

Received: 2015.09.20
Accepted: 2015.10.16
Published: 2016.02.12

Highly Vascularized Primarily Inflammatory Pseudotumor of the Omentum in an Adult Male: A Case Report

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Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Conflict of interest: None declared

Patient: Male, 38
Final Diagnosis: Inflammatory myofibroblastic tumor
Symptoms: Abdominal pain • anorexia • weight loss
Medication: —
Clinical Procedure: Operation
Specialty: Surgery

Objective: Rare disease
Background:

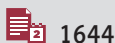
Inflammatory pseudotumors can affect any organ, whereas primary omental tumors are very rare. A few cases have been reported in the literature, all affecting adult patients. They are usually difficult to diagnose preoperatively and pathology remains the criterion standard for diagnosis. Surgical resection is considered the first-line treatment in limited disease, whereas recurrent or metastatic disease is treated by re-excision. There is no role for chemo- or radio-therapy in limited disease. Here, we present a rare case of omental myofibroblastic tumor in an adult male.

Case Report: A 38-year-old healthy man presented to our clinic complaining of lower abdominal pain associated with anorexia and low-grade fever, and he also reported weight loss. His initial hemoglobin was 9.7 g/dl. Magnetic resonance imaging (MRI) showed an enhancing solid mass in the lower abdomen, with close proximity to the appendix and the urinary bladder. The patient was treated successfully with laparotomy and excision of the tumor. Histopathology of the mass revealed spindle cells of vague fascicular pattern. Further immunohistochemical staining showed presence of reaction for CD68, CD34, and ALK. No omental infiltration was noted. No adjuvant treatment was applied and the patient was free of disease after 1-year follow-up.

Conclusions: Omental pseudotumors are a rare pathology. They are usually slowly-growing, circumscribed tumors with a low malignant potential. They have a predilection for children. The overall mortality is reported to be 5–7% in cases with multiple recurrences.

MeSH Keywords: Granuloma, Plasma Cell • Laparotomy • Omentum

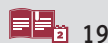
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Background

Inflammatory myofibroblastic tumors (IMTs) are rare benign lesions characterized histologically by fibroblastic proliferation associated with reactive inflammatory cells, mimicking a malignant process [1,2]. They were first described by Brunn in 1939 in the lung, one of their preferred sites [3]. However, they can affect any organ [1,2], with 43% of extrapulmonary IMT being omental in origin [4]. The most common primary omental tumors are leiomyosarcomas, leiomyomas, and GISTs, affecting people in their 5th and 6th decades of life. Omental IMF tumors are even more uncommon [5], with less than 15 cases reported in the world literature [6], and they are extremely rare in older patients [2]. Inflammatory pseudotumors are known by a variety of synonyms, including inflammatory plasma cell granuloma, fibroxanthoma, pseudo-lymphoma, histiocytoma, xanthomatous pseudotumor, pseudosarcomatous fibromyxoid tumor, inflammatory fibrosarcoma, cellular inflammatory pseudotumor, and inflammatory myofibrohistiocytic tumor [5,7,8]. This disease has a predilection for children, with a mean age of 10 years at presentation [9], and children account for most of the cases reported in the literature [5]. Girls appear to be slightly more commonly affected [10].

The etiology is still not very well understood. Some authors attribute it to an exaggerated response to tissue injury or infection (e.g., intracellular *Mycobacterium avium*, *Corynebacterium jejeuni*, *Bacillus sphaericus*, *Coxiella burnetii*, Epstein-Barr virus, *Escherichia coli*, and *Herpes*) [8], and there has been an association with trauma, surgery, tumors [11], it has been recently reported following Wilms tumor [12], presence of fever, elevated erythrocyte sedimentation rate (ESR), hypergammaglobulinemia, and inflammatory cells [11,13]. Others attribute it to an auto-immune mechanism, as there have been reports of associated vasculitis, inferior vena cava thrombosis, sclerosing cholangitis, and retroperitoneal fibrosis [11]. While some believe it is a true neoplasm, others believe that because of its potential for local infiltration, recurrence, and multicentricity, it could be a low-grade sarcoma with inflammatory cells [2].

Case Report

A 38-year-old previously healthy man presented with 3-week history of dull aching lower abdominal pain, anorexia, nausea, decreased PO intake, weight loss (10 kg over 30 days), and nocturnal low-grade fever and sweats. He denied any change in bowel habits, urinary symptoms, history of traveling, recent infectious processes, or trauma. His past medical and surgical history were otherwise insignificant apart from a previous right inguinal hernioplasty. Physical examination revealed low-grade fever of 37.9°C, and a soft, nondistended, nontender abdomen. However, a soft, mobile, suprapubic mass about 7 cm

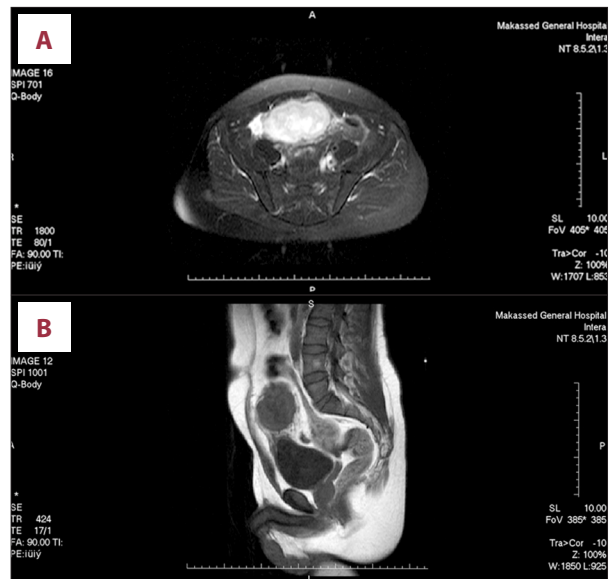


Figure 1. MRI of abdomen and pelvis showed a 10×7×9 cm, highly vascularized, thick-walled, necrotic mass in the lower abdominal cavity (A) with close proximity to the urinary bladder (B).

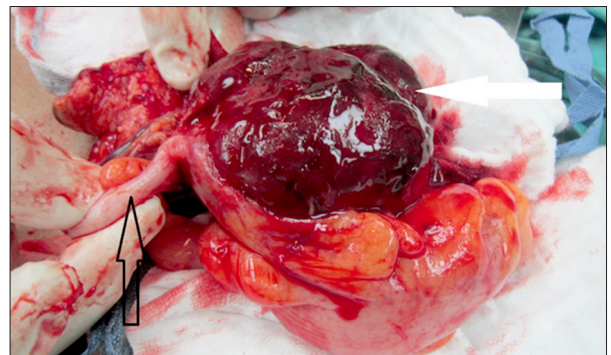


Figure 2. At laparotomy, the mass (white arrow) is located in the midabdomen, and attached to the omentum and to the apex of the appendix (black arrow).

in diameter was palpated, with no palpable lymphadenopathy. His initial lab results revealed hemoglobin 9.7 g/dl, white blood cell count 11.8, segmented 71%, platelets 817, and a high C-reactive protein (CRP) 121. An MRI of the abdomen and pelvis was done and revealed a 10×7×9 cm, thick-walled, necrotic, solid, and enhancing tumor in the anterior pelvic and lower abdominal cavity, adherent to the roof of the urinary bladder, adjacent bowels, and abdominal wall (Figure 1). The initial differential diagnosis was a sarcomatous tumor; therefore, an exploratory laparotomy was done. Intraoperative examination revealed a highly vascular mass, located in the midabdomen, attached to the omentum below the bladder and to the apex of the appendix (Figure 2). A moderate amount of intra-abdominal blood was noticed and sent for cytological examination. The patient underwent resection of the mass, omentectomy,

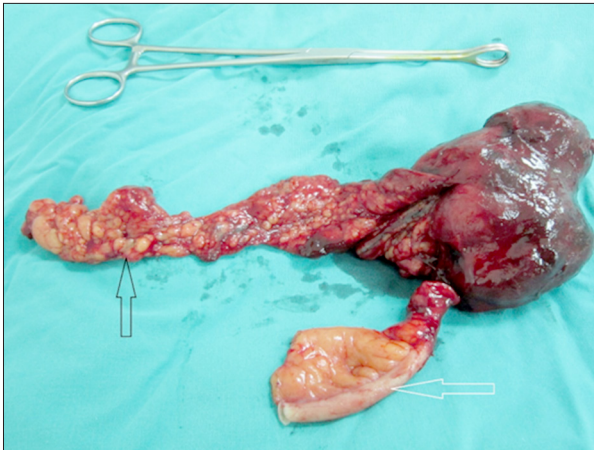


Figure 3. Specimen showing the omental resection with the tumor (black arrow) and resected appendix (white arrow).

and appendectomy (Figure 3). Thorough abdominal exploration showed no further pathology. Cytological examination of the fluid revealed red blood cells with a few scattered inflammatory cells. Histopathological examination of the mass showed a tumor with a mixture of spindle cells, fibroblastic and myofibroblastic, with polymorphous cell infiltrate, prominent rich vascularization, spindle cells of vague fascicular pattern with mild-moderate nuclear irregularities, and scattered rare mitotic figures. Further immunohistochemical staining showed absence of reaction for actin, desmin, EMA, CD117, but a positive CD68, CD34, and ALK (Figure 4). Omental tissue examination showed focal infiltration by the same tumor, and the appendix was normal. Therefore, the patient was diagnosed as having an omental inflammatory myofibroblastic tumor (pseudotumor). Postoperatively, the patient showed marked recovery, with resolution of his symptoms. Upon 1-year routine follow-up, he shows no signs of recurrence.

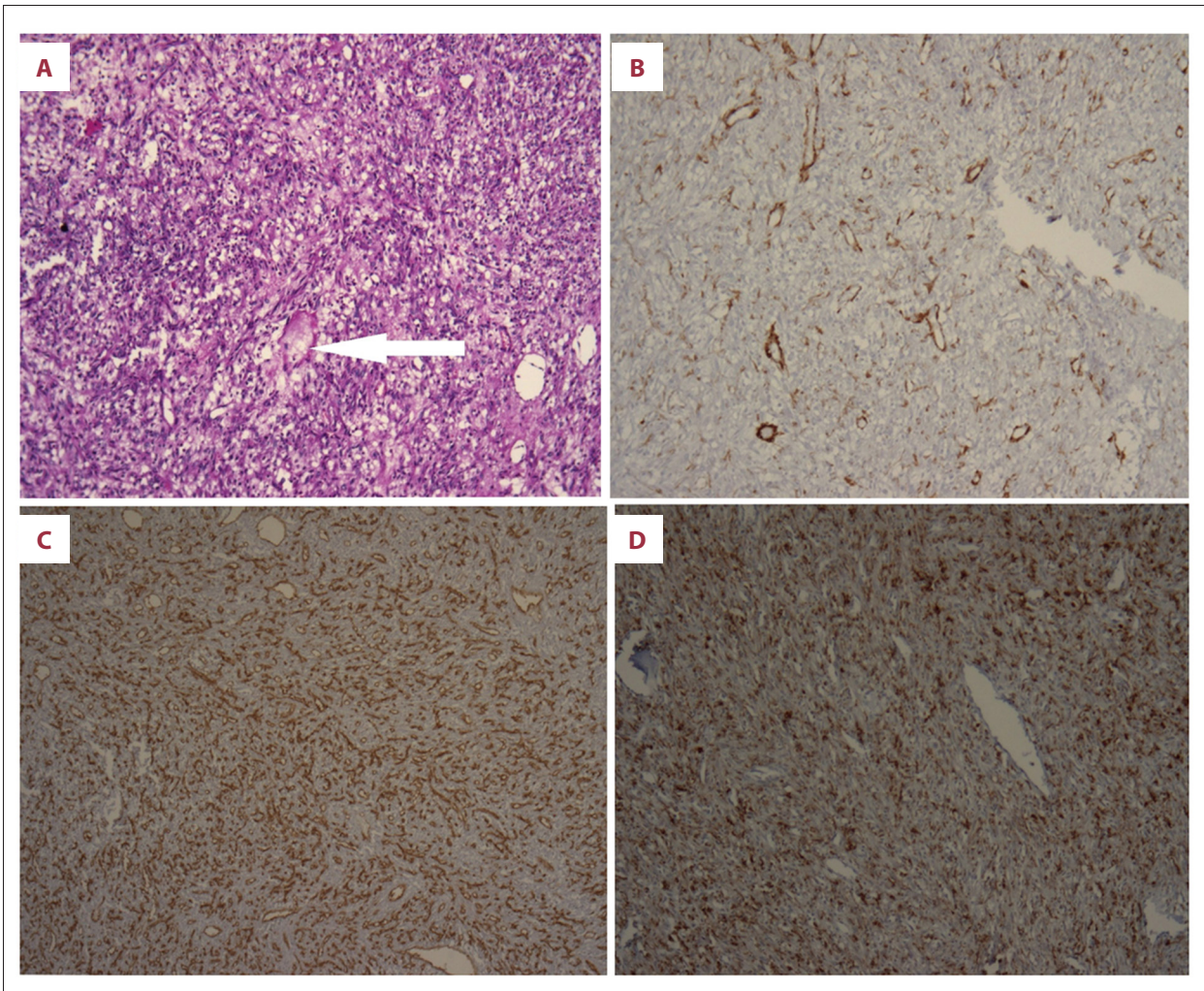


Figure 4. Spindle-shaped cells in short interlacing fascicles with nuclear irregularities (A, white arrow). Immunohistochemical analysis for actin shows negative staining in tumor cells (B) and for CD68 (C) and CD34 (D) positive staining.

Discussion

Pathology shows that IMTs are usually slowly-growing, circumscribed tumors with a low malignant potential [14]. They frequently present as a single, multinodular, firm mass, with a yellow-to-brownish surface, whorled, fleshy-cut surface, destroying the tissue that it invades. It may show focal areas of hemorrhage, necrosis, and calcifications ranging in size from 1 to 20 cm, with a mean size of 7 cm [14]. At surgery, omental-mesenteric disease typically has a malignant appearance, is highly vascular, and has variable adherence [15]. Microscopically, they appear as interlacing fascicles of myofibroblastic slightly atypical spindle cell component associated with chronic inflammatory stroma of lymphocytes/histiocytes/plasma cells with little mitotic activity and pleomorphism [1,15].

Immunohistochemistry shows that strong diffuse cytoplasmic reactivity to vimentin is typical for almost all IMTs, while reactivity to muscle-specific actin (SMA) varies from a focal to a diffuse pattern in the spindle cell cytoplasm. Reactivity to desmin is identified in many cases, while focal cytokeratin immunoreactivity is found in one-third of cases. However, IMTs show no reactivity to myogenin, myoglobin, S100 protein, or CD117. Likewise, TP53 immunoreactivity is rare, and it has been reported in association with recurrence and malignant transformation [16].

A distinctive aspect of IMT is the expression of atypical lymphocyte kinase 1 (ALK-1), which is present in up to 60% of cases, particularly in lesions occurring in young children [11,17]. FISH studies have shown that many IMTs harbor a recurrent clonal chromosomal abnormality involving the 2p23 region, specifically the region containing the ALK gene. The significance of structural changes in neoplastic disease is not known, but it has been implicated in hematologic disorders such as anaplastic large cell lymphoma (ALCL). ALCL and IMT may share an identical gene fusion, but no clinical link has been shown. These data, however, argue against a reactive nature for IMTs and firmly support a neoplastic process [17].

Most patients are asymptomatic, but 15–30% present with constitutional symptoms (e.g., unexplained fever, malaise, and weight loss). Local symptoms might include an incidentally detected abdominal mass as the most common presenting feature, and abdominal pain, but rarely intestinal obstruction, ascites, or vomiting [13]. Laboratory results might show hypochromic, microcytic anemia, thrombocytosis, polyclonal hyperglobulinemia, and/or an elevated ESR [15].

IMTs are usually difficult to diagnose preoperatively because the radiological imaging is characterized by pleomorphism with no characteristic findings [1]. Therefore, radiology is used to determine the respectability of the tumor rather than

differentiation between benign and malignant lesions [5]. They usually appear as well-defined solid iso- to hypo-echoic masses on ultrasound, with prominent vascularity on Doppler US [6]. Computed tomography (CT) scans show a large, irregular, intra-abdominal mass with well-defined to infiltrating margins. There may be associated areas of central hemorrhage, necrosis, and/or fibrosis. However, calcifications and lymphadenopathies are rare [1,9,10,15]. MRI shows the same findings as CT, with intermediate signal intensity on T1-weighted images and high signal intensity on T2-weighted images. T1-weighted images after Gd-DTPA show a variable enhancement pattern, depending on the relative amount of fibrous tissue and cellular material ± 6 [4]. Fine-needle aspiration (FNA) of these lesions usually shows scattered spindle cells and occasional multinucleated giant cells [6]. Although the age distribution, clinical picture, and/or imaging might occasionally give a preoperative clue about the diagnosis, a final histopathological result is the only definitive diagnosis of IMTs. Differential diagnosis includes abscesses, tuberculous granulomas, soft tissue sarcomas, lymphomas, calcifying fibrous tumor, benign fibrous tumors of the mesentery (e.g., mesenteric fibromatosis and sclerosing mesenteritis), and metastasis [5,9,18].

Complete surgical resection is the first-line management in limited disease, whereas recurrent or metastatic disease is treated by re-excision or metastasectomy [5]. There is no role for chemo- or radio-therapy in limited disease [9]. Although there is not yet an expert consensus that non-steroidal anti-inflammatory drugs (NSAIDs) play a role in the management of IMF tumor, there are occasional reports that these tumors (especially unresectable ones) can be successfully resolved with steroids, radiotherapy, chemotherapy, or even (NSAIDs) such as ibuprofen [5], and Cox-2 inhibitors.

Bertocchini et al. demonstrated dramatic response of diffuse abdominal IMTs to adjuvant anti-inflammatory drugs (e.g., ketorolac tromethamine) and chemotherapy (e.g., methotrexate vinblastine followed by ifosfamide, Adriamycin, and ifosfamide alone), an observation which opens a new door in the treatment of not only residual or recurrent IMT, but also for large tumors, which, for anatomical reasons, are difficult to resect [13]. Furthermore, Butrynski et al. presented impressive rates of tumor regression using Crizotinib, an ALK inhibitor, thus suggesting its possible use along with surgery in cases complicated by recurrences or in unresectable IMTs that may respond to Crizotinib, thus, facilitating their complete surgical removal. However, it was noticed that resistance develops in most patients within 1 to 2 years [2,19].

The disease may progress in 3 different ways: spontaneous regression, gradual growth, and rapid enlargement with local invasion. Local or regional recurrence is possible, and metastases are rare [1,9]. Signs, symptoms, and laboratory

abnormalities resolve with resection, and recur with recurrence [4]. Recurrences appear to be more common in extrapulmonary lesions, size >8 cm, locally invasive lesions, >1 organ involvement, lesions showing hyperdiploidy, retroperitoneal location, incomplete excision, atypia, and ganglion-like cells, but not in high tumor cellularity, mitosis, and necrosis. ALK positivity has recently been shown to predict lower risk of metastasis. Recurrences have been reported even after many years. Hence, regular follow-up is essential for early diagnosis of tumor recurrence, and it includes ultrasound, CT, labs (e.g., ESR, Hgb, and mammaglobin). Overall mortality is reported to be 5–7% in cases with multiple recurrences [4,6,11,15].

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Conclusions

Inflammatory myofibroblastic tumors of the omentum are very rare. They mainly affect children and are extremely rare in adults. They are difficult to diagnose preoperatively. Surgical resection using negative margins remains the first-line treatment of resectable intra-abdominal inflammatory pseudotumors.

Conflict of interest

The authors declare that there is no conflict of interest, financial or otherwise, related to the publication of this study or its findings.