Living with Sarcoma

A guide for patients and families

Written by Dr. Pablo Lozano Lominchar

Surgical oncologist · Hospital General Universitario Gregorio Marañón, Madrid

This guide is written for you and for the people beside you. No prior knowledge is needed. Read it at your own pace and take it to your appointment with your questions noted down.

What Is Sarcoma?

A Cancer of Connective Tissue

Sarcoma is a type of cancer that arises from the connective tissues of the body — the structural materials that support and connect all our organs and body parts. These include fat (adipose tissue), muscle, fibrous tissue, blood vessels, nerves, cartilage, and bone. Unlike the more common carcinomas (such as breast, colon, or lung cancer) which arise from epithelial cells lining body surfaces and glands, sarcomas arise from mesodermal tissues — the deep structural fabric of the body.

Sarcomas are rare. They account for approximately 1% of all adult malignancies — around 13,000 new cases per year in the United States, and roughly 5,000 per year in the European Union. Because they are uncommon, expertise is concentrated in specialist centres, and it is strongly recommended that all sarcoma patients are managed by a specialist multidisciplinary team at a reference centre.

The Two Main Categories

There are over 70 distinct subtypes of sarcoma, but they fall into two broad categories:

- **Soft tissue sarcomas (STS):** Arise from fat, muscle, fibrous tissue, blood vessels, nerves, or other non-bony connective tissues. Can occur anywhere in the body, but are most common in the extremities (arms and legs) and the retroperitoneum (the space behind the abdominal cavity).
- **Bone sarcomas:** Arise from bone (osteosarcoma) or cartilage (chondrosarcoma), and are most common in children and young adults. Ewing's sarcoma is another major category affecting bone and soft tissue.

Retroperitoneal Sarcoma: A Special Entity

The **retroperitoneum** is the space at the back of the abdominal cavity, behind the peritoneum, containing the kidneys, ureters, aorta, vena cava, and adrenal glands. Retroperitoneal sarcomas (RPS) are the most common soft tissue sarcomas affecting this space, and they present a unique surgical challenge because of their proximity to major blood vessels and vital organs.

The two most common subtypes of RPS are:

- **Liposarcoma:** Arising from fat tissue. Can be well-differentiated (slow growing, rarely spreads to distant sites) or dedifferentiated (more aggressive, with potential for distant metastasis).
- **Leiomyosarcoma:** Arising from smooth muscle, often from the wall of the inferior vena cava. More aggressive than well-differentiated liposarcoma, with a higher risk of distant metastasis.

🔑 KEY POINT

Retroperitoneal sarcomas often grow to enormous sizes (20–30 cm or more) before causing symptoms, because the retroperitoneal space is large and accommodating. An abdominal mass discovered incidentally on imaging — often performed for an unrelated reason — is one of the most common presentations.

Diagnosis and Staging

Imaging

The standard imaging workup for a suspected retroperitoneal sarcoma includes:

- **CT scan of chest, abdomen and pelvis** with intravenous contrast: evaluates the primary tumour, its relationship to adjacent organs and vessels, and any pulmonary or hepatic metastases.
- **MRI of the abdomen**: superior to CT for assessing soft tissue detail, tumour composition (fat vs. solid), and the relationship to the spine, psoas muscle, and neural structures.
- **PET-CT**: used selectively, particularly for high-grade sarcomas, to detect occult distant metastases.

Biopsy: The Sarcoma Rule

Before any treatment is planned, a tissue diagnosis is essential. For retroperitoneal masses, this is typically obtained by **CT-guided core needle biopsy (CNB)**, performed by an interventional radiologist. The biopsy tract must be planned carefully to avoid contaminating uninvolved tissue planes.

⚠️ PITFALL

Never undergo biopsy or surgery for a suspected sarcoma at a centre that does not have specialist sarcoma expertise. A poorly planned biopsy can compromise subsequent curative surgery. If in doubt, seek a second opinion at a sarcoma reference centre before any intervention.

Grading

Sarcomas are graded (G1, G2, G3) based on their microscopic appearance — specifically the degree of differentiation, mitotic activity, and tumour necrosis. Grade is one of the most important prognostic factors:

- **G1 (low grade):** Well-differentiated; slow growing; low risk of distant metastasis
- **G2 (intermediate grade):** Intermediate behaviour
- **G3 (high grade):** Poorly differentiated; faster growing; significant risk of distant metastasis

TNM Staging (AJCC 8th Edition)

Stage	T	N	M	Grade
IA	T1	N0	M0	G1/Gx
IB	T2, T3, T4	N0	M0	G1/Gx
II	T1	N0	M0	G2/G3
IIIA	T2	N0	M0	G2/G3
IIIB	T3, T4	N0	M0	G2/G3
IV	Any T	N1 or M1	Any	Any

Treatment and Recovery

Surgery: The Cornerstone of Treatment

For localised retroperitoneal sarcoma, surgery is the only potentially curative treatment. The goal is an **R0 resection** — complete removal of the tumour with microscopically negative margins — performed en bloc with any adherent or involved adjacent structures.

The surgical philosophy for retroperitoneal sarcoma has evolved significantly in the past 15 years. The TARPSWG (Trans-Atlantic Retroperitoneal Sarcoma Working Group) consensus recommends **compartmental resection**: rather than removing only the tumour itself, the surgeon removes the tumour together with all ipsilateral structures in the same anatomical compartment, even if they appear macroscopically uninvolved. For a left-sided liposarcoma, this typically means removing the tumour, the left kidney, the ipsilateral psoas muscle, and the left hemicolon.

This aggressive approach reduces local recurrence rates: without compartmental resection, local recurrence affects approximately 50–60% of patients within 5 years. With proper compartmental resection, local recurrence rates fall to 25–35%.

Radiotherapy

Preoperative (neoadjuvant) radiotherapy is used at some centres to reduce the size of the tumour before surgery and to sterilise the margin at greatest risk of positive resection. The evidence for routine preoperative radiotherapy in RPS is still evolving — discuss this option specifically with your multidisciplinary team.

Chemotherapy

The role of adjuvant (post-operative) chemotherapy in retroperitoneal sarcoma is limited and not standard practice for most histological subtypes. Systemic chemotherapy is used for metastatic disease (doxorubicin ± ifosfamide as first-line) and in selected cases of locally advanced or recurrent disease. Specific subtypes (e.g. leiomyosarcoma) may have higher chemosensitivity.

Follow-Up After Surgery

Given the significant risk of local recurrence, follow-up after sarcoma surgery is more intensive than for many other cancers:

- CT of chest, abdomen and pelvis every 3–4 months for the first 2 years
- Every 6 months from year 2 to year 5
- Annual review from year 5 onwards

⚠️ ALARM SIGNS DURING FOLLOW-UP

- Progressive abdominal swelling or fullness
- New or worsening abdominal pain
- Unexplained weight loss
- New onset of back pain (possible retroperitoneal recurrence)
- Unexplained fatigue

Do not wait for your next scheduled appointment — contact your sarcoma team promptly.

Living Well After Sarcoma Surgery

Recovery from retroperitoneal sarcoma surgery typically takes 6–10 weeks for the physical component. However, the psychological adjustment to a sarcoma diagnosis and the associated follow-up burden can take much longer. Many patients benefit from psychological support, oncology social work, or peer support groups — do not hesitate to ask for these resources.

Most patients are able to return to work and their previous activities of daily living within 3–4 months of surgery, provided there are no major complications. Discuss specific activity restrictions with your surgical team at your follow-up appointments.

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Pablo Lozano Lominchar, MD, PhD, EBPSM | HGUGM / UCM | © 2025

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