

VOLUME 42

AUA

# UPDATE SERIES

2023

LESSON 17

## Nonrenal Retroperitoneal Tumors

**Learning Objective:** At the conclusion of this continuing medical education activity, the participant will be able to describe the differential diagnosis of nonrenal retroperitoneal tumors and the appropriate diagnostic and staging evaluation and multidisciplinary management of these tumors and to recognize open surgery as the most appropriate surgical approach to retroperitoneal sarcoma and list 2 separate ureteral reconstructions that might be required.

This AUA Update aligns with the American Board of Urology Module on Oncology, Urinary Diversion, and Adrenal. Additional information on this topic can be found in the AUA Core Curriculum sections on Anatomy & Physiology, and Oncology-Upper Tract.



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**KEY WORDS:** nonrenal, retroperitoneal sarcoma, retroperitoneal tumor

## INTRODUCTION

This review specifically focuses on primary nonrenal retroperitoneal tumors, a rare but diverse group of neoplasms that arise within the retroperitoneum outside of the major organs in this space.<sup>1</sup> Approximately 75% of nonrenal tumors found in the retroperitoneum are malignant; however, they make up only 0.1% to 0.2% of all malignancies. The differential diagnosis of tumors involving the retroperitoneal space is broad, encompassing benign and malignant etiologies. Tumors may be of mesenchymal, epithelial, or germ cell origin. Lymphoproliferative and metastatic lesions can also be found.

## ANATOMY OF THE RETROPERITONEUM

To approach retroperitoneal tumors, a detailed understanding of anatomy is necessary. The retroperitoneum is a complex potential space located between the parietal peritoneum and the transversalis fascia.<sup>2</sup> Broadly, the retroperitoneum can be divided into 3 compartments (lateral, median, and posterior). The median or “vascular” compartment contains the great vessels, while the posterior compartment contains the iliopsoas musculature. The lateral compartment contains 3 separate spaces defined by their relationship to the leaves of the renal fasciae: anterior pararenal, posterior pararenal, and perirenal (Figure 1).

Retroperitoneal organs may be either primarily or secondarily retroperitoneal, depending on their embryological

development. However, retroperitoneal organs are abdominal organs, which are not suspended by mesentery and are incompletely covered by peritoneum. Primarily retroperitoneal organs (kidney, ureter, adrenal gland) are found in the inverted cone-shaped perirenal space, surrounded by the contiguous renal fasciae. Secondarily retroperitoneal organs (ascending/descending colon, second through fourth portions of the duodenum, head and body of the pancreas) started intraperitoneally but migrated posteriorly during development; thus, these are found in the anterior pararenal space. Finally, the posterior pararenal space is mainly composed of fat.

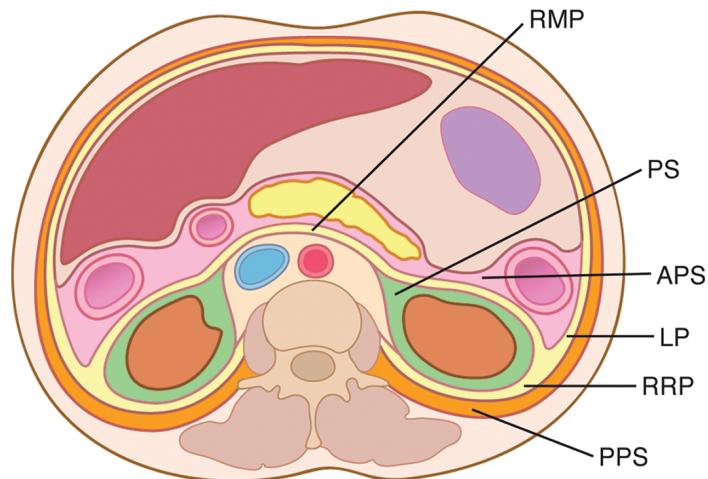
## PRESENTATION OF NONRENAL RETROPERITONEAL TUMORS

Nonrenal retroperitoneal tumors often pose both a diagnostic and management challenge. The retroperitoneal spaces can expand significantly, accommodating a large tumor mass without causing symptoms. These tumors often remain asymptomatic until they grow large enough to compress or invade contiguous structures. Symptomatic patients often present with tumors exceeding 10 cm and may present with abdominal distention and discomfort, early satiety, and a potentially palpable mass. They may also present with obstructive gastrointestinal and urological symptoms due to displacement or direct tumor involvement of organs, with neurological symptoms from compression of lumbar or pelvic nerves, or with both. Smaller, asymptomatic tumors are found incidentally on cross-sectional imaging obtained for unrelated symptoms.

## EVALUATION OF NONRENAL RETROPERITONEAL TUMORS

The full evaluation will be guided by the clinician’s suspicion based on the initial clinical history, physical examination, and imaging. For example, some benign lesions can be diagnosed on the basis of CT findings alone, whereas others may require specific functional imaging studies, such as <sup>18</sup>F-fluorodeoxyglucose (FDG) positron emission tomography (PET)/CT or <sup>68</sup>Ga-DOTATATE PET/CT.<sup>3</sup> **Percutaneous core needle biopsy (CNB)** is an important component of the diagnostic workup for most retroperitoneal masses before formulation of the management plan and prior to initiation of any treatment. Each patient’s evaluation should be undertaken with input and guidance from an experienced, multidisciplinary team.<sup>4-6</sup>

**Imaging.** When a retroperitoneal tumor is identified on cross-sectional imaging, specific imaging features provide insight into the etiology. **For example, well-differentiated liposarcoma (WDLPS) demonstrates a characteristic imaging appearance with a predominantly fatty component displacing (not invading) surrounding structures.** The presence of such features as macroscopic fat, myxoid stroma, calcifications, cystic areas, vascularization, and mantle growth pattern provides additional information for radiological interpretation.



**Figure 1.** Anatomy of the retroperitoneum. Abdominal organs that are not suspended by the mesentery and lie between the abdominal wall and parietal peritoneum are said to lie within the retroperitoneum. Several individual spaces make up the retroperitoneum. These spaces are the anterior pararenal space (APS), posterior pararenal space (PPS), and the perirenal space (PS). The retromesenteric plane (RMP), retrorenal plane (RRP), and lateral conal plane (LP) are also shown. Reprinted with permission from Tirkes et al, *Radiographics*. 2012;32(2):437-451.

**ABBREVIATIONS:** core needle biopsy (CNB), dedifferentiated liposarcoma (DDLPS), <sup>18</sup>F-fluorodeoxyglucose (FDG), positron emission tomography (PET), retroperitoneal sarcoma (RPS), well-differentiated liposarcoma (WDLPS)

If a retroperitoneal sarcoma (RPS) is suspected, staging should be performed<sup>4</sup> with CT of the chest, abdomen, and pelvis with intravenous and oral contrast. Arterial or triple-phase CT imaging may be useful for operative planning. MRI can be considered in select circumstances, such as CT contrast allergy or if better anatomical delineation of specific sites is needed. <sup>18</sup>F-FDG PET/CT can be considered before planned surgical resection of a high-grade retroperitoneal liposarcoma but is not mandatory for staging.

Finally, assessment of the contralateral kidney function is important if the tumor is isolated to a single lateral compartment, which can be estimated with a CT scan or a nuclear medicine scan with split renal differential function.

**Biopsy.** Percutaneous coaxial image-guided CNB (14- to 18-gauge) is considered standard of care for diagnosis, particularly in the case of a suspected RPS.<sup>4-6</sup> Multiple CNBs should be obtained to allow for adequate histological and molecular subtyping by a specialized sarcoma pathologist. Sampling of more solid tumor components (higher perfusion on contrast imaging or high standardized uptake value on <sup>18</sup>F-FDG PET/CT) may limit undergrading. Open and laparoscopic biopsy should not be performed because of the risk of tumor contamination and distortion of surgical planes.<sup>4</sup> If an expert radiologist has determined that the imaging is definitive and there is no plan for preoperative treatment, then a biopsy may be omitted. However, this should only be decided after evaluation by an expert tumor board.

**Patient selection and optimization for receipt of multimodal treatment.** Patients with large retroperitoneal tumors may be deconditioned and/or malnourished at presentation. Malnutrition in the RPS patient has been associated with worse postoperative outcomes, including higher rates of postoperative complications and longer hospitalization following surgery.<sup>7,8</sup> Nutritional status may be assessed by objective measures, such as loss of >5% body weight; arm circumference; albumin, prealbumin, and transferrin levels; and lymphocyte count. Patients may benefit from preoperative oral nutritional supplementation for at least 2 weeks prior to surgery and participation in a prehabilitation program.<sup>9</sup>

## DIFFERENTIAL DIAGNOSIS OF NONRENAL RETROPERITONEAL TUMORS

**RPS.** RPS makes up 10%-15% of soft tissue sarcomas with an annual incidence of 0.5-5 cases per 100,000 population.<sup>4</sup> Prognosis is driven by histological subtype.<sup>1,10</sup> The most common RPS histologies are liposarcoma (50%-65%) and leiomyosarcoma (2%-25%); other histological subtypes also arise within the retroperitoneum but are less common.<sup>4</sup> Percutaneous CNB should generally be performed,<sup>4-6</sup> and pathological diagnosis should be made by a sarcoma expert pathologist, according to the 2020 World Health Organization classification,<sup>1</sup> before multidisciplinary management recommendations are made or treatment initiated.

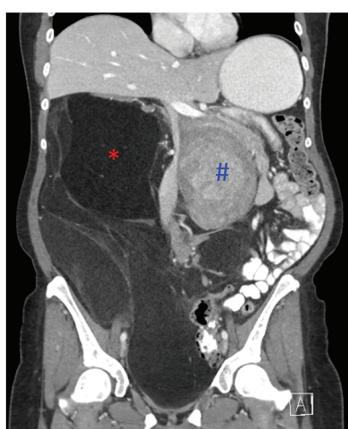
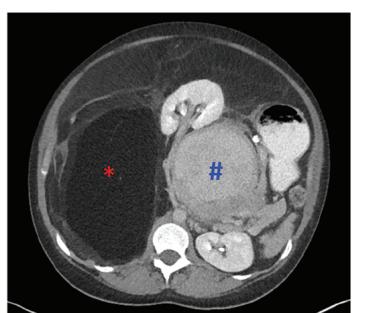
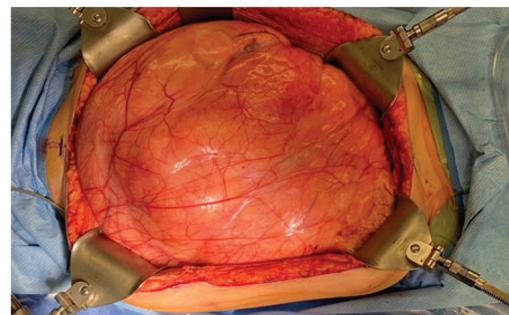
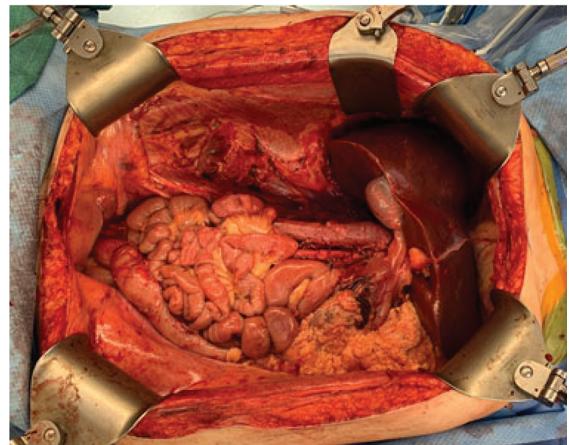
Importantly, most patients benefit from multidisciplinary discussion and management. Due to the rarity and complexity of these tumors, patients with RPS should be referred to sarcoma reference centers and/or within reference networks sharing multidisciplinary expertise

and treating a high number of patients annually.<sup>4-6</sup> Data from the United States<sup>11-13</sup> and Europe<sup>14-18</sup> have consistently shown better outcomes—including reduced risk of early postoperative morbidity, postoperative mortality, local recurrence, and longer overall survival—among patients treated at sarcoma reference centers or reference networks compared to community settings.

The most common liposarcoma subtypes occurring in the retroperitoneum are WDLPS and dedifferentiated liposarcoma (DDLPS). WDLPS and DDLPS share cytogenetic abnormalities and often co-occur in patients; however, they are characterized by different clinical behavior and prognosis.<sup>10</sup> DDLPS often has distinct radiographic and pathological features, which distinguishes it from WDLPS; percutaneous biopsy should be performed of suspected DDLPS component of a mixed WDLPS/DDLPS. **Detection of MDM2 and/or CDK4 amplification can help distinguish WDLPS from benign adipose tumors and DDLPS from other undifferentiated sarcomas.**<sup>1</sup> Myxoid, round cell, and pleomorphic liposarcomas are uncommon. Retroperitoneal liposarcomas may be quite large at presentation, often abutting but rarely invading adjacent retroperitoneal structures and organs (Figure 2). Local recurrence following liposarcoma resection occurs in 35%-60% of patients. **Although WDLPS generally does not distantly metastasize, up to 30% of patients with DDLPS will develop distant metastasis, and thus staging evaluation of patients with DDLPS should include chest imaging.** Although standard-of-care treatment algorithms for retroperitoneal liposarcoma are lacking and there have been no prospective, randomized studies demonstrating a survival benefit of systemic therapy for patients with liposarcoma, systemic treatments may be considered for patients with DDLPS after expert multidisciplinary discussion either in the neoadjuvant or adjuvant treatment setting. In particular, neoadjuvant systemic therapy is often considered for patients with recurrent disease that may be borderline resectable.<sup>4</sup> **Although WDLPS is largely chemoresistant, DDLPS is more chemosensitive, and responses have been observed with doxorubicin-based regimens or gemcitabine-docetaxel.**<sup>19,20</sup> For patients whose DDLPS progresses on these first-line systemic therapies, eribulin and trabectedin have shown clinical benefit.<sup>21,22</sup> **The role of radiation therapy remains a matter of debate, although patients with lower-grade liposarcoma may benefit from preoperative radiation therapy.**<sup>23-25</sup>

Retroperitoneal leiomyosarcoma typically arises from the inferior vena cava (Figure 3), its tributaries, or any small vessel and can also arise from the uterus or gastrointestinal tract. **Distant metastatic disease is more common, affecting up to 60% of leiomyosarcoma patients.**<sup>10,26</sup> Thus, patients diagnosed with leiomyosarcoma should undergo appropriate staging imaging (PET/CT with contrast or CT of the chest/abdomen/pelvis). **For patients with resectable leiomyosarcoma, preoperative systemic therapy should be considered.**<sup>27,28</sup> Preoperative radiation therapy may be helpful in cases in which an “at-risk” margin is a concern.

**Lymphoproliferative neoplasms (benign and malignant).** Lymphoproliferative neoplasms typically demonstrate a mantle growth pattern, characterized by lesions that involve adjacent structures without any signs of infiltration.<sup>3</sup> These

**A****B****C**

**Figure 2.** A 65-year-old woman who presented with abdominal distention and early satiety underwent resection of a large retroperitoneal liposarcoma with well-differentiated (\*) and dedifferentiated (#) components. A, The tumor abuts and displaces the posterior aspect of the head of the pancreas and duodenum, small bowel, and large bowel, and it encases the right kidney and renal hilum. B, Large liposarcoma arising from retroperitoneum. C, Peritoneal cavity after resection of liposarcoma.

**A****B****C**

**Figure 3.** A 64-year-old woman who presented with back pain was diagnosed with (A) a leiomyosarcoma arising from the inferior vena cava-right renal vein confluence. The tumor abuts the aorta (\*), duodenum (\*\*), and psoas muscle and vertebral body (#). B, Patient received preoperative systemic therapy (doxorubicin/dacarbazine) and radiation therapy, with radiographic and pathological response. C, Patient underwent radical resection of right retroperitoneal leiomyosarcoma with en bloc right nephrectomy with inferior vena cava reconstruction using polytetrafluoroethylene interposition tube graft.

may be malignant (lymphoma) or benign (Erdheim-Chester disease, retroperitoneal fibrosis). Percutaneous biopsy is an important tool in initial clinical management.

**Lymphoma is the most common malignant retroperitoneal neoplasm and typically presents as a para-aortic or pelvic mass that involves adjacent structures but does not invade them.**<sup>18</sup>  $^{18}\text{F}$ -FDG PET/CT is typically used for staging. High-grade, aggressive lymphomas (ie, B-cell, nodular sclerosing Hodgkin) display more FDG avidity.<sup>29,30</sup> Molecular subtyping and nodal architecture are fundamental to treatment planning, thus requiring at minimum percutaneous CNB with flow cytometry. On occasion, excisional biopsy of a lymph node may be required for diagnosis or to guide management. Treatment typically comprises chemotherapy  $\pm$  radiation  $\pm$  immunotherapy.

Secondary metastasis to the retroperitoneal nodes may look similar to lymphoma. **Testicular cancer, discussed below, is a common primary cancer that metastasizes to the retroperitoneum.** Other common primary cancer sites that metastasize to the retroperitoneum include esophageal, hepatic, gastric, prostatic, colorectal, cervical/ovarian, and urothelial.

Erdheim-Chester disease is a rare, benign, non-Langerhans cell histiocytosis of unknown origin.<sup>3</sup> It is characterized by multiorgan histiocytic inflammation and commonly affects men ages 40-60 years. In the retroperitoneum, it is typically bilateral, involving the perirenal and periaortic space. Mainstays of treatment include steroids and immunomodulatory and cytotoxic agents.

Retroperitoneal fibrosis also is more common in male patients. Two-thirds of these cases are classified as primary (idiopathic), and the remaining cases are considered secondary, caused by infection, trauma, or malignancy. On imaging, homogenous attenuation of soft tissue surrounding the major vessels  $\pm$  ureters is seen. Ureteral involvement may lead to obstruction. Idiopathic disease is treated with steroids  $\pm$  tamoxifen, whereas secondary retroperitoneal fibrosis is treated by addressing the underlying etiology.

**Germ cell tumors (primary or secondary).** Germ cell tumors typically arise from the testes or ovaries and secondarily involve the lymph nodes in the retroperitoneum through metastatic spread. However, 1%-2.5% may originate in extragonadal locations, typically in midline structures (ie, mediastinum, brain, retroperitoneum).<sup>31</sup> These extragonadal tumors are hypothesized to originate from the arrested descent of primordial germ cell rests during the fourth through sixth weeks of embryogenesis.<sup>32</sup>

Regardless of where they originate, germ cell tumors are classified as either pure seminomas or nonseminomas using serum tumor markers and histopathology.<sup>33</sup> Clinical staging includes cross-sectional imaging (chest/abdomen/pelvis) and assessment of tumor markers and may incorporate the pathological staging of the testicular tumor.<sup>34</sup> Testicular tumors follow a well-defined pattern of spread based on their laterality, referred to as “landing zones.” Tumors that originate in the right testicle most commonly involve the interaortocaval lymph nodes; the next most common sites are the pre- and paracaval lymph nodes. Left-sided testicular tumors usually involve para-aortic and preaortic nodes. Right-sided tumors are more likely to cross over to the contralateral (aortic) side. Primary retroperitoneal germ cell tumors are found in

the midline, and in general, their outcomes are not as good as those of tumors that originate in the testicle.<sup>35</sup> Treatment for retroperitoneal germ cell tumors may include a combination of surgery, radiation, or chemotherapy, depending on the clinical stage. An important subtype of germ cell tumor is the teratoma, a pluripotent tumor that may be either immature (malignant) or mature (benign). On imaging, it is often cystic with areas of fat and calcification.<sup>3</sup> Although considered benign, mature teratomas are resistant to chemotherapy and radiation; can grow rapidly, causing compression of surrounding organs; and can undergo malignant transformation. Therefore, they need to be surgically removed.

**In male patients with retroperitoneal adenopathy, serum tumor markers ( $\beta$ -human chorionic gonadotropin,  $\alpha$ -fetoprotein, lactate dehydrogenase) and testicular ultrasound examination should be performed.** PET imaging is not indicated for the up-front evaluation or staging of testicular cancer.

**Epithelial and metastatic tumors.** Numerous malignancies can secondarily involve the retroperitoneal space. These can arise from retroperitoneal organs (adrenal gland, kidney, pancreas) or result from metastatic spread from other sites in the body. Subtle findings on cross-sectional imaging beyond the scope of this review may be helpful in determining whether a retroperitoneal tumor is arising from a retroperitoneal organ or is a primary retroperitoneal tumor. Percutaneous biopsy should also be considered in these situations.

**Neurogenic tumors (ganglioneuromas).** Ganglioneuromas are rare benign tumors that arise along the sympathetic plexus and within the adrenal medulla and occur in both pediatric and adult patients. These tumors are often identified incidentally or during evaluation of nonspecific symptoms associated with mass effect (Figure 4, A). **Diagnosis of ganglioneuromas can be challenging as they are not associated with specific symptoms and do not have typical imaging characteristics or laboratory findings, and their management can be challenging as their potential for recurrence after resection and for malignant transformation to neuroblastoma remains unclear.**<sup>36</sup> In a recent multicenter retrospective analysis of 328 patients with ganglioneuroma treated at 29 institutions across 5 continents, the median age at diagnosis was 37 years (range 4-79), and 41% of patients presented with symptoms.<sup>37</sup> Malignant degeneration to neuroblastoma was rare (0.9%), and patients who underwent nonoperative management (n=116, 35%) had indolent disease courses. Among patients who underwent surgical resection (n=212, 65%) for symptoms, tumor growth, or concern for malignancy, recurrence was uncommon (n=4, 2%).

**Pheochromocytomas/paragangliomas.** Pheochromocytomas and paragangliomas are tumors that arise from chromaffin cells of the adrenal medulla and from neuroendocrine cells of the extra-adrenal autonomic paraganglia, respectively. Pheochromocytomas and ~40% of paragangliomas release catecholamines, resulting in hypertension and arrhythmia, and up to 40% of these tumors are malignant. Evaluation of a patient with suspected pheochromocytoma or paraganglioma should include measurement of fractionated metanephrenes and normetanephrenes in 24-hour urine or free metanephrenes in plasma; levels 3 times above the upper limit of normal are

diagnostic.<sup>38</sup> An adrenal protocol abdomen/pelvis CT scan is recommended. If metastatic or multifocal disease is suspected, somatostatin receptor imaging (somatostatin receptor PET/CT or somatostatin receptor PET/MRI with <sup>68</sup>Ga-DOTATATE tracer),<sup>39</sup> chest CT scans, and metaiodobenzylguanidine scan should be performed as appropriate. Although pheochromocytomas and paragangliomas may be sporadic, many are associated with inherited genetic syndromes (MEN2A, MEN2B, neurofibromatosis, von Hippel-Lindau syndrome), and patients diagnosed with these tumors should undergo genetic counseling and testing.<sup>38</sup> Surgical resection is the mainstay of treatment for patients with resectable disease, and a minimally invasive approach is appropriate when safe and feasible. Patients may experience significant and potentially life-threatening hypertension intraoperatively. **Careful preoperative preparation with  $\alpha$ -adrenergic blockade, volume repletion, and high-salt diet is required.**

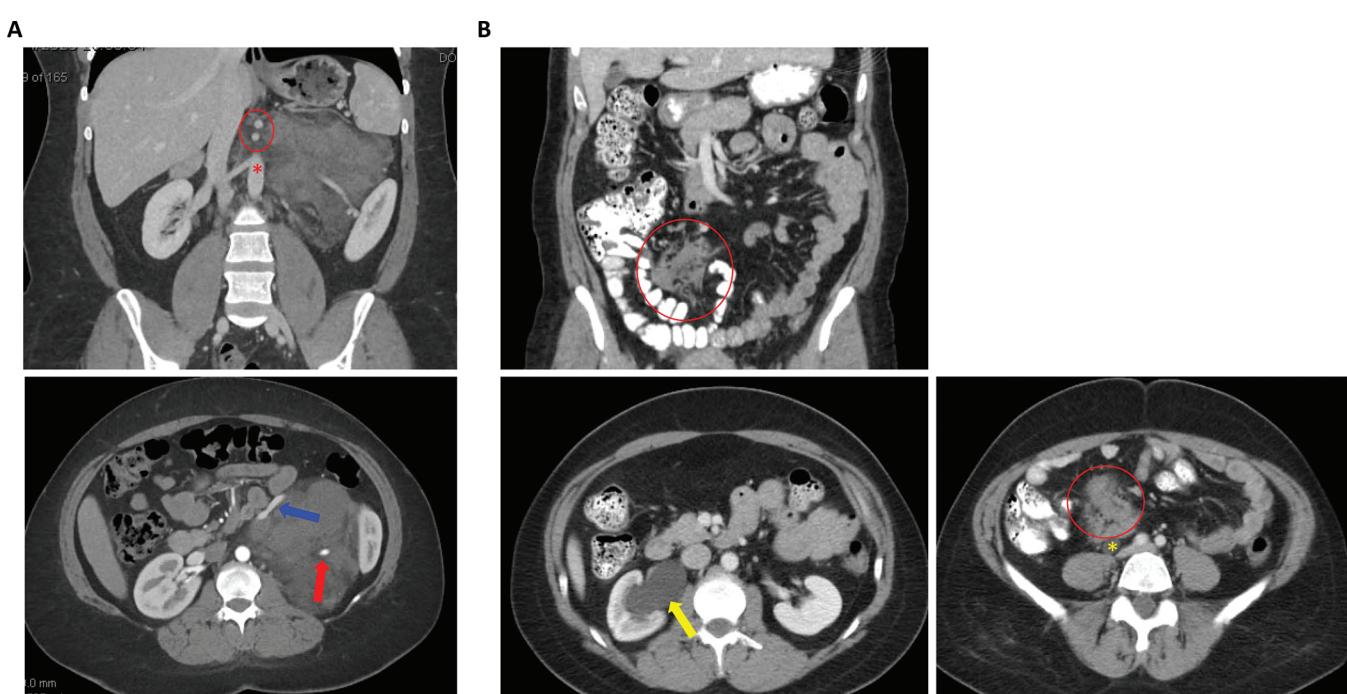
*Desmoid fibromatosis (also known as aggressive fibromatosis).* Desmoid tumors are an aggressive fibroblastic proliferation of differentiated fibrous tissue, which can be locally invasive but rarely metastasize and can cause functional morbidity (Figure 4, B). Desmoids may arise spontaneously or following surgical intervention. Abdominal desmoids may arise in 7.5%-16% of patients with familial adenomatous polyposis; therefore, patients presenting with desmoid tumors should be evaluated for Gardner syndrome/familial adenomatous polyposis.<sup>5,40</sup> Management of desmoid tumors is challenging due to the extent of surgery often required and high local recurrence rate after resection. **All patients should be managed by a multidisciplinary team.**<sup>5</sup> Asymptomatic resectable desmoid fibromatosis can be managed expectantly if the tumors are not located in an area that could lead to functional limitations if the tumor progresses; 20% of desmoid tumors may spontaneously regress.<sup>41,42</sup> If there is progression,

patients can be treated with surgery and/or systemic therapy.<sup>5</sup> For symptomatic patients with tumors causing pain, functional limitation, and/or morbidity, treatment options include surgery and/or systemic therapy. Radiation therapy is not generally recommended for retroperitoneal/intra-abdominal desmoid tumors. Systemic therapy using nonsteroidal anti-inflammatory drugs (sulindac), hormonal or biological agents (tamoxifen), tyrosine kinase inhibitors (imatinib, sorafenib), and cytotoxic drugs (methotrexate and vinblastine, doxorubicin-based regimens) has shown responses in patients with desmoid tumors.<sup>5</sup>

## THE ROLE OF SURGERY FOR NONRENAL RETROPERITONEAL TUMORS

*Preoperative considerations and surgical planning.* Surgery is often required in the management of nonrenal retroperitoneal tumors, and is often complex and requires careful preoperative patient assessment and optimization. Importantly, it also requires engagement and involvement of multiple surgical specialists and multi-team coordination and cooperation during the preoperative surgical planning process, intraoperatively, postoperatively, and during follow-up. Specifically, surgery for RPS requires thoughtful planning and coordination of expertise by a sarcoma surgeon and other surgical subspecialists such as a urologist, vascular surgeon, colorectal surgeon, orthopedic surgeon, and/or plastic surgeon.

From a urological standpoint, stent placement at the start of the surgical procedure to aid with intraoperative ureteral identification and injury recognition may be considered. For patients with ureteral obstruction at presentation of a nonrenal retroperitoneal tumor, nephrostomy tube placement should be performed prior to planned surgical resection.



**Figure 4.** Examples of benign nonrenal retroperitoneal masses. A, Ganglioneuroma abutting and/or encasing the aorta (\*), celiac and superior mesenteric arteries (red circle), and left renal artery (red arrow) and vein (blue arrow). B, Desmoid fibromatosis (red circle) involving the mesentery and retroperitoneum causing right ureteral obstruction, hydronephrosis (\*), and hydronephrosis (yellow arrow).

**Technical considerations of surgery for RPS.** Although surgery is the mainstay of treatment (and the only potentially curative treatment) for resectable RPS, complete resection can be challenging due to the close proximity of these often large tumors to vital structures. Wide, negative margins are often not feasible due to anatomical constraints, and positive microscopic margin is generally assumed. **The goal of surgery is to achieve complete resection of the intact tumor, which may require multivisceral organ resection, and avoidance of piecemeal tumor resection.**<sup>10,43,44</sup>

Although it is common for an RPS to displace a kidney, most sarcomas do not invade directly into the renal parenchyma, and in some cases stripping the renal capsule en bloc with the mass can facilitate renal preservation with negative surgical margins. To do this, the surgeon must open the renal capsule away from the mass and continue stripping the capsule until the kidney is off the mass and the mass is free. **Many liposarcomas involve the renal hilum and/or Gerota's fascia or encase the ipsilateral kidney; nephrectomy is usually needed in these cases (unless the patient has a contraindication for nephrectomy) and is generally well tolerated.**<sup>45</sup>

The ureter is the other structure that is commonly involved. If the upper ureter is involved, transureteroureterostomy is safe and effective. It is vital during transureteroureterostomy to adequately mobilize the donor and recipient side to ensure a completely tension-free anastomosis. With aggressive mobilization of the donor kidney and the recipient kidney to create an iatrogenic horseshoe kidney, very short ureters of 3-5 cm can be successfully reconstructed (Figure 5). The renal capsule or Gerota's fascia of 1 kidney can be sewn to Gerota's fascia of the other kidney to limit traction. If the renal capsule has been completely stripped, a large sheet of oxidized cellulose polymer (eg, Surgicel) can be used as a bolster to anchor a donor kidney in proper orientation for transureteroureterostomy.

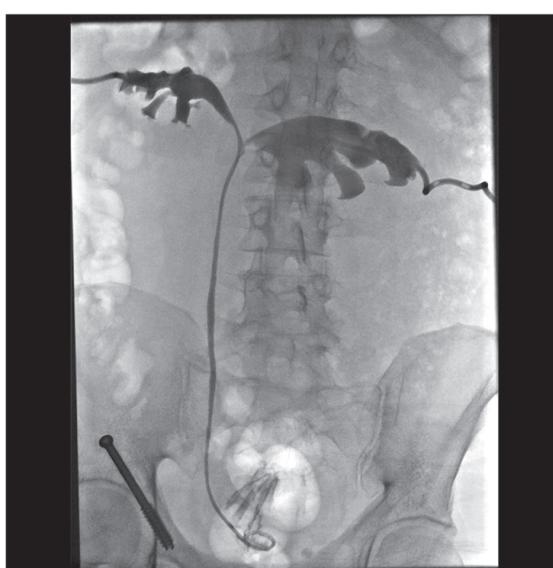
Some large liposarcomas will elevate and stretch the ureter and cause the mid-ureter to become ischemic during resection. The mid-ureter can sometimes be reconstructed with an ipsilateral ureteroureterostomy, which has the greatest success for a ureteral resected length of  $\leq 5$  cm.<sup>46</sup> Mobilizing the kidney downward can facilitate a tension-free ureteroureterostomy. If the lower ureter is involved, a ureteroneocystomy with or without psoas hitch is the preferred approach.<sup>47</sup>

**Open vs minimally invasive surgery.** For patients with a known benign retroperitoneal mass, minimally invasive surgical procedures, including robotic approaches, can be considered depending on the experience of the surgeon and the location and size of the mass.<sup>48</sup>

**However, for patients with RPS, the initial approach and surgical quality of the patient's first operation is critically important for maximizing the chance of cure. If the first operation is inadequate or suboptimal, the recurrence risk is higher and the possibility of cure is significantly reduced as locoregional recurrences are generally incurable.** RPS is almost invariably large at presentation, and it is not uncommon that patients presenting with WDLPS and DDLPS undergo resection of the dedifferentiated (solid) component while the well-differentiated (lipomatous) component is unrecognized as part of the malignancy and is left behind (Figure 6). Incomplete gross resection and tumor rupture are profound predictors of local recurrence and worse oncologic outcomes. **Therefore, a minimally invasive surgical approach is not recommended nor standard of care for RPS.**<sup>4,49</sup>

**Complications of surgery for retroperitoneal tumors.** Early postoperative complications include bleeding/hemorrhage, superficial or deep surgical space infection, pancreatic leak/fistula, deep venous thrombosis/pulmonary embolism, and delayed recognition of ureteral injury. Later in the postoperative period, complications that may develop include bowel obstruction from adhesive disease, incisional

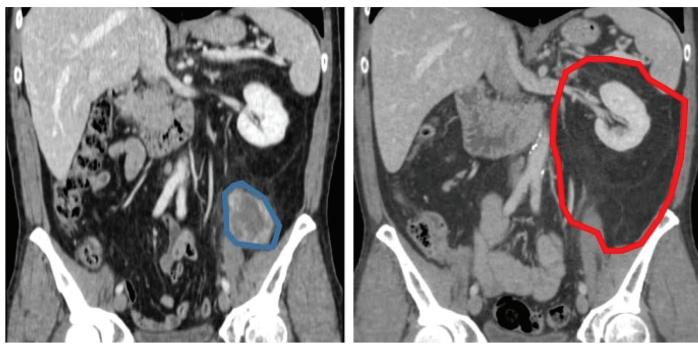
A



B



**Figure 5.** A 41-year-old man who underwent resection of a complex retroperitoneal sarcoma with reconstruction of both the inferior vena cava and aorta. A "horseshoe" type aggressive bilateral renal mobilization was performed to facilitate right-to-left transureteroureterostomy. In the postoperative period, the patient had edema at the transureteroureterostomy anastomosis, requiring bilateral percutaneous nephrostomy tubes. A, Bilateral antegrade nephrostograms showing the altered axis of the renal pelvis bilaterally to facilitate transureteroureterostomy. B, Coronal image from a surveillance CT scan showing donor right kidney and ureter and lower pole of recipient left kidney.



**Figure 6.** CT scan before (left) and after (right) incomplete laparoscopic resection of a retroperitoneal liposarcoma. The dedifferentiated component (blue line) was removed laparoscopically; the well-differentiated component (red line) was in place. Reprinted with permission from Gronchi A, et al, *Ann Surg Oncol*. 2018;25(8):2129-2131.<sup>49</sup>

hernia, ischemic ureteral stricture, and uncommonly, renal insufficiency in those who underwent en bloc nephrectomy.<sup>45</sup> Patients who underwent major vascular reconstruction as part of their operation for retroperitoneal tumor resection should undergo surveillance for vascular graft/vessel thrombosis or stricture at the discretion of the participating vascular surgeon.

Chyle leaks can occur in up to 5%-7% of patients undergoing major retroperitoneal surgical procedures, such as post-chemotherapy retroperitoneal lymph node dissection, due to operative trauma to the cisterna chyli. **Most lymphatic leaks resolve with conservative management consisting of dietary modification, drainage, and supportive care.** However, refractory chyle leaks can be devastating complications requiring prolonged management with total parenteral nutrition, shunting, and reoperation. **Recent data support lymphangiembolization as a viable treatment option for chyle leaks that do not improve with dietary modification alone.**<sup>50</sup>

Venous thromboembolic events after major abdominal oncologic surgery are a major cause of postoperative morbidity, and current guidelines recommended 28 days of prophylaxis with low-molecular-weight heparin, typically enoxaparin, to reduce this risk.

## CONCLUSION

Nonrenal retroperitoneal tumors encompass a broad range of benign and malignant entities. In most cases, percutaneous CNB is an important component of the diagnostic workup in order to guide multidisciplinary treatment decisions. For many patients with nonrenal retroperitoneal tumors (including RPS and desmoid fibromatosis), management should be undertaken by or with input and guidance from an experienced, multidisciplinary team.

## DID YOU KNOW?

- Nonrenal retroperitoneal tumors are rare, accounting for <1% of all malignancies, and most (75%) are malignant.
- The differential diagnosis of nonrenal retroperitoneal tumors is broad and includes both malignant entities (RPS, lymphoproliferative malignancies, germ cell tumors, epithelial tumors, and metastatic lesions) and benign entities (neurogenic tumors, pheochromocytomas/paragangliomas, desmoid fibromatosis).
- Retroperitoneal tumors are often large before they result in symptoms, and initial evaluation should include cross-sectional imaging and percutaneous CNB for histological diagnosis.
- Patients with RPS should be referred to sarcoma reference centers or network facilities (per national and international clinical practice guidelines). Expert multidisciplinary sarcoma care rendered at expert sarcoma reference centers/network facilities is associated with improved postoperative and oncologic outcomes.
- Multivisceral resection, including en bloc nephrectomy, may be required for appropriate surgical management of RPS; tumor rupture, piecemeal resection, and grossly incomplete resection should be avoided.

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## Study Questions Volume 42 Lesson 17

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1. The preferred technique/approach to obtaining a biopsy of a nonrenal retroperitoneal tumor is
  - a. Incisional biopsy
  - b. Percutaneous fine needle aspiration
  - c. Percutaneous core needle biopsy
  - d. Excisional biopsy
2. The most common retroperitoneal malignancy is
  - a. Sarcoma
  - b. Metastatic disease
  - c. Lymphoma
  - d. Pheochromocytoma/paraganglioma
3. The posterior pararenal space contains which of the following?
  - a. Kidney
  - b. Adrenal
  - c. Fat pads
  - d. Tail of pancreas
4. A PET scan is not indicated for the initial evaluation and staging of which of the following tumors?
  - a. Germ cell tumor
  - b. Lymphoma
  - c. Pheochromocytoma
  - d. High-grade liposarcoma
5. A patient is found on routine imaging to have a 10-cm retroperitoneal mass abutting the inferior vena cava and right ureter. Biopsy confirms liposarcoma. Guidelines for the management of retroperitoneal sarcoma include the following surgical approaches with an experienced multidisciplinary surgical team
  - a. Open surgery
  - b. Open or robotic surgery according to surgeon experience and preference
  - c. Open, laparoscopic, or robotic surgery according to surgeon experience and preference
  - d. Extraperitoneal approach by open, laparoscopic, or robotic surgery according to surgeon experience and preference