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ARTICLE

Retroperitoneal sarcomas

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Abstract

Retroperitoneal sarcomas are rare neoplasms. CT or MR imaging is performed in patients with these tumors to detect local extent and distant metastases of the tumor and for preoperative surgical planning. Most sarcomas cannot be characterized as to cell type with CT or MR, with the exceptions being liposarcomas and intracaval leiomyosarcomas. Similarly histological grading cannot be made definitively with imaging alone, the exception being liposarcoma since well differentiated liposarcomas contain more macroscopic fat than do less differentiated liposarcomas. After surgery, follow up imaging with CT or MR and careful scrutiny of the tumor bed and resection site are essential to detect early recurrences, which can often be managed with re-resection.

Keywords: *Retroperitoneal space; neoplasms; CT; MR; radionuclide imaging; sarcomas.*

Incidence and statistics

Retroperitoneal sarcomas are rare tumors accounting for only 1%–2% of all solid malignancies. Of all sarcomas, the majority occur outside of the retroperitoneum. Only 10%–20% of sarcomas are retroperitoneal sarcomas, and the overall incidence is 0.3%–0.4% per 100 000 of the population[1]. The peak incidence is in the 5th decade of life, although they can occur in any age group.

The most common types of retroperitoneal soft tissue sarcomas in adults vary from study to study. However, in most studies, the most frequently encountered cell types are liposarcomas, leiomyosarcomas and malignant fibrous histiocytomas (MFH)[2]. Recently, the frequent diagnosis of MFH in the retroperitoneum has been disputed. With the use of immunohistochemistry, many of these fibrous tumors have now been shown to represent other sarcoma types such as leiomyosarcomas or dedifferentiated liposarcomas[3,4]. For this reason, it is anticipated that the number of these neoplasms that will be considered as MFH will be dramatically reduced in the future.

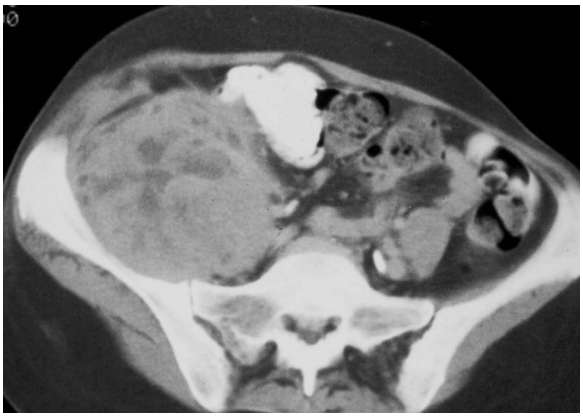
Patients with sarcomas present late, because these tumors arise in the large potential spaces of the retroperitoneum and can grow very large without producing symptoms[5,6]. Moreover, when symptoms do occur, they are nonspecific, such as abdominal pain and fullness, and are easily dismissed as being caused by other less serious processes[7]. Retroperitoneal sarcomas, therefore, are usually very large at the time of presentation.

What the surgeon needs to know

Imaging is important in the diagnostic workup of these patients, being required not only for tumor detection, staging, and operative planning, but also for guiding percutaneous or surgical biopsy of these tumors.

As other neoplastic processes, such as lymphoma and metastatic disease, which are treated differently, may mimic retroperitoneal sarcomas, tissue diagnosis is of paramount importance. Therefore, image-guided and surgical biopsies have a relatively greater role to play in the diagnosis of retroperitoneal sarcomas than is the case for sarcomas elsewhere in the body[8–11].

(a)



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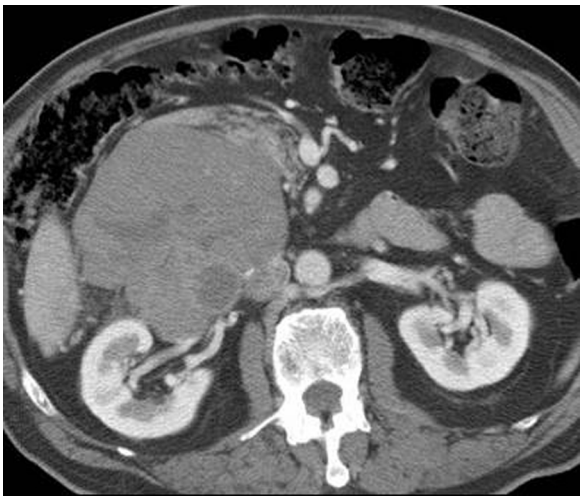


Figure 1 **Contrast-enhanced axial CT shows two large retroperitoneal heterogenous neoplasms with areas of low density due to degeneration or necrosis. These proved to be malignant: a peripheral nerve sheath tumor (a) and retroperitoneal leiomyosarcoma (b), respectively.**

Once the diagnosis is made, the surgical team needs to determine if the retroperitoneal sarcoma can be resected. Therefore one of the first determinations to be made is whether the tumor is localized, its local extent, and also if there is evidence of intra- or extra-abdominal metastatic spread of tumor. The location and size of the tumor, its relationship to adjacent organs, presence or absence of local extension, relationship to and/or involvement of major vascular structures, as well as the presence of normal anatomic variants and anomalies of major abdominal arteries and veins, are all crucial pieces of information that need to be provided. Since resection of one kidney is not uncommon, any radiographic evidence of unilateral renal dysfunction involving the kidney that is not adjacent to the tumor should be relayed to the surgical team. While it may be unavoidable that the patient will be left with a single poorly functioning kidney, the surgeon must be provided with all relevant information prior to attempted tumor resection.

Table 1 Classifications

Histological grade (G)	
G1	Well differentiated
G2	Moderately well differentiated
G3	Poorly or very poorly differentiated
Primary site (T)	
T1	Tumor less than 5 cm in diameter
T1a	Superficial tumor
T1b	Deep tumor
T2	Tumor 5 cm or more in diameter
T2a	Superficial tumor
T2b	Deep tumor
N.B. Retroperitoneal and pelvic sarcomas are classified as deep tumors	
Nodal involvement (N)	
N0	No histologically verified metastases to lymph nodes
N1	Histologically verified regional lymph nodes
Distant metastasis (M)	
M0	No distant metastases
M1	Distant metastases present

In evaluating preoperative imaging studies, the radiologist should be cognizant of two facts: (A) up to 75% of retroperitoneal sarcoma resections involve concomitant resection of at least one adjoining intra-abdominal visceral organ (commonly large or small bowel or kidney); (B) the most common types of vascular involvement precluding resection are involvement of the proximal superior mesenteric vessels or involvement of bilateral renal vessels. Accordingly, since these tumors tend to invade organs with which they are contiguous, such contiguity must be mentioned even in the absence of imaging evidence of gross tumor invasion of these organs. Also, the mesenteric and renal vessels must be carefully examined and their relationship to a mid or upper retroperitoneal tumor described.

Tumor staging

Accurate staging is important as it facilitates determination of appropriate surgery, establishes prognosis, and provides a guide for adjunctive therapy.

The American Joint Committee Staging System (Tables 1 and 2) of extremity soft tissue sarcomas, which is based on the TNM classification, is most commonly used for most retroperitoneal soft tissue sarcomas, although it is better suited for extremity sarcomas[12]. This staging system takes into consideration histological grade, tumor size and depth relative to the superficial muscular fascia, presence or absence of lymph node involvement, and the presence or absence of distant metastases (Table 2). Nearly all retroperitoneal sarcomas are large and >5 cm and are deep to the superficial fascia. Therefore localized retroperitoneal sarcomas are nearly always classified as Stage IIB (large, low-grade, and deep) or stage III (large, high-grade and deep) neoplasms,

Table 2 ***American Joint Committee staging of soft tissue sarcomas***

Stage	Classification	Description
IA	GI, T1, N0, M0	Grade 1 tumor, <5 cm in diameter no regional lymph nodes and/or distant metastases
IB	GI, T2, N0, M0	Grade 1 tumor, 5 cm or more in diameter, no nodes and/or metastases
IIA	G2, T1, N0, M0	Grade 2 tumor, <5 cm in diameter, no nodes and/or metastases
IIB	G2, T2, N0, M0	Grade 2 tumor, 5 cm or more in diameter, no nodes and/or metastases
IIIA	G3, T1, N0, M0	Grade 3 tumor, <5 cm in diameter, no nodes and/or metastases
IIIB	G3, T2, N0, M0	Grade 3 tumor, 5 cm or more in diameter, no nodes and/or metastases
IIIC	G1–3, T1, 2, N1, M0	Tumor of any grade and/or size, with regional involved nodes, but no metastases
IVA	G1–3, T3, N0, N1, M0	Tumor of any grade invading bone vessels/nerves, with/without nodes, no metastases
IVB	G1–3, T1–3, N0, N1, M1	Tumor with distant metastases

with the distinction between these two stages being made only on the basis of histologic grade.

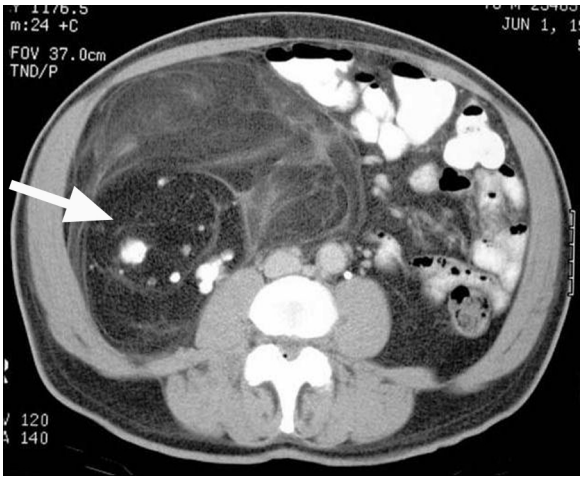


Figure 2 **Liposarcoma. Contrast-enhanced axial CT shows large right retroperitoneal liposarcoma (arrow) composed predominantly of fat but also has areas of soft tissue density and calcific components.**

have had a successful complete resection and also have low-grade tumors have the best survival rates.

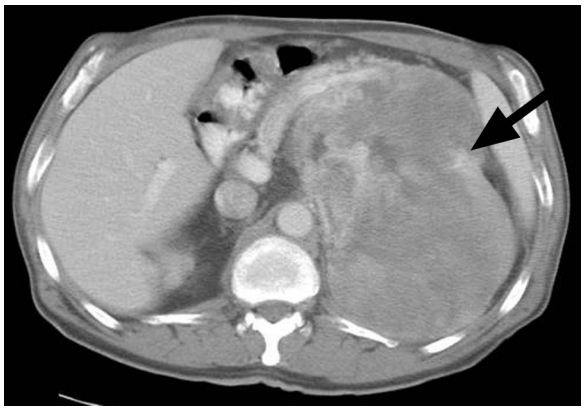


Figure 3 **High-grade liposarcoma. Contrast-enhanced axial CT shows left-sided predominantly soft-tissue density abdominal tumor (arrow) which proved to be a high-grade pleomorphic liposarcoma.**

Role of imaging in sarcoma characterization, grading and prognosis

Imaging cannot be reliably used to predict the cell types of most sarcomas (Fig. 1(a), (b)), with rare exceptions being liposarcoma and intracaval leiomyosarcomas. The presence of macroscopic fat enables one to make the diagnosis of a liposarcoma (Fig. 2). However, not all liposarcomas demonstrate macroscopic fat on imaging (Fig. 3). Some of these tumors may be composed almost entirely of soft tissue and fluid components. In these instances, the tumors cannot be differentiated from other types of sarcomas[13–15]. A tumor within the lumen of the inferior vena cava with expansion of its lumen and enhancing tumor thrombus is pathognomonic of an intracaval leiomyosarcoma (Fig. 4(a), (b))[16,17].

Fortunately, it has been shown that in the vast majority of sarcomas, cell type has no impact on treatment and long-term survival. The major factors that affect survival are the tumor grade and resectability[10,11]. Patients who

Only rarely can imaging predict sarcoma grade and prognosis. The exception is a liposarcoma, and, in general, if a liposarcoma contains mostly fat and very little soft tissue, it is likely to be a low-grade tumor (Fig. 5(a), (b)). However, the converse is not true. A liposarcoma containing a large amount of soft tissue and with little or no macroscopic fat, may be a low-, intermediate- or high-grade tumor. Calcification or ossification within a liposarcoma has been shown to be a poor prognostic feature, often indicating dedifferentiation.

CT is the most commonly used modality for identification, localization, and staging of retroperitoneal sarcomas[5,8,18–20]. The use of magnetic resonance imaging is generally reserved for selected problem solving; such as to address questions regarding vascular invasion, and evaluate problematic indeterminate liver lesions.

More recently PET-FDG imaging has been used in an effort to assess the tumor grade as well as to evaluate patients for tumor recurrence (Fig. 6(a), (b))[21–23].

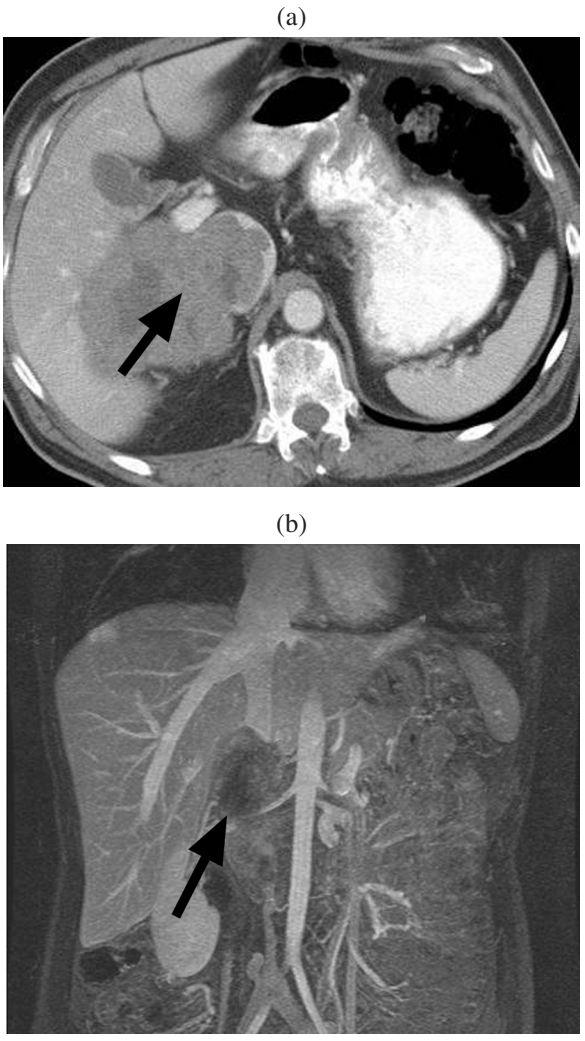


Figure 4 **Intracaval leiomyosarcoma. Large inferior vena caval leiomyosarcoma (arrow) seen on (a) contrast-enhanced CT and (b) coronal contrast-enhanced gradient echo MR image (arrow). Note intraluminal tumor extension on both images.**

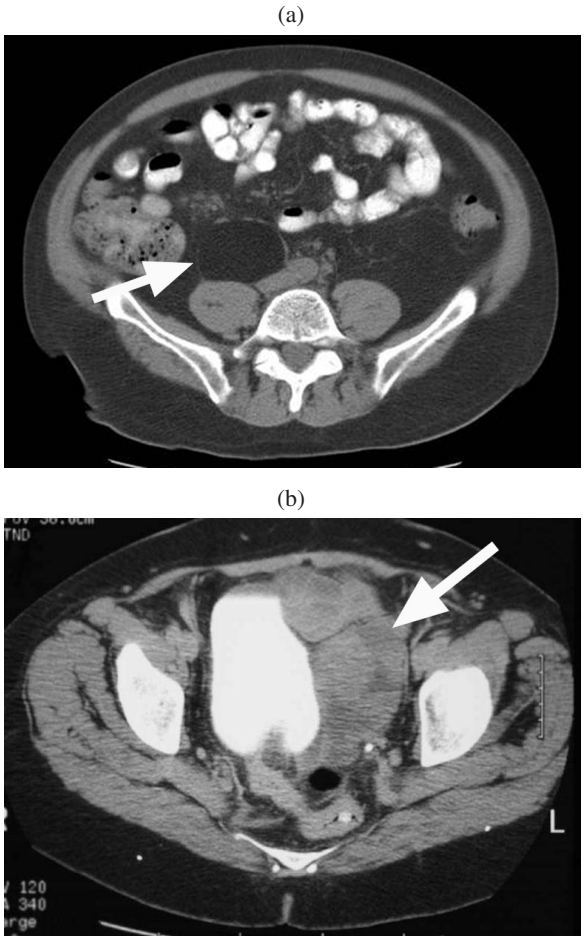


Figure 5 **Well-differentiated low-grade and high-grade liposarcomas. (a) Contrast-enhanced axial CT shows well encapsulated fatty mass (arrow) with no septations or soft tissue component. This proved to be a well-differentiated liposarcoma. (b) Contrast-enhanced axial CT shows a predominantly soft tissue pelvic mass (arrow) which proved to be a high-grade liposarcoma.**

Treatment

The definitive treatment of primary retroperitoneal sarcomas is surgical resection[5]. Chemotherapy and radiotherapy without surgical debulking have rarely been beneficial, when used alone or in combination[5]. Pre-, intra- or post-operative radiotherapy has, however, been of benefit in some patients, but, in most instances, does not improve patient prognosis[5,24–27]. As these tumors are locally invasive, extensive and aggressive local resection of the tumor and any adjacent organs should be performed at the time of presentation. Resection of the tumor en-bloc with adjacent adrenals, kidneys, or segments of small bowel, or colon is often required[28,29]. In a study of 28 patients with liposarcomas, adjacent organ resection was carried out in more than half the cases, with partial or total resection of the kidneys in 60%, colon in 50% and adrenal glands in 35%[27].

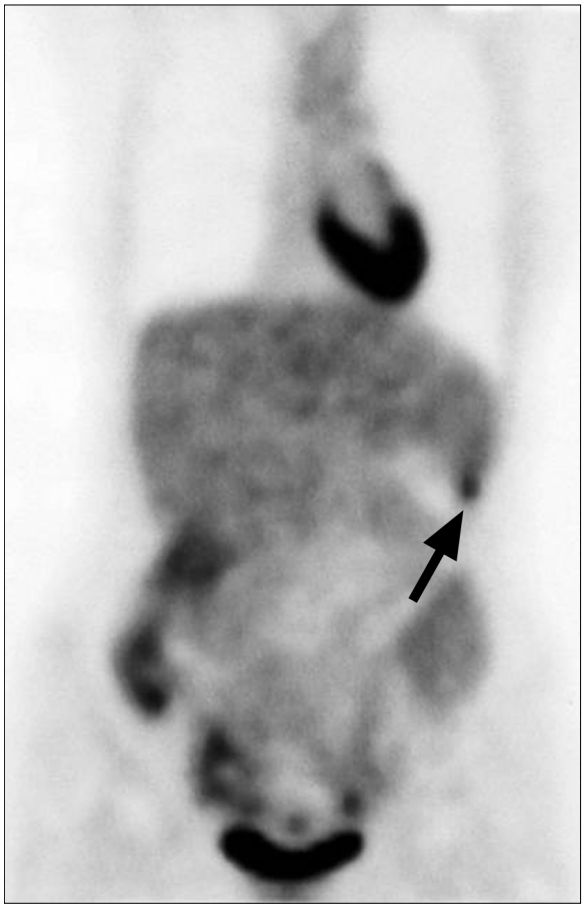
Imaging follow-up

Despite 'complete' resections, 5- and 10-year survival rates are poor, being 51% and 36% respectively[30,31]. This is most often due to local recurrences in the surgical bed. Most tumor recurrences occur within 2 years of initial surgical resection[7,32]. Since subsequent prognosis in these patients is affected by the ability to completely resect the local recurrences, early detection of tumor recurrence is important. When re-resections are performed early, they are successful in up to 90% of the patients[30,31]. Unfortunately, many recurrences are diagnosed late in the course of the disease, leading to incomplete resection, which then leads to re-recurrence in about 50% of patients.

As most recurrences are local, a careful scrutiny of the surgical bed for subtle changes on follow-up imaging

should be made[28–32]. Clinical follow is usually up not helpful as up to 50% of patients are asymptomatic, and if symptoms are present, they are usually nonspecific.

(a)



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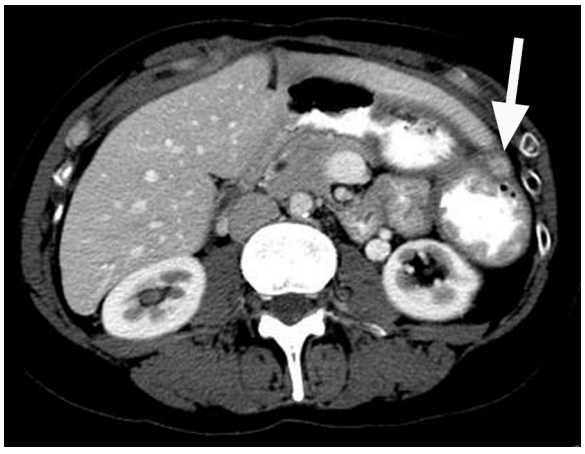
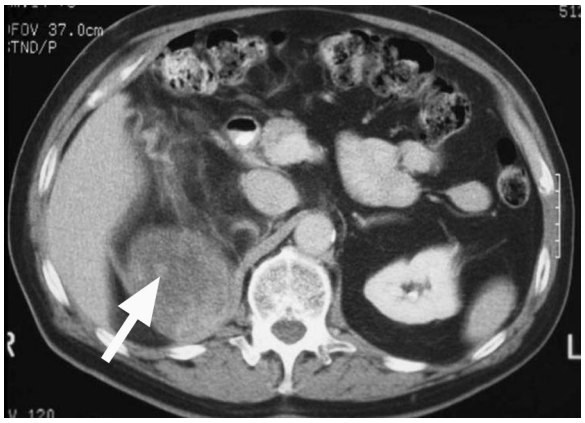


Figure 6 **Metastatic fibrosarcoma. (a) Coronal PET-FDG image shows peritoneal metastatic nodule (arrow). (b) Axial contrast-enhanced CT shows this metastatic nodule (arrow), which was not prospectively identified as it was thought to be part of the left lobe of the liver.**

(a)



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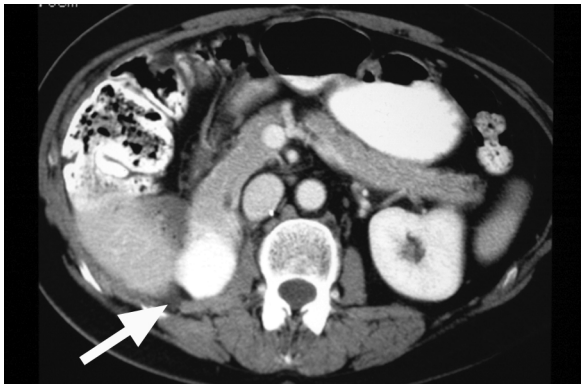

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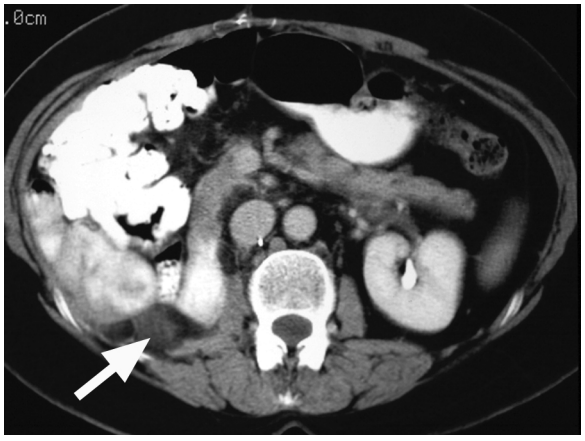

Figure 7 **Recurrent liposarcoma. (a) Contrast-enhanced axial CT shows right retroperitoneal mass (arrow) which as a liposarcoma. (b) On a 6-month post-resection follow-up contrast-enhanced axial CT, there is suggestion of recurrence (arrow). This was interpreted as being indeterminate for recurrence. (c) At 12-month follow-up contrast-enhanced axial CT, the mass (arrow) has shown interval growth and is more obvious. This proved to be recurrent liposarcoma.**

Detection of early local recurrences can be difficult. Soft tissue attenuation recurrences may not be easily distinguished from post-operative scarring/fibrosis in

the surgical bed. Detection of local recurrence in liposarcomas is especially difficulty as, when small, recurrent liposarcomas can be difficult to distinguish from normal retroperitoneal fat on imaging (Fig. 7(a)–(c))[32]. Frequently, closer scrutiny may show that the fat in a recurrent liposarcoma is of slightly higher CT attenuation when compared to normal retroperitoneal fat. Also, at times, recurrent liposarcomas can have different imaging characteristics than that of the primary tumor. In one CT study of fat-containing liposarcomas, four of the eight recurrent tumors did not contain any visible fat[32]. As recurrent tumors are best treated with repeat surgical resection, all tumor-bearing sites should be identified to enable optimal and complete re-resection[32,33].

Regional metastases are also frequent and a thorough search of the draining nodes, peritoneal surfaces, and liver should be made prior to evaluation for distant metastases.

Follow-up imaging is usually performed with CT or MRI with the frequency of follow-up being often dictated by the completeness of the tumor resection, tumor type and grade[34]. One suggested follow up scheme is to obtain imaging at regular intervals (i.e. CT or MRI every 3–4 months for 2 years, then every 4–6 months for 3–5 years, and every 12 months thereafter. Follow up for greater than 5 years is recommended as although most sarcomas (whether high-grade or low-grade) recur within 2 years, marked delay in appearance of recurrent disease is not unusual.

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