall was an exceedingly rare entity and only 4 cases were reported in the English literature [4-7]. Herein, we describe a case of a 49-year-old Chinese man with IMT presented in abdominal wall and we also reviewed the relevant literature to learn more about this rare disease from clinical features to therapeutic approaches.

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Case Presentation

Chief complaints

A 49-year-old Chinese male was admitted to Tongji Hospital (Wuhan, China) with a palpable mass in the right lumber region that had been growing slowly for 2 years.

History of present illness

The patient discovered a touchable mass in the right abdomen 2 years ago without any other discomfort, and the mass gradually enlarged without any other symptoms. He visited Wuhan Xiehe Hospital. CT of the abdomen showed that there was a 49 mm × 22 mm soft tissue density mass on the right side of the abdomen. The enhancement was progressive and obvious and uneven. The boundary was clear, It can be seen that the tortuous and thickened intermuscular blood vessels extend in, which is usually considered as a neoplastic lesion. The patient was referred to our hospital for further investigation and treatment.

History of past illness

There was no history of anemia, fever, malaise, weight loss, or any other systemic symptoms. He had no history of infections, abdominal operations, or trauma to the area.

Personal and family history

The patient denied the history of family genetic disease and alcohol consumption. Had no allergies to food or medicines yet.

Physical examination

Admission to the hospital for physical examination: the patient was well-nourished (the Body Mass Index is 25.4 kg/m^2), afebrile (Temperature 36.7°C) with stable vital signs (Blood Pressure 125/70 mmHg, Pulse 87 bpm). The abdomen is flat and soft, there is no intestinal shape or gastrointestinal peristaltic waves. No tenderness in the whole abdomen and rebound pain. There were no palpable liver and spleen below the costal margin or enlarged superficial lymph nodes. After palpation of the abdomen, we found at approximately 40 mm × 20 mm sized palpabal solid mass on the right upper abdomen without any signs of discomfort.

Laboratory examinations

Basic laboratory examination revealed normal IgG4, IgG4/IgG ratio, white blood cell count and hemoglobin. Tumor markers (CEA, AFP, CA125, CA19-9) were within the normal range. And the rest relative laboratory routine were unremarkable.

Imaging examinations

An enhanced Computed Tomography (CT) of the abdomen demonstrated a 41 mm × 23 mm right upper abdominal wall space-occupying lesion. The mass was well-limited and it is slightly and uniformly strengthened, heterogenous enhancement in portal venous image (Figure 1) and chest radiograph was normal.

Preoperative diagnosis

According to the physical signs and radiographic finds, an initial diagnosis, benign tumor, was made.

Treatment

No medications treatment and invasive procedures have been performed since the discovery of the lesions.

Interventions

The operation was performed. We resected the abdominal wall lesion and segmental obliquus internus and obliquus externus abdominis muscles with clear margins microscopically.

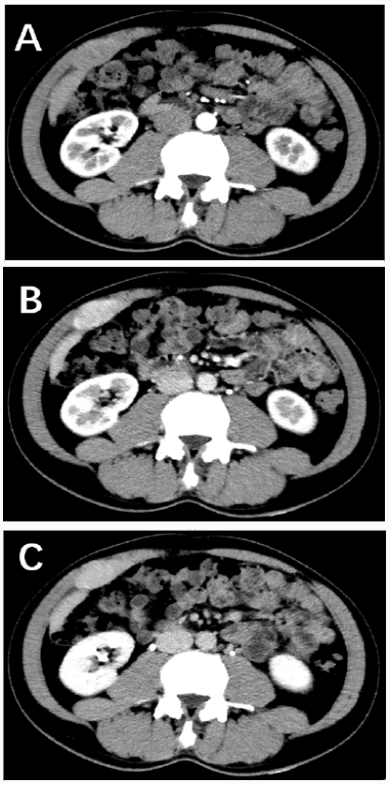

Figure 1: CT image of the tumor at diagnosis. A-C showed CT images of the mass of abdominal wall. A, B and C were contrast-enhanced images obtained in arterial portal venous and delayed phase. The mass was well-limited, heterogeneous enhancement in portal venous phase image.

Hypnology

Macroscopically, we observed a 45 mm × 35 mm sized, Partial hemorrhagic necrosis, grayish-white nodule. Microscopically, histopathologic examination of the resected specimen showed that the tumor was predominantly comprised of spindle cells arranged in myxoid stroma with an inflammatory infiltrate of lymphocytes, plasma cells and histiocytes (Figure 2).

Immunochemistry analyses

An immunohistochemical analysis demonstrated positive staining for vimentin, CD34, D2-40, but negative staining for CD21, CD23, CD1α, Smooth Muscle Actin (SMA), HMB45, desmin, Melan-A, S100, SOX10, Myogenin, STAT6, CD117.

Final diagnosis

Hence, the diagnosis was confirmed, based on the findings above, as inflammatory myofibroblastic tumor of the abdominal wall, with negative surgical margins.

Outcome and follow-up

The patient was well recovered without clinical or radiographic evidence of recurrence after the last 27 months of follow-up period.

Discussion

Inflammatory Myofibroblastic Tumor (IMT) is a distinctive soft tissue neoplasm comprised of myofibroblastic spindle cells with the infiltration of lymphocytes, plasma cells and eosinophils [1]. In

Table 1: Four reported cases of IMT of the abdominal wall and the present case.

No.	Author	Year	Age	Sex	Syndromes	size (cm)	Treatment	Follow-up time	Recurrence
1	Pratap et al. [4]	2007	6	Male	abdominal pain palpable mass	16 × 14 × 12	resection	4 months	no
2	Kaneko et al. [5]	2007	58	Male	abdominal pain palpable mass	5.5 × 5 × 3	resection	13 months	yes
3	Yagci et al. [6]	2010	50	Female	abdominal pain palpable mass	8 × 8 × 6	resection	12 months	no
4	Hernández et al. [7]	2012	37	Male	palpable mass	17 × 12	resection	unknown	unknown
5	The case	2018	49	Male	palpable mass	4.5 × 3.5 × 3	resection	27 months	no

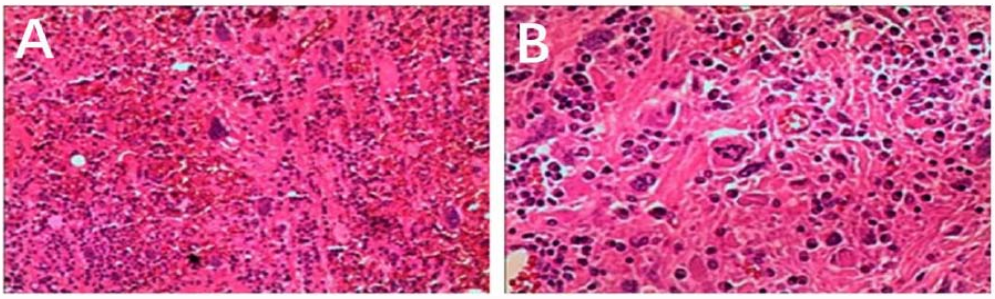

Figure 2: A and B illustrated HE stain of the tumor with the magnification of 100 and 400, respectively. Histologically, tumor was comprised of myofibroblastic spindle cells in an inflammatory stroma of lymphocytes, plasma cells and histiocytes.

1939, Brunn [8] first reported the inflammatory myofibroblastic tumor occurring in the lung. In 1954, Umiker [9] named IMT as an inflammatory pseudotumor because its clinical features and imaging findings mimicked those of malignant tumors. IMT has many synonyms, such as plasma cell pseudotumor, inflammatory pseudotumor, inflammatory myofibroblastic proliferation, inflammatory myofibrohistiocytic proliferation and plasma cell granuloma, due to its histopathological features. IMT shows a predilection for children, adolescents and young adults [1]. There is a slight predominance in females (female:male, 4:3) [1]. IMT could occur in visceral organs and soft tissue throughout the body and the most frequent sites are the lung, mesentery and omentum [10]. Inflammatory Myofibroblastic Tumor (IMT) of the abdominal wall is exceedingly rare. We reviewed the relevant literature on PubMed using the key words "inflammatory myofibroblastic tumor" and "abdominal wall". The search through PubMed listed 47 citations involving 4 patients with primary IMT of the abdominal wall [4-7] (Table 1). Of the 5 reports of IMT arising from the abdominal wall, including the present case, the age at diagnosis ranged from 6 years to 58 years (median age, 49 years old). IMT of the abdominal wall occurred primarily in middle-aged adults other than children or young adults. The predominance of male gender was evident (4 males and 1 female). Abdominal mass and abdominal pains were the most common symptoms. The average tumor size was 10.2 cm. Information about follow-up was available in 4 cases and the average duration of follow-up time was 14 months ranging from 4 to 27 months. One case had a recurrence after 13 months but the other 3 cases did not show recurrence after resection.

Trauma, inflammation, autoimmune disorders, surgery, and infections by the human herpes virus or Epstein-Barr virus could lead to the development of IMT, but the etiology is still unclear [11-13]. The clinical manifestation of IMT is variable based on the anatomical location of the lesions.

Patients generally present with a palpable mass with/without associated symptoms. The nonspecific symptoms can include fever, anemia, malaise, weight loss, abdominal pain, gastrointestinal symptoms, cough, and chest pain [14]. Imaging appearances showed an ill-defined soft tissue mass with heterogenous enhancement on CT and MRI [15-18]. Grossly, inflammatory myofibroblastic tumors may be fleshy, gelatinous, or firm, with a tan or white cut surface [14]. The mean tumor size was 6 cm ranging from 1 cm to >20 cm in the greatest dimension [9]. The clinical presentations and radiologic characteristics of inflammatory myofibroblastic tumor are nonspecific, so the diagnosis is predominately based on the histopathological features.

Histologically, IMT is characterized by myofibroblastic spindle cells with infiltration of plasma cells and lymphocytes. Three histological patterns have been described, as follows: A compact spindle cell pattern; a myxoid/vascular pattern; and a hypocellular fibrous pattern [20]. The myxoid/vascular pattern resembles nodular fasciitis, granulation tissue or other reactive processes, with loosely arranged plump spindle cells in an edematous myxoid stroma, abundant blood vessels and an infiltrate of lymphocytes, plasma cells and eosinophils. The compact spindle cell pattern resembles a variety of spindle cell neoplasms, with a compact fascicular spindle cell proliferation in a collagenous stroma. The hypocellular fibrous pattern resembles a desmoid fibromatosis or scar, with relatively fewer spindle cells in a densely collagenous stroma and scattered inflammatory cells. Immunohistochemically, tumor cells usually show positive staining for vimentin, Smooth Muscle Actin (SMA), desmin and muscle specific actin but negative staining for S-100, CD117, Myogenin [19,21].

The *Anaplastic Lymphoma Kinase* (*ALK*) gene locus on chromosome 2p23 plays a role in the development of IMTs [22,23]. The *Anaplastic Lymphoma Kinase* (*ALK*) gene encodes a *Receptor Tyrosine Kinase* (*RTK*) normally expressed only in nervous system and *ALK* gene rearrangement can lead to inappropriate activation of the *ALK*-receptor tyrosine kinase [24,25]. Approximately 50% to 70% of IMTs presents with *ALK* rearrangements [26,27]. *ALK*-positive IMTs occur more frequently in younger patients [22,27]. However, recent evidence using next-generation sequencing suggests *ALK*-negative IMTs demonstrate the gene fusions involving *ROS-1* (*ROS*

proto-oncogene 1), *RET* (*RET proto-oncogene*), PDGFRβ (Platelet-Derived Growth Factor Receptor β), and NTRK (Neurotrophic Tyrosine Receptor Kinase) [28-31].

Although the spontaneous regression of IMTs has been reported in various organs [32-37], complete surgical resection is recommended as the primary therapeutic approach for most patients with IMTs [38]. Complete surgical resection is the curative treatment and the best method for diagnosis [39]. Incomplete surgical resection is a risk factor for recurrence. If the tumor cannot be completely resected, radiation therapy, chemotherapy, anti-inflammatory agents, such as corticosteroids and Non-Steroidal Anti-Inflammatory Drugs (NSAIDS) can be considered [33,40].

However, there is no standard systemic treatment and different approaches have been used with varying success rate. *ALK* expression provides the opportunity to perform targeted gene therapy on this rare tumor [41]. As an *ALK* inhibitor, crizotinib has been used to treat the *ALK*-positive IMTs in recent case reports [42]. *ALK*-positive IMTs have demonstrated a favorable response to crizotinib [43-46].

Inflammatory myofibroblastic tumor was classified as a distinctive mesenchymal tumor of intermediate biological potential with a tendency for local recurrence (25%) and a small risk of distant metastasis (<5%) [47]. The lung, brain, liver and bone are the most frequent sites of metastasis [10]. Current research on reliable predictors of aggressive behavior in IMTs is limited. Tumor size, necrosis, mitotic activity and cellularity are not related to prognosis [26,48]. The presence of histological atypia, p53 overexpression, ganglion-like cells and aneuploidy pattern may be associated with a more aggressive behavior [48]. The *ALK*-negative tumors have a high risk of metastasis, however, *ALK* gene rearrangement does not seem to be associated with disease recurrence [26,27,49]. The prognosis of IMT is generally good, even in patients with unresectable disease and *ALK*-negative cases [50].

Conclusion

Inflammatory Myofibroblastic Tumor (IMT) arising from the abdominal wall is exceedingly rare. IMT of the abdominal wall occurred primarily in adults other than children or young adults, in contrast to other IMTs. The median age is 49 years old, ranging from 6 to 58 years in the 5 reported cases. The predominance of male gender was evident and the most frequent symptom was abdominal mass. Histopathological examination is helpful in differentiating IMT from other spindle cell lesions that can involve the abdominal wall. Complete surgical resection is recommended as the primary therapeutic approach. Chemotherapy, radiotherapy, anti-inflammatory agents can be considered in patients with unresectable tumor. Crizotinib could be used in *ALK*-positive inflammatory myofibroblastic tumor.

Author Contributions

Conceptualization, Nuerabula. Wujimaimaiti, Yi Wu and Renyi Qin; Investigation, Jingxiong Yuan; Resources and Data curation, Jingxiong Yuan; Writing-original draft, Nuerabula. Wujimaimaiti and Yi Wu; Writing-review and editing, Renyi Qin; Supervision, Renyi Qin. All authors have read and agreed to the published version of the manuscript.

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