



Dorothy Fox. *Neighborhood Gossip*. Watercolor, 30" × 36".

The pathology, prognostic features, and treatment options for localized and metastatic retroperitoneal sarcomas are reviewed.

Retroperitoneal Sarcomas

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Background: *The evaluation and treatment of retroperitoneal sarcomas are challenging because the tumors are relatively rare and frequently present with advanced disease in an anatomically complex location.*

Methods: *We reviewed the literature on experience in the management of retroperitoneal sarcomas, and we present our own experience in the treatment of these tumors.*

Results: *The identification of prognostic factors other than the adequacy of resection has been inconsistent. Due to a lack of associated symptoms, retroperitoneal sarcomas smaller than 5 cm are rare. Computed tomography is the most useful tool in the evaluation of retroperitoneal tumors. Surgery, radiation therapy, and chemotherapy are treatment options, but the most important factor in the treatment of primary tumors is complete surgical resection. The role of neoadjuvant and adjuvant therapies is not defined and should be considered within the context of clinical trials.*

Conclusions: *Early referral of patients with retroperitoneal soft tissue tumors will help to ensure that they will receive the benefits of multidisciplinary evaluation and treatment of their disease and ready access to clinical trials.*

Introduction

Soft tissue sarcomas are rare, with approximately 8,600 new cases diagnosed annually in the United States — less than 1% of all newly diagnosed malignancies.¹ One third of malignant tumors that arise in the retroperitoneum are sarcomas, and approximately 15% of soft tissue sarcomas arise in the retroperitoneum.² Retroperitoneal sarcomas are malignant tumors arising from mesenchymal cells, which are usually located in muscle, fat, and connective tissues. Retroperitoneal sarcomas have varying clinical courses depending on their histologic subtype and grade. The rarity of retroperitoneal sarcomas, combined with the vast array of histologic subtypes, has complicated our understanding of these tumors and impeded the development of effective therapies.

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Abbreviations used in this article: CT = computed tomography, MRI = magnetic resonance imaging, IORT = intraoperative radiation therapy.

In this article, we discuss the pathology and prognostic features of retroperitoneal sarcomas as well as the treatment options for localized and metastatic disease.

Pathology

Sarcomas are believed to develop from mesenchymal stem cells residing in muscle, fat, and connective tissues. The origin of these stem cells is unclear, and occasionally even their mesenchymal derivation is in question (as for nerve or nerve sheath sarcomas, gastrointestinal stromal tumors, and primitive neuroectodermal tumors). Two prevailing theories suggest that mesenchymal stem cells are found in local tissue pools or arise from the bone marrow.³ Approximately half of retroperitoneal sarcomas are high-grade tumors.^{4,13} The most commonly encountered histologic subtypes of retroperitoneal sarcoma are liposarcoma (41%), leiomyosarcoma (28%), malignant fibrous histiocytoma (7%), fibrosarcoma (6%), and malignant peripheral nerve sheath tumor (3%).^{4,9,14,15}

Prognostic Features

Characterizing the heterogeneous group of tumors called sarcomas has been difficult due to the relative rarity of each subtype. Our understanding of the clinical features and natural history of retroperitoneal sarcomas is largely limited to case reports and single-institutional experiences. To study sufficient numbers of patients with sarcomas, these reports generally combined data from patients who were treated over several decades, who underwent markedly different treatment regimens, and who had histologic subtypes with dramatically different clinical behaviors. Despite these limitations, a number of salient features associated with retroperitoneal sarcomas can be identified that are generally supported by studies involving large and often prospectively collected databases.

The identification of prognostic factors other than the adequacy of resection has been inconsistent across studies. Tumor size has not been identified as a predictor of survival since virtually all retroperitoneal sarcomas are larger than 5 cm at presentation. Tumor grade has been reported as a significant factor in some studies, with the weight of evidence supporting shorter recurrence-free and overall survival for patients with high-grade tumors.^{4,14,16-26}

Presentation and Evaluation

Sarcomas that arise in the retroperitoneum most commonly present as an abdominal mass, often without other symptoms. While the median patient age is approximately 50 years, retroperitoneal sarcomas occur at any age and arise equally in men and women. In most cases, retroperi-



Fig 1. — CT scan of a large, high-grade retroperitoneal leiomyosarcoma with lateral displacement and compression of the right kidney.

toneal sarcomas smaller than 5 cm are rarely seen since they usually are not noted by the patient until they are larger in size (Fig 1).^{4,13,16,24-27} When symptoms are present, they relate to the mass effect of the tumor or to local invasion. Early satiety, gastrointestinal obstruction or bleeding, lower extremity swelling, or pain are among the first symptoms leading to the discovery of a retroperitoneal sarcoma.

Computed tomography (CT) is the most useful tool in the evaluation of retroperitoneal tumors. A CT scan allows not only assessment of the tumor's location and its relationship to adjacent organs, but also identification of metastatic lesions in the liver or peritoneal cavity. In addition, characterization of fatty tumors and detection of intraabdominal metastasis are possible with CT scanning of the abdomen. In the pelvis, all of these features of CT are relevant, as well as excellent characterization of bony invasion. A high-quality magnetic resonance image (MRI) can be difficult to obtain, whereas CT is less sensitive to motion artifacts.²⁸ In a review of techniques and interpretation of MRI in the evaluation of retroperitoneum, Granstrom and Unger²⁹ emphasized the importance of axial images in addition to sagittal and coronal views. While MRI has been investigated in the evaluation of specific organs such as the pancreas and adrenal glands, large studies comparing MRI of retroperitoneal sarcomas with CT are lacking. Currently, we rely primarily on CT in the evaluation of soft tissue tumors arising in the abdomen and pelvis.

Once a retroperitoneal tumor has been identified, a number of clinical entities must be considered, including functioning and nonfunctioning adrenal tumors, renal tumors, pancreatic tumors, advanced gastrointestinal carcinomas, germ cell tumors, and soft tissue sarcomas. A detailed history and physical examination can help to distinguish many of these entities and direct further studies. Testicular examination, ultrasonography, and measurements of serum β human chorionic gonadotropin (β -hCG) are indicated in cases of suspected testicular cancer with



Fig 2. — CT scan of a leiomyosarcoma arising between the inferior vena cava and duodenum.

retroperitoneal metastasis. In patients with lymphadenopathy, either core needle or excisional biopsy of enlarged lymph nodes may be diagnostic for lymphoma. When tumors appear to have arisen from the stomach, pancreas, or duodenum, upper gastrointestinal endoscopy with biopsy may be diagnostic. Likewise, colonoscopy with biopsy can be useful in diagnosing tumors arising from the colon. If these diagnoses are ruled out or are considered of low probability and sarcoma is the most likely diagnosis, the role of biopsy is controversial.

Some recommend surgical exploration as the most appropriate next step for a retroperitoneal mass suspected of being a sarcoma.² When the diagnosis may change the preoperative therapy, we perform a percutaneous biopsy. A negative biopsy does not justify a period of observation, and we proceed to surgery. Examples include the use of imatinib mesylate (Gleevec) for gastrointestinal stromal tumors or primary chemotherapy for germ cell tumors or lymphomas. Distinguishing between these diagnoses can be difficult, with nonspecific physical findings and imaging studies.

Treatment of Tumors

Surgery

Retroperitoneal soft tissue sarcomas present challenges that distinguish them from the more common soft tissue sarcomas of the extremities. Difficulty in the management of retroperitoneal sarcomas relates to their large size and the complexity of the retroperitoneal anatomy (Fig 2). Complete margin-negative resections can be difficult to achieve. While the most common site of first recurrence for patients with extremity sarcomas is a distant site, patients with retroperitoneal sarcomas are more likely to develop recurrences within the abdominal cavity. The overall survival for patients with extremity sarcomas is superior to that of patients with retroperitoneal sarcomas. Local failure is evident in nearly 90% of patients who die of retroperitoneal sarcomas, a fact that reflects the large tumor size on presentation, the inability to achieve wide surgical margins, and the limitations of adjuvant radiation and chemotherapy.¹⁷ Local failure continues to occur beyond 5 and 10 years following resection, leading some to estimate that the long-term recurrence rate for resectable retroperitoneal sarcomas exceeds 70%.^{18,30}

As with primary sarcomas of the extremities, surgery is the treatment standard for retroperitoneal sarcomas. Selected reports describing experience in the surgical management of primary retroperitoneal soft tissue sarcomas with no evidence of metastasis are summarized in Table 1. These reports have consistently documented the significance of complete resection of all gross disease in improving local control and disease-specific survival. In most reports, complete excision is achieved less than 70% of the time, with local recurrence occurring in approximately half of patients undergoing complete resection.^{4,5,7-14,19,20,22-24,26} The impact of local recurrence is reflected in diminished overall survival despite attempts at further resections.^{4,16,31} Recurrent disease confers a decrease in the ability to resect all disease and achieve long-term disease-free survival in those who have their recurrence completely resected.

Table 1. — Selected Reports Evaluating Surgical Treatment of Primary Retroperitoneal Soft Tissue Sarcomas

Study	No. of Patients ^a	Complete Resection ^b	5-Year Local Recurrence ^c	5-Year Overall Survival ^d	Study Design
Lewis et al ⁴ (1998)	500	80%	59%	70%	Retrospective review of prospectively collected data
Jaques et al ⁹ (1990)	114	65%	49%	Not reported	Retrospective review of prospectively collected data
Stoeckle et al ⁵ (2001)	165	65%	48%	46%	Retrospective review of National Registry data
Hassan et al ¹⁰ (2004)	97	78%	44%	51%	Retrospective review of case series

^a Total patients in study, including some presenting with recurrent disease.
^b Percent resected with primary retroperitoneal sarcomas.
^c Percent local recurrences among those who had complete surgical resection for primary retroperitoneal sarcoma.
^d Five-year overall survival among those who had complete surgical resection for primary retroperitoneal sarcoma.

With the possible exception of low-grade retroperitoneal liposarcomas, no survival benefit has been observed when incomplete resection is undertaken.^{4,9,15,16,19,32} However, major complication rates are identical for partial and complete resections. Thus, patients undergoing incomplete resection are exposed to the morbidity of the procedure but without the potential survival benefit achieved by their counterparts who undergo complete excision. This emphasizes the need for careful preoperative planning as well as determination of unresectability early in the operative procedure so that incomplete resections are not mandated because the surgeon has passed “the point of no return.”

Retroperitoneal liposarcomas represent a distinct situation that may justify a more aggressive surgical approach, including multiple resections for repeated recurrences and even occasionally incomplete resections. Liposarcomas in this location have been observed to have a lower incidence of distant metastases (7%) than that of other histologic subtypes (15-34%).^{9,14,31} Shibata and associates³¹ observed prolonged survival in patients with partially resected liposarcomas compared with those who had only biopsy. They also reported effective palliation of symptoms in 75% of patients who underwent debulking procedures.

The clinical presentation and imaging evaluation of retroperitoneal sarcomas were discussed previously. The next step is determining tumor resectability. It is often difficult to determine preoperatively if adjacent vascular structures or organs are involved with tumor. In a review by Kilkenny and colleagues,¹⁹ vascular involvement was noted in 34% of patients undergoing resection. When the tumor is near major vessels but routine CT does not resolve whether the vessels are involved, we use MR or CT angiography. Multivisceral resections are required in the majority of cases (63%-86%), most frequently involving the kidney, colon, small bowel, pancreas, and bladder.^{4,9,10,16,19,21,24} Determining whether adjacent organs will be attached to or freely separable from the tumor can be difficult based on preoperative imaging. In planning resection, there is a high likelihood that extensive en bloc resections will be required to achieve complete excision. A retroperitoneal mass should not be surgically treated unless the surgeon is prepared for the magnitude of the resection that may be required.

Radiation Therapy

Several investigators have explored methods to decrease the incidence of local failure following resection. Extrapolating from evidence that supports improved local disease control with the use of radiation therapy for sarcomas of the trunk and extremities, radiation therapy is widely used as an adjunct to surgery in retroperitoneal sarcomas. However, radiation therapy is more problematic in the treatment of retroperitoneal sarcomas. To date, no randomized trial has evaluated the role of adjuvant radiation for retroperitoneal sarcomas.

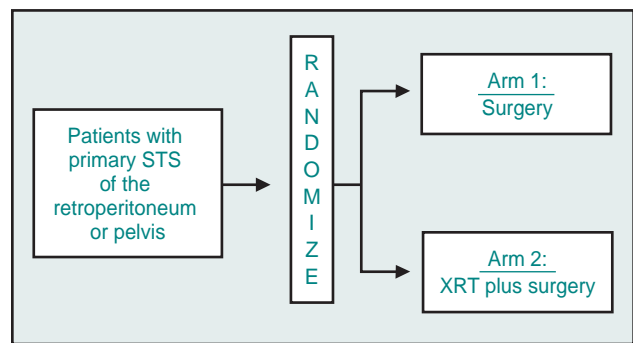


Fig 3. — Trial schema for Z9031, the prospective, randomized, multi-institutional American College of Surgeons Oncology Group study (STS = soft tissue sarcoma, XRT = external-beam radiation therapy).

Proponents of preoperative radiation therapy cite the theoretical advantages of using the tumor’s bulk to displace uninvolved intraabdominal viscera out of the radiation field, thereby decreasing local toxicity and increasing the ability to administer therapeutic radiation doses.^{18,30,33} This approach also allows the target volume to be easily delineated for treatment planning, and treating the tumor prior to surgical manipulation theoretically decreases the likelihood of tumor implantation. Tumor regression in response to therapy may also facilitate resection.

Postoperative external-beam radiation at the doses that are most likely to be effective can be associated with acute and delayed bowel toxicity.^{17,34-36} After the large tumor mass that had been displacing adjacent viscera is removed, the bowel tends to fall into the resection bed where it may become fixed by postoperative adhesions. When postoperative radiation therapy is performed, these fixed loops of bowel are trapped within the radiation treatment volume. If postoperative radiation doses of 50 Gy are utilized, gastrointestinal toxicities are significant.

To assess the role of preoperative radiation therapy for retroperitoneal soft tissue sarcomas, the American College of Surgeons Oncology Group recently opened a multi-institutional prospective randomized trial (Z9031) comparing preoperative external-beam radiation therapy vs surgery alone (Fig 3). This trial will evaluate time to progression and overall survival as primary endpoints. It will also evaluate whether preoperative radiation results in a change in microscopically complete resection rates. Correlative investigations will focus on tumor biology, genetic profiling through gene chip microarrays, proteomics, and gene methylation studies.

Another approach that has been investigated is the use of intraoperative radiation therapy (IORT). This was formerly a complicated approach wherein anesthetized surgical patients were transported to treatment equipment in the radiation oncology department with open abdomens. IORT is now more practically performed, albeit at a limited number of centers with specially designed and shielded operating rooms equipped with built-in radiation devices. With IORT, the resection bed can be directly targeted with a high dose of radiation while nearby radiosensitive tissues

Table 2. — Selected Studies of External-Beam and Intraoperative Radiation Therapy for Retroperitoneal Soft Tissue Sarcomas

Study (year)*	Therapy	No. of Patients	Local Recurrence (%)	External-Beam Radiation Therapy	
				Preoperative	Postoperative
Prospective Randomized Trials:					
Sindelar et al ³⁸ (1993)	IORT	15	40	No	Yes
	No IORT	20	80	No	Yes
Pilot Studies:					
Pisters et al ³⁷ (2003)	IORT	35	Not reported	Yes	No
Bobin et al ⁶⁸ (2003)	IORT	22	50	Yes	Yes
Gieschen et al ⁶⁶ (2001)	IORT	20	17	Yes	No
	No IORT	17	51	Yes	No
Alektiar et al ⁶⁷ (2000)	IORT	32	38	No	Yes
Gunderson et al ⁶⁹ (1993)	IORT	19	15	No	Yes
Willet et al ⁷⁰ (1991)	IORT	12	25	Yes	No

* Studies frequently included primary and recurrent retroperitoneal soft tissue sarcomas. Some patients were also treated with chemotherapy. IORT = intraoperative radiation therapy

are mechanically retracted out of the treatment field. Pisters and colleagues³⁷ demonstrated the feasibility of this approach in patients who were given up to 50.4 Gy of external-beam radiation with concomitant continuous-infusion doxorubicin followed by 15 Gy of IORT. Table 2 presents a summary of selected studies investigating the use of IORT for retroperitoneal sarcomas. The weight of evidence — including one small randomized trial — suggests that IORT increases in-field tumor control but not recurrence-free or overall survival, as disease can recur just outside the treatment field.³⁸

The use of preoperative radiation therapy in combination with postoperative brachytherapy was evaluated by Jones et al³⁹ in a pilot study of primary and recurrent resectable retroperitoneal soft tissue sarcomas that were treated with preoperative external-beam radiation followed by surgery and then by postoperative brachytherapy. Of the 55 patients entered into the study, only 19 ultimately had both preoperative external-beam radiation and postoperative brachytherapy. The authors observed significant toxicity associated with the use of brachytherapy, particularly when used in the upper abdomen.

On the basis of the existing data, IORT and brachytherapy should be considered investigational treatments for retroperitoneal soft tissue sarcomas and should not be administered outside the context of a clinical trial.

Chemotherapy

The role of chemotherapy in the treatment of retroperitoneal sarcomas remains controversial, and the quality of available evidence is variable. The majority of published reports focus on tumors arising in the extremities. In studies that included soft tissue sarcomas at all sites, retroperitoneal sarcomas comprised a small portion of the total number of treated tumors.

The Sarcoma Meta-analysis Collaboration evaluated 14 adjuvant chemotherapy trials that included 1,568 patients

with localized resectable soft tissue sarcomas at all sites.⁴⁰ This analysis revealed a modest improvement (10%) in the recurrence-free survival rate with the addition of adjuvant chemotherapy to surgery. As a result, treatment regimens have been changed to use higher-dosed regimens that have been observed to have higher response rates.⁴¹⁻⁴⁸ However, these studies did not focus on retroperitoneal sarcomas, and it is unknown if we can extrapolate results from studies of soft tissue sarcoma for the extremities or all sites.

Since the use of adjuvant chemotherapy for retroperitoneal sarcomas remains investigational, our preference is to provide adjuvant chemotherapy treatment of soft tissue sarcomas under strict protocol-based regimens or in the context of prospective clinical trials. Preoperative or neoadjuvant chemotherapy may have practical advantages over postoperative chemotherapy, not the least of which is the ability to monitor response (or lack thereof) and alter or terminate therapy in patients who do not appear to be deriving any benefit. However, randomized trials have not directly compared preoperative chemotherapy vs postoperative chemotherapy for retroperitoneal sarcomas.

Local Recurrence

In the absence of metastatic disease, surgery is the choice for locally recurrent retroperitoneal sarcomas. Many studies have shown that a significant number of patients experience prolonged disease-free survival when all grossly evident recurrent disease can be resected. The addition of chemotherapy or radiation in the treatment of locally recurrent disease remains the subject of debate. In the setting of recurrent disease, adjuvant treatment following resection should be considered for all patients who have not previously received chemotherapy. Following resection of one recurrence, subsequent recurrences have progressively diminishing chances of being successfully

resected (Fig 4).⁴ Evidence showing the benefit of third and subsequent resections of retroperitoneal sarcomas is scant and is largely limited to studies of patients with low-grade liposarcomas.³¹ Such aggressive attempts at disease control should be relegated to centers with extensive expertise in the management of retroperitoneal tumors.

Metastatic Disease

Compared with patients with extremity soft tissue sarcomas, those with retroperitoneal sarcomas have a greater tendency for local recurrence and disseminated disease throughout the abdomen. Potter et al⁴⁹ reported that 80% of recurrences occur within 5 years. The treatment of metastatic pulmonary disease in patients with retroperitoneal sarcomas is extrapolated from experience with the treatment of extremity sarcomas and studies that include sarcomas at all sites.

Surgery for Pulmonary Metastases

Either plain film radiography or CT of the thorax can be used to detect pulmonary metastatic disease. Pulmonary metastases from soft tissue sarcomas of all sites are associated with median survival durations of 6 to 12 months.^{50,51} Several reports have shown that resection of even multiple pulmonary metastases (metastasectomies) is associated with prolonged relapse-free survival in a small but significant percentage of patients (probably at least 25%).^{52,55} Several prognostic features associated with long-term survival in patients undergoing pulmonary metastasectomy

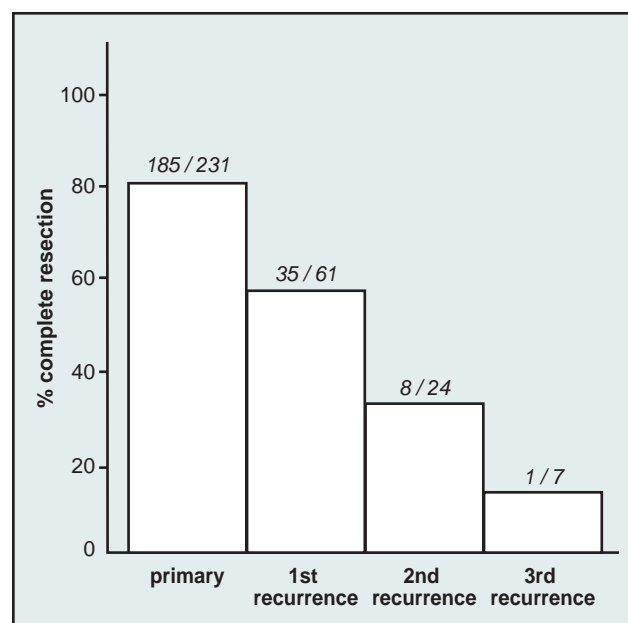


Fig 4. — Rates of complete resection for primary disease and after the first, second, and third recurrences. From Lewis JJ, Leung D, Woodruff JM, et al. Retroperitoneal soft-tissue sarcoma: analysis of 500 patients treated and followed at a single institution. *Ann Surg.* 1998;228:355-365. Modified with permission from Lippincott Williams & Wilkins. <http://lww.com>

have been identified. In an evaluation of soft tissue sarcoma pulmonary metastases, Billingsley et al⁵⁴ observed a more favorable prognosis for patients who had complete resection of all metastases, a disease-free interval of more than 12 months, or a low-grade primary tumor. A relationship between outcome and time interval from initial diagnosis to the development of pulmonary metastases has been observed in multiple studies, although this may be largely or entirely a surrogate for histologic grade of the primary tumor.^{51,53,56} Greater numbers of metastatic nodules and rapid tumor doubling times have been associated with diminished survival following resection of soft tissue sarcoma pulmonary metastases. Reported survival rates following complete resection of pulmonary metastases (sometimes with repeated thoracotomies) range from 25% to 39% at 5 years.^{52,53,55} On the basis of published reports to date, an aggressive approach to resection of pulmonary metastases is warranted.

Surgery for Hepatic Metastases

Resection of soft tissue sarcoma hepatic metastases has also been evaluated. Survival rates following hepatic resection have generally been less than those observed for resection of pulmonary metastatic disease. One series of patients with soft tissue sarcoma who underwent resection of hepatic metastases reported a 100% recurrence rate.⁵⁷ Despite this, the median survival duration was 30 months for patients who had hepatic resection compared with 11 months for those who did not. The inclusion of many patients with what are now recognized to be gastrointestinal stromal tumors in most series describing sarcoma metastases to the liver makes interpretation of these series more difficult.

Chemotherapy for Unresectable Metastatic Disease

The role of chemotherapy in the treatment of unresectable metastatic soft tissue sarcoma has been extensively reviewed,^{41,58} but the use of chemotherapy for retroperitoneal sarcomas is extrapolated from studies that included only extremity sarcomas or sarcomas at various sites. Doxorubicin, ifosfamide, and dacarbazine have all been shown to have significant single-agent activity in the treatment of metastatic soft tissue sarcomas. While published reports of various combinations of available drugs have suggested that combinations are superior to single agents, little evidence from prospective randomized trials is available to support this contention.⁵⁹ One randomized trial is representative: the addition of ifosfamide to doxorubicin increased response rates at the expense of markedly greater toxicity but did not result in any detectable difference in time to progression or overall survival.⁶⁰

There is no consensus regarding the ideal second-line chemotherapy regimen for patients with metastatic disease refractory to combination therapy with doxorubicin and ifosfamide. Higher doses of ifosfamide (in the range of

12 to 14 g/m²) have been associated with objective responses in patients who had failure or progression after chemotherapy with lower doses of ifosfamide, with synovial sarcomas appearing to be particularly responsive to this approach.^{61,62} The toxicity of ifosfamide in this dose range mandates careful patient selection, and it excludes many older patients and those with impaired renal function. Recently, the combination of gemcitabine and docetaxel in specific sequence has been associated with high objective response rates, even in patients with prior doxorubicin and ifosfamide chemotherapy. This regimen appears to be particularly effective for patients with leiomyosarcomas, but responses in other histologic subtypes have also been reported.^{63,64}

Clinical trials remain a highly appropriate option for patients with metastatic soft tissue sarcomas of all histologic subtypes. Novel approaches to clinical trial design are also worthy of exploration, given the multiple potential interactions of drug type, dose, and schedule with histologic subtype and prior treatment.⁶⁵

Surveillance Guidelines

Several considerations arise when deciding the appropriate surveillance plan for a patient following treatment for soft tissue sarcoma. The effects of early detection of recurrences on therapy and outcome vary by anatomic location of the recurrent disease. For retroperitoneal sarcomas, failure occurs primarily within the abdomen and in the liver, and an additional 20% to 30% of recurrences involve the lungs. Thus, it seems appropriate that a surveillance strategy would include physical examination, CT of the abdomen, and chest radiography. Early detection of local recurrence, hepatic metastases, and pulmonary metastases will occasionally result in surgical intervention; it is assumed but not proven that such interventions may prolong survival and improve quality of life.^{4,16,54,55,57} The incidence of recurrence for intra-abdominal and retroperitoneal sarcomas is also highest in the early post-treatment period, and a schedule of evaluation similar to that used for extremity sarcomas would seem reasonable. These sarcoma surveillance strategies remain to be proven in prospective trials but are widely used and almost universally recommended, with some controversy remaining as to the incremental value of chest CT over radiography. The current guidelines from the National Comprehensive Cancer Network for the surveillance of soft tissue sarcomas arising in the retroperitoneum recommend that patients with low-grade disease undergo physical examination with chest/abdomen/pelvis CT every 3 to 6 months for 2 to 3 years, then annually. For those with high-grade disease, patients should undergo physical examination with chest/abdomen/pelvis CT every 3 to 4 months for 3 years, then every 6 months for 2 years, then annually.⁷¹

Conclusions

The evaluation and treatment of retroperitoneal soft tissue sarcomas remain challenging. The most important factor in the long-term success in the treatment of primary tumors is complete surgical resection. It is important that patients with these tumors be evaluated and treated at centers with multidisciplinary treatment planning and expertise in treating these rare tumors. These centers also can invest time and resources into investigations of novel therapies and have access to clinical trials. Local recurrence remains a difficult problem, with increased associated morbidity and psychological stress for affected patients. We hope that with improved education, early referral of patients with retroperitoneal soft tissue tumors will become the norm, and patients will derive the benefits of multidisciplinary evaluation and treatment of their disease.

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