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A Study on Evaluation and Management of Soft Tissue Sarcoma

KEYWORDS: Soft tissue sarcoma, Trucut Biopsy, Metastatic work up .

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ABSTRACT Soft tissue sarcoma are rare interesting tumors, they frequently occur in young individuals and this together with challenges they take to the pathologists in diagnosing them and to the clinicians in treating them generate fascination in them. Soft tissue sarcoma (STS) are rare and unusual neoplasm. In this is clinical study the current concepts are reviewed with regards to etiology, diagnosis and management. The study is aimed at mode of presentation , histological types, investigative modalities and treatment options of patients admitted in our hospital between September 2012 and October2014.

INTRODUCTION

Soft tissue sarcomas are rare and unusual neoplasms accounting for about 1 % of adult human cancer and 15% of pediatric malignancies. These soft tissues neoplasms include all the non-epithelial extraskeletal tissue with the exception of glia of central nervous system, reticuloendothelial system and supporting tissues of various parenchymal organs. Included in these are lesions composed of or derived from fat, fibrous tissue, smooth muscle, skeletal muscle, blood vessels and lymphatics all of which originate from embryonic mesoderm.

Sarcoma (Greek) means 'fleshy growth'. Malignant tumors that arise from the primitive mesoderm are called sarcoma. All the soft tissues sarcomas are grouped together because of their similarities in pathological appearance, clinical presentation and behavior.

Although these tumors develop in any anatomical site, approximately 50% occur in extremities followed in order of frequency by the trunk, retro peritoneum and head and neck.

Finally, recent advances in the use of adjuvant chemotherapy and radiotherapy have made dramatic in roads, in the mortality of these sarcomas to the point that patients with tumor previously uniformly fatal are not only now being cured but also escape mutilating surgery and consequent disability.

In the clinical study, the current concepts are briefly reviewed with regards to etiology, diagnosis and management. The soft tissue sarcomas that were studied at Osmania General Hospital and MNJ Cancer Hospital from September 2012-October 2014 are recorded and analyzed with respect to the incidence, pathology, clinical features, investigations and management. The various aspects are discussed and conclusions drawn.

MATERIAL AND METHODS

About 38 cases of adult patient with soft tissue sarcomas involving extremities, trunk and abdomen which were referred to Osmania General Hospital / MNJ Cancer Hospital for diagnosis and treatment during the period of September 2012 to October 2014 are studied and analyzed. Among 38 patients, 24 are primary,14 are recurrent swellings.Ten cases presented with metastastic disease (Lymphnode metastasis - 8 cases, distant metastasis - 2). Mean duration of presentation is < 6m with average tumor size > 10 cm. All cases are examined clinically and diagnosed by biopsy with FNAC/Trucut/incisional biopsy technique. MRI or CT scan were done to assess local extent of tumor. Metastatic work up is done by X-ray chest or CT Scan or with both. Treatment is given as standard protocol.

RESULTS, ANALYSIS AND DISCUSSION

The Total sex incidence in present study is M:F 1:1.1, which is matching with other studies like Kandel et al G.D. Jonnson et al., showing female bias 1:1.1. But R.C. Ramanandhan, Hashimotos et al, showing male bias with 1.1:1 and Qadir etal 1.2:1. Peak incidence is noticed in between 4th to 5th decade which is comparable with the other studies like Johnson et el, 50+/- 5and kandel et al 55 +/- 8 years

TABLE – 1
AGE INCIDENCE

S. No	Age in years	No. of patients	Percentage
1	0-10	0	0
2	11-20	2	5.2
3	21-30	3	7.8
4	31-40	6	15.7
5	41-50	13	34.2
6	51-60	7	18.4
7	61-70	5	13.1
8	> 70	2	5.2

TABLE – 2
SIZE OF TUMOR

Size in cm	Present Study	Pistel et al (1996) 1041	J.Leavy (1993)321
<5	10 (26.3%)	431 (41%)	114(35.5%)
5-10	12 (31.5%)	293 (28%)	116(36.1%)
> 10	16 (42.1%)	258 (25%)	91(28.4%)

Majority of patients presented with large masses (>10cm. 42.1%) usually cannot be treated with surgery alone. Unlike other studies Pister et al., J Leavy et al, where tumor size at the time presentation is relatively small (>30%). Site of presentation in present study one patient is hav-

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ing multiple neurofibrosarcoma over the thigh, chest and left cheek.The site incidence in our study roughly correspond to other series with most common site being the lower limb (55.2%) then trunk (18.4%), upper limb (13.2%) and abdomen retroperitoneum (10.5%, wall 2.6%). Proximal extremities involved more commonly than distal. The mean duration of presentation in our study is 3m to 1 year, more during first 6m (55.2%) which is corresponding to the standard literature. Retro peritoneal tumors present late. This may be due to asymptomatic nature of the tumors which is corresponding to the standard literature.

TABLE – 3
MODE OF PRESENTATION

Mode of presentation	Present study	Pistel et al 1041
Primary swelling	24 (63.1%)	836 (80%)
Recurrent swelling	14 (36.8%)	205 (20%)
Ulcerative swelling	3 (7.9%)	102 (9.7%)
Pain	11 (28.9%)	196 (19%)
Fever	6 (15.7%)	110(10.5%)
Bleeding from tumor	4 (10.5%)	76 (7.3%)
History of Trauma	4 (10.5%)	52 (4.9%)
Distalneurovascular compression	3 (7.8%)	97 (9%)
Restricted joint mobility	7 (18.4%)	100 (9.6%)

Most of the patients presented with either primary (63.1%), recurrent swelling (36.8%), few having distal neurovascular pressure effects (7.8%). Hisstory of trauma is present in up to 10%, but whether it is the cause or it draws the patient's attention to the tumor is not known. In few cases joint mobility is restricted (18.4%), tumors when they are located near a joint. Most of the patients are anemic. And few also presented with pain and fever. These features are comparable with other studies. The only exception in the present study is the incidence of recurrent swelling is high at the time of presentation.

The incidence of lymph node metastasis is very high (21%) in our study compare to other studies (<11%) (Weinguard et al, Mazeron et al, Fong study). Lymphnode metasis are seen in about 8 among 38 patients, of them 6 are most common histologic types where lymphnode metasis are possible Malignant Fibrous Histocytoma(MFH) – 2/ Rabdo Myo Sarcoma(RMS)-2/Synovial Sarcoma- 1 and Angiosarcoma- 1 and 2 cases of liposarcoma where lymphnode metstasis are relatively uncommon.

Two cases presented with distant metastasis in the lungs1-High grade pleomorphic liposarcoma lower limb2-Angiosarcoma chest wall.The first one is treated with Surgery+Chemotherapy (S+CT), second one was inoperable and treated with Radiotherapy+Chemotherapy (RT+CT) only

The diagnosis of soft tissue sarcoma is confirmed by FNAC in 20/38, Trucut biopsy 17/38 and incisional biopsy in 3/38. The reliability of correct diagnosis including histopathologic type is better with Trucut (76.4%) and incisional (100%) biopsy compared to FNAC (45%). These result comparable with other studies (MSKCC – 86.6, 95.6 & 55.5%) respectively in that order. Overall the sensitivity is more with trucut & incisional biopsy than FNAC.Most common histologic type in our study MFH (26.3%) followed by liposarcoma (15.7%) RMS (13.2%),Malignant Peripheral Nerve Sheath Tumor MPNT (13.2%). Fibrosarcoma (10.5%) and other like Dermato Fibro Sarcoma Protruberance DFSP, synovial sarcoma, extra skeletal chondrosarcoma. Angiosarcoma. Haemangiopericytoma, Clear cell sarcoma. Desmoid tumor, angiolipomas each carrying 2.6%. These are roughly equivalent to other studies. Though the incidence of Leomyosarcomas (LMS) is relatively high in retroperitoneal tumors, no single case is seen in our study. Similarly MFH is relatively less common in the retro peritoneum compare to the other studies. The difference, in these incidences is to be further studied and clarified.

Radiological investigations CT scan and MRI were performed on 28 cases to assess the local extent of tumor as well as to predict resectability and compared with postoperative findings .Thus MRI predicts resectability and local extent better than CT scan. These are comparable with other studies such as Kirby I bland et al. The results have to be further studied based on anatomical location.

The selection of our treatment modalities, apart from other variables that are universally accepted like size, site, histologic type and grade of the tumor, also depends upon the socioeconomic status of the patient and availability of modalities.

TABLE – 4
MANAGEMENT

S.No	Treatment given	Present study
1	Surgery alone	15 (39.4%)
2	S+RT	12 (31.5%)
3	S+RT+CT	8 (21%)
4	S+CT	2 (5.2%)
5	RT+CT	1 (2.6%)
6	Total	38 (100%)

Surgery alone was done in nearly half the cases (39.4%) and in others surgery is combined with either adjuvant RT or CT (57.7%). Overall surgery alone in 15 (39.4%) adjuvant RT in 20 (52.6%), adjuvant CT in 10 (26%) and only RT+CT is given in 1(2.6%) case of Angiosarcoma presented with distant metastasis and poor general condition.Among the surgeries the performed, wide excision is done in more than half of the patients 25/37 (67.6%) and other procedures are amputation 10 (27%), marginal excision in 2 (5.4%). Our amputation rate is reduced to 27% due to development of adjuvant RT and CT compared to previous studies.Most commonly performed procedure in our study is wide excision.

On the whole treatment that is given broadly along the lines indicated by other authors. (J. Leavy et al, 1993, RC Ramanadhan 1998, Mandard et al 1999).

The follow up cases are 26 with median follow period of 6months – 2 years. Total cases of recurrence are 5. Among these 2 are Local recurrence (LR), 2 are distant metastasis (DM), 1 both L.R & DM with mean period of recurrence 6m.Local recurrence is seen in about 2 cases, among those who have been treated with S+RT+CT (total-8) and the cause may be inadequate RT due to poor follow up, and histologic diagnosis in these cases is high grade pleomorphic RMS.Distant metastasis is seen in about 2 cases, those who have been treated with surgery alone (total-15) one case is pleomorphic MFH, a high grade tumor presented with in 6m, another extra skeletal chondrosarcoma which is a high grade tumor present with in 2yrs. In both of these cases it is due to advanced disease at the time presentation, and also they have not taken either R T or CT after surgery, of course the role of the these modalities is to be clarified further.

Both local and distant metastasis seen in one case, who has been treated with surgery alone (total-15) this case is

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retroperitoneal neurofibrosarcoma, a high grade tumor, presented with in 6M. No single recurrence is seen with patients who have been treated with S + RT indicates adequate local control with RT after surgery. In general local control is better achieved with limb sparing S + RT as mentioned in the literature, and also proved in our study. We need better patient education for early presentation and better compliance and development of standardized modality and therapeutic facilities.

CONCLUSIONS

STS though rare are interesting tumors, they frequently occur in relatively young individuals and this together with challenges they take to the pathologists in diagnosing them and to the clinicians in treating them generate fascination in them.

In this study the soft tissue sarcomas occurred with median are group of 30-50 years, with nearly equal incidence in both the sexes with slight female predominance. Average time of presentation is<6m, usually with larger tumor size> 10cm due to asymptomatic nature of the tumor. More than half of the patients present with primary swelling and the role of trauma is still controversial. Most common site is proximal parts of lower extremities with most common histopathology of MFH. Metastasis to regional lymphnodes is very high in our study may be due to high grade large tumors or due to previous surgeries. Two cases presented with distant metastasis in the lungs. The incidence of MFH in the retroperitoneum in our study is high compared to the other studies. It is to be further studied and clarified.

Most commonly performed reliable biopsy procedure is trucut. Incisional biopsy is done where trucut biopsy is not giving adequate tissue sample for histopathologic subtype and grading for the management. FNAC is useful in recurrent swellings where diagnosis is already known. Radiological investigations such as CT and MRI play an important role in predicting resectability of the tumor as well as assessing local extent of the tumor with MRI providing better delineation of soft tissues and neurovascular structures. Though results of the present study are in line with other studies, it needs to be further studied based on anatomical site involved.

Surgery alone is done for low graded, smaller size tumor less than 5cm and wide excision is preferable. Limb sparing surgeries are preferable to radical amputative procedures and if margins are in doubt it is better to supplement with adjuvant radiotherapy with or without chemotherapy. Patients who are unfit for surgery can be tried with RT + CT. The recurrence rate is very high for high grade, large size tumors with positive margins.

In the present study the STS occurring in this region are analyzed and their management outlined. These are found to be in line with other series. However our patients usually present late with advanced diseases where curative measures are not useful and further follow up rate is poor to draw clear conclusion regarding effectiveness of the treatment strategies used. It requires further study to be clarified.

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