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CASE REPORT

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Postoperative recurrence of myxoid liposarcoma of left thigh with pericardial metastasis: A case report

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Abstract

Myxoid liposarcoma (MLS) is a common subtype of liposarcoma in children and adolescents and can occur anywhere in the body. Cardiac metastases from MLS are very rare. We report a rare case of postoperative recurrence of MLS in the left thigh with ectopic and metachronous pericardial metastases. Cardiac metastases from MLS are rare, the prognosis is poor, and long-term follow-up of patients after discharge is necessary.

KEYWORDS

case report, magnetic resonance imaging, metastasis, myxoid liposarcoma, pericardial, tomography, X-ray computed

1 | INTRODUCTION

Myxoid liposarcoma (MLS) is a translocation-related sarcoma. It has a translocation with FUS-DDIT3 or rarely EWSR1-DDIT3 fusion. Some MLS pathology has round cell areas within the tissue that represent histologic progression to high-grade tumors. Round cells account for more than 5% of total cells and have a high malignant and metastatic potential. MLS is a common subtype of liposarcoma in children, adolescents, and young adults and can occur anywhere in the body, mostly in the deeper tissues of the extremities, especially the thighs. MLS has an unusual propensity for extrapulmonary metastases. Clinically, distant metastases to retroperitoneal, abdominal, and bone sites occur in about one-third of MLS patients, but cardiac metastases are very rare. This text reports a case of pericardial metastasis in a patient with MLS originating in the left thigh.

2 | CASE REPORTS

A 53-year-old man with a history of MLS in the left thigh was treated with wide excision at local hospital. No metastases were found anywhere in the patient's body at that time, and no adjuvant therapy was given after surgery. 2 years later, the patient found an "egg"-like swelling above the knee on the left lower extremity, without pain or fever. After tumor resection, histological findings of the resected specimen revealed large number of hyperplastic small vessels in a fibro-mucinous background; the tumor cells are diffusely distributed in the mesenchyme, dense, asteroidal or irregular in shape, with abundant cytoplasm and red staining, a round cell component of 15%, nuclear splitting about 7/10 HPF. Adjuvant chemotherapy with two cycles of epirubicin (60 mg/m^2/d on days 1–2) and ifosfamide (30 mg/m^2/d, on days 1–3), mesna (18 mg/m^2 on hours 0, 4, and 8), and apatinib capsules

(250 mg) was administered. This regimen was repeated every 3 weeks. During the treatment, the patient's condition was stable and received adjuvant radiotherapy at the same time. 2 months after the second surgery, the patient developed chest tightness and wheezing and lower limb edema; he could not lie flat at night. Physical examination reveals an enlarged cloudy heart and audible pericardial fricative sounds. A mass measuring approximately 5.0 × 4.0 cm was palpable in the left groin and a mass measuring approximately 1.8 × 1.5 cm was palpable in the left popliteal fossa, which were tough, painless, and fixed. The tumor markers revealed that tumor-associated antigen 125 level was increased to 192.20 U/ml (normal range 0.01–35 U/ml).

Imaging examinations: (1) Digital radiography (DR) examination shows an enlarged heart shadow with a "pear-shaped" appearance and a cardiothoracic ratio of approximately 0.65 (Figure 1). (2) Ultrasonography (US) shows an enlarged left atrium with a cystic solid component mass measuring approximately 10.2 × 9.9 cm in the pericardial cavity to the left of the heart, squeezing the heart forward (Figure 2). (3) Computed tomography (CT) examination shows a cystic hypodense shadow with well-defined borders on the left edge of the pericardium, with a CT value of approximately 22 HU, and no significant enhancement on enhancement. The left ventricle was compressed, and the left ventricular myocardium was thickened compared to the right ventricular myocardium (Figure 3). Left pericardial cyst is considered. (4) Cardiac magnetic resonance (CMR) examination shows a mass measuring approximately 10.9 × 7.3 × 10.4 cm (LR×AP×SI) in the pericardial region outside the lateral wall of the left ventricle was clearly demarcated from the myocardium, and no significant infiltration was seen. The mass shows isointensity on T1-weighted images (T1WI), slightly mixed hyperintensity on T2-weighted images (T2WI), and significantly inhomogeneous high signal on fat-saturated T2WI, with speckled low signal seen locally. Delayed enhancement scan shows a strip of flocculent soft-tissue density shadow with heterogeneous mild enhancement within the lesion (Figure 4). The left ventricular chambers were compressed and diastolic motion was limited, with a reduction in end-diastolic and end-systolic volumes of 43.48 and 10.88 ml, respectively. (5) Lower extremity magnetic resonance imaging (MRI) examination shows irregularly shaped mass measuring approximately 6.3 × 5.7 × 6.4 cm (LR×AP×SI) with still clear borders in the left inguinal region. The mass shows low signal on T1WI and significantly inhomogeneous high signal on fat-saturated T2WI. The lesion was heterogeneously enhanced on enhancement scan and was poorly demarcated from the left external iliac artery. A round-like mass measuring approximately 2.4 × 3.0 × 3.0 cm (LR×AP×SI) was seen on the left posterior femur, the mass shows low signal on T1WI, and mixed hyperintensity on fat-saturated T2WI, irregular strips of low signal are seen within it, edge undercooling, enhancement shows significant inhomogeneous strengthening (Figure 5). (6) Positron emission tomography (PET) examination shows large lamellar hypodense shadow in the left pericardium with slightly concentrated radiological distribution, SUV_{max} about 2.6, CT value about 16 HU. Multiple hypodense masses were seen in the left inguinal region and left popliteal fossa with slightly dense radiological distribution, SUV_{max} about 2.9, larger size about 3.4 × 5.4 cm, CT value about 26 HU (Figure 6).

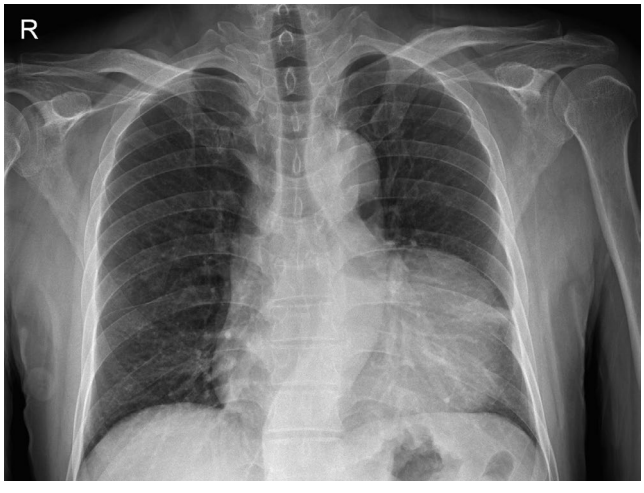

FIGURE 1 Digital radiography shows an enlarged heart shadow with a "pear-shaped" appearance and a cardiothoracic ratio of approximately 0.65

Patient undergoes CT-guided pericardial mass puncture, the pathological tissue after puncture shows a dense arrangement of homogeneous round-forming lipocytes and large, circular, ring-like adipocytes on light microscopy, and pathological nuclear schizophrenia was common. Immunohistochemistry shows AE1/AE3 (-), Vimentin (+), SMA (-), Desmin (-), CD34 (vascular+), S-100 (-), INI-1 (+), Caldesmon (-), CD68 (-), and Ki-67 (10%+). Clinician diagnosed MLS pericardial metastasis in combination with immunohistochemistry and previous medical history (Figure 7).

The patient and his family refused to undergo pericardial surgery and continued with the previous adjuvant regimen for the 3rd cycle of chemotherapy, along with liver-protective, stomach-protective, and immune-boosting drugs. At the end of chemotherapy, the patient's condition stabilized and the patient and his family requested to be discharged, and the patient died at 5 months of follow-up after discharge.

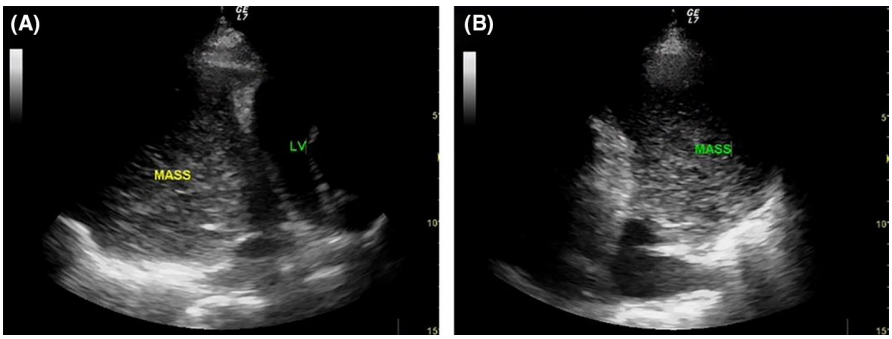

FIGURE 2 Ultrasonography shows an enlarged left atrium with a cystic solid component mass measuring approximately 10.2 × 9.9 cm in the pericardial cavity to the left of the heart (A), squeezing the heart forward (B)

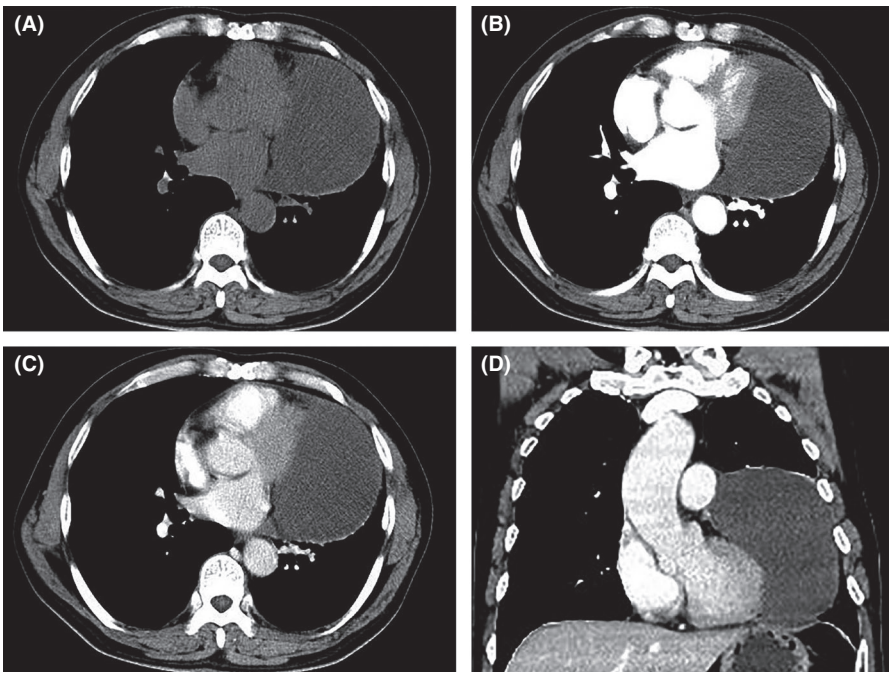

FIGURE 3 Computed tomography scan shows a cystic hypodense shadow with well-defined borders on the left edge of the pericardium, with a CT value of approximately 22 HU (A); arterial phase shows no significant enhancement of the lesion (B); venous phase shows thickening of the left ventricular myocardium compared to the right ventricular myocardium (C); venous phase coronal image shows left ventricular compression with lesion wrapping around the heart (D)

3 | DISCUSSION

Liposarcomas are the second most common soft-tissue sarcoma; there are four subtypes of liposarcoma confirmed by WHO.[1] MLS accounts for about 5% of all soft-tissue sarcomas and 15%–20% of all liposarcomas. It is usually located in deep soft tissue of the limbs, and more than half of the cases are seen in the thigh muscles.[2] MLS is a translocation-related sarcoma and is characterized by translocation with FUS-DDIT3 or EWSR1-DDIT3 fusion, as a result of the t (12;22) or t (12;16) balanced translocation on the 12q13.3 locus.[3] The fusion protein produced by these fusion genes acts as deregulated and activated transcriptional factors; it stimulates the proliferation of tumor cells.[2-4] Estourgie et al. [5] reported that 55% of MLS patients with metastatic disease had extrapulmonary metastases. MLS usually metastasizes to the retroperitoneum, abdominal wall, abdominal cavity, and bone, but cardiac metastases are very rare.[5-7] To date, only about 30 or so cases of cardiac metastases in MLS patients have been reported, including pericardial.[8] It has been reported in the literatures that the tendency to metastatic spread in extrapulmonary sites was attributed to an affinity for adipose tissues. The number of pericardial adipocytes is small compared to that within the soft tissues of the extremities, retroperitoneum, etc., suggest that MLS transfer is not predicated on the enrichment of the tissue with adipose tissue cells. The time interval between primary lesion and cardiac metastasis is relatively long, ranging from 1 to 25 years.[8] This patient had a short delay between the first postoperative recurrence of the tumor and cardiac metastasis. Only 10% of patients with cardiac metastasis have been reported to show some symptoms.[9] Cardiac metastases from liposarcoma often present as congestive heart failure or a heart murmur; these signs and symptoms are associated with impaired myocardial contraction or pericardial tamponade due to tumor invasion of the myocardium.[3] The patient in this case presented clinically with chest tightness and wheezing and lower extremity edema, and also had a recurrence of MLS in the left groin and

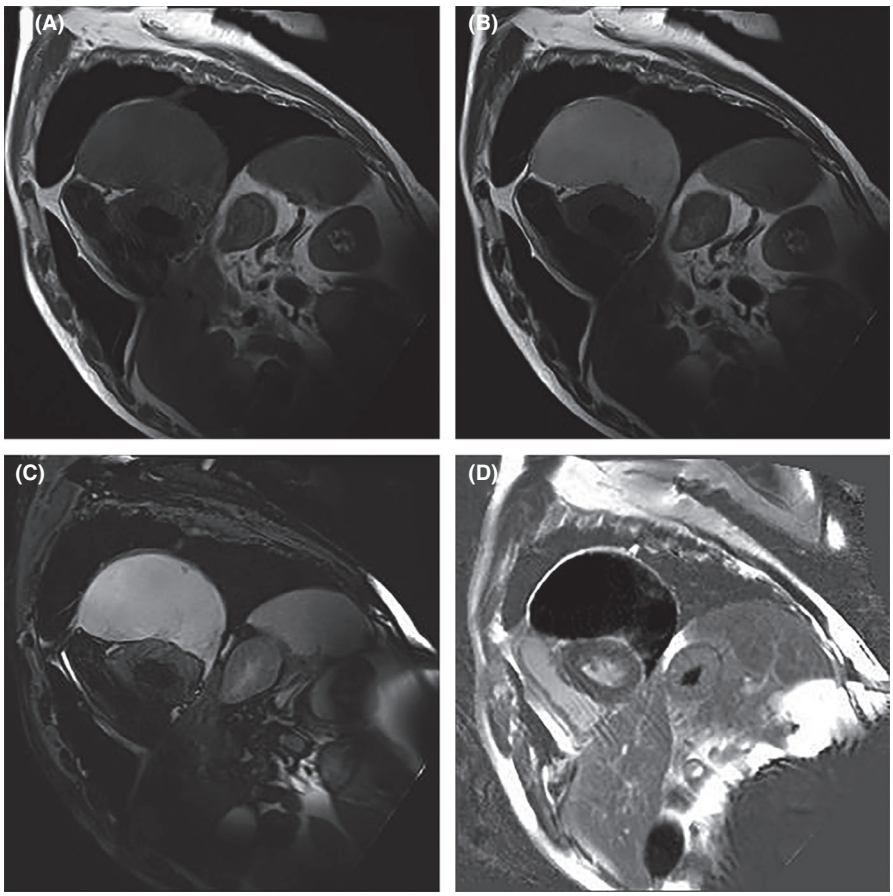

FIGURE 4 Cardiac magnetic resonance shows the lesion is isointensity on T1WI (A); T2WI in the sagittal plane shows the lesion is slightly mixed hyperintensity (B); significantly inhomogeneous high signal on fat-saturated T2WI, with speckled low signal seen locally (C); delayed enhancement scan shows a strip of flocculent soft-tissue density shadow with heterogeneous mild enhancement within the lesion (D)

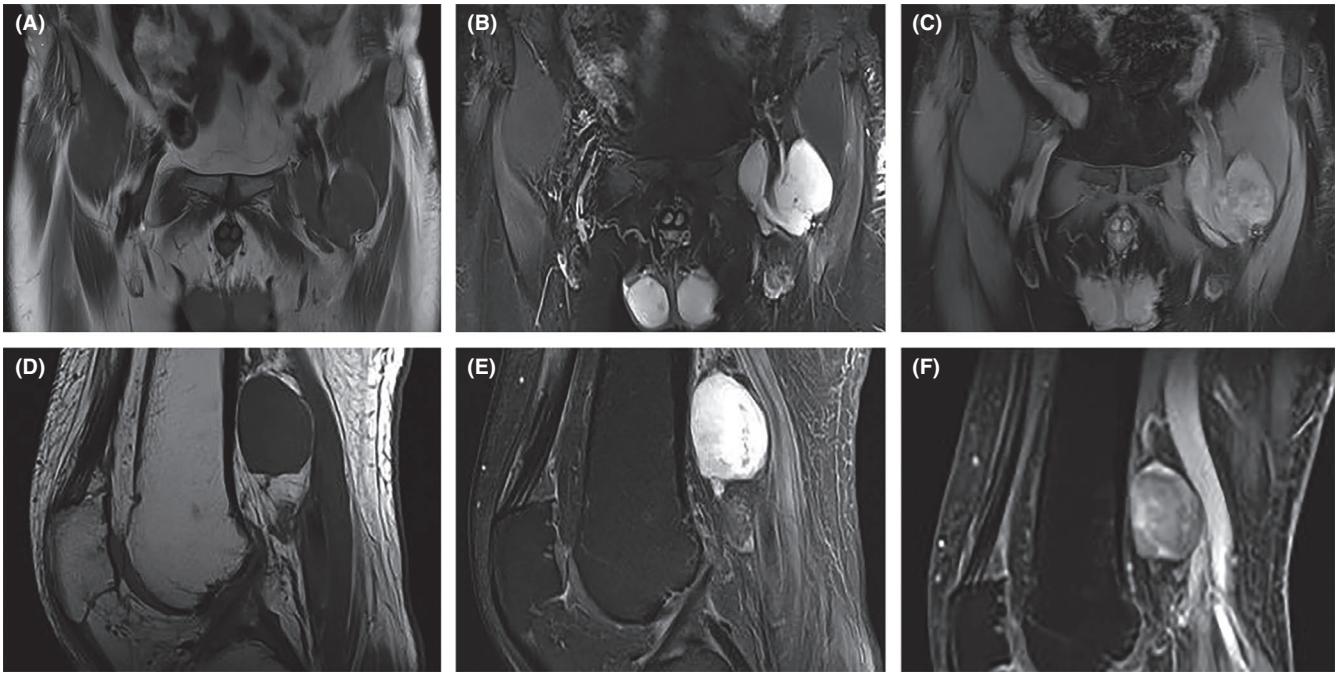

FIGURE 5 Magnetic resonance imaging shows the lesion in the left inguinal region is low signal on T1WI (A); the lesion shows significantly inhomogeneous high signal on fat-saturated T2WI with still clear borders (B); the lesion shows inhomogeneous enhancement after enhancement (C); the left posterior femoral lesion shows hyposignal on T1WI (D); the lesion shows mixed hyperintensity on fat-saturated T2WI, irregular strips of low signal are seen within it, edge undercooling (E); the lesion shows significant inhomogeneous strengthening after enhancement (F)

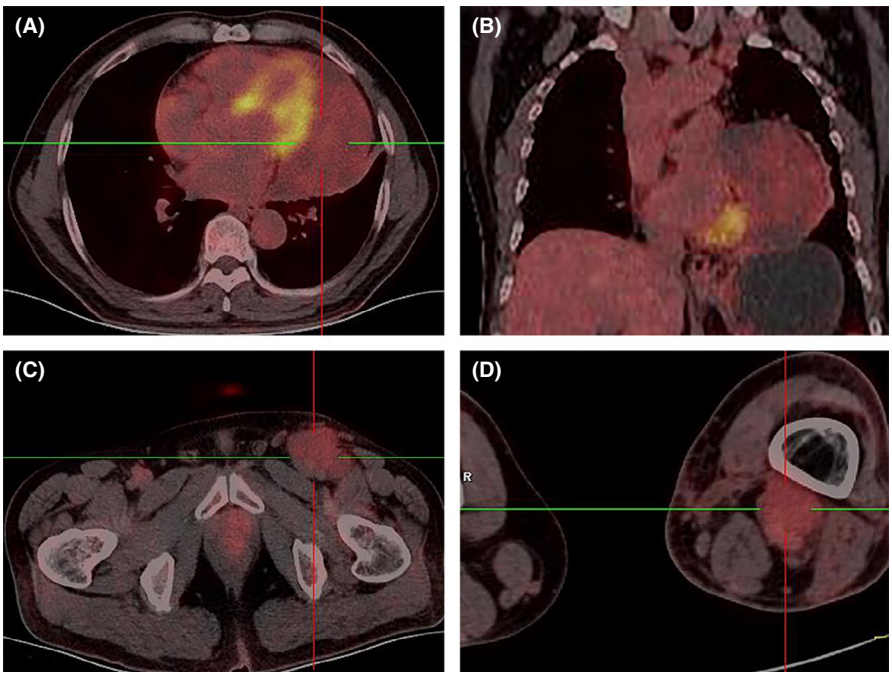

FIGURE 6 Positron emission tomography examination shows large lamellar hypodense shadow in the left pericardium with slightly concentrated radiological distribution, SUV_{max} about 2.6, CT value about 16 HU (A, B); a hypodense mass shadow with a slightly dense radiological distribution was seen in the left inguinal region, measuring approximately 3.4 × 5.4 cm (C); a hypodense mass shadow was seen in the left popliteal fossa with a slightly dense radiological distribution, SUV_{max} about 2.9 (D)

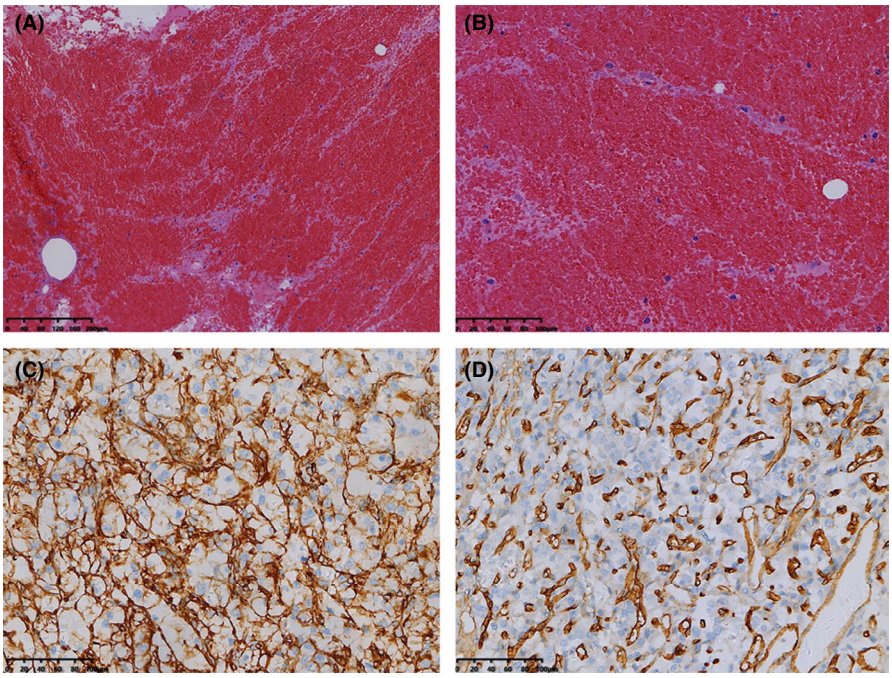

FIGURE 7 Hematoxylin-eosin (HE) staining shows a dense arrangement of homogeneous round-forming lipocytes and large, circular, ring-like adipocytes on light microscopy, and pathological nuclear schizophrenia was common (magnification, A × 100; B × 200; A, B); immunohistochemical staining revealed Vimentin positivity (Envision, ×200; C); immunohistochemical staining revealed high CD34 positivity (Envision, ×200; D)

left popliteal fossa, which made it difficult to identify the possibility of cardiac metastasis in the patient. Early diagnosis of cardiac metastases is not adequate based on the patient's physical examination, US and CT alone.

On histopathological examination, the MLS can be seen under light microscopy to consist of uniform, round or ovoid, small lipogenic cells with an interstitium rich in fine venous structures.[2] The cytoplasm and nucleus of some tumor cells are pushed to one side by the contained lipid droplets and appear as imprinted adipoblasts. Some tumors have round cell areas that represent histologic progression to high-grade tumors. Histologically, low-grade MLS has a better prognosis, with a 5-year survival rate of 90%, and high-grade MLS with a round cell component >5% has a relatively low 5-year survival rate of 60%.[10,11] The fatty component of MLS is its special sign, and histologically, it is predominantly mucinous with minimal fat content; therefore, this case shows a hypodense mass

of indeterminate nature with well-defined borders on US and CT, which was not easily distinguishable from a common mucinous cyst. The high soft tissue contrast and resolution of MRI scans, supplemented by fat suppression techniques to detect a small fat component in the lesion, is key to the diagnosis. The case shows isointensity on T1WI, slightly mixed hyperintensity on T2WI, and significantly inhomogeneous high signal on fat-saturated T2WI, with speckled low signal seen locally. It is evidence of the presence of adipose tissue nodules within them, pathologically areas of high adipoblast aggregation, provides an important imaging basis for the diagnosis of MLS. Delayed enhancement scan shows a strip of flocculent soft-tissue density shadow with heterogeneous mild enhancement within the lesion. In contrast, true cystic masses are only mildly reinforced in the peripheral cyst wall and are generally not reinforced internally, thus distinguishing them from pericardial cysts. Myocardial thickening on CT images in this case, but no infiltrative tumor growth was found on CMR, and the lesion was clearly demarcated from the myocardium. The clinical symptoms of chest tightness and wheezing in this patient were related to compression of the myocardium or surrounding great vessels caused by the expansive growth of the tumor, which corresponded to a decrease in left ventricular end-diastolic volume and end-systolic volume on CMR. The whole-body MRI and PET-CT are the most reliable modalities for surveillance of all likely sites of extrapulmonary metastases. In this case, the pericardial metastasis of MLS has low uptake on PET.

Surgery is the mainstay of treatment for limited MLS, wide local excision of the tumor with a minimum margin of 3 cm in consideration of their tendency to occur locally.[12] When a cardiac metastasis is found incidentally, it is usually incurable, this may be because the poor control of the primary lesion and systemic dissemination that make excision ineffective.[7,8] For patients with locally advanced or metastatic MLS, systemic therapy is usually used. MLS is radiosensitive, whether preoperative, intraoperative, or postoperative radiotherapy.[13]

4 | CONCLUSIONS

In summary, this report described a patient with MLS in the thigh, accompanied by two recurrences after surgery and ectopic and metachronous cardiac metastases. Cardiac metastases from MLS are rare, the prognosis is poor, and long-term follow-up of patients after discharge is necessary. CMR is useful to clarify the diagnosis of cardiac metastases, to define the anatomical relationship between the metastases and cardiac tissue and to determine myocardial activity and whether the myocardium is invaded. The whole-body MRI and PET-CT are the most reliable modalities for surveillance of all likely sites of extrapulmonary metastases.

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The study was approved by the Institutional Review Board at the First Affiliated Hospital of Zhengzhou University.

CONFLICTS OF INTEREST

The authors declare that they have no competing interests.

AUTHOR CONTRIBUTIONS

HW served as a principal author. LL, and JG served as supervisor and author.

CONSENT

Participant gave written consent to participate in the study.

DATA AVAILABILITY STATEMENT

All the data are provided with this case report.

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