Clinical Case Report

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Imaging findings of inflammatory myofibroblastic tumor from the greater omentum

One case report

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Abstract

Rationale: Inflammatory myofibroblastic tumors (IMTs) are rare neoplastic lesions with benign tendency. Even more rare are IMTs from the greater omentum (GO-IMT). A GO-IMT is easily misdiagnosed as other malignant tumors before operation; thus, clinicians need to be familiar with its imaging findings. Here, we report the imaging findings of a GO-IMT patient presenting with a pelvic mass.

Patient concerns: Ultrasound of the IMT in the pelvic cavity showed a hypoechoic mass. A computed tomography (CT) scan showed a nearly circular soft tissue mass with a clear border and heterogeneous density, and the surrounding tissues were pushed and compressed. Contrast-enhanced CT showed severe persistent enhancement in the lesion edges and mural nodules, but not in the central necrosis.

Diagnoses: Histopathology and immunohistochemistry confirmed that the mass was a GO-IMT.

Interventions: The tumor was resected after preoperative preparation.

Outcomes: No recurrence or metastasis was found during a short-term follow-up.

Lessons: The GO-IMT is an inferior epigastric mass in the periphery of the bowel, and is usually well-demarcated without calcification or lymphadenopathy. Contrast-enhanced CT showed a heterogeneous hypervascular mass where the center necrosis, the edge of the tumor, and the mural nodules can be partially reinforced.

Abbreviations: CT = computed tomography, GO-IMT = greater omentum inflammatory myofibroblastic tumor, IMT = inflammatory myofibroblastic tumor.

Keywords: CT, greater omentum, imaging, inflammatory myofibroblastic tumor

1. Introduction

The inflammatory myofibroblastic tumor (IMT) is a rare stromal tumor with intermediate biological potential.[1] The pathological features of an IMT are myofibroblastic-type spindle cells with varying degrees of plasma cell/lymphocyte infiltration, which is called an inflammatory pseudo tumor, inflammatory myofibroblastic proliferation, xanthomatous pseudo tumor, or plasma cell granuloma.[2,3] An in-depth pathology study has confirmed that the IMT is a solid tumor with occasional recurrence or metastasis.[4,5] An IMT may occur in almost any part of the body, from the central nervous system to the limbs, and has nonspecific clinical manifestations.[2] An IMT has diverse imaging findings, including a soft tissue mass with a clear or blurred boundary and marginal infiltration, accompanied by different proportions of inflammation and fiber components.[1] The imaging findings of IMTs are often confused with those of malignant tumors,[6] so clinicians need to be familiar with the imaging findings of different IMTs.

An IMT usually occurs in the lung, and the occurrence of an extra pulmonary IMT in the greater omentum is rare. So far, about 20 cases of IMTs originating from the greater omentum (GO-IMT) have been reported, but only half of these cases involve imaging findings.[2,7–17] We report a case of ultrasonography and computed tomography (CT) manifestations of a GO-IMT in a woman presenting with a pelvic mass, and review previous literature to summarize the imaging features of GO-IMTs.

1.1. Consent

The clinical and imaging data were obtained with the patient's signature for use in the publication of scientific research papers.

2. Case report

A 25-year-old female patient, who was found to have a pelvic mass during a physical examination after pregnancy and who experienced abdominal distention and discomfort in the lower abdomen for more than 1 month, came to our hospital to

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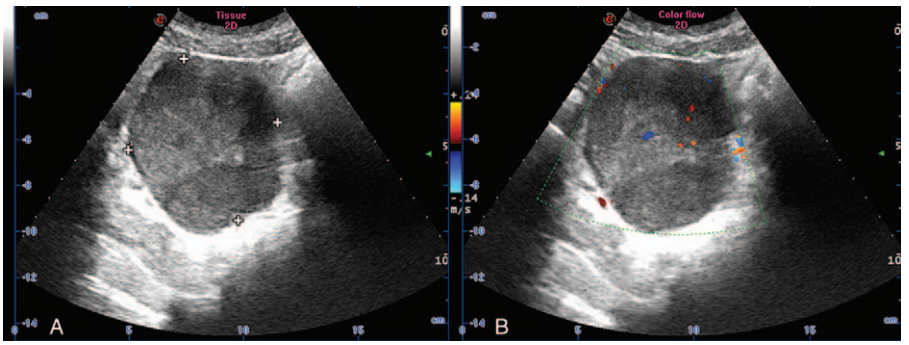

Figure 1. Ultrasound showed a hypoechoic mass. The mass had a clear boundary (A) and blood flow signals (B).

continue treatment after inducing abortion for the surgical removal of the pelvic mass. She had no abdominal pain, nausea, vomiting, or other symptoms. She had undergone an operation of the right lower limb for a trauma more than 10 years ago. She denied alcohol or tobacco habits or a history of drug addiction. During palpation, a movable soft tissue mass was felt at the entrance of the pelvis. Routine tests of blood, liver, and kidney functions were not significantly abnormal. Blood tumor markers (eg, CEA, CA199, CA125) were not significantly elevated. Ultrasound examination showed a hypoechoic occupancy (about $6.6 \times 7.4\,cm^2$) in the pelvis with a clear boundary and blood flow signals (Fig. 1). CT scan showed a nearly circular soft tissue mass with a clear border and heterogeneous density, whereas the surrounding tissues were obviously pushed and compressed. Contrast-enhanced CT scan showed that the lesion edges and mural nodules were significantly enhanced in the arterial phase, but not in the venous phase or the center necrosis area (Fig. 2). No obvious enlarged lymph nodes were found around the lesion or in the pelvic cavity. A small amount of effusion was found in the pelvic cavity.

The tumor was resected under laparoscopy after the completed preoperative preparation. A mass from the greater omentum measuring about $6.0 \times 6.0 \times 7.0\,cm^3$ was found in the uterus-rectum fossa. This mass had a smooth surface and no adhesion to the surrounding organs, and the womb and the double appendix had no special case, with a small amount of pale yellow effusion in the pelvis. The greater omentum along the greater curvature of the stomach contained several small nodules, liver exploration was unremarkable, and no other abdominal and pelvic tumors were found. The greater omentum mass and nodules were resected. Intraoperative fast frozen pathological examination showed that the lymph nodes were reactive hyperplasia. The resected mass was grayish white with a complete capsule, and, under microscopy, the spindle tumor cells were arranged in bundles while a number of inflammatory cells infiltrated the stroma (Fig. 3). Immunohistochemistry showed ALK (+), CD117 (partial +), Desmin (partial +), CD34 (vascular +), SMA (+), DOG-1 (partial +), Ki-67 (5%, +), CK (+), S-100 (−), BcL-2 (−), and CD99. The final pathological diagnosis was GO-IMT. The patient recovered after surgery and no recurrence was observed in the short-term follow-up.

3. Discussion

The etiology and incidence of IMTs are still unclear, but some cases are reported related to trauma, surgery, and inflammation.[1] In this case, although the patient has a history of lower limb trauma and related surgical treatment, the occurrence site is different from the IMT in the pelvic cavity. The incidence rates of IMTs are not significantly different between sexes, but are especially high in children and young people, since the average age of onset reported by Coffin is 9 years old.[2] Moreover, the onset age of abdominal IMT in children and young people is delayed to 14 and 25 years old, respectively.[10,18] The onset age of GO-IMT ranges from 1.5 to 38 years, with a mean age of 13 years.[7–17] The clinical symptoms of IMTs vary with the occurrence sites, but the clinical symptoms of GO-IMTs are nonspecific, including low fever, abdominal mass, and gastrointestinal symptoms.[7–17] Laboratory tests also lack specificity and show negative tumor markers.[12] The case presented in this report occurred during pregnancy, which was different from previous reports. Typical IMTs generally have good prognosis, but special attention should be paid to the highly aggressive variant of IMT, epithelioid inflammatory myofibroblastic sarcoma (EIMS).[19] This atypical IMT occurs in the mesentery

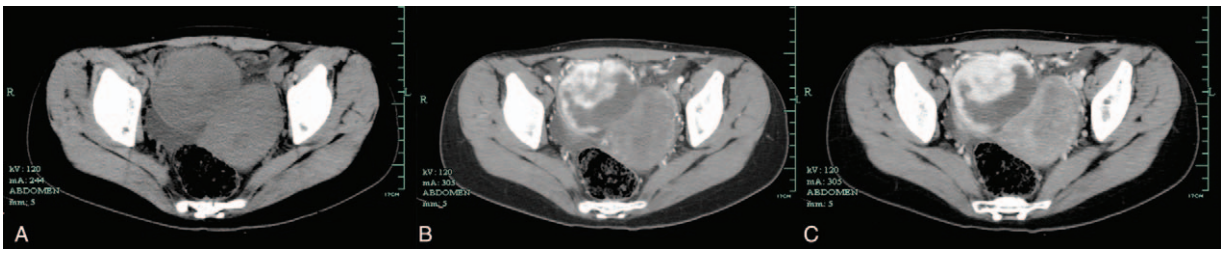

Figure 2. Abdominal unenhanced and contrast-enhanced CT scan showed a nearly circular soft tissue mass. Uniform density was observed in an unenhanced CT scan (A), and lesion edges were enhanced in arterial phase (B); the lesion edges were constantly enhanced and obvious mural nodules could be seen in venous phase (C) and delay period (D). CT = computed tomography.

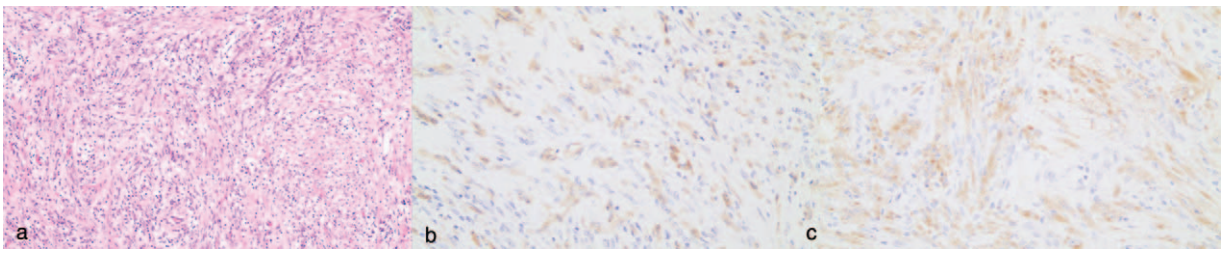

Figure 3. Histopathological examination showed the spindle cells arranged in bundles. A large number of inflammatory cells infiltrated the stroma (H&E × 100; A). Immunohistochemical staining with SMA (B) or ALK (C) was positive.

or omentum of male adults above age 40, and has the pathological features of epithelioid morphology and nuclear membrane or perinuclear anaplastic lymphoma kinase (ALK).[19,20] A recent study showed the pathogenesis of EIMS includes ALK fusion mechanisms and fusion with the RRBP1 gene.[21] Though no genetic test was performed, this was the case of omental IMT. EIMS was not considered in this case due to the lack of epithelioid morphology in pathological tissues.

The occurrence sites of GO-IMT are not fixed, which locate in the peritoneal cavity and the pelvic cavity around the intestine, under the abdominal wall.[7–17] GO-IMTs can present as a solitary nodule, nearly circular masses, or with an irregular shape, and occasionally appear in multiples.[7–17] The lesions of GO-IMTs are usually larger, with sizes from 3.6×5.5 to $11.5 \times 15.0\,cm^2$.[7–14,16,17] Ultrasonic examination shows that the lesions are homogeneous or inhomogeneous hypoechoic lobulated masses,[7–10,12,13] accompanied by blood flow.[9,10,13] CT examination shows that the lesions are often soft tissue masses with a clear boundary, uniform or nonuniform density, necrosis, and occasionally a thin coating.[7–11,13–15] On contrast-enhanced CT scans, the lesions show heterogeneous enhancement[7,11,13,14] or edge enhancement.[8,15] One case of an omental IMT with magnetic resonance imaging findings shows hypointensity in T1-weighted imaging and hyperintensity in T2-weighted imaging.[16,17] The GO-IMT in this case had clear borders and was manifested on the contrast-enhanced CT as a hypervascular mass, similar to previous reports. However, in the current case of the IMT, there is necrosis in the lesion center and mural nodules are obvious, which is different from other reports. The imaging differential diagnosis of GO-IMT include malignant tumors such as metastatic tumor, peritoneal carcinomatosis, sarcoma, or lymphoma.[10,13] It should be noted that IMT can have simultaneous involvement in the mesentery and greater omentum.[7,8] Coffin found that more than 40% of peritoneal IMTs occurred in the mesentery and/or greater omentum, whereas a small portion accumulated in both areas, and IMTs in these areas could be multiple.[2] Therefore, when imaging a mass that is considered to be an IMT of the mesentery or greater omentum, it is necessary to carefully observe for multiple lesions and simultaneous involvement. A GO-IMT can be seen in a small to large number of ascites, without calcification or being accompanied by enlarged lymph nodes.[7–17] A recent study confirmed that there is no calcification in abdominal cavity or pelvic cavity IMTs, which has a different occurrence site from other IMTs.[22] Therefore, IMTs should be included in the differential diagnosis of masses in the mesentery and greater omentum, especially when no calcification is present.

Both solitary and multiple GO-IMTs can be resected by surgery with good prognosis.[7–11,13,14,16,17] Chemotherapy is commonly used for unresectable GO-IMTs, including adriamycin and ifosfamide, which can benefit partial cases.[12] However, the prognosis of EIMS is poor, as it may easily induce relapse or death after surgery.[19,20] Therefore, with a deep understanding of the molecular tumorigenic mechanisms of IMT, targeting drugs may be an expected choice for treating invasive IMTs.[23]

4. Conclusions

Greater omentum inflammatory myofibroblastic tumors usually manifest as inferior epigastric masses around the intestine, and can be multiple and necrotic, but without calcification or enlarged lymph nodes. Contrast-enhanced CT shows a hypervascular mass and, when the tumor has center necrosis, it can be partially enhanced on the edges and occasionally has mural nodules. The prognosis of a GO-IMT is type-dependent.

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