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Review Article

Soft Tissue Sarcoma: Clinical Perspectives & Unchanged Survival Over Time

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ABSTRACT

Soft tissue sarcoma is very rare type of malignancies. Over the last few years, there is a advance understanding in the pathology. Now, over the period the understanding of the clinical behavior has been bringing to the great extinct. For the treatment of the group of heterogenous tumor, there was the concern that there was no often recognized care for the patients in the India.

Certain tissue causes the cancer diseases in bone or muscle and this type is called sarcoma. There are the two types of sarcoma. That are soft tissue sarcoma & osteo-sarcoma. Osteo-sarcoma is always developed from bone and the soft tissue sarcoma is developed by the soft tissue like blood vessels, fibrous tissue, muscle, fat., nerve, or deep skin tissue. This is very rare or exceptional case of the cancer because it mainly effects the adjoining parts of the body such as fibrous tissue, blood vessels, muscles, fat or any of the supporting tissue of the body. In this type of sarcoma many other body parts get affected due to the attachment with the connective tissue. After the parts get affected due to the attachment they randomly start dividing themselves and hence, they create the lumps. The size of the lump increases due to the internal pressure. Which cause the disruption in neighbor organs of body. This type of the cancer is very rare. Among adult About 1% & among the children 16% is affected by the soft tissue sarcoma. Different types of genes also have the role in inducing the STS. Such as N-RAS; cyclic AMP dependent kinase number 2(CDKN2); Mouse double minute 2 homolog (MDM2); protein 53 or tumor protein 53 (p53); retinoblastoma protein 1(RB1). To identify the diseases symptoms, biopsy is done or CT scan or MRI is worked out in order to pursue the patients treatment and increase the survival rate of the subjects chemotherapy & radiations treatment are done.

INTRODUCTION:

STS (Soft Tissue Sarcoma) are the group of lesions which are more often subtle in presentation. And they have recognized as the malignant or the aggressive behavior. Carcinomatous tumors By standards are such as cancers of the bowel breast, lung, prostate, & kidney[1]. Each type of the cancer has the frequency of 160,000 per year in the United States. This soft tissue sarcoma is comparably less found in the country which is probably less than, 5000 cases annually approaching. There are 7 benign soft tissue tumor. They are lipomas, schwannomas, fibromas or myxomas. These are mostly confused with malignant tumor in some of the cases. Because of this sometimes-excessive treatment is given to the patient or some times less treatment is given. Soft tissue sarcoma have the 7% of the overall malignant tumor present all over the world. It mostly occurs in 15 to29 years old people[2]. The group of different histotype include the highly heterogeneous*(Table:1)*. And these were characterized by the aggressiveness & propensity to metastize in the local area. This RMS mostly occurs in the older age rather than in children & it was characterized by the high responsiveness of the radiotherapy & chemotherapy.

The treatment for this disease is dramatically changed as the new innovations and the medication is identifies for the subjects suffering from the STS. The past 30 years the findings are going on the STS. The change in the cure is about 40-70%.Adults having the RMS have the less favorable outcome than the in children. But after some time the prognosis have improved treatments. Malignancy have the different type of grades to the different tumor like foe adult type sarcoma. And these are localized to the extremities. In this type of tumor , there is only a surgery option as a treatment[3]. The roles of adjuvant therapies are not cleared till now. The prognosis is related to the local invasiveness, feasibility of surgical resection, & clearly, to histological grade, &,tumor size, the presence of metastases.

Soft tissue sarcoma is the tumor which is commonly found in the adolescents and young adults(*Table:2*). Probable as the positional halfway b/w the pediatric small round cell tumors like RMS. the most typical adult sarcomas regarding responsiveness to chemo-therapy. Therefore; the entities present in the tumor of heterogeneous group are not rare in adolescents & young adults. The treatment of these patient look like that they are complex & necessarily multidisciplinary[4-5]. These types of patient need adequate expertise.

It is an important task to emphasize that adolescents & young adults should receive good treatments within selected & experienced institutions that enroll subjects into clinical trials[6-8]. Cooperation b/w pediatric oncologists & adult oncologists are needed to define the good treatment options for adolescents & young adult subjects. age. Although, by the age doctor prognostic the factors of the soft tissue sarcoma, age of the patient is not the important task to know but the subjects histology & the tumor biology and there characteristics are important than the age. the part the certain histotype of soft tissue sarcoma would identify in the children, adolescents or adults. Cooperative studies are done to identify or investigate the new molecular related therapies and the new therapies which are introduced. There may be the chromosomal translocation found in the soft tissue sarcoma[9-12]. This is mostly found in the children over the adults. This type of tumor spread in the whole body not at the particular site and it appears to be the painless lump which slowly increases its size. Benign or malignant can be the type of diverse group of tumors. Malignant soft tissue tumors grow in an uncontrolled manner and can invade adjacent tissue and metastasis around the body. This can occur in any age with the different types symptoms &mainly affect the lower lymph. Benign tumor does not spread to the different parts but grow continuously at the particular site and block the particular organ function[13].

Table1: Soft tissue sarcoma affected site in the body.

Region	% Affected
Lower extreme	10%
Peritoneum membrane and wall	15%
Chest Wall	15%
Upper Extreme	45%
Head and Neck	15%

Table2 : Soft Tissue Sarcoma By Histologic Type

Historic Type	Total Subjects (%)
Malignant fibrous histiocytoma	4.4%
Clear cell sarcoma	1.0 %
Angiomatous/ vascular sarcoma	2.9%
Rhabdomyosarcoma	6.8%
Giant cell sarcoma	0.9%
Small cell sarcoma	0.9%
Kaposi sarcoma	35.4%
Liposarcoma	4.6%
Lelomyosarcoma, fibrosarcoma	6.4%
Malignant peripheral nerve sheath tumor	3.8%
Spindle cell sarcoma	1.0%
Epitheloid sarcoma	1.48%
Alveolar soft part sarcoma	1.8%
Dermatofibrosarcoma, including protuberans	14.8%
Ewing sarcoma/PNET	4.9%
Desmoplastic small round cell tumor	0.4%
Synovial cell sarcoma	6.3%
Chondrosarcoma(soft tissue)	0.5%

Table3: survival rate for different stages in soft tissue sarcoma

Sites & Stages	Survival Rate (observed for 6 years)
Regional site (near the lymph node)	58%
Distant site	26%
Localized and primary site	73%
Stage1	94%
Stage2	81%
Stage3	86%

Normal soft tissue has the relationship with the soft tissue tumor to make the lesion arise in relation to elastic tissue, smooth muscles, blood vessels, fascia, fat, fibrous capsule and nerv[14-17]. These tumors have they function on the normal tissue which are adjacent to symptomatology of the tumor(*Table:3*).

Malignant bone tumor does't occur frequently but it is easily diagnosed because it has the relation to bony structure and some limited no. of the cells of origin like marrow element, principal osseous and cartilaginous[18-20].

Table4: Major types of soft tissue sarcoma found in adults

Site of Origin	Type Of Sarcoma	Location In The Body
Fibrous tissue	Malignant fibrous	Legs
Blood vessels	Kaposi's sarcoma	Legs; Trunk
Lymph vessels	Lymphangiosarcoma	Arms
Synovial tissue	Synovial sarcoma	Legs
Peripheral nerves	Neurofibrosarcoma	Arms; Trunks

Histology-Diagnosis.

WHO classification is determined to make the histological diagnosis's according to it only. They determine the grade & stage of the tumor(*Table:4*). Grade is always provided base on the recognized system in all the case where it can be possible[21-25].

Adjuvant Radiotherapy.

The standard treatment for all intermediate or high-grade soft tissue sarcomas is postoperative radiotherapy (*Table:5*). This approach enables the preservation of function with alike local control rates and survival to radical resection (i.e., compartmental excision/amputation) [23-26]. Adjuvant irradiation is not necessary for subjects which have undergone a compartmental resection or amputation assuming that the margins are clear. 60 – 66Gy in 1.8 – 2 Gy fractions is the referenced postoperative radiation dose [27]. The technique employed is usually a 2-phase using shrinking field; 50 Gy to the initial larger volume seconded by 10 - 16 Gy to a smaller volume [28, 29]. Sometimes this dose may need to be decreased if the field involves critical structures (for example the brachial plexus). Attention is drawn to the VORTEX clinical trial in extreme soft tissue sarcomas [27]. This randomized clinical trial, which is currently recruiting in the UK, is checking for differences between the standard 2-phase conventional radiotherapy technique with a single phase to a smaller tissue volume, in an attempt to avoid normal tissue & hence improve subsequent limb function without compromising local control.

Neo-Adjuvant Radiotherapy.

Studies of pre-operative radiotherapy in limb sarcoma deduced associations with increased postoperative complications in comparison with the standard postoperative treatment but less late toxicity (reflecting the lower pre-operative dose of 50 Gy compared with the postoperative dose of 66 Gy and a smaller treatment volume), with equal tumor control [30]. Pre-operative radiotherapy is not used routinely in the United Kingdom, but may be used in certain situations where the size of the radiation field required for post-operative treatment is likely to be associated with significant late morbidity, or when the tumor is of borderline operability and pre-operative radiotherapy is judged to have the ability of rendering the tumor operable [31–33]. Attributed to some radiosensitive histological subtypes, such as myxoid liposarcoma, pre-operative radiotherapy may be particularly beneficial, given the degree of tumor shrinkage that can be garnered. The standard regimen for pre-operative radiotherapy is 50 Gy, in 1.8 - 2 Gy fractions, followed by surgery approximately 6 weeks after completion of radiotherapy. Extended radiotherapy (10 - 16 Gy) may be introduced post-operatively, if tumor margins are positive34-36.

Table5: Soft tissue sarcoma grouped by chemosensitivity

RELATIVE CHEMOSENSITIVITY	EXAMPLES OF SOFT TISSUE SARCOMAS
Chemosensitive	1. Round cell liposarcoma
	2. Uterine leiomyosarcoma
	3. Synovial sarcoma myxoid/
Moderately chemosensitive	1. Scalp and face angiosarcoma
	2. Desmoplastic small round cell tumor
	3. Angiosarcoma 4. Peripheral nerve sheath tumours
	5. Leiomyosarcoma
	6.Pleomorphic Rhabdomyosarcoma malignant
	7. Epithelioid sarcoma
	8. Myxofibrosarcoma
	9. Pleomorphic liposarcoma
Relatively chemo-insensitive	1. Endometrial stromal sarcoma
	2.Clear cell sarcoma
	3. Dedifferentiated liposarcoma
Chemoinsensitive	1.Alveolar soft tissue sarcoma
	2.Extrasekeletalmyxoid chondrosarcoma

Neo-Adjuvant Chemotherapy.

Pre-operative chemotherapy is considered for the subjects having large high grade tumors which are considered borderline resectable by the MDT, though the data for this is limited. The demographics of the patient must be taken on the account like his/her age or any of the comorbidity he/she have. There is a broad variation in chemosensitivity in different types of the histological subtypes[37].

CLOSING REMARKS:

Competent imaging, predictive immunological & genetic studies, improved surgery, & newer methods of adjunctive & neo adjunctive treatment should result in improvements in outcomes for subjects with these tumors . There are different type of soft tissue sarcoma found and they effect the various body parts & the name given to those sarcoma are according to the part they are affecting. Most of the time tissue part is affected in the body. Young aged & adults are affected with this sarcoma along with the evolvement of the painless lump. Various genes are also involved in inducing soft tissue sarcoma & they are as RB1, CDKN2, P53, N-RAS. In addition to the known immunoreactivity for CAM5.2 and EMA the positivity for CK7 and 34BE12 for small proportion cases is also observed. Research work is still under process and this soft tissue sarcoma has basically four stages (stage 1; stage 2; stage 3; stage 4).patient only survive for the 5 years. If the patient is diagnosed in the early time then it may have the cure but the percentage of the curity is very low. Probability of the recurrence of sarcoma is very low but even then it can occur. This type of the soft tissue sarcoma has the painless lump ; therefore many of the detection has to be done. Some of them are: biopsy, immunohistochemistry, cytogenetics, reverse transcript. If the patient has gone under the surgery then chemotherapy is also done to cure it. Many of the researcher are till on the track to find out the better solutions for the soft tissue sarcoma so, that it can cure easily and affectively.

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