

CAR/CSAR Practice Guideline on Retroperitoneal Sarcomas (RPS)

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Abstract

Retroperitoneal sarcomas (RPS) are a heterogeneous family of neoplasms. The Canadian Association of Radiologists Retroperitoneal Sarcoma Working Group was established to provide recommendations for the imaging evaluation and initial workup of suspected RPS in the context of the Canadian healthcare system. The recommendations, which are based on available literature and expert consensus, provide guidance on the imaging pathway, biopsy approach and surveillance for RPS.

Résumé

Les sarcomes rétro-péritonéaux (SRP) constituent une famille hétérogène de néoplasmes. Le Groupe de travail de l'Association canadienne des radiologistes sur les sarcomes rétro-péritonéaux a été créé afin de formuler des recommandations relatives à l'évaluation par imagerie et au bilan initial des SRP présumés dans le contexte du système de santé canadien. Ces recommandations, fondées sur la littérature scientifique et le consensus d'experts, fournissent des directives sur le parcours d'imagerie, les techniques de biopsie et la surveillance des SRP.

Keywords

retroperitoneal neoplasms, sarcoma, diagnostic imaging, image-guided biopsy, multidisciplinary team, practice guidelines

Background and Rationale

Retroperitoneal sarcomas (RPS) comprise 12% to 15% of soft tissue sarcomas, with an incidence of 0.5 to 1 cases/100 000 per year.^{1,2} These tumours are often large at time of detection and typically require multivisceral resection for definitive management, with wide margins difficult to achieve due to relationships with critical structures.^{3,4}

Sarcomas tend to grow to a very large size in the retroperitoneum before symptoms develop (eg, palpable mass, abdominal pain, nerve pain).^{5,6} Some RPS, such as liposarcomas and leiomyosarcomas, have characteristic imaging appearances, but the differential diagnosis may remain broad.⁷ Failure to recognize RPS can lead to inappropriate management with devastating consequences, such as incomplete resection or contamination of the peritoneal cavity with tumour. These surgical misadventures can render a potentially curable situation incurable, underscoring the importance of accurate diagnosis and specialized management from the outset.⁸

To date, there have been no standardized Canadian guidelines for the diagnostic approach to retroperitoneal masses. The lack of consensus on appropriate imaging and biopsy techniques can lead to delayed or suboptimal multidisciplinary

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Table 1. Consensus Recommendations for Diagnostic Imaging, Biopsy, Referral and Surveillance of Suspected Retroperitoneal Soft Tissue Masses.

Diagnostic imaging

1. Multiplanar contrast-enhanced CT of the abdomen and pelvis is recommended as the standard initial investigation. Following diagnosis, and if appropriate, CT chest is recommended for staging.
2. Additional imaging beyond the initial CT should be performed at the discretion of the surgical oncologist or MDT based on clinical context.
3. Bone scan, head CT and brain MRI are usually not required for staging.
4. MRI and 18F-FDG PET/CT imaging should not delay a biopsy or referral to a sarcoma specialist.

Biopsy

5. Tissue diagnosis before treatment is recommended, with biopsy requested by a surgical oncologist or other sarcoma specialist following imaging review.
6. A percutaneous image-guided coaxial core biopsy using a retroperitoneal approach is recommended, targeting the most hyper-enhancing area of the solid-appearing component. CT guidance is most commonly used, but US can be used as an alternative if the targeted area is visible and accessible.
7. Transperitoneal biopsy is generally avoided and should only be considered after additional consultation with a surgical oncologist.
8. Collection of a minimum of 4-6 cores is recommended and should be informed by the preferences of the local pathologist.
9. A needle gauge of 14-18 G is recommended.

Referral

10. New diagnoses should be reviewed by an MDT in a specialized sarcoma referral centre.
11. The MDT should include expertise in surgical oncology, radiology, pathology, medical and radiation oncology.

Surveillance

12. Following resection, contrast-enhanced CT of the chest, abdomen and pelvis should be performed at 3 to 6-month intervals for the first 5 years, and annually thereafter for high-grade tumours. The length of annual surveillance should be at the discretion of the surgical oncologist. Lower-grade tumours may require less intensive surveillance.

management. To support best practices in imaging, the RPS working group has developed recommendations for diagnostic imaging and imaging-guided biopsies to facilitate definitive diagnosis, management and surveillance. These guidelines, based on a review of existing literature and a Delphi consensus process, are intended to serve as a reference for radiologists and multidisciplinary care teams involved in the care of RPS patients. A sample reporting checklist is provided in Appendix Table A1.

Methodology

The Canadian Association of Radiologists Retroperitoneal Sarcoma Working Group is composed of radiologists (JY, JG, AH, AC, JD, JB, BA, SC) and surgical oncologists (AM, TH, AD, SB) from across Canada, with experience in retroperitoneal imaging and management. To determine the consensus statements and recommendations, the working group used a modified Delphi consensus process to complete two rounds of questionnaires rating 13 statements across the categories of diagnostic imaging, biopsy, referral and follow-up (Table 1).

Statements were rated anonymously for agreement on a 7-point scale. Statements that received ratings of 6.0 to 7.0 from at least 60% of respondents were considered to have reached consensus in Round 1. Statements with ratings between 3.0 and 5.9 were carried forward to the second round, incorporating suggested revisions based on feedback from the working group. The same criteria for agreements and disagreements were applied in the second round. Following both

rounds of voting, the working group convened to review all the statements and confirm consensus.

Following the completion of the Delphi process and finalization of the recommendations, an independent pathologist (TN) reviewed the final consensus statements. This author did not participate in the anonymized voting rounds but provided formal clinical review, revisions and approval of the final text prior to publication.

The consensus statements apply for retroperitoneal masses of non-visceral origin (Table 2).^{9,10} The guidelines apply to suspected or possible sarcomas, and do not apply to neoplasms originating from solid organs (eg, renal angiomyolipomas), some of which may only be excluded after biopsy. Visceral sarcomas are excluded as they often require a different diagnostic approach.

Role of Imaging in Retroperitoneal Sarcoma Diagnosis

Computed tomography (CT) of the abdomen and pelvis with intravenous iodinated contrast is considered the best imaging modality for the diagnosis and workup of RPS.^{8,9,11-15} The administration of contrast aids in determining tumour origin, identifying enhancing solid components for biopsy targeting, delineating anatomic relationships for surgical and radiation planning, and recognizing anatomical variants.^{5,8,16,17} To optimize its utility, thin slices (3mm) and multiplanar reformats are suggested.^{18,19} Oral contrast is not necessary to help characterize retroperitoneal masses or to

Table 2. Masses Included and Excluded From the Scope of These Guidelines.

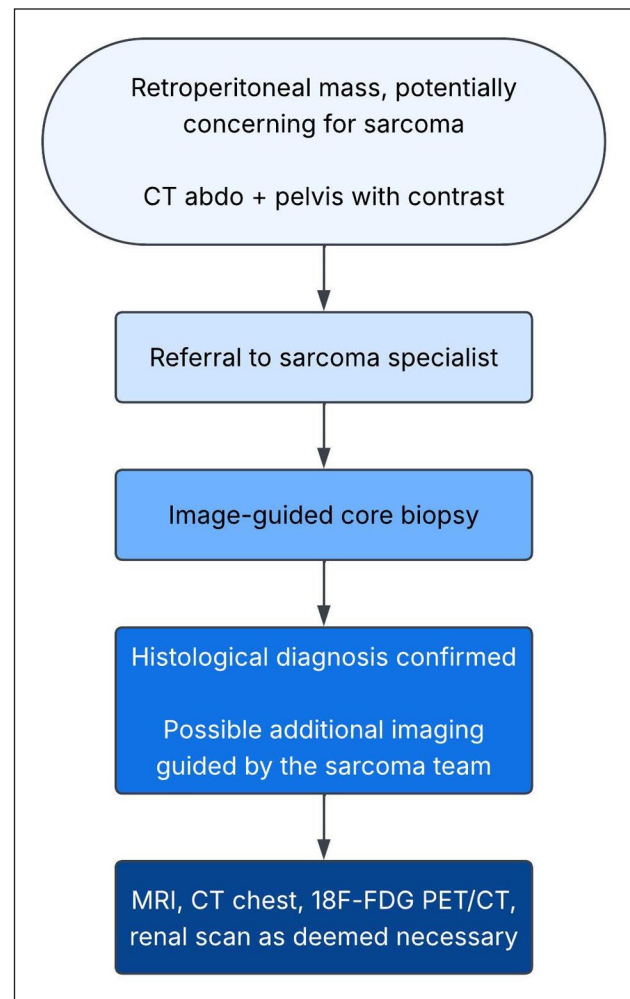
Include	Exclude
Lipoma	Benign entities such as benign peripheral nerve sheath tumour/schwannoma, ganglioneuroma, lymphangioma and angiomyolipoma.
Liposarcoma	Desmoid type fibromatosis and aggressive angiomyxoma.
Leiomyosarcomas, most commonly arising from major veins or ureter	Gastrointestinal stromal tumours, visceral sarcomas such as those arising from the gut or its mesentery, uterine leiomyosarcoma, and other gynecologic sarcomas, prostatic sarcoma and paratesticular/spermatic cord/inguinal canal sarcoma.
Unclassified/undifferentiated pleomorphic sarcoma of the psoas	Ewing family sarcomas, desmoplastic small round cell tumour, alveolar/embryonal rhabdomyosarcoma, sarcoma arising from teratoma, sarcomatoid carcinoma (carcinosarcoma), clear cell sarcoma and radiation-associated sarcoma.
Synovial Sarcoma	Adrenal cortical carcinoma, paraganglioma and malignant pheochromocytoma
Myxofibrosarcoma	
Solitary fibrous tumour	
Malignant peripheral nerve sheath tumour	

Source. This list was derived from Swallow et al.¹⁰ *Management of Primary Retroperitoneal Sarcoma (RPS) in the Adult: An Updated Consensus Approach from the Transatlantic Australasian RPS Working Group.*

help delineate their relationship with adjacent structures. Biphasic/hypervascular protocol is often only indicated in instances of vascular encasement, as requested by the surgical oncologist.

Any additional imaging beyond the initial CT should be performed at the discretion of the surgical oncologist or multidisciplinary team (MDT) based on clinical context (Figure 1). In contrast to extremity soft tissue masses, routine abdominal magnetic resonance imaging (MRI) is not required for adequate characterization of RP soft tissue masses. Following diagnosis, and if appropriate, a staging CT chest is recommended to evaluate thoracic involvement.^{9,10,12,13,15} For example, up to 50% of IVC leiomyosarcomas may present with metastases at diagnosis, with the commonest sites being lung (15%) and liver (17%).²⁰ Bone scan, CT head and brain MRI are usually not indicated for routine staging.^{9,10} Renal scan for functional assessment of the contralateral kidney may be considered in cases of equivocal perfusion on CT or in patients with previously documented severe hypersensitivity reaction to intravenous iodinated contrast agents.^{10,21,22}

MRI and 18F-FDG Positron Emission Tomography (PET) CT modalities should not delay biopsy or referral to a sarcoma specialist, although they may be considered prior to treatment at the discretion of the surgical oncologist and/or multidisciplinary tumour board. Possible indications for MRI include staging in patients with contrast allergies and assessing anatomic relationships and tumour extension, such as into the neural foramina, sciatic notch and inguinal canal.^{6,9} In addition, MRI can delineate the extent of peritumoral edema prior to initiating radiation therapy.²³ In the event of equivocal CT findings (sclerotic bone lesions, borderline-enlarged lymph nodes), 18F-FDG PET/CT can help rule out distant metastatic disease in high-grade masses. Although 18F-FDG PET/CT can help differentiate high-grade from low-grade lesions at primary diagnosis, it is inadequate in distinguishing

**Figure 1.** Imaging workflow for RPS.

Note. CT abdomen and pelvis is the main imaging modality for diagnosis. MRI and PET should not delay biopsy and referral to a sarcoma specialist.

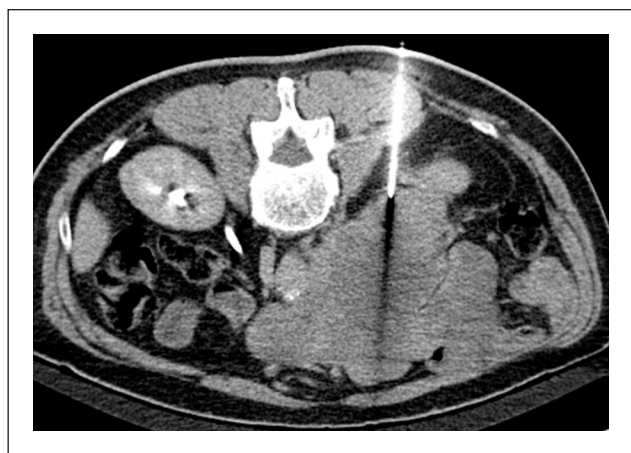


Figure 2. Seventy-five years old male high-grade dedifferentiated liposarcoma with posterior solid component targeted via retroperitoneal approach for core biopsy with a 17G coaxial needle.

low-grade masses from benign pathologies and is not consistently reliable for the evaluation of recurrence.^{24,25}

Referral to Specialist Sarcoma Centres

Multiple professional societies strongly recommend early referral to an experienced multidisciplinary team in a sarcoma specialist centre.^{10,12,13,15,17,26,27} Management of primary RPS at these centres is associated with improved short-term post-operative outcomes, reduced risk of recurrence, and improved long-term survival.²⁸⁻³⁰ These centres are more likely to have accurate histologic diagnosis by an experienced pathologist, multidisciplinary discussions of all cases, and more opportunities for clinical trial participation. A 2021 prospective study found that MDT discussion affected treatment plan in 50% of patients.³¹ Another study with data spanning 7 years reported twofold lower odds ratio of death when patients were managed at a sarcoma specialist centre with MDT.²⁸ The MDT should include at least one surgical oncologist, radiologist, pathologist, medical oncologist and radiation oncologist with expertise in the treatment of RPS.³² Retroperitoneal masses should be referred to the appropriate surgeon so that they can be investigated and managed appropriately.

Minimum institutional volumes to define a specialist sarcoma centre is an unsettled question, with disparate thresholds applied globally due to the rarity of RPS and geographic considerations. The American National Cancer Database has identified 13 RPS resections as the minimum annual volume associated with improved long-term survival.³³ Consensus guidelines from the 2021 Transatlantic Australasian RPS working group specify that a minimum of 10 to 20 RPS annual cases is required for an institution to be considered a dedicated RPS centre.^{10,33} In comparison, the NHS England Sarcoma Service specification recommends an annual case volume of 24 new primary cases.^{13,34} Similar to the Spanish

Group for Research in Sarcomas, the Canadian Working Group reached a consensus to omit a specific institutional number for this paper.¹²

Biopsy Considerations

Image-guided biopsy is strongly recommended as the standard of care. It enables consideration of neoadjuvant therapy and surgical planning according to histologic grade and subtype.^{9,12,15,17,35-37} Biopsy should be undertaken in the pre-treatment phase, since treatment-related changes can complicate pathological diagnosis and grading. There are exceptional cases when biopsy may not be performed (eg, no planned treatment, not technically feasible), but such a decision should be discussed with a surgical oncologist.^{8,9,13,17,36}

Percutaneous co-axial core needle biopsy is the preferred method of histological sampling, following imaging review.^{9,17} The risk of needle track seeding is minimal (0.5%) and is not a reason to forgo biopsy.³⁸ Furthermore, using a co-axial system allows multiple samples via a single percutaneous access track. Fine-needle aspiration/cytology should be avoided as it is rarely diagnostic and can result in treatment delays.^{9,10} Surgical biopsy should also be avoided as it increases the risk of peritoneal cavity contamination from tumour rupture, lacks image guidance to reliably target high-grade areas, and distorts tissue planes, making subsequent resection more technically challenging.^{9,10}

Most retroperitoneal masses can be biopsied safely using a retroperitoneal approach, which has a low early complication rate of 3.4%.³⁸ The retroperitoneal approach tends to be the safest method to minimize the risk of peritoneal seeding, bowel injury and patient discomfort.^{39,40} It is important to carefully plan the optimal biopsy route to minimize risk to critical retroperitoneal structures (aorta, inferior vena cava, renal hilum) and viscera, while being limited by the vertebra, sacrum and iliac crest. Transperitoneal biopsy should only be undertaken after additional consultation with a surgical oncologist. For heterogeneous masses, the biopsy should target the region most likely to be high-grade on pathology, which is typically the most hyper-enhancing area of the solid-appearing component (Figure 2).^{41,42} Biopsy of a deeply located tumour is most commonly performed under CT-guidance, but ultrasound can be used if the tumour is superficial, easily accessible, and visible.

The needle gauge and the number of passes impacts the diagnostic pathologic yield.⁹ A typical range of 4 to 6 cores and 14 to 18-gauge needles is recommended for morphological and molecular diagnosis, in accordance with local pathologist preferences.^{9,15,17,26} Biopsy specimens are generally fixed in 4% buffered formalin and subsequently embedded in paraffin blocks. Detection of MDM2 amplification by immunohistochemistry (IHC) and/or fluorescence *in situ* hybridization (FISH) is helpful for the diagnosis of well-differentiated and dedifferentiated liposarcomas.⁴³⁻⁴⁵ At minimum, two 18G cores are required; however, some institutions may prefer

more tissue samples (eg, six 14G cores), particularly for diagnosing well-differentiated liposarcoma, given lower core yield and evolving subclassification for prognostication.⁴⁶ Centres employing other methods for histological diagnosis may require more tissue using a larger calibre needle or additional cores. Given the variation in practice across Canadian institutions, this RPS Working Group recommends that radiologists consult with their local pathology departments on institutional preferences prior to biopsy.

Follow-Up

The likelihood and time interval of recurrent disease depends on numerous factors, including tumour size, histological subtype, tumour grade, completeness of resection and receipt of neoadjuvant radiotherapy.^{4,17,18,47,48} Following curative-intent resection, locoregional recurrence may occur in up to 35% of patients and is associated with 75% of RPS-related deaths.⁴⁹ There is a lack of published data on optimal follow-up of surgically treated patients. Most societies recommend follow-up with a contrast-enhanced CT of the chest, abdomen and pelvis at 3 to 6-month intervals for the first 5 years, and annually thereafter, although the appropriate duration of surveillance has not been established.^{9,12,13,17,50} Lower-grade tumours may not require as frequent follow-up imaging. For patients with well-differentiated or low-grade dedifferentiated liposarcomas, CT chest may not be routinely required for surveillance.^{12,49}

Conclusion

Retroperitoneal sarcomas are a rare and heterogeneous group of neoplasms. Prompt recognition by the radiologist is required to ensure appropriate diagnostic imaging, biopsy and referral to a specialized sarcoma centre with a multidisciplinary team. This guideline provides a set of recommendations from a multidisciplinary working group on diagnostic imaging, biopsy planning and surveillance imaging in the Canadian setting.

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Appendix

Table A1. Sample Checklist for Reporting.

Tumour characteristics

Size in 3 dimensions

Composition – solid, fat, septa, mixed, necrotic, calcifications

Location in the retroperitoneal space – *abutment and direction of displacement of viscera (pancreas, spleen, kidney, splenic flexure, duodenojejunal flexure, uterus and adnexa)*

Enhancing components (targets for biopsy)

Resection considerations/preoperative planning

Vascular anatomy

Conventional? Identify any variants for major arteries and veins

Vascular involvement

Encasement and compression – celiac axis, porta hepatis (if left sided), superior mesenteric vessels, IVC, gonadal, iliac, lumbar vessels

Feeding vessels

Adjacent collateral veins

Extension: Note special structures involved at tumour margins

Psoas, iliopsoas extent of contact

Inguinal canal [femoral nerve (*consider MRI*)]

Sciatic notch

Obturator foramen

Sacroiliac

Vertebral foramen (neural foramen, nerve root, central canal)

Diaphragmatic hiatus

Visceral invasion

True invasion present? (rare, high-risk feature)

Invasion of parietal wall

Contralateral kidney:

Normal/abnormal. State any abnormalities

Other viscera:

Normal/abnormal. State any abnormalities

Bones

Normal/abnormal. State any abnormalities

Lymphadenopathy: consider alternate diagnosis if present

Complications

Hydronephrosis

Nerve compression (femoral nerve)

Bowel obstruction

Intestinal perforation

Pulmonary embolism

Management

Recommend evaluation by a multidisciplinary sarcoma team

Follow-up

No evidence of recurrent or metastatic disease

Evidence of recurrent or metastatic disease