TITLE:

"A SURVEY OF SOFT TISSUE SARCOMAS AT KENYATTA NATIONAL HOSPITAL OVER A SIX YEAR PERIOD JANUARY 1991 TO DECEMBER 1996"

DISSERTATION SUBMITTED IN PART FULFILLMENT FOR AWARD OF THE DEGREE OF MASTERS OF MEDICINE IN SURGERY

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TABLE OF CONTENTS . . . . . PAGE

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DEDICATION

This work is dedicated to my dear wife Margaret and my two daughters Angela and Doreen.

DECLARATION

This dissertation is my original work and has not been presented for a degree in any other University.

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Signed.....

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SUMMARY

Soft tissue sarcomas are a heterogenous group of neoplasms accounting for 1% of all malignancies and 5% of childhood malignancies according to Western Series, while locally they account for about 3.8% of all malignancies. Eighteen (18) different histological types were recorded compared with over 30 types recognised world wide so far. This study presents a review of 187 patients whose histological diagnosis of soft tissue sarcomas was made at Kenyatta National Hospital and subsequently treated between 1991 and 1996.

The peak incidence of soft tissue sarcomas is in the first decade of life with more than half of these tumours occurring during the first three decades. Mean age of occurrence is 29.5 years and male female ratio is 1.2:1 in this study.

About 31 new patients are seen annually with soft tissue sarcomas. There is no evidence of decrease nor increase in the incidence of these cases. The commonest presenting complaint is painless swelling (85%) and our patients present late with 71% presenting with tumours greater than 5 cm.

Of the 178 patients whose site of tumours was recorded at the Cancer Registry, 52 (29.3%) were in the lower limbs, 39 (19.1%) were in the trunk, 39 (19.1%) head and neck, 20 (11.3%) upper

limb visceral, 16 (8.9%) retroperitoneal, 14 (7.8%) genitourinary, 6 (3.4%) and intrathoracic 2 (1.1%). Extremities therefore took 40.6% which is inkeeping with Memorial Sloan Kettering Cancer Centre Series (40-50%) [1]. Retroperitoneal tumours are slightly less than in other series 7.8% cf 15-17%, head and neck were more in our series 19.1% cf 5-8% in Western Series [2].

The five commonest histological types in this series were: Rhabdomyosarcoma 15 (20.8%), fibrosarcoma 13 (18.1%), liposarcoma 12 (16.7%), dermatofibrosarcoma 6 (8.4%) and malignant fibrous histiocytoma 5 (6.9%).

Rhabdomyosarcoma was the predominant tumour below 20 years of age 54%, followed by fibrosarcoma (14%) and then atypical tumours (9%). After the age of 40 years, malignant fibrous histiocytoma 23%, fibrosarcoma 15% and liposarcoma were the commonest in that order. Eighty five (85) patients had their tumours graded with 24 (53%) having high grade tumours, 9 (20%) intermediate grade and 27% low grade sarcomas.

Diagnostic methods used were fine needle aspiration cytology 20%, excisional biopsy 35%, incisional biopsy 45%, CT scan 5% abdominal ultrasound 7% The high incidence of incisional biopsy is explained by the late presentation of the patients and lack of core needle biopsy facilities, No conclusive diagnosis was

provided by FNAC and other biopsies had to be taken.

Surgery was the mainstay of treatment offered 12 out of 85 patient (14%) underwent enucleation/shelling out which is wrong treatment, 19 (22.3%) wide excision, 6 (7%) amputation, 10 (11.8%) radiotherapy alone, 15 (18%) surgery and radiotherapy, 2 (2.3%) surgery and chemotherapy and 3 (3.5%) a combination of the three modalities. Enucleation, chemotherapy alone, radiotherapy alone or radiotherapy and chemotherapy alone had poor results with 2 years recurrence rate of 50%, 98%, 70% and 50% respectively with 3,4,3,4 patients dead within 2 years from the disease respectively. Wide excision and amputation and when combined with adjuvant therapy had best results. Of 19 patient treated with wide excision alone, 7 had greater than 2 years recurrence free period (37%), 10 had less than 2 years recurrence free period (53%), while only 2 were dead from disease within 2 years (10%), while all the six who underwent amputation were all alive and free of disease by the end of 2 years.

Size of the tumour, grade and site were noted to be prognosis determinants. Visceral tumours had the worst prognosis with 5 of the 6 patients dead within 2 years (8%), while 1 had no change in disease progress (17%), while extremity sarcoma had the best prognosis with 6 out of 10 (60%) getting greater than 2 years recurrence free period, 2 (20%) got recurrence within 2 years and 2 (20%) died within 2 years.

Seven out of 22 (32%) of those with high grade tumours had recurrence within 2 years, 6 (27%) had recurrence free period > 2 years, while 9 out of 12 (41%) were dead within 2 years from sarcoma as compared to those with low grade tumours where 3 out of 13 (23%) with recurrence within 2 years, 8(81%) recurrence period greater than 2 years and only 1 dead (7.6%) within the same time span of the patients with low grade tumours less than 5 cm, all (100%) had no recurrence by the end of 2 years while for the 9 with tumours greater than 5 cm, 3 out of 9 (33%) had recurrence within 2 years, 4 out of 9 (44.4%) had recurrence period greater than 2 years and 2 out of 9 (22%) were dead within 2 years. These differences were treated statistically and found to be significant for the grade but insignificant for size.

The main recommendation from this series is to sensitize our people in the need to report early when they notice lumps in the body:

- To discourage any temptations to do enucleation of these tumours
- To emphasize to the pathologists to give grades of tumours at histology and to be more aggressive surgically even when dealing with retroperitoneal tumour.

Planned adjuvant therapy should always be instituted and need to have more studies especially prospective ones carried out in order to come up with set down protocols for the management of these neoplasms.

INTRODUCTION

The relatively small number of case seen and the great diversity in histopathologic presentation anatomic site and biologic behaviour have made a comprehensive understanding of these disease entities extremely difficult [11]. These tumours are however ideal prototype to demonstrate the important role of multidisciplinary management.

Local literature on this subject matter is lacking. This series is therefore aimed at being an eye opener on the issue by high lighting the extent of the problem and our approach to its management.

With a figure of 3.8% of all malignancies seen at Kenyatta National Hospital, these malignancies cannot therefore be ignored. Great Britain and American series have found this problem constituting about 1% of all malignancies [1,2,3,10,15].

These malignancies have been found to be prevalent in the first, fifth and sixth decades of life [1]. Though this series have shown the prevalence to be in the first two decades of life.

The histological types seen compares quite well with types elsewhere in the world with rhabdomyosarcoma, fibrosarcoma, liposarcoma, malignant fibrous histiocytoma, dermatofibrosarcoma

and leiomyosarcoma being the commonest [1,3,15].

The grade of the tumour size at presentation, completeness of excision and site of tumour have been found to be strong determinants of prognosis of these tumours [1,9,12,30]. We cannot therefore forget to emphasize on the need to treat these tumours within the earliest time possible and also the need for wide excision with negative margins where possible. These tumours should always be graded inorder to determine the aggressiveness of treatment. Although surgery remains the mainstay of the treatment of these neoplasms, the need for multimodality-multidisciplanary approach to their management should be emphasized. Radiotherapy and chemotherapy have been found to be of help in some histological types such as rhabdomyosarcoma and also as adjuvant treatment [6,16,21,25].

Inadequacy of our health facilities, poor infrastructure and prevalence of poverty in our society has resulted in late presentation to hospital which outrightly affect our treatment results. With only one radiotherapy facility available country wide, overbooking has resulted in delays in administering this treatment with some patients missing it all together.

More studies should follow this one so that at the end of this day we can have set down policies and protocols in the management of these malignancies institutions such as the Memorial Sloan

Kettering Cancer Centre have soft tissue sarcoma committees with various specialists involved meaning that the patient gets the best possible treatment depending on the nature of the individual disease.

AIMS AND OBJECTIVES

1. To highlight the epidemiological pattern of presentation of patients with soft tissue sarcoma with regard to age, sex, histology and clinical pathological profile of these neoplasms.

2. To identify the magnitude and trend of soft tissue sarcoma over the study period.

3. To study the management modalities applied for the treatment of these tumours and to recommend future approach in dealing with these tumours.

RATIONALE

To date there are no local series done on this subject matter hence the need for this study in order that it can act as an eye opener on this topic.

The study will draw more interest in this subject and increase the awareness of medical fraternity in relation to soft tissue sarcoma so that in future collaborated management can be availed to these patients.

MATERIALS AND METHODS

The study was carried out at Kenyatta National Hospital. This was a descriptive study covering a period of 6 years from January 1991 to December 1996. A further period of 2 years was included in order to evaluate the outcome of patients who had been seen even in 1996.

Only those patients whose diagnosis was confirmed by histology at Kenyatta National Hospital and entered in the National Cancer Register were included some of whom were subsequently managed at Kenyatta National Hospital. The sample size was determined by the study period.

The relevant records of patients with soft tissue sarcoma were

tissue sarcoma were reviewed after approval of the study proposal by the Kenyatta National Hospital Ethical and Research Committee

Records scrutinised included in-patient files from the Central Medical Records, data from the Cancer Registry and Histopathology Department. Information was obtained regarding age, sex, histological diagnosis, size of tumour, site, diagnostic method used and management modality given and outcome of treatment at 2 years duration after. Data was collected using tally sheet and analysed using the statistical computer programme.

LITERATURE REVIEW

Soft tissue saracomas are a relatively rare group of malignant neoplasms that arise from extraskeletal connective tissue. However some tumours of neuroectodermal origin such as malignant schwanoma are included. They account for 1% of the adult malignancies and about 5% of all childhood tumours [1,2] in these series which included all age groups from 0 to over 70 years they accounted for 3.8% of all malignancies.

The age adjusted incidence for the United Kingdom is 1/100,000 which is about 1200 cases reported annually while in the US the incidence is approximately 6,000 cases annually [1,2,6].

The relatively small number of case and the diversity in histopathological presentation anatomical site and biological behaviour have made a comprehensive understanding of these disease entities extremely difficult. It is clear that soft tissue sarcoma diagnosed early is eminently curable. When diagnosed at a time of extensive local disease or metastatic disease it is rarely curable.

AETIOLOGY

Most soft tissue sarcomas have no clearly defined aetiology although multiple associated or predisposing factors have been identified. These are:-

1. **Genetic predisposition**

   As seen in Von-Recklinghausen's disease, Li-Fraumeni syndrome, Retinoblastoma and Familiar polyposis coli, Genetic predisposition to malignancy is well established in the form of an autosomal dominant gene in 8 - 9% of children with soft tissue sarcomas [35]. Jan C. Liang and Donald P. Pinkel in 1989 demonstrated presence of mutant chromosomes in a cancer prone family members. A follow up of Li-Fraumen. Families with childhood rhabdomyosarcoma found additional cancers in some of the patients including soft tissue sarcomas [69].

2. **Ionising Radiation**

   Post irradiation soft tissue sarcomas have been identified as far back as 1922 [1]. A present study by Bradt M.S. and Gaynor J.J. showed that patients receiving radiation therapy for breast and cervical cancer subsequently developed malignant fibrous histiosarcoma, angiosarcoma and lymphangiosarcoma in 2% of the number studied [3]. Rufus J.

Mark and Joseph Poem reviewed patients with soft tissue sarcomas and found a 0.03 - 0.8% risk of development of post irradiation soft tissue sarcomas on long follow up. [18].

3. Chemical exposure

Chemicals such as phenoxacetic acid, chlorophenols (in herbicedes) thorotrast, vinyl chloride and arsenic have resulted in the incidence of these tumours among agricultural industry workers. Marianne N. Prout and Hugh L. Davis Jr demonstrated three case of haemongiopericytoma of bladder after PCV exposure [11].

4. Lymphoedema has long been associated with development of lymphangiosarcoma. The most well recognised association is with the post-mastectomy post-irradiation lymphoedematous arm described by Steward and Treves [8]. Similar advance sarcomas have been seen following filarial infection and chronic lymphoedema. In these series, only one patient had lymphangiosarcoma and there was no associated disease identified.

5. Patient with HIV infection and renal transplant patients are known to have high incidence of soft tissue sarcomas. Kaposis sarcoma though not classified under these sarcomas is a very frequent tumour among these patients.

Distribution

Soft tissue sarcomas can occur in any site throughout the body. Fifty percent (50%) of all soft tissue sarcomas however appear in the extremities, 35% in the lower extremities and 15% in the upper extremities, while trunk takes up 10% Retroperitoneal 14%, visceral 15%, other sites such as head and neck 11% [1,3].

Overall the most common histopathologic subtypes are malignant fibrous histiocytoma, liposarcoma, leiomyosarcoma and fibrosarcoma with rhabdomyosarcoma being the commonest tumour below 20 years of life. A study done at Memorial Sloan Kettering Cancer Centre between 1982 and 1995 demonstrated liposarcoma 290/2678, MFH 230/2678, fibrosarcoma 150/2678 and leiomyosarcoma 90/2678 to be the commonest extremity tumours, leiomyosarcoma 130/2678 and liposarcoma 180/2678 commonest retroperitoneal tumours, leiomyosarcoma 280/2678 as predominant visceral STS while liposarcoma, leiomyosarcoma MFH, fibrosarcoma in the trunk [1].

Fibrosarcoma has a peak in the second and third decade, malignant fibrous histiocytoma fifth and sixth and seventh decade, leiomyosarcoma fourth and sixth decade while liposarcoma had uniform distribution in the third to seventh decade of life [1].

Clinical presentation

Most patients will have a history of progressive painless swelling until the lesions are quite large. There are very few symptoms and when they occur they are caused by pressure or direct invasion of adjacent structures which result in pain oedema or neurological deficit. Other systems will depend on organ system involved.

Those tumours in extremities present early so do those in the head and neck, but those in the retroperitoneum and viscera present late with abdominal pain and backache [2]. These has a bearing on the outcome of treatment as size of tumour has been shown to be a prognostic factor.

Diagnosis

Differential diagnosis of soft tissue sarcomas include non-neoplastic conditions such as haematomas, chronic abscess, benign neoplasms such as lipomas, neurofibromas and harmatomas. Other malignant tumours eg. lymphoma and metastatic carcinoma and sarcoma of bone are other differentials. It is therefore important to biopsy all soft tissue lumps that persist or grow.

Tissue Diagnosis

Core needle biopsy is the procedure of choice and can be done in out patient set up under local anaesthesia. The primary thrust of

biopsy is to obtain adequate tissue for definitive histopathologic confirmation, to evaluate grade and to identify prognostic factors that would alter the approach to definitive treatment in the main, for lesions that are less than 5cm particularly those that are superficial, excisional biopsy is the preferred approach. Cone needle biopsy has an accuracy greater than 95%

Fine needle aspiration cytology

Fine needle aspiration cytology has been examined by a number of authors, but is usually confined to the confirmation of recurrence rather than for primary diagnosis.

Incisional biopsy

This has an accuracy of almost 100% in experienced hands as adequate tissue is made available and hence other parameters like grade are possible to determine. The incision should be placed in such away that subsequent excision of the tumour will include the biopsy incision.

Frozen section

In some institutions frozen section is relied upon as the diagnostic tool of choice. For many institutions including ours, this is however not available. Heslin M., Lewis J. and Woodruff at Memorial Sloan Kettering Cancer Centre showed frozen section to provide adequate tissue in 94% Corrent diagnosis in 88%

correct grade 65% and correct histology in 47% as compared to incisional biopsy which showed adequate tissue-100%, correct malignancy 100%, correct grade 95%, correct histology 86% while core needle biopsy provided adequate tissue in 92% Correct malignant in 95%, correct grade in 85% and correct histology in 73% [1]. These shows that in order of superiority, incisional biopsy comes first, followed by core needle biopsy and then frozen section.

Imaging

Imaging studies such as ultrasound, CT scan, MRI are important especially in retroperitoneal and visceral tumours in establishing the relationship of these neoplasms to other organs and neurovascular bundles in anticipation of definitive treatment.

Pathology

Macroscopically, sarcomas are of variable appearances and consistency depending on their tissue of origin. They enlarge in a centrifugal manner compressing normal surrounding tissue and appear to be encapsulated. This pseudocapsule is actually composed of an inner compressed rim of normal tissue and an outer rim of oedema and newly formed vessels. Fingers of tumour can be extended into and through the capsule giving rise to satellite lesions. The pseudocapsule pauses a big draw back during surgery as the surgeon might be deceived to think that the tumour is

shellable, hence, leading to early recurrence. The fragile blood vessel lacking in basement membrane and only supported by neoplastic cells result in bleeding and early blood borne metastatic. In these series 12/85 (14.1%) were treated by enucleation with a very poor outcome in terms of local control.

The histological classification of soft tissue sarcoma is based on the morphological resemblance of tumour to normal tissue. To date about 30 different histological types have been identified. Recent advances such as immunohistochemistry electron microscopy and cytogenetics are important in identifying cell types.

The following is a pathological classification of soft tissue sarcomas depending on their tissue of origin. Examples of the histological subtypes are also given.

1. **Sarcomas of fibrous and fibrohistiocytic tissue**
   - Dermatofibrosarcoma protuberance
   - Fibrosarcoma
   - Malignant fibrous histiocytoma

2. **Sarcoma of muscle tissue**
   - Leiomyosarcoma
   - Rhabdomyosarcoma
   - Epithelioid leiomyosarcoma

3. Sarcoma of adipose tissue

- Liposarcoma

4. Sarcoma of the blood vessels and lymphatics

- Haemangiosarcoma
- Epithelioid haemangiosarcoma
- Lymphangiosarcoma

Note that kaposis sarcoma is not classified here. According to the WHO classification of diseases it is classified as a special sarcoma. There is a controversy regarding it's tissue of origin.

5. Sarcoma of the synovium

- Synovial sarcoma
- Malignant giant cell tumour of tendon sheath

6. Sarcoma of the peripheral nervous system

- Malignant schwanomas
- Primitive neuroetodermal tumours
- Malignant granular cell tumour

7. Sarcoma of the pleuripotential mesenchyme

- Malignant mesenchynoma

8. Sarcoma of the unknown histogenesis

- Extraskeletal Ewings sarcoma
- Alveolar soft part sarcoma
- Epithelioid sarcoma

- Clear cell sarcoma
- Desmoblastic small cell tumour

As mentioned earlier the most common subtype are malignant fibrous histiocytoma, liposarcoma, fibrosarcoma, rhabdomyosarcoma and leiomyosarcoma. Liposarcoma, MFH, tendosynovial sarcoma and fibrosarcoma are the commonest extremity sarcoma, leiomyosarcoma being the commonest visceral sarcoma, Liposarcoma, fibrosarcoma and rhabdomyosarcoma being commonest retroperitoneal sarcomas while desmoids, liposarcoma and myogenic sarcomas are the commonest trunk and chest wall tumours [1,2].

Grading

Grading of these tumours depend on cellularity, differentiation pleomorphism, mitotic rate, extent of necrosis and amount of stroma. They are basically graded into high, intermediate and low grade. This is as recognised by the American Joint Commission on Cancer (AJCC) [19]. A binary system as used in Memorial Hospital is also used-(high versus low) [20].

In different trials, a grade eg. high grade had been defined differently at different centres, making comparison of results between trials hazardous due to subjectivity. Many Western series have showed the grade to be a major predictor of treatment outcome and is therefore a major determinant of the mode of treatment to be offered ie. to give or not to give adjuvant

therapy [9,22,23].

These tumours are basically graded into high, intermediate and low grade [22].

- G_1 - Low grade
- G_2 - Intermediate grade
- G_3 - High grade

Staging

Several staging systems have been proposed. In the AJCC/VICC. System TMN classification has been modified to a GTNM system where G is for the tumour grade.

- T - primary tumour
- T_1 - tumour < 5cm
- T_2 - tumour ≥ 5cm

- N - regional lymphnodes
- N_0 - no metastases to nodes
- N_1 - histologically verified metastases [22].

To lymph nodes

- M - distant metastases
- M_0 - no metastases
- M_1 - distant metastases present

Stage grouping

Stage IA	G_1	T_1	N_0, N_0
Stage IB	G_1	T_2	N_0, N_0
Stage IIA	G_2	T_1	N_0, N_0
Stage IIB	G_2	T_2	N_0, N_0
Stage IIIA	G_3	T_1	N_0, N_0
Stage IIIB	G_3	T_2	N_0, N_0
Stage IVA	Any G, any T		N_1, N_0
Stage IVB	Any G, any T		N_1, N_1

Other staging systems such as Enneking staging system only applies to extremity sarcomas.

Stage I - Low grade no metastases

stage II - High grade no metastases

Stage III - Either grade with metastases

Each grade is classified as an A or B depending on whether the tumour is confined to one compartment or extends beyond it.

Treatment

The mainstay of treatment for all soft tissue sarcomas of extremity and trunk is surgical resection. Even for retroperitoneal and head and neck as complete resection as possible should be undertaken. The issue of debate is how extensive that surgical resection should be and whether it should be preceded or followed by adjuvant therapy. Wide enbloc

resection is used most often. Historically attempts to resect all muscle bundles from origin to insertion have been supplanted by the encompassing resection aiming at obtaining 2cm of all uninvolved tissue in all directions. This is often unrealistic, however, because the limiting factor is usually neurovscular juxtaposition or occasionally bony juxtaposition.

Soft tissue sarcomas rarely involve skin, so, major skin resection should be limited in situation of primary or recurrent tumours where skin is involved, or the tumour is so extensive that the skin is involved, then free flap or rotational flap closure is necessary especially where subquent radiotherapy will be used. The most extensive resection is clearly amputation. This should be only rarely be indicated in soft tissue sarcoma at present time, limb sparing operation are possible in at least 95% of patients as was demonstrated by Shiu M.H. et al [24].

Experience over the last 25-30 years at Memorial Sloan Kettering Cancer Centre (MSKCC) indicated that by 1995, the 50% amputation rate in the 1960's is only 5% Amputation should thus be reserved for tumours not able to be resected by any other means, without evidence of metastic disease and the propensity for good long term functional rehabilitation. Often these are patients with large low grade tumours with considerable cosmetic and functional deformity who can be rendered symptom-free by a major amputation [26,27].

A lot of work has been done comparing limb sparing surgery versus amputation. In a series by Rosenberg S.A., Tepper J., Glastein E. et al (1978) showed that local recurrence can occur after limb sparing operation where resection was combined with adjuvant radiotherapy and chemotherapy as compared with less incidence of recurrence when amputation alone is used, but there was no impact on long term survival [27].

The same issue has been addressed at the National Cancer Institute in the US where a randomised trial was done and data available over a 10 year period shows that although local recurrence is greater in those undergoing limb sparing operation plus irradiation compared with amputation survival overall is not different [26].

Sarcoma of the retroperitoneum and viscera pause a big challenge in their management. This is due to their inaccessibility for wide excision and the fact that radiation therapy would have major bad effects on other organs. These factors however should not preclude attempt at resection.

Complete resection is usually possible in 60% to 70% of patients. In a series by Jaques D.P., Coitdg, Hadjv S.I. et al [27]. Although nephrectomies were done in 46% of cases, the kidney itself was rarely involved 2%, nevertheless the encompassment of the kidney and involvement of the hilar renal vasculature make

the resection of the kidney on occasions technically necessary. The overriding principle is not to be reticent about resection of adjacent organs should they be involved by tumour but conversely not to resect organs not involved if they are not the limiting factor in the margins, for example the resection of a kidney when in fact the venacava is the closest margin makes no oncologic sense. Overall the use of debulking for recurrences is rarely of significance in terms of long term survival. More extended resections do not seem to improve local recurrence or survival. The concept should therefore be that unless palliation can be achieved operation should be reserved for those patients whom complete resection is at least possible, if not possible, the basis for unresectability is usually the presence of peritoneal implants or extensive vascular involvement [1].

The size is a prognostic factor for outcome in terms of both local recurrence and subsequent metastatic disease, the approach to lesions can be varied. For small lesions less than 5cm correct surgical resection is usually sufficient adjuvant therapy being reserved for only those with recurrent lesions. Given the high risk of recurrence and of systemic disease for lesions larger than 10cm that are high grade these patients are candidates for investigational approaches especially neoadjuvant radiotherapy and chemotherapy. All patients with lesions larger than 5cm should be considered for adjuvant radiotherapy as a proven method of limiting local recurrence [12].

Radiation therapy

Radiation therapy is generally used as a surgical adjuvant. External ream irradiation has been the most commonly applied treatment although there has been increasing interest in adjuvant branchy therapy. Radiation therapy can also be given in preoperative fashion. Rarely for unresectable tumours or a medically inoperable patient, radiation is used alone.

Historically, amputation was considered the standard local treatment for soft tissue sarcoma or the extremities although this treatment provided excellent local control, the adverse impact on quality of life was obvious. Several groups therefore initiated a treatment policy of conservative surgery followed by high-dose postoperative external beam radiation therapy.

Suit [12,13] reported local control of 82% in a cohort of 100 patients treated with either radiation alone or radiation delivered after conservative surgery while Lindberg [14] found that a useful limb was maintained in more than 80% of cases with a local recurrence rate of 19% and 33% respectively for intermediate and high grade tumours, less than 5cm and greater than 5cm. The same results were collaborated by Karakousis and colleagues who have prospectively treated extremity sarcomas with conservative resection. In their series patients whose surgical margins were negative for more than 2cm, no further treatment was given while patients with less adequate margins were given

postoperative external beam radiation routinely and they found 93% local control for these patients as compared to 83% for surgery alone group [28]. All these series taken together demonstrated that excellent local control is obtained with limp-sparing resection followed by postoperative external beam irradiation for soft tissue sarcomas of the extremity and superficial trunk.

Adjuvant brachy therapy

This method only treats the tumour bed with no attempts made to treat large margins overlying skin, scar or drain site. After complete surgical resection, after loading catheters are implanted to cover the surgical bed and are sutured in place by catgut sutures. The wound is closed over the catheters with a drain interposed between the catheters and the wound closure. the radioisotope iridium iodine^{125} is loaded on the sixth postoperative day. At Memorial Sloan Kettering Cancer Centre a prospective randomised trial showed a local control of 90% for brachy therapy versus 69% for surgery alone. This mode of treatment is at times combined with external beam radiation with good results. It minimises the damage to surrounding tissues thus reducing the destruction of the normal tissues, hence, better functioning of the treated part of the body [29,30].

This treatment has been tried in the inaccessible retroperitoneum with good results. Preoperative radiotherapy also has some role

in some situations. Suit and coworkers have reported 5 year actuarial local control of 90% for patients treated with preoperative radiotherapy [5]. There is however a large wound complication rate in this mode of treatment.

Chemotherapy

Some histological types such as rhabdomyosarcoma especially the embryonal type have been shown to have good response to chemotherapy. Cyclophosphamide, vincristine, doxorubicin, actinomycin D, ifosphanide and nethotrexate have been used in combinations. By and large chemotherapy has no effect on overall survival, but studies have shown some effects on local control of the disease where surgery and chemotherapy cannot be offered for various reasons and age does not prohibit, then chemotherapy should be given. At the National Cancer Institute US. Patients treated with doxorubicin and followed up for a period of seven years following resection, the five year disease free survival was 75% for those who received chemotherapy as compared to 54% for the non-chemotherapy group, but the difference in overall survival was not significant. Additional studies utilising combination therapy with doxorubicin, cyclophosphamide and methotrexate and involving bigger number of patients alluded to the previous results with 7 year actuarial survival of 56% versus 43%, but again no effect on distant metastases free survival non overall survival [1].

A similar study utilising the same drugs was done at the foundation Bergonie including 59 patients and showed a 5 year survival for chemotherapy treated group of 90% as compared to 50% for the non chemotherapy group. All these studied and many others have shown that chemotherapy has some benefit on survival [1]. Hyperthemia has also been tried to enhance the effects of chemotherapy with some improved results [1].

In summary low grade tumours less than 5cm can be treated adequately by complete resection alone. Post operative radiotherapy would be added only if there was concern about margins for sarcomas of the hand, foot, head and neck. Wide margins are difficult to obtain. Also tumours in the retroperitoneum for low grade lesions 5cm or larger, the standard practice would be complete resection followed by postoperative external bean irradiation. For high grade lesions adjuvant radiation strategy should be brachy therapy. This in good hands provide high local control, excellent functional outcome and low cost in cases where the adequacy of the margin is questionable or where technical factors preclude a good implant supplemental external beam irradiation is added where not feasible then external beam irradiation can be used alone. Radiotherapy alone should rarely be used for definitive management. However, in situations where surgery is not possible, or are only marginally resectable, either preoperative radiotherapy or chemotherapy, brachy therapy with or without postoperative external beam

irradiation would be combined with surgical procedure.

Recurrences are treated with repeat resections followed by adjuvant radiotherapy and chemotherapy. Metastasectomy has been done with reasonable effects on disease free survival.

Prognosis

Overall it has been noted that tumour grade, size, site and location in relation to deep fascia are prognistic factors with 5 year survival rates of stages I, II, III and IV being 80%, 65%, 45% and 10% respectively, sarcoma of the extremities followed by those of trunk while visceral and retroperitoneal sarcomas have the poorest prognosis. Of the cases seen with soft tissue sarcoma annually a little more than 50% will go on to die from the disease.

RESULTS

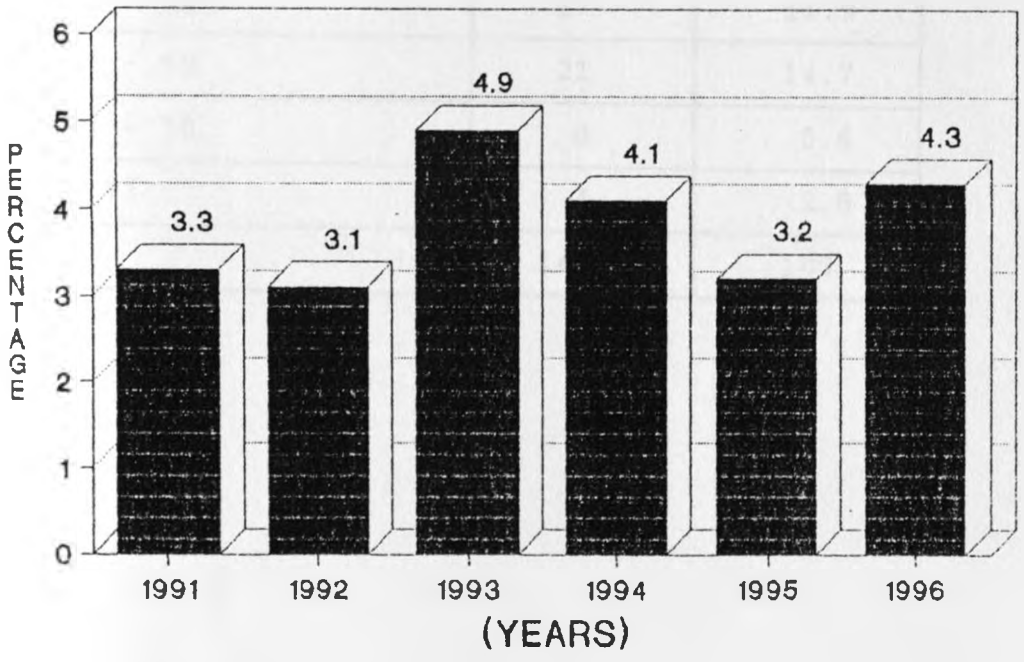
Table 1 shows total number of soft tissue sarcomas per every year of study and percentage of total malignancies, the highest number of cases was in 1993 with 48 patients and the lowest in 1997 with 23 patients

Total number of patients was 187 constituting 3.8% of all malignancies.

TABLE 1:

Year	Malignancies	STS	%
1991	907	30	3.3
1992	734	23	3.1
1993	975	48	4.9
1994	598	25	4.1
1995	793	25	3.2
1996	828	36	4.3
Total	4835	187	3.8

FIG I: SOFT TISSUE AS A PERCENTAGE OF TOTAL MALIGNANCIES



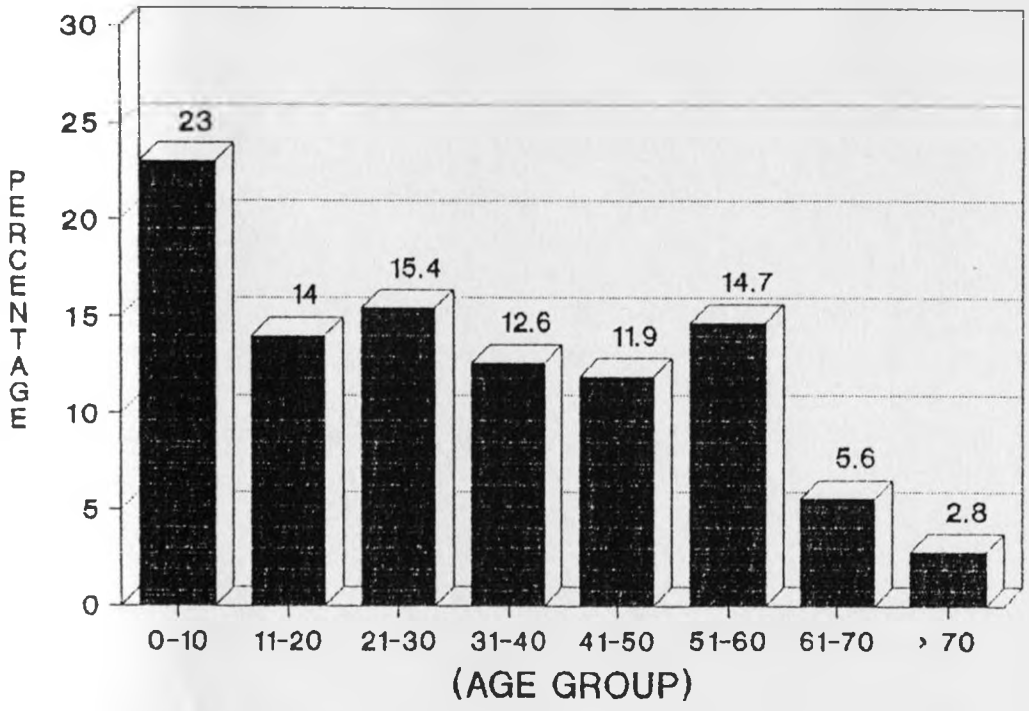
Mean age was 29.5 years

147 patient had their ages documented in the records.

TABLE 2: AGE DISTRIBUTION OF SARCOMAS

Age group	n	%
0 - 10	33	23.0
11 - 20	20	14.0
21 - 30	22	15.4
31 - 40	18	12.6
41 - 50	17	11.9
51 - 60	21	14.7
61 - 70	8	5.6
≥ 71	4	2.8
Total	143	100

FIG. 2: AGE DISTRIBUTION OF SARCOMAS



The most afflicted age group was 0 - 10 years, 23% of all sarcomas with 32% of cases occurring at the age of 20 years and below. There were no very large difference in number of cases seen in the other age groups except after 60 years of age where only 8.4% of these sarcomas were seen.

Table 3 shows histological types of Soft tissue sarcoma seen at Kenyatta National Hospital, their number and percentage of the total number of sarcomas seen.

TABLE 3:

Histological type	n	%
Rhabdomyosarcoma	50	26.7
Fibrosarcoma	34	18.2
Pleomorphic sarcomas	19	10.2
Liposarcoma	18	9.6
Leiomyosarcoma	16	8.6
Dermatofibrosarcoma	11	5.9
Malignant fibrous histiocytoma	11	5.9
Spindle cell sarcoma	7	3.7
Haemangiopericytoma	4	2.1
Heamangioblastoma	3	1.6
Alveolar soft part sarcoma	3	1.6
Angiosarcoma	2	1.1
Synovial sarcoma	2	1.1
Malignant mesenchymoma	2	1.1
Neurofibrosarcoma	2	1.1
Ganglioneuroblastoma	1	0.5
Lymphangiosarcoma	1	0.5
Dermoid tumour	1	0.5
Total	187	100

A total of 18 different histological types of soft tissue sarcoma ware seen. The commonest seven soft tissue sarcomas in order of frequency were rhabdomyosarcoma with 20%, fibrosarcoma with 18.2%, pleomorphic sarcomas with 10.2%, liposarcoma 9.6%, leiomyosarcoma 8.6%, dermatofibrosarcoma 5.9% and malignant fibrous histiocytoma 5.9%

Table 4 shows the sex distribution of the common types of soft tissue sarcomas

TABLE 4:

Histological type	Male	Female	Ratio
Fibrosarcoma	17	15	1.1:1
Rhabdomyosarcoma	23	23	1:1
Leiomyosarcoma	5	10	1:2
MFH	7	3	2.3:1
Liposarcoma	7	8	1.1:1
Dermatofibrosarcoma	8	3	2.7:1
Pleomorphic sarcoma	10	9	1.1:1

Liposarcoma, fibrosarcoma, rhabdomyosarcoma showed no significant difference in the sex distribution of this tumours, while malignant fibrous histiocytoma showed a prepoderance for male 2.3:1, dermatofibrosarcoma also ratio 2.7:1 while leiomyosarcoma showed a preponderance for female ratio 2:1.

Table 5 shows the age distribution of the different histological types of soft tissue sarcomas.

TABLE 5:

Histological type	0-10	11-20	21-30	31-40	41-50	51-60	61-70	>70	Total
Rhabdomyosarcoma	27	12	5	3	1	1		1	50
Dermatofibrosarcoma	1	1	2	2	4	1			11
Fibrosarcoma	1	5	9	8	6	5		1	34
MFH			1	3	2	1	4	1	11
Angiosarcoma		1					1	1	2
Haemangiosarcoma		1							1
Haemangiopericytoma	2								2
Haemangioblastoma	1	2	1						4
Liposarcoma	2	1	3	1	3	5			15
Leiomyosarcoma	2	1	2	4	4	2	1		16
Pleiomorphic sarcoma	2	4	2	3	1	3		2	19
Synovial cell sarcoma		1	1						2
Spindle cell sarcoma	1	1	1	2	2				7
Malignant mesenchymoma							1	1	2
Neurofibrosarcoma			1		1	2			4
Alveolar soft part sarcoma		1	2						3
Lymphangiosarcoma			1						1
Desmoid	1								1
Total	40	31	30	26	24	21	6	6	185

Below 20 years of life the commonest soft tissue sarcoma was rhabdomyosarcoma which accounted for 54% of all tumours seen in this age group, fibrosarcoma 8.8%, pleiomorphic sarcoma 14% After 40 years of age, the three most common tumours were fibrosarcoma 8(15%), MFH 12(23%), Liposarcoma 7)13.2%). No single case of MFH was seen below the age of 20 years.

Table 6 shows the distribution of the soft tissue sarcomas in the body.

TABLE 6:

Site	n	%
Lower limbs	52	29.3
Trunk	34	19.1
Head and neck	34	19.1
Upper limbs	20	11.3
Visceral	16	8.9
Retroperitoneal	14	7.8
Genitourinary	6	3.4
Intrathoracic	2	1.1
Total	178	100

Forty point six percent (40.6%) of the sarcomas were in the extremities, trunk 19.1%, head and neck 19.1%, visceral 8.9% and retroperitoneal 7.8%

Of the tumours in the lower limb 61% were in the thigh with liposarcoma, fibrosarcoma, rhabdomyosarcoma, MFH being the commonest in that order.

Of the tumour in the head and neck 2 of them were intracranial, haemangioblastoma.

FIG 3: DISTRIBUTION OF SOFT TISSUE SARCOMAS IN THE BODY

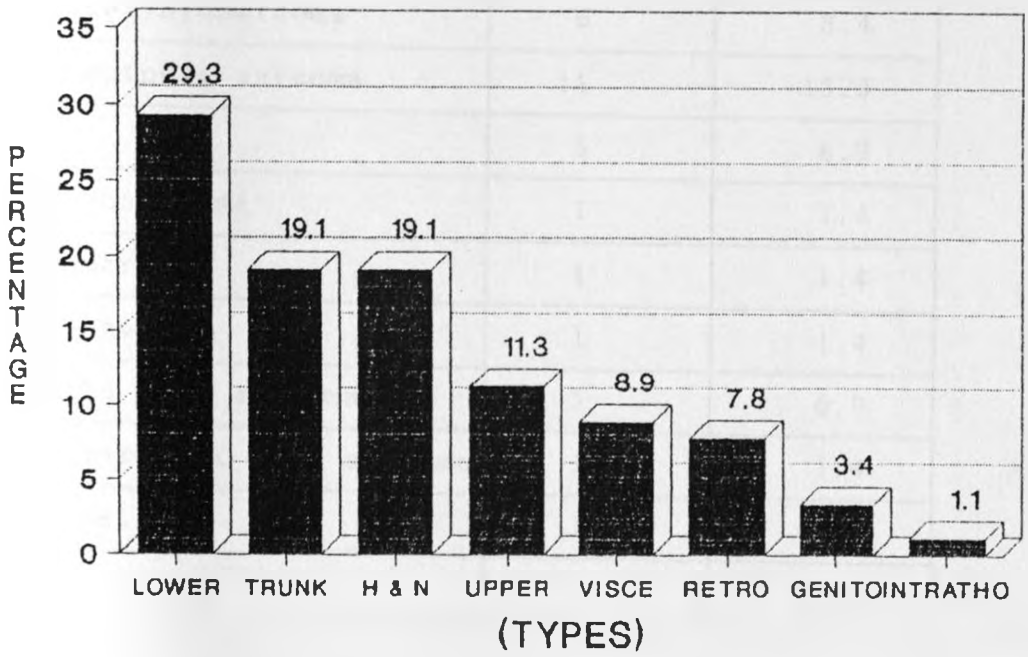


Table 7 shows distribution of the histological types of soft tissue sarcomas seen in the extremities.

TABLE 7:

Histological type	n	%
Rhabdomyosarcoma	15	20.8
Fibrosarcoma	13	18.1
Liposarcoma	12	16.7
Dermatofibrosarcoma	6	8.4
Pleiomorphic sarcoma	11	15.3
MFH	5	6.9
Leiomyosarcoma	1	1.4
Angiosarcoma	1	1.4
Mesenchymoma	1	1.4
Spindle cell sarcoma	5	6.9
Alveolar soft part sarcoma	1	1.4
Endothelioma	1	1.4
Total	72	100

The commonest extremity sarcoma was rhabdomyosarcoma 20.8% followed by fibrosarcoma 18.1%, liposarcoma 16.7% pleiomorphic sarcoma 15.3%. MFH 6.9% and DFS 8.4%

Table 8 shows distribution of the histological types of soft tissue sarcomas seen in the head and neck.

TABLE 8:

Histological type	n	%
Rhabdomyosarcoma	22	65.0%
Fibrosarcoma	4	11.7
Haemangioblastoma	3	8.8
MFH	2	5.8
Malignant mesenchymoma	1	2.9
Neurofibrosarcoma	1	2.9
Pleiomorphic tumour	1	2.9
Total	34	100

The commonest head and neck tumour was rhabdomyosarcoma 56% followed by fibrosarcoma 11.7% The 3 cases of haemangioblastoma were intracranial.

FIG 4: DISTRIBUTION OF SOFT TISSUE SARCOMAS OF HEAD AND NECK

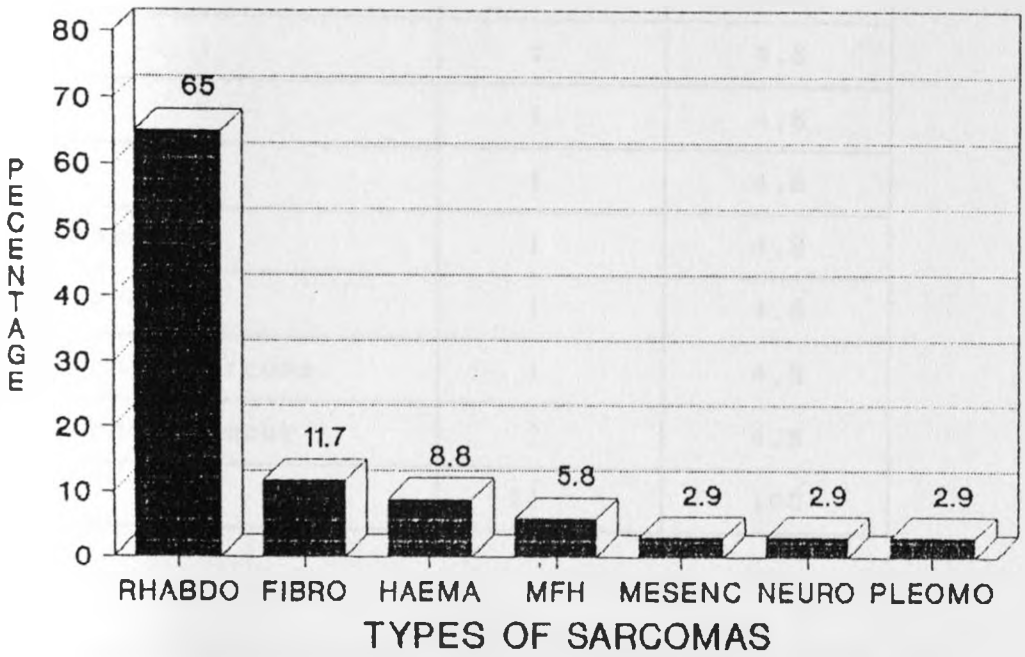


Table 9 shows histological distribution of sarcomas seen in the trunk.

TABLE 9:

Histological type	n	%
Fibrosarcoma	9	42.9
Rhabdomyosarcoma	2	9.6
Neurofibrosarcoma	2	9.6
MFH	1	4.8
DFP	1	4.8
Desmoid	1	4.8
Liposarcoma	1	4.8
Leiomyosarcoma	1	4.8
Spindle cell sarcoma	1	4.8
Pleiomorphic tumour	2	4.8
Total	21	100

The commonest trunk sarcoma were fibrosarcoma 42.9% and rhabdomyosarcoma 9.6% and neurofibrosarcoma.

Table 10 shows distribution of histological types of soft tissue sarcomas seen in the retroperitoneal

TABLE 10:

Histological type	n	%
Leiomyosarcoma	1	11.1
MFH	1	11.1
Ganglioblastoma	1	11.1
Liposarcoma	4	44.4
Fibrosarcoma	2	22.2
Total	9	100

FIG 5: DISTRIBUTION OF SARCOMA SEEN IN RETROPERITONEUM

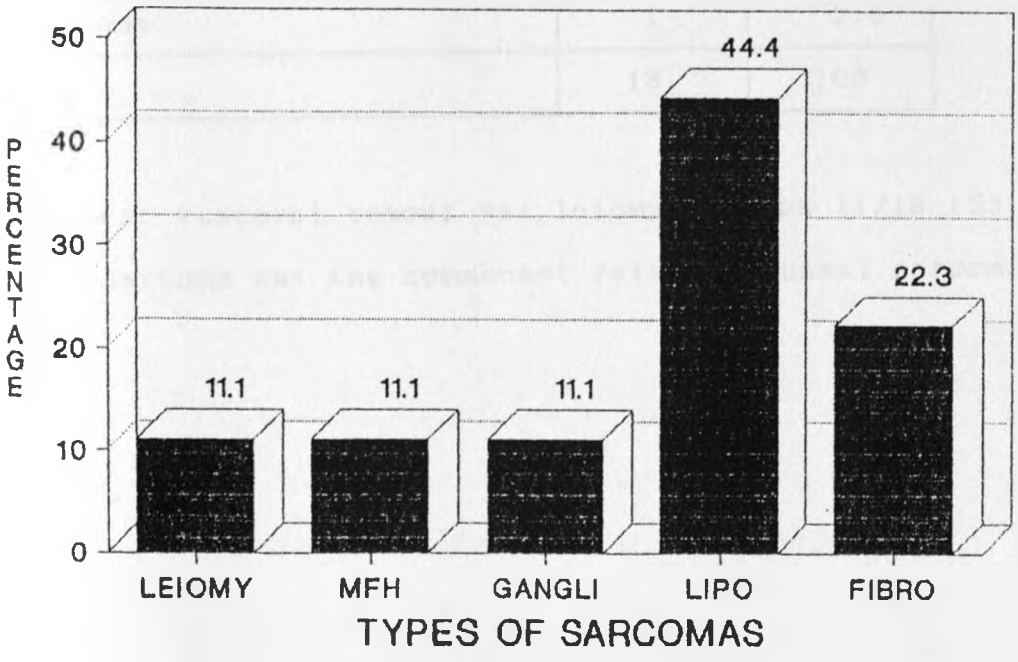


Table 11 shows distribution of histological types of soft tissue sarcoma seen in the viscera

TABLE 11:

Histological type	n	%
Leiomyosarcoma	10	55.5
Rhabdomyosarcoma	5	27.7
Pleiomorphic sarcoma	1	5.6
MFH	1	5.6
Fibrosarcoma	1	5.6
Total	18	100

The commonest visceral tumour was leiomyosarcoma 11/18 (55.5%) while liposarcoma was the commonest retroperitoneal sarcoma 4/9 (44.4%).

Table 12 shows the grades and sizes of sarcomas seen at Kenyatta National Hospital.

TABLE 12:

Grade	Tumour size .	n	%
Low	< 5cm	4	30.8
Low	5 - 10cm	6	46.2
Low	> 10cm	3	23.0
Intermediate	< 5cm	1	11.1
Intermediate	5 - 10cm	3	33.3
Intermediate	> 10cm	5	55.6
High	< 5cm	2	9.0
High	5 - 10cm	5	23.0
High	> 10cm	15	68.0

Out of 85 patients whose records were traced at the records department. 45 were graded while 40 were not graded. Twenty four (24) 53% were high grade 9(20%) were intermediate while 13(27%) were low grade.

FIG. 6: DISTRIBUTION OF TUMOUR GRADES

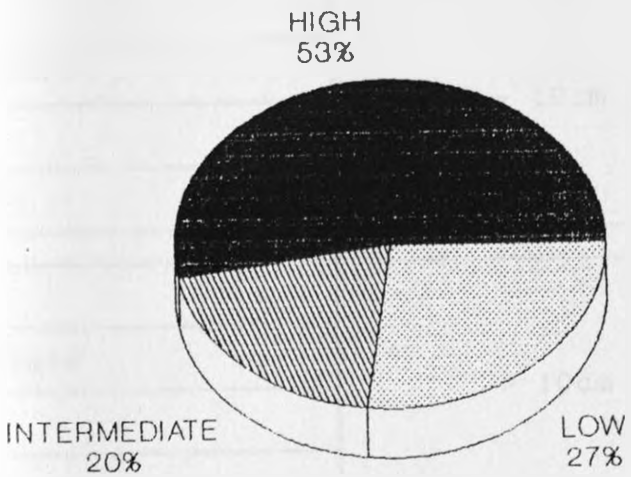


Table 13 shows the size of tumours at presentation, their number and percentage.

TABLE 13:

Grade	Tumour size	%	Total %
Low	< 5cm	4	19
Intermediate		1	
High		2	
Ungraded		9	
Low	5 - 10cm	6	36
Intermediate		3	
High		5	
Ungraded		16	
Low	> 10cm	3	45
Intermediate		5	
High		15	
Ungraded		15	

Sixteen (16) patients 19% presented with tumours less than 5cm. 30 patients (36%) presented with tumours 5-10cm in size while 38 patients (45%) presented with tumours greater than 10cm in size. Seventy one percent (71%) of the patients therefore presented late with tumours greater than 5cm in size.

FIG. 7: SIZE OF TUMOURS AT PRESENTATION

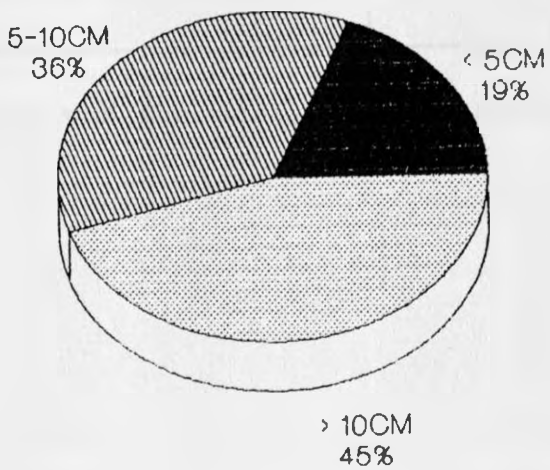


Table 14 shows the diagnostic methods used and the number of patients in which the various methods were used.

TABLE 14:

Diagnostic method used	n
FNA-C	15
Excisional biopsy	20
Incisional biopsy	30
Core needle biopsy	0
C & R	10
Ultrasound	17
CT scan	5

Incisional biopsy was the most utilised method of diagnosis.

FIG. 8: DISTRIBUTION OF DIAGNOSTIC METHODS USED

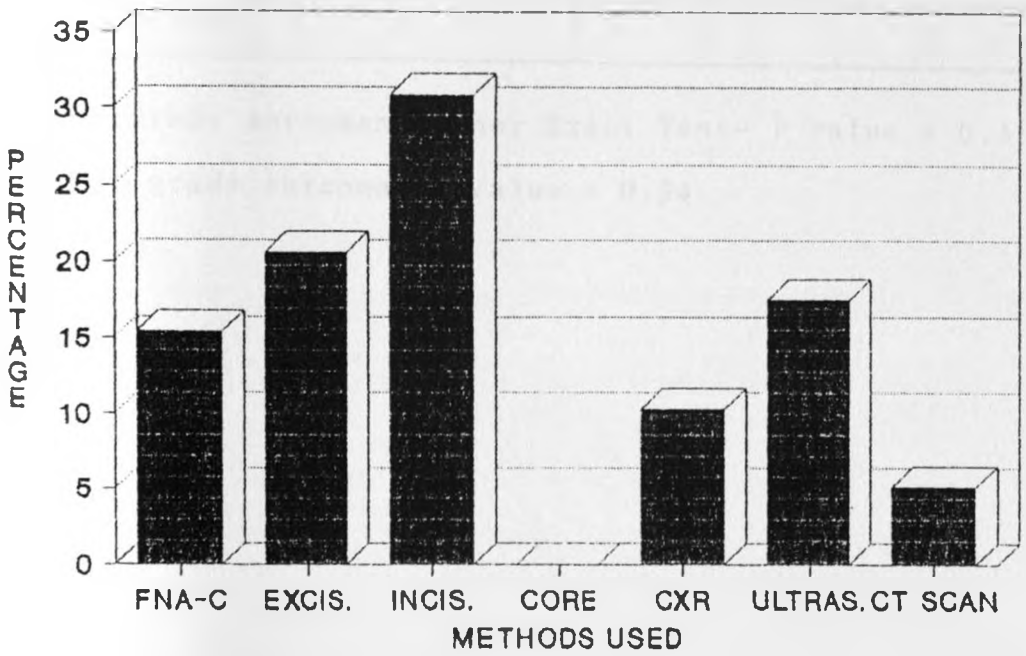


Table 15 shows the relationship between size of tumour and results of treatment.

TABLE 15:

Grade	Size	Less than two years recurrence free period	Greater than 2 years recurrence free period	Dead within two years	Total
Low	< 5cm	0	4	0	4
	> 5cm	3	4	2	9
High	< 5cm	2	0	0	2
	> 5cm	6	5	9	20

For low grade sarcomas Fisher Exact Test- P value = 0.46, while for high grade sarcomas P value = 0.34.

Table 16 shows the relationship between grade of tumour and result of treatment.

TABLE 16:

Grade	Less than 2 year recurrence free period	Greater than 2 year recurrence free period	Dead within 2 years	Total
Low	2	8	1	11
Intermediate	5	4	1	10
High	7	6	9	22

X^2 = 5.6. P value = 0.02

Size

Of the 13 patients who had low grade sarcoma 4 had tumours less than 5cm at presentation, while 9 had tumours greater than 5cm. In the group with less than 5cm, all 4 had a recurrence free period greater than 2 years, none was dead within 2 years. In the group with tumours greater than 5cm (9 patients), 3 had recurrence within 2 years, 4 had recurrence free period greater than 2 years and 2 were dead within 2 years from pulmonary metastases.

Of the 22 patients who had high grade tumours, 2 presented with tumours less than 5cm and both of them had a recurrence free period of less than 2 years after resection while 20 had tumours greater than 5cm and 6 of them had recurrence free period less than 2 years (27%), 5 had recurrence free period greater than 2 years (22%) and 9 of them were dead within 2 years from metastatic disease (40%).

Grade

Of the 22 patients who had high grade tumours, 7 had recurrence within 2 years (32%), 6 had a recurrence free period greater than 2 years (27%) and 9 of them were dead within 2 years (40%). Of the 13 patients with low grade tumours, 3 had recurrence within 2 years (23%), 8 had recurrence free period greater than 2 years (61%) while only 1 died with 2 years from the disease (7.7%). High grade tumours were found to have a significant worse

prognosis than low grade tumours (P = 0.02). Of the 9 patients with intermediate grade, 5 had a recurrence free period of less than 2 years (55.5%) while 4 had a recurrence free period greater than 2 years (44.5%) after resection while none was dead within 2 years.

Site of tumour and prognosis

Patients who had high grade tumours that were greater or equal to 5cm at presentation had their records evaluated to determine outcome of the 6 patients with intra-abdominal or intrathoracic tumours. Five (5) were dead within 2 years while 1 had no change in disease state though still alive by 2 years. For the extremity group 10 patients had their records examined of which 2 were dead from disease within 2 years. Six (6) had recurrence free period greater than 2 years. For the trunk tumours 1 was dead within 2 years and the other had a recurrence within 2 years.

For the head and neck tumours, of the 2 patients in this category, 1 had recurrence within 2 years while 1 had a recurrence free period of greater than 2 years.

Intra-abdominal tumours therefore had worst prognosis while extremity tumours had the best prognosis. Patients with low grade tumours were not evaluated due to their small number-5 in all.

Table 17 shows the different modalities of treatment applied and the outcome.

TABLE 17:

Mode of treatment	Greater than 2years disease free state	Less than 2 years recurrence free state	No change	Dead within 2 years	Total
Shelling/ enucleation	2	6	1	3	12
Wide excision	7	10		2	19
Amputation or radical resection	6				6
Radiotherapy alone		3	4	3	10
Surgery and radiotherapy	4	5	1	4	15
Radiotherapy and chemotherapy	1	1			2
Chemotherapy alone	1		4	4	9
Surgery and chemotherapy	2				2
Surgery Chemotherapy/radiotherapy	1	2			3

Of the surgical methods utilised enucleation had the poorest results with 50% of patients having recurrence within 2 years, 8.3% dead within 2 years and only 33% having a recurrence free period greater than 2 years, while amputation had the best result with 100% recurrence free period of greater than 2 years. Wide incision treatment resulted in 53% of recurrence free period less

than 2 years. 37% got recurrence free period greater than 2 years while 10% were dead within 2 years from the disease. Radiotherapy alone, chemotherapy alone and radiotherapy and chemotherapy without surgery had very poor results. For those who were managed with surgery and radiotherapy, 27% had recurrence free period greater than 2 years, 6% showed no change in disease process, 27% were dead within 2 years while 40% had recurrence free period less than 2 years.

Mortality

Thirteen percent (13%) of the 85 patients whose record were analysed were dead from the disease and all died from pulmonary metastases. Eighty one percent (81%) of them had high grade tumours while 9.5% had low grade sarcoma and 9.5% had intermediate grade sarcoma. All of those dead had presented with tumours greater than 5cm.

DISCUSSION

Soft tissue sarcomas remain a relatively rare group of neoplasms, series done in the Western world have found these malignancies to account for about 1% of all malignancies and about 5% of childhood malignancies.

This series between 1991 and 1996 at Kenyatta National Hospital found these neoplasms to account for 3.8% of all malignancies with 187 cases documented during the study period and an average of 31 case annually.

Kaposis sarcoma which is classified under special sarcomas and skin neoplasms by the WHO classification of diseases was excluded from this study. No evidence of an increasing trend of these neoplasms was demonstrated.

There is no significant difference in incidence between male and female ratio 1.2:1. Some histological type however such as malignant fibrous histiocytoma and dermatofibrosarcoma are more common in males ratio 1.3:1 and 2.7:1 respectively. This is in accord with findings of Rooser B. et al and Mcpeak C.J. et al who found ratios of 2:1 and 3:1 respectively. At Kenyatta National Hospital the peak incidence of these tumours is in the first decade of life 40/185 (22%) with more than 50% of these neoplasms occurring in the first three decades of life, mean age of

occurrence being 29.5 years. Only 12/185 (6.4%) of these tumours were found after 60 years of age. Memorial Sloan Kettering Cancer Centre series found 15-20% in this age group which can be explained by our lower life expectancy [1,2].

Eighty five percent (85%) of the patients presented with painless swelling and 71% presented with tumours greater than 5cm. A study of distribution by size of 2678 patients done at Memorial Sloan Kettering Cancer Centre found 33% of patients presenting with tumours less than 5cm, 33% with lesions greater than 10cm. Being a painless swelling and poor access to health facilities by our people might explain the late presentation in our set up.

In these series 40.6% were in the extremities, 19.1% trunk, 19.1% head and neck, 8.9% visceral and 7.8% retroperitoneal, 3.4% genitourinary and 1.1% intrathoracic. Extremities presented an almost similar proportion to those obtained in various Western world series [1,2,10,21]. Head and neck slightly more in our series 19.1% as compared to 5-8% in the Sloan Kettering Memorial Cancer Centre institute of the US. Retroperitoneal sarcomas took a smaller proportion 7.8% as compared to 15-17% in the same series.

The commonest histological types in this series were rhabdomyosarcoma 20.8%, fibrosarcoma 18.1%, liposarcoma 16.7%, dermatofibrosarcoma 8.4% and malignant fibrous histiocytoma 6.9% which is in the agreement with the findings elsewhere in the

world [1,2,10,13,21].

Rhabdomyosarcoma was the predominant sarcoma below 20 years of age followed by fibrosarcoma. After the fourth decade. MFH. fibrosarcoma and liposarcoma were found to increase in frequency.

The commonest extremity sarcoma was rhabdomyosarcoma 20.8% Other common types were fibrosarcoma 18.1%. fibrosarcoma 16.7%. malignant fibrous histiocytoma 6.9% and dermatofibrosarcoma 8.4%

Fibrosarcoma and rhabdomyosarcoma were the commonest head and neck sarcomas 11.7% and 6.5% respectively while fibrosarcoma 42.9%. rhabdomyosarcoma 9.6% and neurofibrosarcoma 9.6% were the commonest trunk sarcomas. Leiomyosarcoma was the commonest visceral sarcoma (56%) while liposarcoma was the commonest retroperitoneal sarcoma 44.4% Similar distribution was found by Enzinger F.M. et al at St. Louis Morsy in 1988.. Sixty percent (60%) of the extremity sarcomas were in the thigh with liposarcoma being the commonest type.

Most clinicians currently consider grade and size of tumour to be acceptable predictors of outcome of treatment. The most useful and critical piece of information the pathologist can provide to the clinician after establishing the diagnosis of sarcoma is the histologic grade.

In this series only 85/187 (45%) had the grade of their sarcomas provided by the pathologist. Fifty three percent (53%) were high grade, 20% intermediate and 27% low grade. The grade should determine our approach to management and hence lack of grading might have resulted in under-treatment of some of our patients.

Costa J. et al., Trojani M. et al. and Myhre Jensen O. et al in different series all showed grade to be a determinant of survival after surgical and adjuvant treatment of soft tissue sarcomas [9,20,22].

Diagnostic methods used in this series were, fine needle aspiration cytology in 20% of the cases, excisional biopsy in 35%, incisional biopsy in 45% of the cases, CT scan in 6% and Ultrasound in 7% if cases. The latter methods of diagnosis were used in intra-abdominal lesions.

The high prevalence of utilisation of incisional biopsy could be explained by late presentation of patients in this series where 71% presented with lesions already greater than 5cm in diameter and unavailability of core needle biopsy needles at Kenyatta National Hospital. No conclusive histological diagnosis was provided by FNAC and all had incisional or excisional biopsies performed after to confirm histologic type. Other centres have used this latter diagnostic method to detect recurrence. FNAC requires highly experienced cytologists for meaningful

interpretation of this slides. These specialists are in very few numbers at Kenyatta National Hospital. Frozen section was not utilised as the facilities are not available locally. Heslin M. et al at Memorial Sloan Kettering Centre showed incisional biopsy to be the most superior followed by core needle biopsy and lastly frozen section by providing correct histology in 86%, 73% and 50% respectively [1].

Surgery remains the mainstay of treatment with radiotherapy and chemotherapy being only adjuvant. The latter two modalities can only be applied alone where resection of whatever nature is impossible. Surgery is always aimed at obtaining tumour free margin in all planes. David P.. Jaques et al at Memorial Sloan Kettering Cancer Centre in a study of management of retroperitoneal sarcomas showed that extent of resection determined prognosis of these sarcomas. In their study patients with possible complete resection had a median survival of 60 months compared to 24 months for those who underwent partial resection and 12 months for those with unpresentable tumours [41]. These findings were echoed by Patrick C. McGrath et al at Massey Cancer Centre, Varginia. WHO found a 5 year survival of 78% for those undergoing complete resection as compared to that of 8% for those undergoing partial resection or biopsy only. However despite an improvement disease-free survival, there is still a high recurrence rate (60-80%) even after complete resection and hence the need for adjuvant therapy.

In this series surgery was the mainstay of treatment. Radiotherapy and chemotherapy were instituted either as adjuvant treatment or in retroperitoneal sarcomas where no attempt at resection was made. Fourteen percent (14%) of the patients were treated with enucleation, 22.3% wide excision, 7% with amputation, 11.8% with radiotherapy alone, 18% with surgery and radiotherapy, 2.3% radiotherapy and chemotherapy alone, 1.5% chemotherapy alone, 2.3% surgery and chemotherapy and 3.5% a combination of chemotherapy, radiotherapy and surgery. No guide lines are given as to why these modalities of treatment were chosen and it was noted that adjuvant therapy was instituted havehazardly. It is an established fact that enucleation has no role at all in management of sarcomas. It might have been an error of inexperience while in developed countries there are well established cancer centres with sarcoma management teams, this is not the case locally, overbooked radiotherapy facilities at Kenyatta National Hospital (being the only public functional radiotherapy unit) and the prohibiting cost of chemotherapy drugs might also have affected the administration of this treatment.

In management of extremity sarcomas the trends are shifting from amputation to limb sparing operations which are minimising resection and instituting radiotherapy either in the form of Brachytherapy or external beam radiation. This had reduced amputation rates from 50% in the 1960s to 5% currently. Shiu Man H. and coworkers at Memorial Sloan Kettering Cancer Centre

obtained local control in 70% of patients in their study and 5 year disease free state of 60% after limb function preserving surgery and Brachytherapy [42].

Good results have also been obtained with external beam radiation but with more complications such as fibrosis of intact muscle group [40]. Intra-operative radiotherapy has also been used in western countries.

Several studies have been carried out to investigate the effect of radiation alone on control of sarcomas. Joel E. Tepper et al [43] found that although neither local control nor the long term survival are very good when radiation alone is utilised, but the 33% actuarial local control and the 25% 5 year survival indicated that such a treatment is worth delivering as a substantial core can result. McNeer et al and Windeyen et al found similarly encouraging results [37,38]. No local studies available to date on this subject matter.

Chemotherapy has been found to have impressive benefits in treatment of rhabdomyosarcoma and Ewing sarcoma where 5 year disease free survival rates have increased from 10-25% to 50-70% [44]. It is clear in our series that no clear treatment protocols exist locally and the treatment given depends on the individual doctors. Most of the time radiotherapy was given after failure of initial resection treatment.

In our series enucleation, chemotherapy alone, radiotherapy alone or radiotherapy and chemotherapy alone had poor results with 2 year recurrence rate of 50%, 98%, 70% and 50% respectively, with mortality rates of 25%, 80%, 20% and 44% respectively within 2 years. Wide excision and amputation and when adjuvant therapy was offered produced the best results, with 37% recurrence free rate at 2 years. 53% had recurrence within 2 years while mortality within same period was 10%, while amputation had 100% disease free survival at the end of 2 years. The high recurrence rate with wide excision of 53% could be explained by the fact that most patients had high grade tumours with big size at presentation. Again this study did not consider the tumours at different sites separately.

With regard to site, visceral and retroperitoneal tumours had the worst prognosis with 83% of the patients dead within 2 years, while extremity sarcomas had the best prognosis with 60% getting greater than 2 years recurrence free period and only 20% dead within 2 years. These findings correlate well with western series, but our visceral and retroperitoneal sarcomas show a worse outcome. The approach in western set up is more aggressive with resections being more extensive at time even involving some organs encroached on by the sarcomas [1,45,46]. Thirty two percent (32%) with high grade sarcomas had recurrence (local) within 2 years, 27% had a recurrence free period greater than 2 years and a mortality of 41% within 2 years as compared to low grade

sarcomas where 61% had a recurrence free period greater than 2 years, 23% had recurrence within 2 years and mortality of 7.6% was obtained within 2 years. The difference in the outcome was statistically significant (P = 0.02). Costa J. et al and Trojan M. et al and Mayre Jensen et al in different series showed grade of sarcoma to be an important prognostic factor which is in agreement with our series [20,22,23].

In evaluation of size and prognosis 100% of patients with sarcomas less than 5cm had no recurrence by the end of 2 years, while those greater than 5cm had a 33% recurrence rate within 2 years, 44% recurrence free rate by end of 2 years and mortality rate of 22% within the same period. Statistically no significant difference was obtained)P = 0.34). These result might have been affected by the small number of patients.

By and large our approach to the management of these neoplasms is wanting and there is need to focus our attention to them in order to come up with clearly set down protocols to their approach. Collaboration between surgeons, pathologists, oncologists, physiotherapists and nurses should be put into place in order to have a comprehensive multidisplinary and multimodality care of these patients.

Conclusions

1. Soft tissue sarcomas are a relatively rare group of neoplasms accounting for 3.8% of all malignancies at Kenyatta National Hospital.

2. Painless swelling is the most common presenting complain leading to late presentation at Kenyatta National Hospital.

3. Although overall there is no sex preponderance to these tumours, MFH and dermatofibrosarcoma are more common in males than in famales.

4. At Kenyatta National Hospital, these neoplasms are more common in the younger people. Peak age being first decade of life.

5. Below 30 years of age rhabdomyosarcoma and fibrosarcoma are the commonest sarcomas with rhabdomyosarcoma taking up 54% of sarcomas below 20 years of age.

6. The commonest 7 soft tissue sarcomas in order of frequency age, rhabdomyosarcoma, fibrosarcoma, liposarcoma, leiomyosarcoma, dermatofibrosarcoma and malignant fibrous histiocytoma which is no different from western world.

7. Commonest site in the body is the extremities as for other part of the world, but there is less of retroperitoneal and visceral sarcomas locally.

8. Just as for the western world, myogenic sarcomas (rhabdomyosarcoma to be precise), fibrosarcoma, liposarcoma, dermatofibrosarcoma and MFH are the commonest extremity and trunk sarcomas, liposarcoma the commonest retroperitoneal sarcoma while leiomyosarcoma is the commonest visceral sarcoma, while rhabdomyosarcoma and fibrosarcoma are the commonest in the head and neck.

9. Majority of the sarcomas are not graded by our pathologists which is a shortcoming as grade is an important outcome determinant.

10. Incisional biopsy is the most utilised method due to late presentation and lack of core needle biopsy needles.

11. Surgery is rarely applied in the management of sarcomas of the retroperitoneal which explains our poor treatment results, our approach is more conservative as compared to the western world.

12. Grade, size and site of tumours are important factors in determining prognosis.

13. Lack of facilities make adjuvant therapy inadequate in our set up, hence inappropriate and delayed treatment is normally given.

14. Wrong surgical management (enucleation) is used in a significant proportion of patients.

15. There is no clear policy and protocols and approach to these neoplasms locally at Kenyatta National Hospital.

16. Collection and analysis of data is occasionally made difficult by incomplete records and untraceable files.

Recommendations

1. In view of the relative rarity of these neoplasms and the painless nature of the initial lesion and hence late presentation, awareness of both community and medical fraternity should be increased so that early detection and treatment is instituted.

2. All lumps in the body should be investigated with urgency.

3. Temptation to enucleate these neoplasms should be discouraged at all costs in order to improve on our results.

4. Trucut/core needle biopsy needles should be made available to the clinicians.

5. Pathologist should be reminded to always grade the tumours.

6. Multidisciplinary approach to the management of these neoplasms is recommended.

7. There is a need to be more aggressive in our approach to the surgical management of these tumours with a bigger emphasis on retroperitoneal and visceral tumours.

8. Although facilities are lacking a more prompt and adequate adjuvant therapy of these neoplasms should be put in place.

9. There is a need to improve on our record keeping especially easier retrieval, and clinicians should remember to document all the necessary biodata and findings.

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APPENDIX I

Data collection format.

1. Patients serial number_____ IPNO._____

2. Age

   0-10

   11-20

   21-30

   31-40

   41-50

   51-60

   61-70

   > 70

3. Sex

4. Clinical presentation

   -Painless swelling

   -Painful swelling

   -Ulcerate mass

   -Loss of function

   -Abdominal mass

   -Abdominal pain

   -Metastasis

   -Others (specify)

5. Size of the tumour where given

   < 5cm in diameter

   5-10cm in diameter

   > 10cm in diameter

6. Grade

High grade

Intermediate

Low grade

7. Site

Lower limbs

Upper limbs

Trunk

Head and Neck

Retroperitoneal

Visceral

Others (specify)

8. Diagnostic method

FNAC

Core needle biopsy

Incisional biopsy

Excisional biopsy

Imaging:

Soft tissue X-ray

C&R

Ultrasound

CT scan

9. Histological type

Dermatofibrosarcoma

MFH

Fibrosarcoma

Leiomyosarcoma

Rhabdomyosarcoma

Haemangiosarcoma

Lymphangiosarcoma

Haemongiopericytoma

Synovial sarcoma

Malignant schwanoma

Primitive neuroectodermal tumour

Malignant mesenchynoma

Alueolar of soft part sarcoma

Spindle cell sarcoma

Neurofibrosarcoma

Desmoid

Pleomorphic sarcoma

Others

10. Treatment

Surgery alone

Enucleation

Wide excision

Radical excision

Amputation

Surgery + radiotherapy

Surgery + radiotherapy + chemotherapy

Radiotherapy alone

Radiotherapy + chemotherapy

Chemotherapy alone

Symptomatic treatment

11. **Result of treatment**

Recurrence

Greater than 2 years of disease free period

Less than 2 years of local recurrence or metastasis free period

Dead within 2 years from the disease.

No change in disease process