Review Article

Surgical management of truncal soft tissue sarcoma and other selected soft tissue neoplasms

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Abstract: Soft tissue sarcoma (STS) is a heterogeneous entity comprising only 1% of all adult cancers that has received considerable attention since it was initially described after the 1st century as "fleshy" by Claudius Galenus. Nick-named the forgotten cancer, more than 100 histologic subtypes have been identified making treatment paradigms extremely complex. A key principle in the management of truncal STS is a defined multi-disciplinary team consisting of several providers. In most instances, surgery is the cornerstone of treatment. This overview will focus on the management of truncal sarcoma from a surgical perspective that will entail several points of consideration including histologic subtype, degree of differentiation, margin status as well as necessity of reconstruction; it will also include discussion of other unique soft tissue neoplasms relevant to the breast and abdominal wall.

Keywords: Soft tissue sarcoma (STS); surgical management; breast angiosarcoma; abdominal wall desmoid

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Introduction

Soft tissue sarcoma (STS) is a heterogeneous entity comprising only 1% of all adult cancers (1) that has received considerable attention since it was initially described after the 1st century as "fleshy" by Claudius Galenus (2). Nick-named the forgotten cancer, more than 100 histologic subtypes have been identified making treatment paradigms extremely complex (3). John Hunter, however, in the 17th century was the first to declare the sine qua non of surgical management—wide resection (4). Although most commonly encompassing the extremities, STS can originate from the head/neck, trunk, retroperitoneum, or visceral organs such as the gastrointestinal, gynecological, and genitourinary tracts (5). Koniaris and Sola have proposed combining both truncal and retroperitoneal STS together, since outcome may be similar (6). However, others such as Nathan (7) and Perez (8) have disputed this notion, underscoring that these are two distinct anatomic sites with unique challenges. Truncal STS is usually detected earlier than their retroperitoneal counterpart given the more superficial location but is often subjected to reconstructive circumstances. This overview will focus on the management of truncal sarcoma from a surgical perspective that will entail several points of consideration including histologic subtype, degree of differentiation, margin status as well as necessity of reconstruction; it will also encompass discussion of other unique soft tissue neoplasms relevant to the breast and abdominal wall.

Presentation

Bordered by the clavicle superiorly and the groin inferiorly, the trunk entails the chest, abdomen, and back. STS in this region comprises upwards of 20% amongst all cases (9). Upon evaluating a patient with a putative STS, several elements need to be queried such as duration of tumor, rapidity of growth, presence of pain, family history of genetic disorder and personal history of radiation. On examination, several factors should be discerned such as degree of mobility, proximity to bone, size of mass, prior biopsy site, and quantity of lesions (10).

Diagnosis

Although attaining cytological analysis via fine needle aspiration is occasionally used as the initial modality of diagnosis in STS, it is not recommended (11,12). As much as 10% of patients labeled with a STS has been misdiagnosed; in approximately 25% of cases, the histologic subtype has been misclassified. Image-guided core biopsy when available is the gold-standard and paramount in devising a therapeutic plan; it mitigates the risk of injury to the neural or vascular structures. A blind biopsy may not adequately sample the index area as the tumor could be heterogeneous; the goal would be to ascertain the site that is most likely to harbor a high-grade component (13). Incisional biopsy should be conducted if the core biopsy is equivocal (14). The extent should be minimal to deter dissemination. The area of the biopsy should be noted and the tract ultimately excised, especially if the STS is elucidated to be high-grade (15). This will reduce the chance of recurrence (16).

Histology

STS of the trunk most commonly metastasizes to the lung; however, histologic subtype may influence the pattern of spread. The most common include undifferentiated pleomorphic sarcoma, liposarcoma, and myxofibrosarcoma (17). Others such as epithelioid sarcoma, rhabdomyosarcoma, angiosarcoma, synovial sarcoma, and clear cell sarcoma invoke a higher rate of nodal propensity; the myxoid/round cell liposarcoma variant has a peculiar predilection for the retroperitoneum, subcutaneous tissue, and other fat bearing areas including the marrow within the spine (18).

Imaging

Imaging is essential in the evaluation of truncal sarcoma patients. Although ultrasound can help distinguish a solid mass from a cystic lesion, MR is recommended to provide the anatomic information regarding relationship of the primary tumor to surrounding structures such as bone, muscle, or nerve; it can also appropriately characterize the lesion to guide the most optimal site of tissue analysis (19). CT of the chest (in most instances) and/or of the abdomen/pelvis (in selected situations) are necessary in determining presence or absence of metastatic disease. A more recent trend is the utilization of PET scan as an adjunct to predict grade (20) and as a surrogate marker for response to therapy (21). Although investigational, when the decision to operate has been made, preoperative lymphoscintigraphy/single-photon emission computed tomography scan on the day of surgery may be considered for the previously mentioned subtypes with a lymphatic propensity i.e., epithelioid sarcoma, rhabdomyosarcoma, angiosarcoma, synovial sarcoma, and clear cell sarcoma (22).

Staging

The American Joint Committee on Cancer (AJCC) 8th edition staging classification for STS has for the first time been segregated according to anatomic site (23). Lesions that arise from the superficial trunk are categorized with the extremity. Additionally, the size of the primary tumor has been modified with T1 <5 cm, T2 5–10 cm, T3 10–15 cm, and T4 >15 cm. The schema is as follows: stage IA T1 N0 M0 G1 T2 >5 to ≤10 cm; stage IB T2, 3, 4 N0 M0 G1 T3 >10 to ≤15 cm; stage II T1 N0 M0 G2, 3 T4 >15 cm; stage IIIA T2 G2, 3; stage IIIB: T3, 4 G2, 3; stage IV Any T, G N1 or M1 (24).

Prognosis

Prognosis of truncal STS from an anatomic standpoint, are typically felt to be intermediate between extremity (most favorable) *vs.* head/neck (least favorable). This conclusion was based on a landmark series published by investigators at Memorial Sloan-Kettering. A 12-year nomogram was devised after analyzing 2,163 patients that amongst other factors incorporated age, size, depth, grade, and histologic subtype (25). A large historical study found that the overall 15-year survival was 59.5% (26). Although prior models incorporating these traditional data have been effective, other tools that are demonstrating promise in predicting outcome in high-risk STS patients that have received chemotherapy include the Sarculator (27).

Treatment

A key principle in the management of truncal STS is a defined multi-disciplinary team consisting of several providers (28). This ensemble should include pathologists, radiologists, medical oncologists, radiation oncologists, surgical oncologists, and reconstructive surgeons (29). When feasible, a dedicated tumor board discussion inclusive of these specialists in concert with the relevant slides and images should occur on a routine basis. Verifying the

diagnosis and considering other factors including family history of genetic syndromes such as Li-Fraumeni or von Recklinghausen as well as personal history of radiation may affect options in management. If either of these germline disorders are suspected, comprehensive evaluation by a geneticist is crucial. The knowledge of prior radiation likely excludes it as a part of the therapeutic armamentarium but also portends a more aggressive biology (30). Surgery with *en bloc* resection of additional skin or soft tissue is the cornerstone treatment for STS of the trunk and the essence of a wide margin cannot be over emphasized (31). Although controversy exists as to what defines an adequate margin, several cardinal rules should be implemented (32). It is imperative that the tumor pseudocapsule not be disrupted and the neurovascular bundle not be sacrificed in the absence of direct invasion (33). Although there are many similarities in the surgical management of truncal STS with the extremities as exemplified by their combined NCCN guideline algorithm, factors that need to be contemplated include the neighboring structures e.g., underlying thoracic and peritoneal cavities as well as the extensive surface area. Due to the proximity, in some instances *en bloc* resection of underlying ribs for chest wall tumors and muscle for abdominal wall tumors is necessary for high grade lesions, especially when there is invasion of periosteum and fascia respectively, given their more aggressive biology (34). Moreover, it should be emphasized that some histologic subtypes tend to be extremely infiltrative and warrant a more radical resection irrespective of degree of differentiation; this is pertinent for both myxofibrosarcoma (35) and dermatofibrosarcoma (36). Subsequently, once the tumor has been resected, collaboration with a reconstructive surgeon is essential to provide adequate coverage of the defect. A temporary apparatus such as the Vacuum Assisted Closure device may be a useful conduit in cases when the margin status is tenuous and ill-defined; this is especially relevant in an anticipated complex reconstruction (37). Options for definitive closure including skin graft, tissue flap, and mesh implementation have been demonstrated to be beneficial (38). The skill of the plastic surgeon can influence operative candidacy of the truncal STS patient as an aggressive resection without appropriate reconstruction will impact outcome. This is especially true if adjuvant therapy will need to be administered. Studies by Hussain *et al.* (39) and Suresh *et al.* (40) have highlighted the importance of the plastic surgeon in the role of the STS patient. The investigators strongly demonstrated the improved clinical benefit observed when priority was placed on reconstruction. The ultimate guiding principle is to achieve a negative margin and preserve functional status. Furthermore, limiting morbidity is paramount. The occurrence of a postoperative complication in a large study analyzing 546 STS patients including those involving the trunk revealed an adverse survival (41). The data incorporating chemotherapy as well as radiotherapy in extremity STS perioperatively can generally be applied to the trunk but suffer from practice patterns that are highly variable. In fact, a National Comprehensive Cancer Network guideline review determined that radiation therapy for STS involving the trunk and extremity is underutilized (42). Although prior studies did not report consistent administration of combined modality administration, there is an evolving trend in the implementation of neoadjuvant chemoradiation for high grade STS of both the trunk and extremity (43).

Surveillance

Like other locations, truncal sarcomas can recur locally, regionally, or systemically. Factors that determine frequency of follow-up include stage classification, tumor grade and histologic subtype. At the minimum, patients should have a clinical encounter every 3 to 6 months for the first 2 years, and biannually thereafter for up to a 5-year time-frame.

Special circumstances

Soft tissue neoplasms that may be encountered on the trunk include breast angiosarcoma and abdominal wall desmoid.

Angiosarcoma of the breast can arise *de novo* or develop from iatrogenic causes including radiation. It originates from the endothelial lineage. Epidemiologically, those with primary breast angiosarcoma are younger, often diagnosed in their forties while those with secondary breast angiosarcoma are older, typically identified in their seventies (44). Other than disparities in age at presentation, they share very similar biologic behaviors and therapeutic principles. Body in 1987 was the first to underscore the association of radiation induced breast angiosarcoma (45). It is a rare occurrence, with an estimated frequency of <1 per 1,000 breast cancer patients. In the era of breast conservation therapy, this histology is becoming more commonplace (46).

Mechanistically, edema of the breast has been implicated as an inciting factor for angiogenesis with radiation likely contributing genomic alterations (46). A large-scale

analysis of breast cancer patients from the Surveillance, Epidemiology, and End Results (SEER) Program incorporating ancillary data determined that the risk of development was more than 25-fold in those who received radiation in comparison to those who did not (47). Unlike other radiation-associated sarcoma, the latency period is typically less than 10 years (47). The clinical manifestations include multifocal violaceous discoloration of the skin and frequently a concomitant mass (48). Depending on the presenting signs, diagnosis is either confirmed via punch biopsy of the skin or core biopsy of the tumor. MRI in lieu of mammography may be a useful tool in the management, although a high index of suspicion by the clinician is essential (49).

The mainstay of treatment is mastectomy with excision of all radiated field (50). Due to the subsequent defect after extirpation, a skin graft or tissue flap may be required (51). Adjuvant therapy with either external beam radiation (52) or chemotherapy is typically not incorporated due to lack of efficacy (53). However, postoperative proton beam therapy is being examined as a potential adjunct (54). Systemic options that have demonstrated some benefit in the locally advanced or metastatic settings include cytotoxic agents such as paclitaxel as well as multi-kinase agents such as pazopanib; some reports have also highlighted beta blockade with propranolol as part of a metronomic regimen (55-57). Despite promise for an expanding armamentarium, prognosis is poor and is plagued by a high local recurrence rate as well as a prohibitive risk of distant relapse; median overall survival is approximately 1–4 years (58,59). Early detection is paramount as a recent study conducted by investigators from England elucidated that tumor size >5 cm was associated with an adverse outcome (60).

Although angiosarcoma of the breast is unique, comprising <1% of all breast malignancies, it is an evolving entity that must not be overlooked given the large proportion of breast cancer patients who have already received radiation in the antecedent decades.

Despite some controversy, desmoid should be considered a malignant entity although it lacks the potential to metastasize. However, it can be locally infiltrative but at times behave in an unpredictable manner. It originates from the mesenchymal lineage and hence is also known as aggressive fibromatosis. Epidemiologically, desmoid is rare and seen in only 3% of all soft tissue tumors (61). Desmoid can be associated with Familial Adenomatous Polyposis i.e., FAP in up to 5% of cases but typically emerges in sporadic fashion. In non-germline tumors, alterations in beta-catenin has been demonstrated to be a driving event in the development of desmoid (62). Estrogen may play a role in the pathogenesis as the majority of patients is of the female gender. Other inciting events include trauma or surgery. Epidemiologically, desmoid most commonly afflicts patients between 25 to 35 years of age (63). Although subset locations include extra-abdominal (60%) and intraperitoneal cavity (15%) such as mesentery, the abdominal wall (25%) will be emphasized (64).

The clinical manifestations are quite similar to STS of the trunk and can present as a painless lump; furthermore, the finding of a mass in the vicinity of a cesarean section scar should raise the suspicion of desmoid (65). When feasible, tissue confirmation via image-guided core biopsy is essential. MRI should be obtained to determine the depth of the tumor (66). It can also be utilized to assess degree of cellularity; low collagen content has been linked with increased rapidity of growth (67).

Traditionally, the mainstay of treatment was resection which encompassed segment of fascia and musculature; closure of the defect was either primary or incorporation of mesh. Although excision with a negative margin was once the desired goal, this concept is now recognized as controversial since recurrence rates in many instances between a positive microscopic margin and negative margin have been equivalent (68). Prior studies reported a 10% risk of recurrence (69). Wilkinson detailed a 92% success rate in 50 patients with no recurrence after 5 years (70) However, a recent trend has been a more pragmatic approach. Expectant management was utilized in a large trial consisting of 106 patients with abdominal wall desmoids with more than 25% experiencing spontaneous regression by 3 years (71). Of the remaining patients, only approximately 15% required surgical intervention. However, the investigators found that tumor size >7 cm predicted a higher likelihood of intervention with pharmacotherapy or eventual operative intervention.

Other modalities that have historically been used included external beam radiation, especially for margin positive disease. While some studies have demonstrated benefit (72), it has not exhibited efficacy in other trials (73). Due to the lack of consistent benefit coupled with the added risk of radiation enteritis and secondary malignancy, radiation is not routinely recommended.

Other options of treatment include nonsteroidal therapy e.g., sulindac, antiestrogen therapy e.g., tamoxifen, chemotherapy e.g., doxorubicin, and targeted therapy e.g., imatinib (74,75). A recent report by a Brazilian group

detailed their nonoperative pathway of 5 abdominal wall desmoid patients, with 80% achieving a partial response (76). The authors endorse a schema for abdominal wall desmoid that has been proposed by a European Sarcoma Task Force. No intervention is advised unless there is symptomatic or progressive disease. Initial management should be hormonal, followed by chemotherapy, and then surgical intervention with negative margins. Increasing utilization of other targeted agents include sorafenib, which is a multi-kinase inhibitor against the VEGFR2/PDGFRB/RAF cascade (77) and nirogacestat which is a gamma secretase inhibitor against the Notch pathway (78). A phase III clinical trial implementing sorafenib has demonstrated increased progression free survival. The promise of nirogacestat has facilitated creation of a phase III trial; there is belief that mitigating Notch will reverse activation of B-catenin. It is hopeful that future studies exploring these agents will provide more insight into the management of this challenging neoplasm.

Conclusions

The management for STS of the trunk including other neoplasms such as breast angiosarcoma and abdominal wall desmoid accentuate the absolute requirement for a multi-disciplinary approach. Given the rarity and complexity, a definitive diagnosis by a qualified sarcoma pathologist is vital prior to commencing any intervention. In many instances, surgery should be considered the cornerstone of therapy with the ultimate goal of achieving tumor free margins while preserving functionality.

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Footnote

Conflicts of Interest: The author has no conflicts of interest to declare.

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