

Low-grade myofibroblastic sarcoma with abdominal pain, a stuffy nose, hearing loss, and multiple cavity effusion: a case report and literature review

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Abstract

Low-grade myofibroblastic sarcoma (LGMS) is a rare, low-grade, malignant tumor and is mainly composed of myofibroblasts with varying degrees of differentiation. LGMS results in considerable diagnostic difficulty. We report a case of LGMS that occurred in multiple organs, including the diaphragmatic pleura, and head and neck region. A 34-year-old man was hospitalized in 2014 after coughing and shortness of breath for 10 days, and abdominal distension, abdominal pain, and bilateral lower extremity edema for 4 days. Before this admission, he had an abdominal tumor diagnosed in 1994 and 2003, a nasopharynx tumor in 2010, and a temporal lobe tumor in 2013. All tumors were resected surgically and the diagnosis was atypical fibrous histiocytoma and atypical fibrous xanthoma. Before surgeries for these tumors, no positron emission tomography-computed tomography (PET-CT) or whole-body scans were performed, and after surgery, there was no follow-up. After thoracoscopy and PET-CT after the most recent admission, the patient was diagnosed with LGMS with metastasis to the bone, nodes, and thoracic and abdominal cavities. The patient was discharged with albumin infusion treatment. Although LGMS is rare, it is potentially serious. Therefore, clinicians should be aware of such disease and make an early diagnosis and perform close follow-up.

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Keywords

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Introduction

Low-grade myofibroblastic sarcoma (LGMS) is a rare, low-grade, malignant tumor. These tumors are derived from mesenchymal tissue and are mainly composed of myofibroblasts with varying degrees of differentiation. LGMS occurs more often in adults and predominantly in men.¹ Because of the feature of low malignancy of LGMS, it is easily confused with inflammatory myofibroblastic tumor, leiomyoma, and fibromatosis, which results in considerable diagnostic difficulty. This disease occurs mainly in the head and neck region, and rarely invades multiple organs.^{2,3} We report here a rare case of LGMS that occurred in multiple organs, including the diaphragmatic pleura, and head and neck region. Because surgery and radiotherapy are not suitable for patients with multiple metastases, novel systemic treatments are required to improve the cure rate.

Case presentation

A 30-year-old man was hospitalized on 14 February 2014 after coughing and shortness of breath for 10 days because of multiple serous cavity effusions, including bilateral pleural effusion and peritoneal effusion.

In 1994, the patient had a laparotomy in the People's Hospital of Fengdu County because of abdominal pain. During the operation, fat substances were discovered, but no pathological examination was performed. In 2003, the patient underwent resection of a benign tumor in the left

lower abdomen, but did not receive a post-operative pathological examination. On 13 December 2010, the patient was admitted to the Department of Oto-rhino-laryngology, Xinqiao Hospital, Third Military Medical University, for recurrent epistaxis, nasal resistance, painless and bloody nasal discharge, and hearing loss of the right ear for 5 months. Nasopharynx computed tomography (CT) suggested a space-occupying lesion on the right side of the nasopharynx. Video laryngoscopy indicated a smooth bilateral nasal mucosa, deviated septum, and a nascent object in the right nasal cavity. Surgery was performed during hospitalization and a pathological examination suggested atypical fibrous histiocytoma. On 11 January 2011, the patient was admitted to the Department of Neurosurgery, Xinqiao Hospital of the Third Military Medical University because of a right-sided stuffy nose for half a year and hearing loss in the right ear for 2 months. A nasopharynx biopsy suggested that the patient had an atypical fibrous tissue tumor. Cranial magnetic resonance imaging showed a space-occupying lesion in the right temporal lobe and a pathological examination showed histiocytosis and granulomatous lesions. Surgery was performed during hospitalization. A postoperative pathological examination suggested atypical fibrous xanthoma in the right nasal cavity and right temporal area. He had no follow-up or other treatment.

The patient was a pharmacist with a 10-year smoking history of two packs of

cigarettes per day. He had been married for 10 years and had a daughter and a son. His parents were closely related (i.e., crossed-cousin marriage). The patient had a younger sister who was healthy.

Ten days before hospitalization, the patient began to suffer from coughing and dyspnea. Four days before hospitalization, the patient began to experience abdominal distension, abdominal pain, and bilateral lower extremity edema. Pleural effusion puncture was performed and the contents of pleural effusion showed the following: protein level of 25.2 g/L; leukocyte count of $3604 \times 10^6/L$; 17% multinucleated cells; 77% monocytes; and 6% mesenchymal cells. These findings suggested the presence of exudate. Additionally, the adenosine deaminase level in pleural fluid was 8 IU/L. His liver function parameters were as follows: total protein, 40.3 g/L, albumin, 26.2 g/L, and pre-albumin, 85.2 g/L. With regard to tumor markers, CA125 was 195.7 U/ml and the other markers were normal.

The patient underwent thoracoscopy and white nodules were found on the pleura of the left chest (Figure 1a). A biopsy showed nodules with a tough texture. The blood vessels of the parietal pleura and visceral pleura were tortuous with a smooth surface. Based on a postoperative pathological examination, the patient was diagnosed with LGMS (Figure 1b). Positron emission tomography-CT showed a thickened peritoneum and omentum, elevated fluorodeoxyglucose (FDG) metabolism, and a maximal standard uptake value (SUV) of 11.9, which were indicative of metastasis (Figure 2). The patient also had multiple sites of uneven bone density throughout the body (including areas of bone destruction), increased FDG metabolism, and a maximal SUV of 8.2, which were also indicative of metastasis. Moreover, the patient showed enlarged lymph nodes in the right side of the neck, para-sternal region, and abdominal cavity, as well as enhanced FDG metabolism and

maximal SUVs of 1.9, 3.7, and 10.5, respectively. The patient also showed effusion on both sides of the pleural cavity and in the pelvic abdominal cavity.

The patient was diagnosed with LGMS, which occurred on diaphragmatic pleura associated with systemic multiple bone metastasis, pleural metastasis, peritoneal metastasis, and systemic multiple lymph node metastasis. Because the patient's parents were closely related, a chromosomal examination was performed, but did not show any abnormalities. The patient had lost the chance for having surgery and the disease was considered to be insensitive to chemotherapy. Therefore, the patient was discharged and symptomatic and supportive treatment with albumin infusion was provided.

Written informed consent for participation and publication was obtained from the patient. The associated clinical trial obtained ethics approval from the Ethics Committee of the Second Affiliated Hospital, Military Medical University. A copy of the consent form is available for review by the Editor of this journal.

Discussion

LGMS is a rare, low-grade malignant tumor, with few reports on this condition.^{4,5} Tumors of LGMS are derived from mesenchymal tissue and are mainly composed of myofibroblasts with varying degrees of differentiation. Because LGMS has low malignancy, it is easily confused with inflammatory myofibroblastic tumor, leiomyoma, and fibromatosis, which results in considerable diagnostic difficulty. The patient examined in this study had been diagnosed with atypical fibrous histiocytoma and histiocytosis and granulomatous lesions. The patient was diagnosed with LGMS only in the third histopathological examination. To diagnose LGMS, it needs to be distinguished from the following

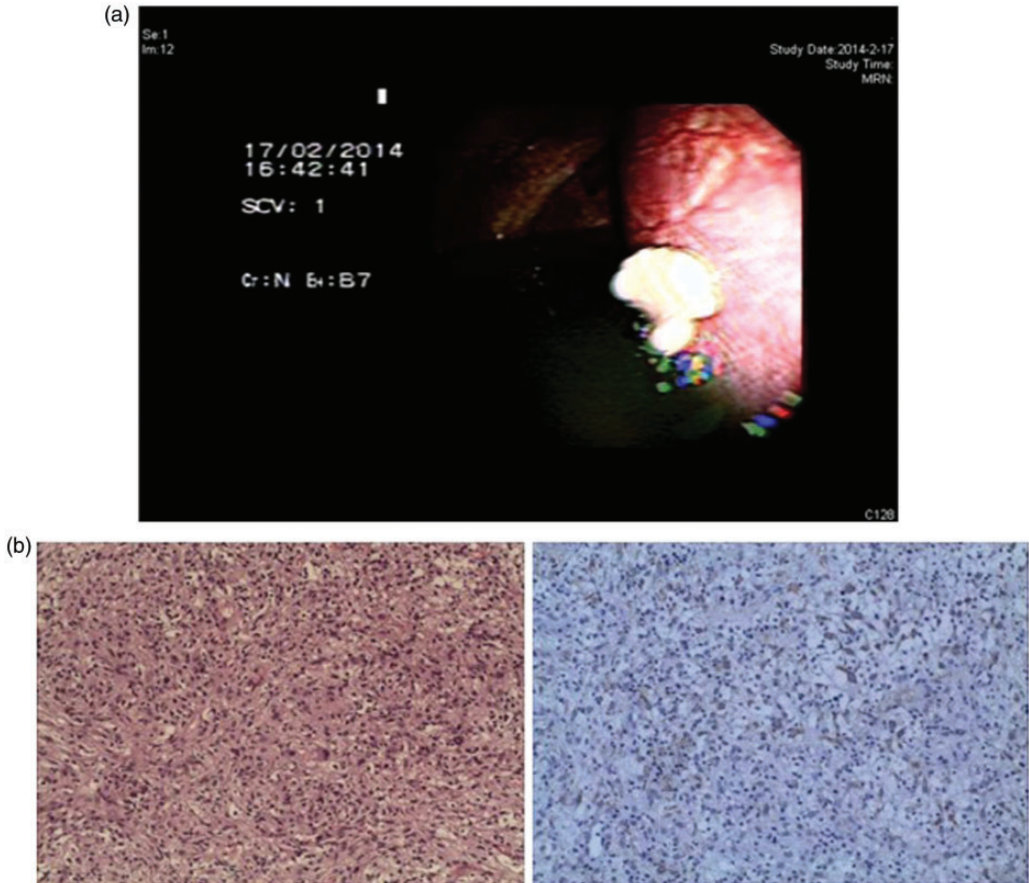


Figure 1. (a) A thoracoscopic examination shows white nodules with a tough texture and tortuous blood vessels with a smooth surface on the chest pleura. (b) A postoperative pathological examination shows a hypercellular area containing proliferation of myofibroblasts with hyperchromatic enlarged and pleomorphic nuclei on a background of abundant intercellular collagen. There is no infiltration of inflammatory cells (200 \times). An immunohistochemical examination shows positive staining of tumor cells for smooth muscle actin, suggesting low-grade myofibroblastic sarcoma (200 \times).

diseases. First, LGMS should be differentiated from inflammatory myofibroblastic tumor. This type of tumor and LGMS have properties of myofibroblasts and may show a pathological mitotic count. However, under a microscope, inflammatory myofibroblastic tumor displays apparent infiltration of inflammatory cells, while LGMS shows no infiltration of these cells, including lymphocytes and plasma cells. Second, LGMS should be identified from

fibromatosis. Lesions of LGMS feature many myofibroblasts, but their major arrangement consists of parallel wide bundles with a slightly wavy appearance and apparent surrounding infiltration covering the skeletal muscle. Additionally, these fibroblasts are closely related to tendons and fascia.⁶ Third, LGMS should be identified from leiomyoma. Tumor cells show an abundant eosinophilic fibrous cytoplasm and clear boundaries, and have a relatively

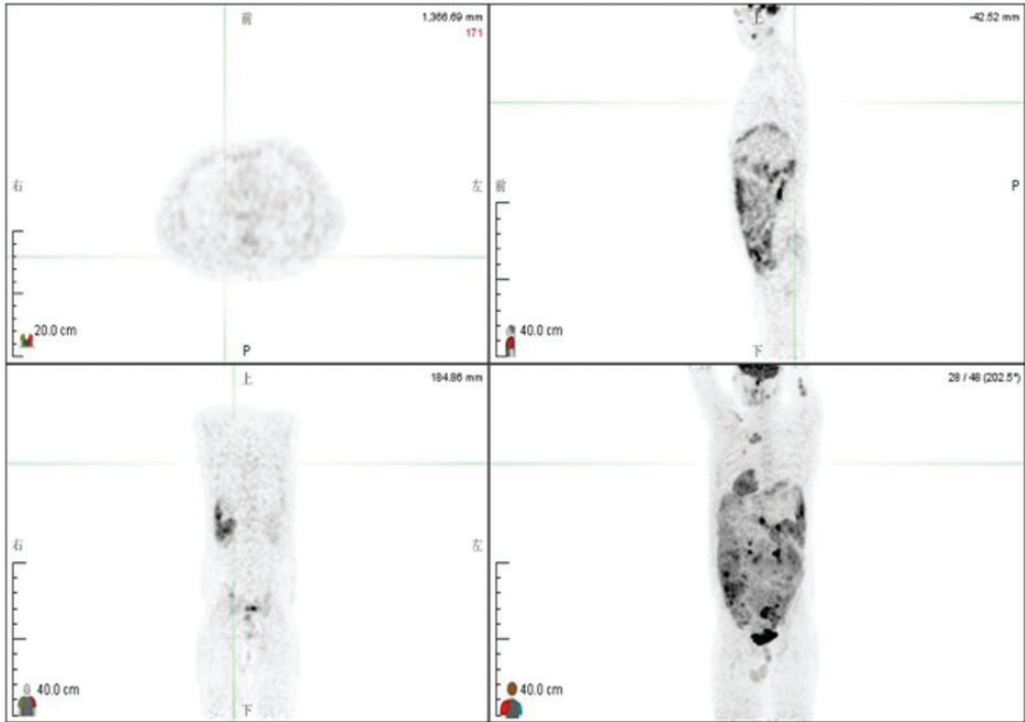


Figure 2. Positron emission tomography-computed tomography shows elevated fluorodeoxyglucose metabolism in a thickened peritoneum and omentum, with a maximal standard uptake value of 11.9. There is also elevated fluorodeoxyglucose metabolism in multiple sites of uneven bone density, with a maximal standard uptake value of 8.2. Additionally, fluorodeoxyglucose metabolism is elevated in enlarged lymph nodes in the right side of the neck, parasternal region, and abdominal cavity, with maximal standard uptake values of 1.9, 3.7, and 10.5, respectively. Positron emission tomography-computed tomography also shows effusion in bilateral sides of the pleural cavity and in the pelvic abdominal cavity. All of these findings indicated metastasis.

regular arrangement (weave-like pattern) in the smooth muscle, while the nuclei show a cigar-like appearance.

LGMS displays no prominent clinical features and has the most common symptom of gradually enlarging and painless masses. Although certain patients with LGMS may have localized painful tumors underneath the skin or fascia, most tumors reside in deep soft tissue. Some tumors have clear boundaries, but in most cases, these tumors display irregular infiltration along the connective tissue gaps and can invade into the muscle or bones. This type of

tumor typically grows locally and rarely undergoes metastasis.⁶ However, in our patient, the tumor metastasized in the pleura and peritoneum, as well as into the lymph nodes and multiple bones, resulting in tumors of different sizes of diameters of 1.5 to 12 cm.

LGMS occurs most often in adults, and predominantly in men. This disease occurs mainly in the head and neck region, especially in the tongue, although lesions in the abdominal cavity, groin, distal femur,⁷ sacrum, pubis, palm, breast, skin, and heart have also been reported. Our patient

had tumors occurring successively in the nasal cavity and temporal area and on the diaphragmatic pleura.

LGMS has a good clinical course and favorable prognosis. Surgery is the main treatment method for LGMS and generally has a good postoperative prognosis. However, this tumor has a propensity for local recurrence, although its distant metastasis is rare. As such, a widely accepted surgical approach is to perform extended resection and close follow-up to prevent recurrence because multiple episodes of recurrence can augment the malignancy. LGMS is not sensitive to radiotherapy and chemotherapy. However, local radiotherapy can allow patients to have a longer survival period.⁸ Therefore, for patients who are suitable for resection, surgery is the favored option.⁹ Because our patient suffered from multiple metastases in the pleura and peritoneum and in several bones, he was suitable for neither surgery nor radiotherapy and could only be provided with symptomatic and supportive treatment. To date, this patient is still alive.

In conclusion, LGMS has a low incidence and rarely invades multiple organs. Because surgery and radiotherapy are not suitable for patients with LGMS and multiple metastases, novel systemic treatments are required to improve the cure rate.

Abbreviations

LGMS, low-grade myofibroblastic sarcoma; CT, computed tomography; FDG, fluorodeoxyglucose; PET, positron emission tomography; SUV, standardized uptake value

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Authors' contributions

Wu XL, Zheng Y, and Fan BJ analyzed and interpreted the data of the patient. Li SY collected the data of the patient. Zhou CX and Guo L were major contributors in writing the manuscript. All authors read and approved the final manuscript.

Availability of data and material

Not applicable.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

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