Retroperitoneal Liposarcoma

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1 Introduction

Liposarcoma (LS) is a rare malignant tumor of mesenchymal origin, while retroperitoneal liposarcoma (RPLS) refers to liposarcoma arising from the abdomen and pelvic retroperitoneal adipose tissue. According to Windham and Pisters (2005), liposarcoma accounts for less than 1% of systemic malignant tumors. RPLS is the most common type (41%) of retroperitoneal soft tissue sarcoma, followed by leiomyosarcoma and malignant fibrous histiocytoma. Vijay and Ram (2015) reported that RPLS accounts for 12–40% of all systemic LS. Age of onset for RPLS is 55–75 years old. RPLS is slightly more common in men than in women, without significantly racial difference. A total of 10,782 cases of retroperitoneal tumor were reported between 1998 and 2007 in China, with a ratio of 1.3:1 for men to women, with an average age at onset of 41.8 years old, consisting of 68.3% retroperitoneal tumor and 11.6% liposarcoma (n = 1246). Among 119 cases of retroperitoneal liposarcoma, the ratio of male-to-female incidence was 1.9:1, and the median age at onset was 58 years old (range, 19–82). Retroperitoneal liposarcoma is extremely rare, with complex clinical manifestations and pathological types, thus making it very challenging to understand this disease and explore effective treatment.

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2 Etiology

The pathogenesis of retroperitoneal liposarcoma remains unclear, which is possibly associated with inheritance, genetic variation, and environmental factors.

2.1 Environmental Factors

1. Environmental carcinogens. Phenoxy acid herbicides, chlorophenols, and contaminant 2-, 3-, 7-, 8-TCDD may be related to retroperitoneal sarcomas.
2. Radiation exposure. It is commonly seen in patients undergoing radiotherapy. About 0.03–0.8% of patients who receive radiotherapy have been reported to develop retroperitoneal sarcoma.

2.2 Immunosuppressant

Both immune deficiency and immunosuppressive drugs are associated with the pathogenesis of retroperitoneal soft tissue sarcoma. It is

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reported that patients with systemic lupus erythematosus developed diffuse infiltrative retroperitoneal mucinous liposarcoma after treated with steroid hormone for 13 years, suggesting critical involvement of immunosuppressant in retroperitoneal liposarcoma.

2.3 Genetic Factors

Individuals with family history of lipoma or liposarcoma are more susceptible to developing retroperitoneal liposarcoma. Retroperitoneal liposarcoma has been reported to occur successively in two compatriots with family history of malignant fibrous histiocytoma.

2.4 Other Factors

The change in levels of insulin receptor and postreceptor in adipose tissue and decrease in biological activity of insulin may be involved in retroperitoneal liposarcoma.

3 Pathogenesis and Pathobiology

The pathogenesis of retroperitoneal liposarcoma remains unclear and may be related to the molecular mechanisms below:

3.1 Mechanism of MDM2-p53

MDM2 gene (human homologue of the murine double-minute type 2) located at 12q13-15 region shows constant amplification in well-differentiated liposarcoma (Vassilev et al. 2004). MDM2 is a p53-specific E3 ubiquitin ligase and principal cellular antagonist of p53, acting to limit the p53 growth-suppressive function in unstressed cells. In healthy body, the precise balance between p53 and MDM2 guarantees the normal proliferation and differentiation of tissue cells. If MDM2 is overamplified, p53 activity is inhibited, resulting in uncontrollable cell proliferation. This may be related to the pathogenesis of retroperitoneal liposarcoma.

3.2 Mechanism for Prune-nm23-H1

Prune, the human homologue of *Drosophila* prune gene, located in 1q21-23, encodes a protein that can bind to nm23-H1 (nucleoside diphosphate kinase) to downregulate its activity (Forus et al. 2001). The nm23-H1 may inhibit cell proliferation and tumor metastasis. The balance and precise coordination between prune and nm23-H1 expression present in healthy human bodies; in contrast, overexpression of prune gene is found in liposarcoma, with downregulation of nm23-H1 activity. This may be one of the molecular mechanisms responsible for the pathogenesis of liposarcoma.

4 Clinical Manifestation

As RPLS grows in an occult manner, most patients with RPLS in the early stage are asymptomatic. RPLS is often manifested as painless mass and has grown into a very large size before being detected. Symptoms and signs may not be obvious until the tumor has grown large and compressed adjacent organs or tissues.

4.1 Clinical Symptoms

Retroperitoneal tumors that grow in the loose connective tissue space of the retroperitoneum usually don't cause obvious symptoms when they are small. When they grow to a larger size, the tumors may result in symptoms by compressing and invading blood vessels, nerves, or other vital organs.

4.1.1 Space-Occupying Symptoms

As they are often large in size when detected, retroperitoneal liposarcomas may cause abdominal swelling, fullness, and feeling of heaviness by compressing and displacing adjacent organs.

Large retroperitoneal tumors contribute to an increased intra-abdominal pressure, which pushes outward through the weak site in the abdominal wall, resulting in abdominal or inguinal hernia.

4.1.2 Oppressive and Obstructive Symptoms

A large tumor may cause symptoms by compressing vital organs, nerves, and blood vessels in retroperitoneum. It often compresses the abdominal aorta, inferior vena cava, superior and inferior mesenteric veins, and other major blood vessels. The oppression of duodenum can cause proximal (high) or distal (low) small bowel obstruction (SBD).

1. Pain, which may be divided into abdominal pain and lower backache (lumbago). It is usually dull and occasionally sharp and colic. Approximately 40–70% of patients exhibit this symptom, resulting from the compression of surrounding tissues, organs, and nerves by the tumor. Lower backache with unilateral or bilateral radiation to the lower extremities may be caused by the oppression or violation of the lumbar plexus or sacral plexus nerve root.
2. Oppression of the gastrointestinal tract can lead to abdominal fullness after eating, anorexia, nausea, vomiting, abdominal distension, and constipation. Patients of 4–35% present with abdominal distension. Some patients develop intestinal obstruction, loss of appetite, and weight loss in advanced stage, indicating high suspicion of malignant retroperitoneal tumor. Additionally, oppression and displacement of the kidneys, ureters, or bladder can commonly occur and produce urinary tract symptoms, such as frequent urination, urgent urination, hematuria, dysuria, oliguria, and even anuria. Azotemia may occur as a result of bilateral ureteral compression.
3. Lower extremity edema and ascites. Tumor can cause obstruction of blood reflux by compressing the inferior vena cava or portal vein, thus leading to unilateral or bilateral lower extremity edema and ascites.
4. Incontinence and lower extremity paralysis. These symptoms can mainly be attributed to the compression of spinal cord as a result of invasion of intervertebral foramen.

4.1.3 Systemic Symptoms

Loss of appetite, weight loss, and fever are common. These symptoms mainly result from toxins produced by necrotic tissue, metabolites, and cachexia.

4.2 Signs

Most of these tumors can be incidentally palpable by patients. Generally, they won't consult a doctor until the mass has grown to a very large size and affected their daily lives or grown rapidly during a short period of time.

In physical examination, abdominal mass can be usually palpable with or without mild tenderness. Other signs include abdominal distension or tension, splenomegaly, lymphadenopathy, and cachexia. No specific signs have been identified. Retroperitoneal tumors generally do not move with respiration. Special attention should be paid to the mobility and hardness of the mass, which can serve as an indicator for resectability. Hard and fixed (to the abdominal wall or pelvic wall) mass is commonly suggestive of malignant tumor, teratoma, or hamartoma, whereas soft and flexible mass is commonly suggestive of lipoma or liposarcoma.

5 Diagnosis and Staging

5.1 B-Ultrasound

B-ultrasound is a noninvasive and inexpensive technique used to detect the size, number, location, cystic vs. solid nature of tumor(s), as well as relationship with adjacent organs and blood vessels. It has been widely applied in preoperative examination and postoperative follow-up. Due to its insensitivity to soft tissues, B-ultrasound can rarely identify LS. Liposarcoma presents as a large mass, in circular or irregular lobulated shape, with rough margin, and unevenly distributed moderate to strong echoes.

5.2 Examination

CT as the first choice for the diagnosis of RPLS allows precise localization and qualitative assessment. High-resolution and clear images produced by CT are able to objectively display the location and boundary of RPLS, its relationship with surrounding organs, as well as tumor-induced compression and displacement of surrounding organs and major blood vessels. CT plays a valuable role in determining the origin of retroperitoneal tumor, but an unideal role in demonstrating the extent of tumor invasion in adjacent organs. An extremely careful analysis of preoperative CT scan findings would help localize the tumor and predict the pathological nature of LS, which is vital to preoperative assessment of resectability and development of surgical strategy. CT findings vary with pathological types of retroperitoneal liposarcoma, thus providing reference to preoperative interpretation of pathogenesis.

5.2.1 Well-Differentiated Liposarcoma

CT usually depicts fat-density mass, accompanied by multiple floccules at slightly higher density and widely distributed cord-like separation, with a clear boundary between the tumor and surrounding tissue, without significant infiltration (Lahat et al. 2009). For lipoma-like liposarcoma, CT exhibits predominantly the density of adipose tissue, with thickened irregular soft tissue septa, which frequently are enhanced with soft tissue component after IV contrast administration. Sclerosing LS displays a mass of soft tissue density similar to muscle and is moderately enhanced after the injection of contrast media. Due to the lack of intratumoral fat content, sclerosing LS is easily misdiagnosed as other soft tissue sarcomas.

5.2.2 Myxoid Liposarcoma

Myxoid liposarcoma is rich in mucus matrix. Although it is a solid tumor, CT scan shows a cystic mass of uniform low-density (in-between normal fat density and muscle density), mildly progressive mesh-like or sheetlike delayed enhancement, non-enhanced cystic component, and small bifurcated vessels in cystic lesions (Singer et al. 2003).

5.2.3 Round Cell and Pleomorphic Liposarcoma

CT displays a substantially uniform solid soft tissue density inside the tumor, similar to skeletal muscle, with punctate calcification, little mature fatty component or only a small amount of mucus component inside the tumor, and frequent necrosis. Moderate to high enhancement is observed after the injection of contrast medium, with irregular necrotic tissue and non-enhanced necrotic foci. Round cell LS can only be distinguished from pleomorphic LS by histopathological findings rather than imaging.

5.2.4 Dedifferentiated Liposarcoma

Pathologically, dedifferentiated liposarcoma is characterized by the coexistence of well-differentiated and poorly differentiated liposarcoma. CT finding of well-differentiated area is similar to that of lipoma-like liposarcoma, while dedifferentiated sarcoma varies with tissue components, similar to that of primary sarcoma composed of such elements (Lahat et al. 2009). The presence of focal nodule of soft tissue density and water-like low-density area in lipoma-like tissue suggests dedifferentiated liposarcoma.

5.3 MRI Examination

With high resolution, MRI exhibits greater sensitivity than CT in distinguishing normal soft tissue from liposarcoma. Moreover, the lesion can be scanned from all directions, and the distribution and involvement of blood vessels of retroperitoneal tumor can be clearly presented by MRI without contrast medium. Overall, MRI is essential to the diagnosis of invasion of the abdominal aorta, inferior vena cava, or other structures. MRI plays critical roles in determining retroperitoneal liposarcoma subtype and clinical stage, depicting tumor's relationship with surrounding normal tissue, as well as assisting in preoperative diagnosis and development of surgical approach.

5.3.1 Well-Differentiated Liposarcoma

The fat content accounts for >75% of well-differentiated liposarcoma tissue, while the non-fat content is generally manifested as nodule or mass separated by low-intensity signals, with the septa ranging from small to large in size. Lipomatous liposarcoma shows higher intensity signal relative to fat in the abdominal cavity on T1WI. Fatty components in the tumor display low-intensity signals on fat-suppressed T1WI sequence, whereas iso- and low-intensity signals on fat-suppressed T2WI sequence. Sclerosing LS containing dense fibrous components exhibits interlaced low-intensity signals on both T1WI and T2WI (Barile et al. 2002).

5.3.2 Myxoid Liposarcoma

This type of liposarcoma that is rich in water-like components exhibits signal with an intensity similar to that of water on MRI images. As myxoid liposarcoma often contains different amount of adipose tissue, T1WI and T2WI show amorphous, cluster- or line-like high intensity signals, which are helpful for the diagnosis (Francis et al. 2005). Fibrous septa in mucus matrix show low-intensity signal on T2WI. MRI scan shows cystic lesions and progressive mesh-like enhancement of myxoid component after the injection of contrast medium, indicating the presence of solid lesion (Kim et al. 1996).

5.3.3 Round Cell Liposarcoma

Round cell liposarcoma (Song et al. 2007) exhibits slightly lower signal relative to muscle tissue on T1WI and slightly higher signal on T2WI, which is significantly enhanced after the injection of contrast medium. Due to less conspicuous myxoid matrix and capillaries compared to myxoid liposarcoma, the round cell liposarcoma displays lower-intensity signal on T2WI than myxoid liposarcoma. Intratumoral hemorrhage and necrosis most easily occur in round cell liposarcoma, with mixed signals on MRI.

5.3.4 Pleomorphic Liposarcoma

Pleomorphic liposarcoma is a rare and highly aggressive tumor. MRI exhibits aggressive margin of the lesion, whereas burr-like or halo-like change in the margin of the lesion after the injection of contrast medium. There are no characteristic signals on MRI due to the lack of fat component, so it is difficult to differentiate this tumor from other retroperitoneal soft tissue tumors (Song et al. 2007).

5.3.5 Dedifferentiated Liposarcoma

Dedifferentiated liposarcoma is characterized by coexistence of well-differentiated with poorly differentiated tumor components. It exhibits classic MRI findings, namely, a well-defined abrupt boundary between adipose and soft tissue components due to unknown reason (Tateishi et al. 2003). Lipoma-like well-differentiated liposarcoma displays signal intensity similar to other liposarcomas; in contrast, poorly differentiated tissue displays low signal relative to muscle on T1WI and heterogeneously high-intensity signal relative to muscle on T2WI (Kim et al. 2010).

5.4 Ultrasound-Guided Percutaneous Biopsy

If the diagnosis can't be made depending on image findings, ultrasound-guided percutaneous biopsy is recommended as an alternative strategy. However, percutaneous biopsy is contraindicated in patients who have undergone surgical removal as it may cause RPLS metastasis, so the final diagnosis still relies on pathology and histology after surgery. Percutaneous biopsy is indicated for patients who intend to receive preoperative radiotherapy/chemotherapy, who can't undergo tumor resection, or who have experienced hematogenous dissemination.

5.5 Renal Dynamic Imaging

If the CT scan suggests the presence of renal invasion, renal dynamic imaging should be performed to determine the bilateral renal function, in order to assess the potential resectability of the unilaterally involved kidney.

5.6 Intravenous Urography

Intravenous urography can clearly display the shape and function of the kidneys and ureters, so it plays a role in the diagnosis of large retroperitoneal mass, especially those which compress the ureter and kidneys.

5.7 Staging of Retroperitoneal Liposarcoma

5.7.1 Clinical Staging

Currently used staging system for soft tissue sarcomas is the TNM system developed by the American Joint Committee on Cancer (AJCC) (2010, 7th Edition). In this system, clinical staging is based on histology, size, depth, lymph node, and distant metastasis.

T Staging (Primary Tumor T)

Tx Primary tumor cannot be assessed
T0 No evidence of primary tumor
T1 Tumor ≤5 cm in maximum diameter
T1a Superficial tumor
T1b Deep tumor
T2 5 cm tumor, >5 cm in maximum diameter
T2a Superficial tumor
T2b Deep tumor

(Superficial tumor is located exclusively above the superficial fascia without invasion of the fascia; while deep tumor is located in any of the following: exclusively beneath the superficial fascia, superficial to the fascia, with invasion of or through the fascia, or both superficial yet beneath the fascia. Retroperitoneal, mediastinal, and pelvic sarcomas are all classified as deep tumors.)

N Staging (Regional Lymph Nodes N)

Regional lymph nodes that cannot be assessed
No regional lymph node metastasis
Regional lymph node metastasis
M staging (distal metastasis)
M0 No distal metastasis
M1 Distal metastasis
G Histologic grade (G)
GX Grade that cannot be assessed
G1 Grade 1 well differentiated
G2 Grade 2 moderately differentiated
G3 Grade 3 poorly differentiated

5.7.2 Pathological Classification and Grading

Since 2000, liposarcoma has been classified into four categories by WHO according to immunohistochemistry (IHC) and molecular and cytogenetic characteristics based on conventional histopathological findings: (a) nonclassic liposarcoma/high-grade differentiated liposarcoma, (b) well-differentiated liposarcoma/dedifferentiated liposarcoma (WDLPS/DDLPS), (c) myxoid/round cell liposarcoma (MLPS), and (d) pleomorphic liposarcoma (PLPS).

Pathological grading of LS is currently determined by the grading system of soft tissue malignant tumor, which has been recently modified by the French Federation of Cancer Centers Sarcoma Group (FNCLCC) with a new scoring classification method. Histologic grading is calculated as the total score for three parameters, including tumor differentiation, degree of necrosis, and mitotic count in the new classification system.

Differentiation: (a) score, sarcomas closely resembling normal adult mesenchymal tissue (e.g., well-differentiated LS); (b) score 2, sarcomas with confirmed histologic typing (e.g., myxoid liposarcoma); and (c) score 3, embryonal and undifferentiated sarcomas and sarcomas of uncertain types.

Mitotic Figures

a. 0–9/10 HPF score 1
b. 10–19/10 HPF score 2
c. ≥20/10 HPF score 3

Tumor Necrosis (Under Microscopy)

a. Score 0: No necrosis
b. Score 1: ≤50% tumor necrosis
c. Score 2: >50% tumor necrosis

Grading system: grade 1, total score of 2–3 points; grade 2, total score of 4–5 points; and grade 3, total score of 6–8 points. The classification is considered more objective and scientific.

The accuracy of histologic grading directly predicts the prognosis of patients with RPLS.

Histologic Grade
Grade 1: Total score 2–3
Grade 2: Total score 4–5
Grade 3: Total score 6, 7, and 8

6 Treatment

6.1 Surgical Treatment

The mainstay therapy for RPLS is complete surgical excision. Epidemiology studies on retroperitoneal tumor in mainland China have found complete resection rate of 66%, partial resection rate of 18.2%, exploratory biopsy rate of 7.9%, and combined organ resection rate of 7.8%. High postoperative recurrence rate is the most challenging problem. To completely remove the tumor and reduce recurrence rate, the resection margin should be as far away from the accessible and visible border of the tumor as possible, without residual capsule. If there is involvement of adjacent organs and blood vessels, en bloc resection should be performed, including removal of the partial gastrointestinal tract, kidney, liver, abdominal wall, and inferior vena cava. Common surgical approaches include total resection, palliative resection, and debulking/cytoreductive surgery.

6.1.1 Gross Total Resection

Gross total resection is defined as a complete resection without any visible tumor residue, namely, R1resection. Some scholars (Strauss et al. 2010) reported no significant difference in postoperative survival rate and recurrence rate during between R0 and R1 resection in follow-up studies on patients with retroperitoneal liposarcoma. R1 resection has therefore become a major goal worth pursuing. Attention should be paid to identify the false capsule during the resection, and the tumor should be removed completely beyond the false capsule to avoid tumor residue and rupture, which is vital to reduce postoperative tumor recurrence rate and prolong recurrence interval. After the removal of the tumor, the surrounding adipose tissue should be also dissected as possible to reduce the risk of recurrence. In principle, no adipose tissue residue should be left around the surgical area.

Retroperitoneal liposarcoma is often closely related to surrounding organs. If it is impossible to separate adhesions, combined organ resection may be necessary to achieve R1 resection. Combined organ resection rate is reported to be 57–83% (Neuhaus et al. 2005). The organs most common to be jointly resected during the surgical removal of retroperitoneal liposarcoma are the kidneys, followed sequentially by the spleen, intestine, stomach, and pancreas. During surgery, if the tumor is closely attached to surrounding major blood vessels and difficult to separate, firstly the majority of the tumor body should be resected to obtain better surgical field, followed by the residual tumor tissue. Forced separation may cause tear of the major vessels, which should be avoided as possible.

6.1.2 Palliative Resection

Palliative resection is defined as visible lesion residue of less than 1 cm in size when complete removal of gross tumor is impossible. If complete resection is not indicated for an individual patient, palliative resection should be performed as possible in order to decrease the symptoms, alleviate obstruction and compression of organs, protect organ function, prolong survival time, and improve quality of life. After the palliative resection, supplementary comprehensive actions should be taken to control residual tumor in an attempt to prolong survival time and ensure quality of life. Common techniques include internal radiation generated by radioactive nuclide, sustained release of chemotherapeutic drugs, and embedment of intraperitoneal chemotherapy pump.

6.1.3 Debulking/Cytoreductive Surgery

Debulking/cytoreductive surgery is defined as large residues in the main body of the lesion with visible residue of greater than 1 cm in size. It is commonly performed in patients who have

undergone multiple recurrence reoperations, as tumor resection is challenging due to abnormal anatomic structure, critical involvement of multiple organs, or distant metastasis. During debulking (cytoreductive) surgery, wound hemostasis is a major concern. It is relatively easy to stop bleeding in low-grade malignant tumors but not in high-grade liposarcoma with abundant blood vessels. In this setting, subtotal resection should be considered. The smaller the residual wound, the better the hemostatic effect.

6.2 Radiotherapy and Chemotherapy

The effect of radiotherapy and chemotherapy in patients with RPLS remains controversial due to the fact that RPLS is insensitive to both radio- and chemotherapy. Further large-scale controlled trials are guaranteed to prove the exact role of radiotherapy and chemotherapy in improving survival rate and reducing relapse rate.

Patients with retroperitoneal liposarcoma should receive radiotherapy after partial resection. Some patients with a large-volume tumor which is challenging to directly remove may undergo preoperative radiotherapy to shrink the tumor volume and reduce the difficulty in surgery. Well-differentiated mucinous liposarcoma growing slowly is relatively sensitive to radiotherapy, whereas poorly differentiated high-grade malignant liposarcoma growing rapidly is resistant to radiotherapy.

Only a few chemotherapy drugs are currently effective against RPLS, of which anthracyclines (doxorubicin, epirubicin) and ifosfamide are the most important first-line agents. A recent phase II randomized trial (Gortzak et al. 2001) compared the therapeutic effect of doxorubicin/ifosfamide neoadjuvant chemotherapy with surgery with surgery alone in patients with soft tissue sarcoma. The 5-year disease-free survival rate was 56 and 52%, respectively; the overall survival rate was 65 and 64%, respectively; therefore, no significant improvement was observed in the prognosis of patients. A chemotherapy drug, trabectedin, has been approved by the European Agency for Evaluation of Medicinal Products (EMEA) for the treatment of patients with advanced-stage myxoid/round cell liposarcoma who do not respond to anthracycline and ifosfamide.

7 Efficacy and Prognostic Factors

Surgery is the most effective therapeutic approach for RPLS. Surgical resection rate has been reported to be 71.4–88.8% in literatures (Neuhaus et al. 2005). RPLS is characterized by a high rate of local recurrence, and the recurrence interval is gradually shortened with the increased infrequency; however, distant metastasis rarely occurs. The high recurrence rate of RPLS may be attributed to the following factors (Hassan et al. 2004):

a. Large-volume tumor with complex surrounding anatomic structure, as well as invasion of the surrounding vital organs and blood vessels, making it very challenging to perform a complete resection
b. Multifocal tumors with tiny foci that are easily ignored by surgeons during surgery
c. Tumor implantation metastasis caused by surgical procedures
d. Positive pathological findings of tumor margin that is considered “completely resected” by surgeons with the naked eyes.

Although the complete surgical resection rate has been greatly improved in recent years, there is no significant reduction in the postoperative recurrence rate, which maintains at 41–71% (Porter et al. 2006).

The prognosis of RPLS closely correlates with histologic type and involvement of adjacent organs, namely, the lower the degree of differentiation, the worse the prognosis. For LS, the 5-year overall survival rate was 40–60%, and the 3-year survival rate was 73% and 43%, respectively, in those who received complete vs. incomplete surgical resection. The presence of residual tumor cells on the endoscopic resection margin (whether R0 or not) has no effect on

overall survival; however, whether or not all visible (with the naked eyes) tumors have been removed (whether R1 or not) has significant effect on overall survival of patients. In order to achieve R0 resection and reduce local recurrence rate, combined organ resection should be performed.

Myxoid and round cell liposarcoma most commonly occur in 40–50 years, with metastasis in more than one third of patients (Blair et al. 1998). These tumors often metastasize to the retroperitoneal cavity, trunk, limbs, bones, and other regions where fat is distributed (Antonescu et al. 2000), with a 3-year survival rate of 100% and 33%, respectively (Schwab et al. 2007). Pleomorphic LS is an invasive high-grade tumor, which occurs at the median age of 55–65 years old with equal female-to-male incidence (Gebhard et al. 2002). Metastasis to the lungs (90%), bone (8%), and liver (1%) can be observed in 30–50% of patients with pleomorphic liposarcoma. Most patients die quickly of metastatic disease, with an overall mortality rate of 40–50%, whereas a 5-year survival rate of 25–60% (Zagars et al. 1996). Although tumor invasion is also common in dedifferentiated liposarcoma, its overall course is longer than that of pleomorphic liposarcoma. Well-differentiated and myxoid liposarcomas are considered to have a favorable prognosis, with 3-year survival rate of about 90% (Hornick et al. 2004).

Postoperative patients should be actively followed up. For example, CT or B-ultrasound examination once every 6 months for 3 years is recommended for early detection of tumor recurrence.

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