8

Management of Truncal Sarcoma

Paul Sugarbaker

OVERVIEW

Sarcomas of the head, neck, trunk and breast are biologically similar to, and behave like, the soft-tissue tumors found in other anatomic areas. In the past, and now in the present, radical surgical resection with negative margins is the only reliable treatment for these sarcomas.[1,2] The opportunity to use chemotherapy, surgery, and radiation therapy in selected patients as a multimodality approach may improve the likelihood of local control and long-term disease-free survival. Added experience with radiologic evaluation of patients to accurately define the anatomic location of the tumor, more definitive pathology to assess the biological aggressiveness of the lesion, and more conservative wide excisions has allowed patients to retain function and cosmesis. In addition, the development of reconstructive surgical techniques has made it feasible to reconstruct large surgical defects.

GENERAL CONSIDERATIONS

Soft-tissue sarcomas are relatively rare in the head and neck. They account for 4–15% of all soft-tissue sarcomas and less than 10% of all neoplasms at this site.[1,2] However, it is necessary to consider sarcoma in the differential diagnosis in order to avoid serious errors in treatment. Histologic classification, tumor size and tumor grade are the major prognostic factors related to local control and survival.[3–4] In addition, distant disease, most commonly to the lungs, is greatly influenced by these major prognostic factors.

Truncal sarcomas (chest and abdominal wall sarcomas) are also uncommon, accounting for about 20% of the soft-tissue sarcomas.[5–7] The histologic subtype has less prognostic significance, for wide local resections with negative margins are more likely to be achieved. Special reconstruction techniques may often be required. Extensive chest wall and abdominal wall resections can be performed with low morbidity and mortality.[3,4] Specialized radiotherapy techniques may be required to treat inadequate margins of resection. These cancers have a high local recurrence rate and local disease control is the major goal of therapy.[6]

Metastatic malignancy must always be considered in dealing with soft-tissue masses of the trunk since metastatic disease occurs with about the same frequency as do primary tumors. With truncal sarcoma there is a predominance of soft-tissue tumors over the less frequent chest wall bone sarcomas.

Breast sarcomas account for only 1% of breast malignancies. Treatment requirements are a wide excision with generous negative margins of resection. Inability to achieve a negative margin of excision carries an extremely guarded prognosis.[8,9]

CLINICAL FEATURES AND DIAGNOSIS OF HEAD, NECK, TRUNK AND BREAST SARCOMAS

A painless mass is the initial symptom for a great majority of head and neck, trunk and breast sarcomas.[1] Other clinical signs and symptoms produced by sarcomas are related to invasion or pressure effect that the tumor exerts on the structures in these locations. The awareness of the physician that any soft-tissue mass may be a sarcoma is the most important factor in prompt diagnosis.[1] This must always be confirmed by an appropriate biopsy.

A history of prior irradiation or hereditary synromes (Von Recklinghausen's disease, Gardner's syndrome) must be considered (Table 8.1). When dealing with a sarcoma a thorough physical and neurological examination should be performed. Plain films may be very useful prior to biopsy.

Table 8.1 Conditions related to an increased sarcoma incidence

Carcinogenic agents
Phenoxyacidic acid
Chlorophenols
Polyvinyl chloride
Thorotrast
Arsenic
Radiation therapy
Hereditary syndromes
Li–Fraumeni syndrome
Retinoblastoma
von Reckinghausen's disease
Gardner's syndrome

In performing the biopsy the surgeon must avoid several pitfalls. The skin incision or "true cut needle" tract must not jeopardize subsequent skin flaps. Hematoma must be avoided for it may propel sarcoma cells with red blood cells into and along soft-tissue planes. With these constraints in mind the biopsy must be adequate for pathological study. Necrotic tissue, hematoma or inflammatory tissue may be submitted rather than tumor tissue. A frozen section to confirm that cancerous tissue has been obtained with the biopsy may be helpful if the tissue specimen seems inadequate.

If biopsy reveals a sarcoma, plain radiographs of the anatomic site are often helpful in defining the extent of disease. Computerized tomography (CT) and frequently magnetic resonance imaging (MRI) are available for finer delineation of the tumor and to assess the extent of its local involvement. Angiography is of value in selected cases. A computerized tomogram of the chest is necessary to look for metastatic disease.

Resectability should not be based exclusively on the radiologic findings. On many occasions the only way to assess the extent of the sarcoma is by a surgical exploration to determine resectability.

HEAD AND NECK SARCOMAS

The head and neck sarcomas include a wide variety of histologic types of malignancy. Generous biopsy and accurate anatomic localization by CT are necessary prior to surgical intervention, radiotherapy and/or chemotherapy.

Histopathologic Types of Head and Neck Sarcomas

Aggressive Fibrous Lesions

These are neoplasms that arise from cells of the fascia or fibroblasts. The aggressive fibrous lesions include aggressive fibromatosis, dermatofibrosarcoma protuberans, fibrous histocytoma and desmoid tumors. Although these locally malignant diseases present a broad spectrum of invasive capabilities, they are usually nonmetastasizing lesions. Fibrosarcoma, however, differentiated or undifferentiated, implies the potential for metastasis. Because many of these lesions are slowly progressive, and seldom if ever metastasize, some authors do not consider desmoid tumors and dermatofibrosarcoma protuberans as true sarcomas. Others consider aggressive fibromatosis as low-grade fibrosarcomas. Patients with low-grade fibrosarcomas or dermatofibrosarcoma protuberans have a longer survival than patients with higher-grade tumors, but may eventually succumb to cancer or require major amputation though metastatic potential is very low. Surgical resection is the treatment of choice. Adjuvant radiation therapy may play a role in order to obtain local control when positive margins of resection are left behind. Devastating problems may arise when these sarcomas progress along the brachial plexus into the cervical neural foramina.

Malignant Fibrous Histocytoma (MFH)

These are often aggressive lesions with a poor prognosis in spite of multimodal treatment. They can occur within the treatment field of previous radiotherapy for other disorders. Adjuvant treatment is of limited value but worthy of consideration in selected patients. These tumors – along with fibrosarcomas, aggressive fibromatosis, and dermatofibrosarcoma protuberans – account for about 50% of head and neck sarcomas.[1]

Adult Rhabdomyosarcoma

This sarcoma of skeletal muscle lineage is the most common malignant soft-tissue tumor of the head and neck in children, the orbit being a very frequent location. Rhabdomyosarcoma in adults is usually located in the pterygoid region. Prognosis is worse in adults as compared to children because the adult rhabdomyosarcoma is much less responsive to chemotherapy and to radiotherapy. Adult rhabdomyosarcoma tends to be locally invasive, spread along tissue planes, and metastasize hematogenously and lymphatically early in its course. Adult rhabdomyosarcoma is unique among soft-tissue sarcomas in that lymphatic metastases are present in up to 50% of cases. Radiation therapy and chemotherapy play a major role in treatment, and surgery is often used only for diagnosis purposes. Occasionally surgery may be used to resect persistent tumor after maximal chemotherapy or chemoradiation therapy has been employed. The margins on these operations employing "consolidation surgery" are usually minimal or absent. Surgical resection is the initial treatment option for rhabdomyosarcomas if negative margins of resection can be achieved.

Leiomyosarcoma

The sarcomas of smooth muscle lineage are relatively common in the head and neck. They arise from cells of blood vessels or from an undifferentiated mesenchymal cell. Surgical resection is the treatment option and the recurrence rate is as high as 75%.

Hemangiopericytoma

This is a rare neoplasm of capillaries and the pericytes around and between the blood vessels. Children have a better outcome, and an adverse prognosis increases with age. Local recurrence rate is about 40% but this tumor results in distant metastasis in only about 10% of patients. Clinical outcome is closely related to the site of origin because this will usually determine the adequacy of the margins of resection. Treatment is surgical resection if feasible.

Angiosarcoma

This is an uncommon tumor in the head and neck, with the scalp being the most frequent site. It is more commonly seen in older patients. Lymph node metastases are present in 25–50% of cases. A characteristic feature of this neoplasm is its clinically undetectable diffuse infiltration far beyond the visible limits of the primary lesion. Prognosis is poor and surgical resection along with radiation therapy and chemotherapy are usually the options for management.[1]

Neurofibrosarcoma

These tumors arise from Schwann cells; this histologic type occurs as a sarcomatous degeneration of a neurofibroma in patients with Von Recklinghausen's disease. Clinical signs and symptoms may occur within the distribution of the large nerve that is involved. Superficial lesions have a better prognosis than deeper ones. These tumors are frequently locally recurrent, and when hematogenous spread occurs are uniformly fatal. Wide surgical excision and radiation therapy for inadequate margins of resection are the treatment options of choice.

Liposarcoma

This is also a rare neoplasm in the head and neck. This sarcoma arises from lipoblast cells and not from pre-existing lipomas. Although the well-differentiated liposarcoma of the neck has a good prognosis, tumors arising in the oral cavity or the oropharynx have a poor outcome. The incidence of local recurrence is high unless complete resection is achieved. Surgical excision with negative margins of resection is by far the best approach if this can be achieved.

Other rare sarcomas in the head and neck region include synovial sarcoma, alveolar soft-part sarcoma, and chondrosarcomas (Table 8.2).

CLINICAL FEATURES AND DIAGNOSIS OF CHEST WALL AND TRUNCAL TUMORS

Malignant chest wall tumors comprise approximately 50% of all primary tumors at this location. Malignant tumors are more likely to produce overt signs and symptoms (painful enlarging mass) than are benign neoplasms.[3,7,8] Accurate and early diagnosis is the first step in successful management of any primary chest wall tumor, since types of treatment vary for each malignancy. These tumors are seen in males twice as frequently as in females. An accurate clinical history of prior radiotherapy or a hereditary syndrome, a physical examination along with a proper biopsy, remain the standard methods for diagnosis.

Histopathologic Types of Chest Wall and Truncal Sarcomas

Histologic types are similar to those more commonly seen in the extremity. Fibrosarcoma and malignant fibrous histocytoma are the most frequent soft-tissue sarcoma in this location and chondrosarcoma is the most common chest wall bone sarcoma.

Table 8.2 Histologic types of sarcomas of the head and neck

Fibrosarcoma	Synovial cell
Malignant schwannoma	Angiosarcoma
Dermatofibrosarcoma protuberans	Hemangiopericytoma
Aggressive fibromatosis	Rhabdomyosarcoma
Malignant fibrous histiocytoma	Leiomyosarcoma
Liposarcoma	Neurofibrosarcoma

Chondrosarcomas

Chondrosarcoma is the second most common tumor of bone after osteogenic sarcoma, and comprises about 20% of all primary bone tumors. Chondrosarcoma usually appears in middle-aged or older adults. The pelvic girdle (27%), chest wall (ribs and sternum (15–20%)), femur (23%), and humerus–scapula (16%) are the most frequent sites of origin. These sarcomas arising from cells of cartilage lineage are one of the most common primary malignant tumors of the chest wall. Costochondral junction and sternum are the frequent locations.

Chondrosarcomas may be of primary (65%) or secondary (35%) origin, depending on whether they arise *de novo* or from a pre-existent benign cartilage tumor. Repeat surgical resections for tumor recurrences usually precede malignant transformation. Chondrosarcomas may also be induced by previous irradiation (9%).

Clinical presentation is usually as a slow-growing painful mass, and presence of symptoms is generally of value to differentiate these tumors from their benign counterparts. Tumor also has a role to discriminate chondroma from chondrosarcoma of the chest wall.[10] Radiologic findings may show soft-tissue involvement and bone destruction. Multiple radiologic features have been described, but probably the presence of irregular calcifications throughout areas of radiolucency with trabeculation, and the presence of the Codman's triangle (long bones) are the most significant radiologic signs. CT and MRI are excellent tools for an accurate and complete anatomic staging. Angiography is useful if vascular involvement is in question.[10]

Guidelines for Management and Treatment Options for Chondrosarcomas

A special problem in management arises in caring for patients with chondrosarcomas. These lesions have a great heterogeneity in terms of grade of malignancy in different parts of the primary tumor mass. Frequently a biopsy from one portion of the sarcoma will show a well-differentiated lesion suggesting a good prognosis and no need for wide margins of excision. Another area may be widely undifferentiated; of course the prognosis and selection of treatment options is determined by the most aggressive histology that is seen.

Because of this prominent heterogeneity a unique management plan is recommended for both extremity and truncal chondrosarcoma. After a radiologic diagnosis is made the entire tumor is curetted from its normal bone and soft-tissue margins. Liquid nitrogen is used to treat the cavity and extend the margins. Bone chips are used to loosely fill the empty space. Free margins of excision that would be achieved by a

resection are not attempted. If a low-grade I or IIa tumor is diagnosed by a complete histopathologic searching of the specimen, no further treatment is required. If a grade IIb or grade III focus of chondrosarcoma is found then a definitive resection with generous negative margins of excision are required. The prognosis with grade III lesions, mesenchymal chondrosarcoma, and clear-cell chondrosarcoma is guarded.

Plasmocytoma

This is a relatively frequent neoplasm since 25–30% of all primary malignant tumors in the chest wall are of this type (solitary plasmocytoma). Signs and symptoms of a generalized disease such as fever, weakness and anemia may be present since myeloma is also a systemic disease. Increased serum proteins are a common finding. Surgery must be used only for diagnosis purposes. Systemic chemotherapy is the treatment option of choice. Radiation therapy is useful for symptomatic pain relief.

Osteogenic Sarcoma

Typically osteosarcoma of the chest wall is a rapidly growing malignancy of the pediatric patient. Osteosarcoma that occurs in the chest wall presents a poor prognosis (20% 5-year survival). Pulmonary metastases occur early in the course of the disease. Chest wall osteosarcoma usually presents as a painful expanding mass and the characteristic radiological appearance is "sunburst-like." Wide local en-bloc resection is the treatment of choice after aggressive systemic chemotherapy has caused maximal shrinkage of the primary tumor. The prognosis remains very poor but is undoubtedly improved when there is an objective response to chemotherapy.

Ewing's Sarcoma

This neoplasm is usually seen in the pediatric patient. Although it usually arises in long bones, the chest wall (ribs) can also be affected.[8] It is discussed with the pediatric sarcomas.

Desmoid Tumors

These are rare neoplasms in the chest wall and some authors consider them as benign tumors while others consider them as low-grade fibrosarcomas because of the aggressive local fibrosis they show. Women are more commonly affected. Recurrence rate is very high if complete resection is not achieved. Wide resection is the treatment option for primary and recurrent disease.

Giant-cell Tumors of Bone

Giant-cell tumor of bone is a non-metastasizing lesion that is locally aggressive. If inadequately treated it may recur and eventuate in a fully malignant sarcoma with definite metastatic potential. Giant-cell tumor of bone is histopathologically well characterized by multinucleated giant cells uniformly dispersed throughout a well-vascularized tissue made up of plump, spindle or ovoid cells. It represents about 5% of all primary bone tumors. These tumors are more common in the third and fourth decades of life and women are slightly more affected than men. Secondary aneurysmal bone cyst arising in a giant-cell tumor of bone has been confirmed.[11]

More than 75% of giant-cell tumors are located around the articular end of a long tubular bone. Giant-cell tumors of the sacrum are second in incidence to chordoma at this site. Vertebral giant-cell tumors are uncommon but may occur. Multiple giant-cell tumors of bone that appear at different sites in the same patients are rarely seen. Malignant transformation may occur after multiple or even solitary local recurrence of a previously benign giant-cell tumor. A malignant sarcomatous growth will occur at the site of a previously documented benign giant-cell tumor in approximately 5% of patients. These sarcomatous changes usually occur after radiation therapy of a previous benign lesion and are considered to be radiation-induced malignancies. Fibrosarcoma, malignant fibrous histiocytoma, and osteogenic sarcoma are the most common lesions.

A primary malignant giant-cell tumor of bone may occur in 2% of these lesions. It is defined as a newly diagnosed sarcoma in which a histologically identifiable benign giant-cell tumor component is present. The sarcomatous component is usually a malignant fibrous histiocytoma or fibrosarcoma.

Physical findings relate to the involved bone and are nonspecific. They include pain, swelling, tenderness, and limited motion. Tumors located in the spine and sacrum often present with neurologic disturbances.

Radiologic studies reveal lesions arising in the axial skeleton that appear as ill-defined lytic areas lacking specific radiographic features. Sacral lesions even of considerable size may not be recognized as giant-cell tumors because of lack of consistent features. The radiographic appearance of giant-cell tumors cannot differentiate between benign and malignant forms. The angiographic assessment of giant-cell tumors reveals marked tumor vascularity. Size, extent of the lesion, and soft-tissue involvement can be evaluated from an angiogram. Contrast-enhanced CT is a useful noninvasive diagnostic method. Giant-cell tumors involving the bony pelvis are well defined by CT scan, especially if intrarectal air and contrast are used. MRI correctly

defines the size and extent of soft-tissue extension but rarely adds information to the CT findings. Radionuclide bone scanning of giant-cell tumor of bone is of limited diagnostic value.

Treatment of tumors in the sacrum or vertebral bodies is difficult because the lesions are advanced at the time of diagnosis and are less accessible for total removal. Radiotherapy is often required for better management and local control. Total sacrectomy by piecemeal excision of tumor from around nerve roots and/or radiotherapy may provide a long-term cure of tumors at this location. Complete removal is the treatment option of choice when feasible for giant-cell tumors of the pelvis, sacrum, or spine. Curettage or local excision is often all that is possible surgically using radiotherapy for incomplete removal, recurrence or nonresectability. Vertebral giant-cell tumors are more common in the lumbar region. Extensive intralesional excision and arthrodesis with or without radiotherapy achieves the best results. Marginal en-bloc excision is advised only for well-defined lesions not involving the posterior arch or soft tissues. Autogenous bone grafting may be necessary to fill the surgical defects. Curettage and cryosurgery are alternative therapies in some cases.[11]

These tumors show a high local recurrence rate of approximately 30% within the first 2 years and about 50% in 5 years when the curettage approach is employed. About 20% of giant-cell tumors, even in the total absence of histologic malignancy, invade the cortex and directly extend into adjacent soft tissues.

Other malignant tumors which may be present in the chest wall include: fibrosarcoma, angiosarcoma, rhabdomyosarcoma, neurosarcoma, and liposarcoma (Table 8.3).

Histopathologic Types of Abdominal Wall Sarcomas

Two histologic groups are the most frequent tumors of the abdominal wall. Desmoid tumors (low-grade nonmetastasizing fibrosarcomas) and fully malignant soft-tissue sarcomas (rhabdomyosarcoma, fibrosarcoma, leiomyosarcoma, liposarcoma, synovial sarcoma, malignant fibrous histiocytoma, etc.) with malignant potential for metastasis.

Table 8.3 Histologic types of soft-tissue and bone sarcomas of the chest wall

Fibrosarcoma	Ewing's sarcoma
Malignant fibrous histiocytoma	Osteogenic sarcoma
Chondrosarcoma	Hemangiosarcoma
Rhabdomyosarcoma	Plasmocytoma
Lipsarcoma	Leiomyosarcoma
Neurofibromatosis	Desmoid tumors

Desmoid Tumors of the Abdominal Wall

Desmoid tumors often recur after inadequate resection. These are usually slow-growing tumors with no tendency for metastatic spread, although they can cause death from local effects. Prognosis is good and long-term survival is the normal outcome. Wide resection is the optional treatment.

Fully Malignant Soft-tissue Sarcomas of the Abdominal Wall

These rapidly growing tumors recur and metastasize with great frequency. Prognosis is poor, as well as long-term survival. Wide en-bloc resection including full-thickness abdominal wall when deeply located is the option of choice. Adjuvant chemotherapy is advisable to prevent distant disease but definitive studies in this regard are not available.

Histopathologic Types of Breast Sarcomas

Mammary sarcomas are malignant neoplasms of the breast which arise from mesenchymal tissues.[9] They comprise a wide range of tumors including lymphomas, cystosarcoma phyllodes, and the entire spectrum of connective tissue tumors. The most common histologic types are malignant fibrous histocytoma, fibrosarcoma, and liposarcoma. Angiosarcoma occurs disproportionately more often in the breast than at other sites in the body. The term stromal sarcoma describes the group of mammary sarcomas other than lymphoma, angiosarcoma, and malignant cystosarcoma. However, this term should be used for lesions that arise from the specialized stroma of the breast. Intermediate and high-grade sarcomas constitute the majority of breast sarcomas and are certainly capable of metastasizing systemically.[9]

PROGNOSIS OF HEAD AND NECK, TRUNK AND BREAST SARCOMAS

The most important prognostic factors affecting survival are discussed below. Estimates of survival are presented in Tables 8.4 and 8.5.

Histologic type

Differences in survival can be attributed to the variable local behavior and metastatic potential of the different histologic types. However, each histologic type of sarcoma presents a wide range of aggressiveness for

Table 8.4 Long-term survival of head and neck sarcomas

Study	*Year*	*5-year OS (%)*	*10-year OS (%)*
Figueiredo *et al.*	1987	38	32
Farr *et al.*	1981	32	–
Freedman *et al.*	1989	68	60
Greager *et al.*	1984	54	28
Weber *et al.*	1986	40	30

Table 8.5 Long-term survival of chest wall sarcomas

Study	*Year*	*5-year OS (%)*	*10-year OS (%)*
Gordon *et al.*	1990	66	56
Greager *et al.*	1987	51	34
Perry *et al.*	1989	65	59
Paerolero *et al.*	1985	57	49

local invasion and distant metastasis. For this reason each of these soft-part and bone tumors must be considered individually. Prognosis is assessed through the sarcoma grade for that particular histologic type.

Histologic Grade

Differentiation grade (low- vs. high-grade) also correlates with survival and determines to a large extent local control and distant metastasis. The pathologist considers necrosis as the most reliable histologic finding by which to determine grade. Other considerations are mitotic activity and nuclear atypia.

Tumor Size

Tumors equal or larger than 5 cm usually have a poor prognosis, in contrast, tumors smaller than 5 cm have a better outcome. Size will be of great significance for head and neck sarcomas whereas tumor size for chest wall sarcomas does not seem to be such a strong prognostic determinant.

Response to systemic chemotherapy may vary with tumor size

Margin Status

The adequacy of the surgical resection is an important prognostic factor. Selecting an adequate margin of resection for a lesion with a particular biological aggressiveness without unnecessary sacrifice of function is a most difficult but necessary task. Positive surgical margins regardless of adjuvant therapies constitute the single greatest factor leading to local recurrence. The facility of performing a satisfactory wide excision is obviously dependent on tumor location.[1] Margin status affects both disease-free survival and overall survival. Liquid nitrogen has been used to improve the margin of resection on low-grade bone tumors, with excellent results. Low-grade chondrosarcoma and giant-cell tumors are commonly treated in this manner. Radiation therapy has traditionally been the treatment option to reduce the likelihood of local recurrence with narrow margins of resection

Metastasis

Presence of metastasis has a significant adverse effect on survival. Metastasis is by far most common to the lungs except for visceral sarcoma where the first capillary bed draining the primary tumor is the liver.

Prognosis Regarding Breast Sarcoma

Adverse prognostic factors for breast sarcomas include high histologic grade, and high mitotic rate. Tumor size seems not so important to outcome. Breast sarcomas are usually detected earlier than tumors at others sites with a median size of 4 cm.

TREATMENT OPTIONS FOR HEAD AND NECK, TRUNK AND BREAST SARCOMAS

Wide local en-bloc resection with adequate margins is the treatment of choice. This usually results in local control. A high rate of local recurrence is to be expected if wide excision is not sought or cannot be achieved. Low-grade tumors do not usually require adjuvant therapies.

Lymphadenectomy is not routinely performed unless nodes are clinically involved. Nodal involvement and distant spread is most commonly seen in high-grade tumors.

Adjuvant treatment with both radiotherapy and chemotherapy are often recommended in selected patients in an attempt to achieve local control and prevent distant disease. Indications are microscopic or gross positive margins of resection and narrow margins with large high-grade tumors.

Treatment Options for Head and Neck Sarcomas

Cosmetic and functional results are important in head and neck sarcomas and microscopic disease can often

be controlled with radiotherapy, avoiding a radical resection. The role of adjuvant radiation therapy is to sterilize microscopic extensions beyond the resection margins. Radiotherapy improves local control and survival over results achievable with surgery alone.

Chemotherapy is not effective as a single-modality treatment but may help control micrometastatic disease in the lungs and augment the local control provided by radiation therapy. Unfortunately, even aggressive multimodality therapy may fail to control primary disease. Local control and distant metastasis remain a therapeutic challenge for improving survival in high-grade tumors.[1]

Treatment Options for Chest Wall Sarcomas

En-bloc surgical resection with negative margins by using strict surgical oncologic techniques continues to be the most fundamental treatment of most tumors of the chest wall.[8] This is usually effective for local control in low-grade tumors.

The surgical resection of major chest wall tumors requires special preoperative planning with consideration of the overall condition of the patient, the status of the overlying skin, the anticipated skeletal defect and the available options with respect to reconstruction and support.[8] With appropriate preoperative attention to skin and soft-tissue coverage plus skeletal support, resection and reconstruction of major chest wall tumors can be accomplished. Reconstructive techniques available to deal with the resulting defects are presented below.[5] Full-thickness resection of the chest wall is advised for all high-grade sarcomas. Previous biopsy sites must be included. An aggressive surgical approach also applies for recurrences since local relapse and pulmonary metastases remain the most common sites of treatment failure[4–6] (Figure 8.1).

Adjuvant radiation therapy and chemotherapy are selectively used for high-grade sarcomas. They are routinely employed for residual disease and positive or equivocal margins of resection in an attempt to gain local control and prevent distant disease.[4] Innovative multimodal techniques for induction chemotherapy and perioperative radiotherapy are needed to optimize modern sarcoma management (Figure 8.2). However, it must be emphasized that adjuvant chemotherapy and radiotherapy has been used in randomized and nonrandomized studies for patients with high-grade sarcomas, with disappointing results.[12–14] For truncal sarcomas chemotherapy seems to improve disease-free survival.[13] Radiotherapy is mainly used for local control, although benefits similar to those seen with extremity sarcoma have not been reported. More clinical research is required to improve survival, and to better define the precise indications for use of chemotherapy and radiotherapy for head and neck, truncal and breast sarcomas. Elective lymphadenectomy is not

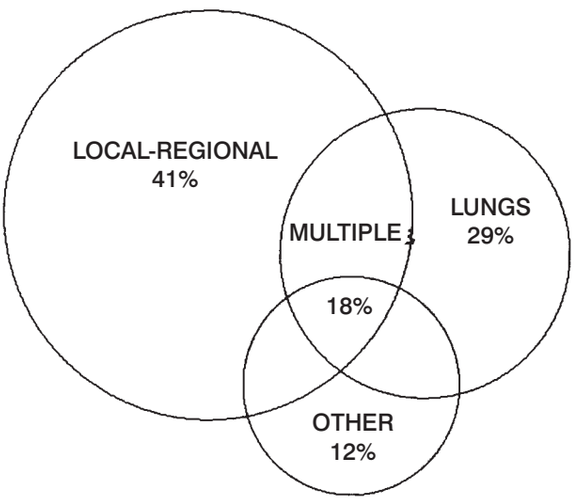

Figure 8.1 Truncal sarcomas. Patterns of recurrence.

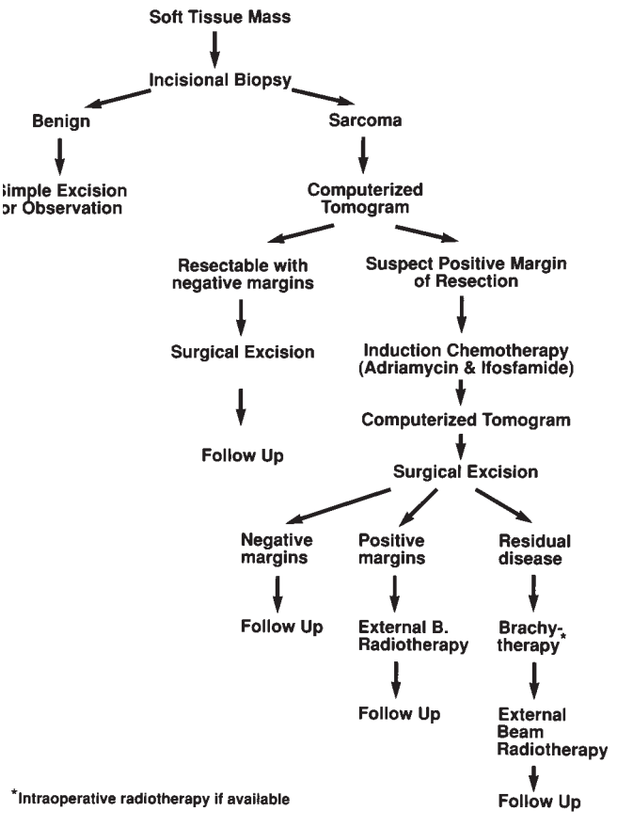

Figure 8.2 Clinical pathway for management of head and neck, trunk, and breast sarcomas.

routinely performed. However, it should be considered in certain high-grade types such as rhabdomyosarcomas, synovial cell sarcoma, and epithelioid sarcoma if these malignancies are immediately adjacent to the axillary nodes. Inguinal nodes are spared to avoid lower-extremity edema.

Treatment Options for Breast Sarcomas

Breast sarcoma is recognized as a separate entity from the more common breast carcinoma, and differences in the behavior of the two tumors must be kept in mind when planning treatment. The strategy used must be based on the knowledge of the biologic behavior of the tumor, including its pathways of spread. Contemporary multimodal approaches include excision of the soft-tissue sarcoma with adjuvant radiation and chemotherapy. However, excellent local control is possible with mastectomy alone if adequate margins are achieved.[9] The failure to establish local control is associated with poor prognosis. Axillary lymphadenectomy is indicated only when tumor is palpable within the nodes or when technically necessary to achieve local control of the primary tumor. It must be emphasized that surgery remains the most important treatment option for most mammary sarcomas. Some sarcomas require simple mastectomy for adequate treatment while others may need only wide local excision. If simple mastectomy does not provide sufficient clearance one must pursue radical mastectomy in order to obtain local control.[9]

Adjuvant therapy with estrogen antagonists, and other hormone manipulations, currently have no place in the treatment since these tumors do not appear to display hormone receptors.

General Guidelines for Wide Excision of Head, Neck, Trunk and Breast Sarcoma

The steps required to complete the surgical resection of a head and neck, truncal and breast sarcoma are as follows:

1. Obtain and display in the operating theater a CT of the lesion and of the lungs to help determine operability for cure and the anatomic site(s) that will be the least margin of dissection. If magnetic resonance images or angiograms were required, they must also be visible to the surgeon during the progress of the dissection.
2. Excise the skin approximately 1 cm around the biopsy site and dissect skin flaps back to the furthest extent of the palpable lesion.
3. Determine as early as in the operation as is possible whether the patient is resectable for cure. If the patient is found by surgical exploration to be inoperable for cure (gross residual disease) minimize the quality-of-life sacrifices and the cost that will result from the treatment.
4. Be prepared to use intraoperative radiotherapy or brachytherapy to definitively treat small areas of gross residual disease.
5. Avoid removal of lymph node groups (axillary or inguinal) surrounding the sarcoma mass unless they are encompassed by the dissection or are palpably involved by tumor.
6. Use electrosurgical dissection with complete muscle relaxation to divide muscle and fascia 1–2 cm beyond the palpable extent of the tumor. Repeatedly assess the adequacy of the margins during the dissection. Avoid resection of entire muscle bundles unless they are directly adjacent to tumor.
7. Tag with sutures divided muscles, tendons, or fascia that may be used in soft-tissue coverage, stabilization or reconstruction.
8. Dissect the least margin of resection by electroevaporation using ball-tip electrosurgery and a smoke evacuator. This will minimize the spill of sarcoma tumor emboli and gain a 1–2 mm margin as a result of heat necrosis.
9. If a spill of sarcoma tumor emboli into the operative field occurs irrigate copiously with a 1% peroxide. If the sarcoma dissemination occurred within the pleural or peritoneal cavity use intracavitary chemotherapy.[15]
10. Generously mark out the extent of the dissection with metal clips to facilitate radiotherapy should it be necessary.
11. Reconstruct and/or stabilize the operative site by myodesis, marlex or prolene mesh, myocutaneous flaps, autologous or allogenic fascia.
12. Meticulously close the subcutaneous tissue and skin over generous closed-suction drainage.
13. Obtain a CT of the operative site approximately 1 month after surgery to use as a baseline examination that documents postoperative changes.

TECHNICAL ASPECTS OF RECONSTRUCTION

The operative management of massive chest or abdominal wall malignancies presents an infrequent but formidable surgical challenge. Massive chest or abdominal wall resection for malignancy must be pursued aggressively whenever these lesions are encountered. Designing a surgical strategy which provides adequate margins and immediate reconstruction is critically important. The age of the patients and the locations or size of the lesions are not significant factors. These operations can be performed safely, and the benefits to patients justify this extensive surgery.

The use of muscle pedicle flaps has greatly enhanced the surgeon's ability to deal with large chest or abdominal wall defects. Pectoralis major or latissimus dorsi muscles, for instance, are equally effective in providing full-thickness coverage.[16,17] The surgical axiom that stabilization of the chest is mandatory before closure may not be infallible. A plastic mesh is appropriate in many instances. However, defects can often be closed depending on size and location, by moving in appropriate muscle flaps, without the use of synthetic material or natural tissues (fascia lata). Most patients will not show evidence of pulmonary compromise and prosthetic material will be obviated.

Prosthetic Materials

When a patient requires full-thickness chest or abdominal wall resection for malignant tumors, careful preoperative planning is crucial, not only to ensure a negative margin, but also in the production of a physiologically and cosmetically acceptable reconstruction at the conclusion of the procedure.[8] Chest and abdominal wall immediate reconstruction sometimes requires the use of synthetic prosthetic devices to cover large defects in order to preserve physiologic functions and prevent paradox. Where no musculofascial or musculocutaneous flaps are available, or when considered inappropriate or unnecessary, a different sort of autogenous tissue (omentum, fascia lata), meshes, or other inert material (methyl methacrylate) can be used as long as full-thickness skin cover is assured. Crucial to the use of any synthetic material is that it must be placed under tension when sewn into place.[8] Infection, prior radiation therapy, need of postoperative radiotherapy or chemotherapy, and foreign-body intolerance must be taken into account when considering these reconstruction techniques. Marlex mesh, Prolene mesh, Gore-Tex sheets, and methyl methacrylate are examples of prosthetic materials available for use in individualized situations.

Myocutaneous Flaps Commonly Used for Head and Neck, Chest or Abdominal Wall Reconstruction after Wide Resections for Tumors

1. *Sternocleidomastoid flap.* This is an excellent flap for the middle aspect of the face or the lower face. It is frequently used for intraoral and pharyngeal lining as well as a potential bone graft (along with clavicle) for mandibular reconstruction. It has also been employed to reconstruct the orbit.
2. *Pectoralis major flap.* This is probably the most commonly used flap in head and neck reconstruction and is usually employed for external resurfacing of skin at these sites, coverage of orbital defects, intraoral and pharyngeal lining, and as a carrier for rib and skin in mandibular repair. Reconstruction of the esophagus has also been done. In addition, transposed pectoralis major muscle has been employed for closure of chest wall defects. Single or bilateral pectoralis major muscle flaps with skin grafts are particularly useful for coverage of large anterior chest wall defects (Figure 8.3).
3. *Latissimus dorsi flap.* This versatile flap can be used in head and neck reconstruction for defects of the posterior neck and shoulder as well as intraoral and pharyngeal repair. The latissimus dorsi flap is particularly suited for use in closure of large midline trunk defects (front and back) and especially for breast reconstruction after mastectomy. Latissimus dorsi will also reach and cover the upper abdomen just below the costal margin, and lateral abdominal wall. However, this flap is of limited use for abdominal wall reconstruction (Figure 8.4).
4. *Trapezius flap.* This is a less widely used flap than pectoralis major flap for head and neck reconstruction; it is commonly used as a secondary option in patients in whom pectoralis major muscle flaps have failed to provide adequate coverage. The trapezius muscle flap has also been used to close upper anterior or posterior chest wall defects.

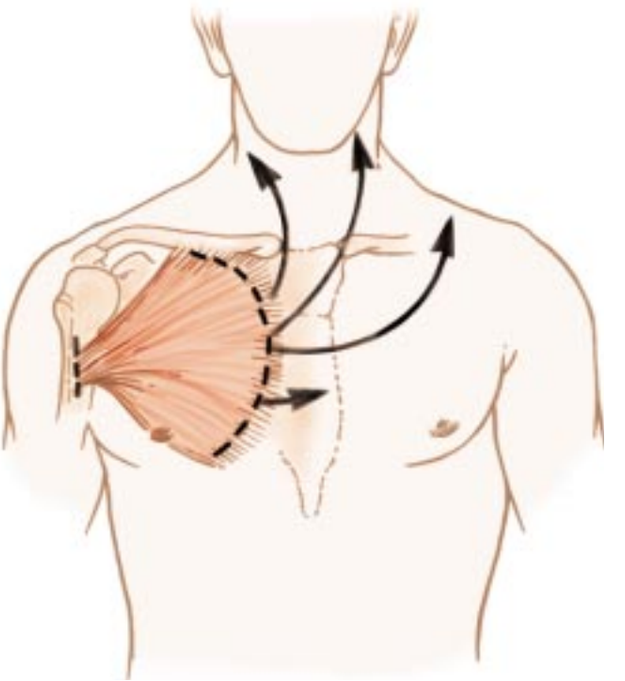

Figure 8.3 Pectoralis major musculocutaneous flap.

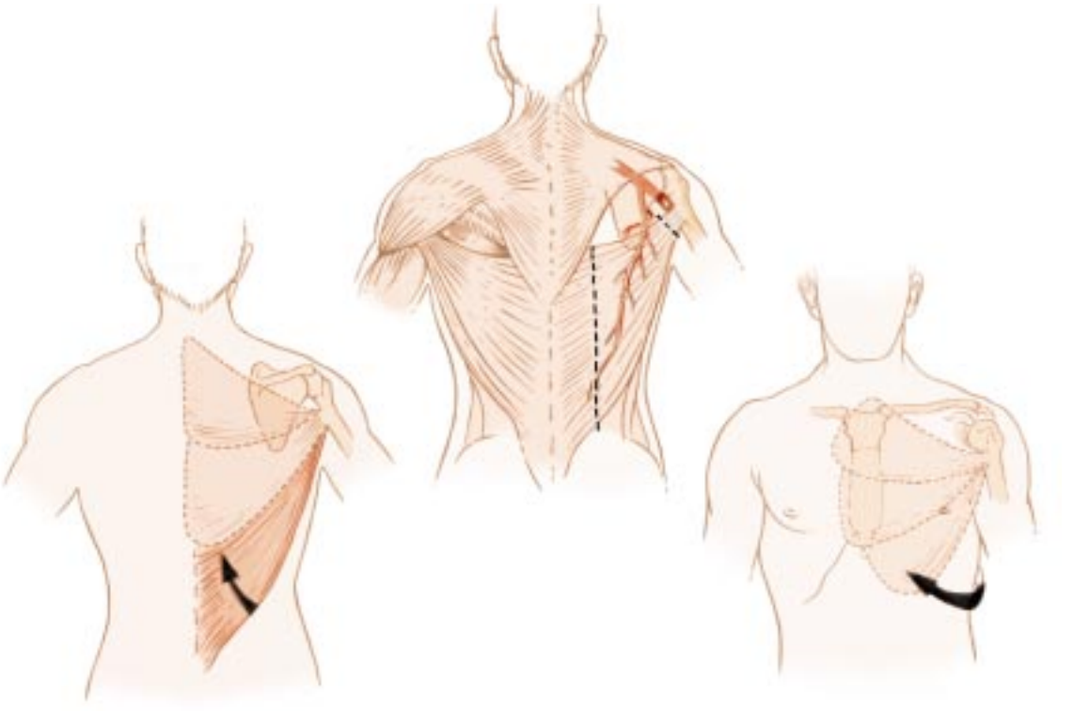

Figure 8.4 Latissimus dorsi musculocutaneous flap.

5. *External oblique flap.* The external oblique muscle flap is mainly used to cover small upper or lower full-thickness abdominal wall defects, although it is not so widely employed as the pectoralis major or latissimus dorsi flaps in chest wall reconstruction or even abdominal wall reconstruction because of its limited use. However, it can certainly provide full-thickness tissue for abdominal wall integrity and soft-tissue coverage and is also useful for correction of midline hernia defects.
6. *Tensor fascia lata flap.* Tensor fascia lata flap is one of the most useful flaps for reconstruction of abdominal wall full-thickness defects. It can be used as a musculofascial or extended musculocutaneous flap. In general, the use of tensor fascia lata flap is limited to the lower half or two-thirds of the abdominal wall. On the other hand, tensor fascia lata myocutaneous free flap is the best means of providing chest wall stability and coverage of large, potentially contaminated chest wall defects (Figure 8.5).
7. *Rectus femoris flap.* This is an excellent and reliable flap that will provide soft-tissue coverage for the abdominal wall. It can be elevated with the fascia lata to reconstruct muscular and fascial defects. It is mainly a second-choice flap if a tensor fascia lata flap is not available. The rectus femoris flap is a more muscular flap and some functional loss in the lower extremity may result after its use.
8. *Rectus abdominis and transverse rectus abdominis muscle flap.* The rectus abdominis flap is often used when considering abdominal wall reconstruction. This unit may provide skin and soft-tissue coverage but does not contribute to maintenance of abdominal wall integrity because of muscle denervation. The use of the superior half of the rectus abdominis muscle (based on the inferior epigastric artery) does not excessively weaken the abdominal wall, because of the strength of the anterior and posterior rectus sheath (Figure 8.6, left). In contrast, the use of the inferior portion of the rectus muscle (based on the superior epigastric artery) may lead to weakening of the lower abdominal wall because of the weak posterior rectus sheath (Figure 8.6, right). The transverse rectus abdominis muscle flap is an excellent flap for full-thickness chest wall defects and allows for adequate fascial reconstruction of the abdominal wall.
9. *Inferior gluteal thigh flap.* Large tumors deep in the pelvis may require extensive sacrifice of perineal skin and soft tissue. After an abdominoperineal resection, recurrence in the perineal incision may require wide

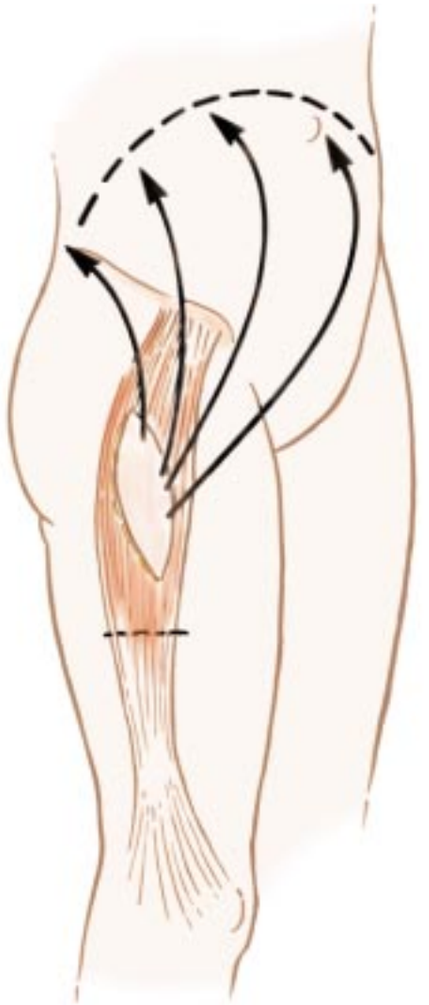

Figure 8.5 Tensor fascia lata musculocutaneous flap.

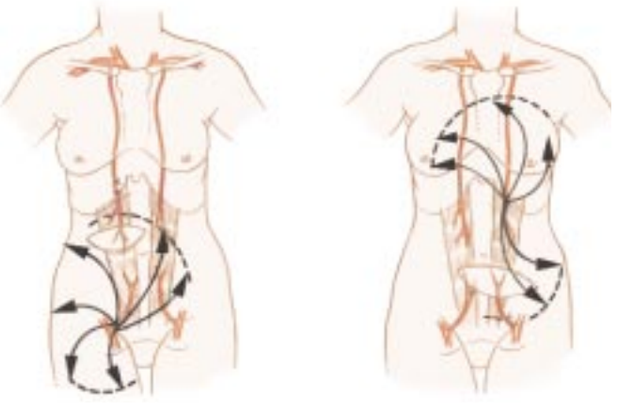

Figure 8.6 Superior (left) and inferior (right) rectus abdominis muscle flap.

re-excision. If radiation therapy was used the tissues will be immobile and failure of perineal wound healing will be common. Several techniques have been developed to transplant fresh tissue into the perineal defect to allow pelvic integrity and function to be restored. The inferior gluteal thigh flap provides a generous volume of tissue to be transferred into the perineal defect (Figure 8.7).

FOLLOW-UP

Patients undergoing surgical resections for head and neck, chest wall or abdominal wall sarcomas should be followed up at regular intervals during the first 5 years. All patients should be seen every 3 months for the first 2 years after surgery and at 6-month intervals up to 5 years. Complete physical examinations, blood chemical evaluations and chest X-ray films should be obtained at each visit. In addition, chest and abdominal CT must be performed every 6 months for the first 3 years. Bone scans can also be obtained in selected patients on a yearly basis.

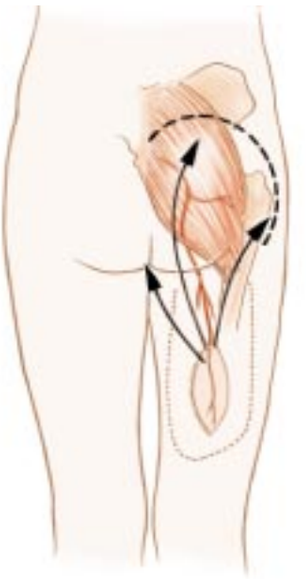

Figure 8.7 Inferior gluteal thigh flap.

References

1. Greager JA, Patel MK, Briele HA, Walker MJ, Das Gupta TK. Soft tissue sarcomas of the adult head and neck. Cancer. 1985;56:820–4.
2. Lawrence W, Donegan WL, Natarajan N, Mettlin C, Beart R, Winchester D. Adult soft tissue sarcomas: a pattern of care survey of the American College of Surgeons. Ann Surg. 1987;??:349–59.
3. Gordon MS, Hadju SI, Bains MS, Burt ME. Soft tissue sarcomas of the chest wall. Results of surgical resection. J Thorac Cardiovasc Surg. 1990;101:843–54.
4. Perry RR, Venzon D, Roth JA, Pass HI. Survival after surgical resection for high-grade chest wall sarcomas. Ann Thorac Surg. 1990;49:363–9.
5. Ramming KP, Carmak Holmes E, Zarem HA, Lesavoy MA, Morton DL. Surgical management and reconstruction of extensive chest wall malignancies. Am J Surg. 1982; 144:146–51.
6. Potter DA, Glenn J, Kinsella T *et al*. Patterns of recurrence in patients with high-grade soft tissue sarcomas. J Clin Oncol. 1985;3:353–66.
7. Weisenburger TH. Overview of malignancies of the chest wall and pleura. In: Roth JA, Ruckdeschel JC, Weisenburger TH, editors. Thoracic Oncology. Philadelphia: WB Saunders, 1989:543–5.
8. Pass HI. Primary and metastatic chest wall tumors. In: Roth JA, Ruckdeschel JC, Weisenburger TH, editors. Thoracic Oncology. Philadelphia: WB Saunders; 1989; 546–65.
9. Pollard SG, Marks PV, Temple LN, Thompson HH. Breast sarcoma: a clinicophathologic review of 25 cases. Cancer. 1990;66:941–4.
10. Huvos AG. Chondrosarcoma. In: Bone Tumors. Diagnosis, Treatment and Prognosis, 2nd edn. Philadelphia: WB Saunders; 1991;343–93.
11. Huvos AG. Giant-cell tumor of bone. In: ???????, editor. Bone Tumors. Diagnosis, Treatment and Prognosis, 2nd edn. Philadelphia: WB Saunders; 1991;429–67.
12. Blackledge G, van Oosteromn A, Mouridsen HT *et al*. The place of chemotherapy in the management of soft tissue sarcoma: experiences of the EORTC soft tissue and bone sarcoma group. Clin Oncol. 1989;1:106–9.
13. Lawrence W, Neifeld JP: Soft tissue sarcomas. Curr Prob Surg. 1989;26:759–827.
14. Glenn J, Kinsella T, Glatstein E *et al.* A randomized, prospective trial of adjuvant chemotherapy in adults with soft tissue sarcomas of the head and neck, breast and trunk. Cancer. 1985;55:1206–14.
15. Sugarbaker PH. Intraperitoneal Chemotherapy and Cytoreductive Surgery. Manual for Physicians and Nurses, 3rd edn. Grand Rapids, MI: Ludann Publishers, 1998.
16. Cohen M. Musculocutaneous flap closure of chest wall defects. In: Nyhus LLM, Baker RJ, editors. Mastery of Surgery, 2nd edn. Boston: Little, Brown; 1992;339–41.
17. Pairolero PC, Arnold PG. Thoracic wall defects: surgical management of 205 consecutive patients. Mayo Clin Proc. 1986;61:557–63.