

**HISTOPATHOLOGICAL STUDY OF MALIGNANT SOFT TISSUE TUMORS**Vinitha Samartha<sup>1</sup>, Shreya Hegde<sup>2</sup>, Zulfikar Ahmed<sup>3</sup>, Umaru N<sup>4</sup>**HOW TO CITE THIS ARTICLE:**

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**ABSTRACT: BACKGROUND:** Soft tissue tumors are a highly heterogenous group of tumors that are classified on a histogenetic basis. They are broadly divided into benign and malignant types. Benign tumors far outnumber the malignant ones. **AIM:** To know the incidence in different age groups, sex incidence, site specific distribution and their grading wherever possible. **MATERIALS AND METHODS:** Thirty five soft tissue tumors were received in the Department of Pathology of a medical college hospital, private nursing homes and hospitals in and around Mangalore. All specimens were received in 10% formalin along with a requisition letter which contained information about the name, age, sex, occupation, present and past history, relevant radiological and laboratory investigation results of the concerned patients. The specimens were then subjected to gross description and appropriate tissue sections were given. The microscopic features were studied with routine haematoxylin and eosin stained paraffin sections and were classified according to the New WHO Classification of malignant soft tissue tumors. **RESULTS:** Out of the 35 cases, 33 adults and 2 infants of soft tissue tumors studied, 19 were males and 16 were females. The most common age group of presentation was 51-60 years. Most of the tumors presented between 0-10cms in size. Liposarcoma and malignant fibrous histiocytoma were the commonest tumors diagnosed among the 35 cases, comprising 7 cases each. **CONCLUSION:** Malignant soft tissue tumors are rare tumors that occur anywhere in the body, but most commonly present in the lower extremities. They occur in adults usually. However rhabdomyosarcoma are common among the children. Malignant soft tissue tumors most commonly occur in the extremities and histopathological study is one of the important diagnostic methods. Ancillary techniques should be judiciously used wherever necessary.

**KEYWORDS:** Malignant soft tissue tumors, Histopathology.

**INTRODUCTION:** Soft tissue is defined as a complex of non-epithelial extraskeletal structures of the body exclusive of the reticuloendothelial system, glia and supportive tissue of the various parenchymal organs. They are broadly divided into benign and malignant types Benign tumours far outnumber the malignant ones.<sup>1</sup> Soft tissue sarcomas, compared with carcinomas and other neoplasms are relatively rare and constitute fewer than 1% of all cancer.<sup>2</sup>

They present most commonly as an asymptomatic mass originating in an extremity but can occur anywhere in the body, particularly the trunk, retroperitoneum, or the head and neck.<sup>3</sup>

For any large soft tissue tumour in which the possibility of malignancy exists, the proper initial diagnostic procedure is to obtain material through biopsy or fine needle aspiration.

Microscopic evaluation of Hematoxylin and Eosin stained sections remains the standard technique for the diagnosis of these tumours and is sufficient in the majority of cases. However, there are special techniques that have been successfully applied to increase diagnostic accuracy; these include conventional special stains, electron microscopy, immunohistochemistry, cytogenetics and molecular methods. After the tumour has been accurately classified histopathologically and with the help of ancillary techniques, it can be treated properly.<sup>4</sup>

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**MATERIALS AND METHODS:** The materials for the present study were soft tissue sarcoma specimens received at Department of Pathology of a medical college hospital, private nursing homes and hospitals in and around Mangalore between June 2009 to May 2011. All specimens were received in 10% formalin and relevant clinical information was obtained. The specimen were then subjected to gross description and adequate sampling by appropriate tissue section. The microscopic features were then studied with routine Hematoxylin and Eosin stained sections.

**RESULTS:** The present study is a prospective study done for a period of 2 years from June 2009 to May 2011. Clinical details and investigations were recorded. Out of the 35 cases studied, 4 were received as incisional biopsies, the rest of them being excisional biopsies. Gross and microscopic features were studied in detail and the findings were then analysed.

**HISTOLOGICAL TYPES OF SOFT TISSUE SARCOMAS:** The histological types of malignant soft tissue tumours encountered in our study as per the main heads of the WHO Classification are shown in Figure 1.

Malignant Fibrous Histiocytoma and Liposarcoma were the commonest tumours consisting 7 each out of 35 cases. The least common were Clear Cell Sarcoma, Leiomyosarcoma, Synovial Sarcoma, Malignant Hemangiopericytoma, Alveolar Soft Part Sarcoma and Extraskeletal Osteosarcoma (One case each)

Most of the cases were seen in the age group of 51-60 years. The lowest age in which tumour found was a 25 day old baby girl who was diagnosed of Infantile Fibrosarcoma. The oldest individual was a 70 year old man who was diagnosed to have Malignant Fibrous Histiocytoma. In the study, we found that males had higher incidence of tumours than females.

Among the various sites, lower extremity was the commonest site, followed by the upper extremity and posterior chest wall. The head and neck region were relatively rare sites.

**MODE OF PRESENTATION OF CLINICAL FINDINGS:** Incidence of clinical findings among the 35 patients are shown in Table 2. Among the gross findings, on cut surface the tumours were solid in most of the cases, while cystic presentation was very uncommon.

**FNLCC (FEDERATION NATIONALE DES CENTERS DE LUTTELE CANCER SARCOMA) GRADING:** Grading by FNLCC showed a high incidence of Grade 1 tumours in malignant fibrous histiocytoma and liposarcoma. When compared with all the other tumours in the present study, Grade 2 tumours were the commonest.

**LIPOSARCOMA:** This was one of the commonest soft tissue sarcoma in our study. The age range of the patients with this tumour were 28-52 years. In the present study this tumour was more common in females. Histopathologically most of the tumours were of myxoid type (3 cases), followed by well differentiated (2 cases), dedifferentiated (1 case) and round cell liposarcoma (1 case). Tumours belonging to Grade 1 were 4 cases, Grade 2 were 2 cases and Grade 3 was 1 case.

**FIBROSARCOMA:** In our study we had 4 cases of fibrosarcoma out of which 3 were females and 1 was a male patient. The age range they belonged to was from 25 days to 60 years. The 25 day old baby

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girl was diagnosed to have infantile fibrosarcoma. The rest of the cases were diagnosed to be Conventional Fibrosarcoma. 2 of the cases were Grade 2 and the remaining 2 cases were Grade 1.

**MALIGNANT FIBROUS HISTIOCYTOMA:** Malignant Fibrous Histiocytoma was the other commonest tumour in the study. Among the 7 cases only 1 case was a female while the remaining 6 were males. The age range was between (43- 70 years) with the commonest location being the right lower extremity and the upper back having equal with incidence of 3 cases each. Out of the 7 cases 4 cases belonged to Pleomorphic type, while 3 cases belonged to myxoid type of MFH histopathologically. Majority of the cases were of Grade 1 (4 cases), followed by Grade 3 (2 cases ) and Grade 2 (1 case).

**LEIOMYOSARCOMA:** We had 1 case of Leiomyosarcoma, who was a female patient aged 30 years. She presented with a mass in the perineal region which was a Grade 2 Leiomyosarcoma.

**MALIGNANT HEMANGIOPERICYTOMA:** 1 case of malignant hemangiopericytoma was reported, which was a 35 year old female. She presented with a mass on the nape of the neck and it was a Grade 1 tumour.

**RHABDOMYOSARCOMA:** Out of the 35 cases, 3 cases were reported as Rhabdomyosarcoma. The age range was from 3 months to 56 years. The most common location was the forearm. 1 case presented in the left extraorbital region. We had one case each of Embryonal, Alveolar and Pleomorphic Rhabdomyosarcoma. 2 cases were of Grade 2, while 1 case was of Grade 3.

**EXTRASKELETAL CHONDROSARCOMA:** 2 cases of Extraskeletal chondrosarcoma were reported. Both the patients were males in the age range of 42 – 55 years. Both the patients presented with a swelling on the right lower extremity. Histopathologically both of them were myxoid chondrosarcomas and belonged to Grade 1 and 2 tumours according to the FNLCC grading system.

**EXTRASKELETAL OSTEOSARCOMA:** 1 case of extraskeletal osteosarcoma was reported. The patient was a 14 year old female who presented with a swelling on the right leg. The ESOS was a grade 2 tumour.

**SYNOVIAL SARCOMA:** 1 case of Synovial Sarcoma was diagnosed who was a 45 year old female patient. The patient presented with a swelling on the right foot. The tumour belonged to Grade 2.

**ALVEOLAR SOFT PART SARCOMA:** 1 case of Alveolar Soft Part Sarcoma was diagnosed. The patient was a 33 year old female who presented with a swelling in the left leg which was a Grade 2 tumour.

**CLEAR CELL SARCOMA:** In the present study, we came across a 56 year female patient with complaints of swelling in the right forearm. She was diagnosed of Clear Cell Sarcoma which was Grade 2.

**PNET/EXTRASKELETAL EWING'S SARCOMA:** 4 cases were reported. Among the 4 cases, 3 were males and 1 was a female patient. The patients ranged in their age from (26 – 51 years). All cases

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were located on the left side. 2 cases were on the lower extremity, followed by 1 case on forearm and 1 case on shoulder region. 2 of the cases belonged to Grade 2, while remaining 2 cases belonged to Grade 3.

**MALIGNANT PERIPHERAL NERVE SHEATH TUMOUR:** 2 cases were reported, 1 male patient(35 years), 1 female patient(36years). Both the cases presented on the right thigh and belonged to Grade 2.

**DISCUSSION:** A total of 35 cases of malignant soft tissue sarcomas were studied. Out of these 35 cases, the most common tumours were liposarcoma and malignant fibrous histiocytoma (comprising 7 cases each). The incidence of various malignant soft tissue tumours were compared with the studies by Tsujimoto et al<sup>1</sup> and by Rosenthal et al.<sup>2</sup> Malignant fibrous histiocytoma was the commonest tumour in these studies. Synovial Sarcoma was a common tumour in the study by Tsujimoto et al<sup>1</sup> as compared to its rarity in the other studies.

**AGE INCIDENCE:** The age incidence of the tumours in our study were compared with other studies. In the study by El- Jabbour et al,<sup>3</sup> the most common age incidence of tumours was between 60–69 years, while in that of Tsujimoto et al,<sup>1</sup> it was between 30–39 years. In our study we found that most of the tumours occurred in the the fourth and the fifth decade with an equal incidence between 40–49 years and 50–59 years of age.

**INCIDENCE OF ANATOMIC SITES:** The incidence of tumours at various sites in the present study were compared with the studies by Potter et al,<sup>4</sup> Buhari et al<sup>5</sup> and is shown in Table 3. The studies are similar to each other with lower extremity being the most common site. The least common site was head and neck, in 2 studies, namely, Potter et al and the present study. However, in Buhari et al,<sup>5</sup> the retroperitoneum was the least common site of occurrence of tumour.

**LIPOSARCOMA:** The present study was compared with Harry et al.<sup>6</sup> The 2 studies showed similarity with the occurrence of histopathological types of liposarcoma. Myxoid liposarcoma is the commonest liposarcoma while pleomorphic liposarcoma being the rarest.

**RHABDOMYOSARCOMA:** Comparison of rhabdomyosarcoma in the present study was done with other studies namely, Ulutin et al<sup>8</sup> and Hawkin's et al.<sup>9</sup> The studies show a male predominance. In Ulutin et al and Hawkin's et al the most common site of occurrence of tumour was the trunk. However in the present study the most common occurrence was in the extremities. In the studies by Ulutin et al and Hawkin's et al, embryonal rhabdomyosarcoma was the commonest type.

**MALIGNANT PERIPHERAL NERVE SHEATH TUMOUR (MPNST):** The present study had 2 cases of MPNST. They were compared with the studies done by Gabhane et al<sup>10</sup> and Nthumba et al<sup>11</sup> and shown in Table 4. In the present study and the study done by Gabhane et al,<sup>10</sup> lower extremity was the most common site. In the study done by Nthumba et al, trunk was the most common site. As compared with that of Gabhane et al<sup>10</sup> and Nthumba et al,<sup>11</sup> Nthumba et al<sup>11</sup> had a higher incidence of association of MPNST with neurofibromatosis.

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**CONCLUSION:** Malignant soft tissue tumours are rare tumours of mesenchymal origin. They are predominantly a tumour of adulthood with rhabdomyosarcoma being more common among children. Soft tissue sarcomas predominate in males. Most of the tumours present only as a swelling. Soft tissue sarcomas can occur anywhere in the body, but most of them originate in the extremities, trunk, retroperitoneum or the head and neck. Liposarcomas and malignant fibrous histiocytomas are among the common histological types of soft tissue sarcoma.

The rare histologic types are alveolar soft part sarcoma, malignant hemangiopericytoma, extraskeletal osteosarcoma, leiomyosarcoma and synovial sarcoma. Microscopic evaluation is most useful for the diagnosis of these tumours and is sufficient in the majority of cases.

Histologic grading is an important prognostic factor and indicator of metastatic risk in adult soft tissue sarcomas.

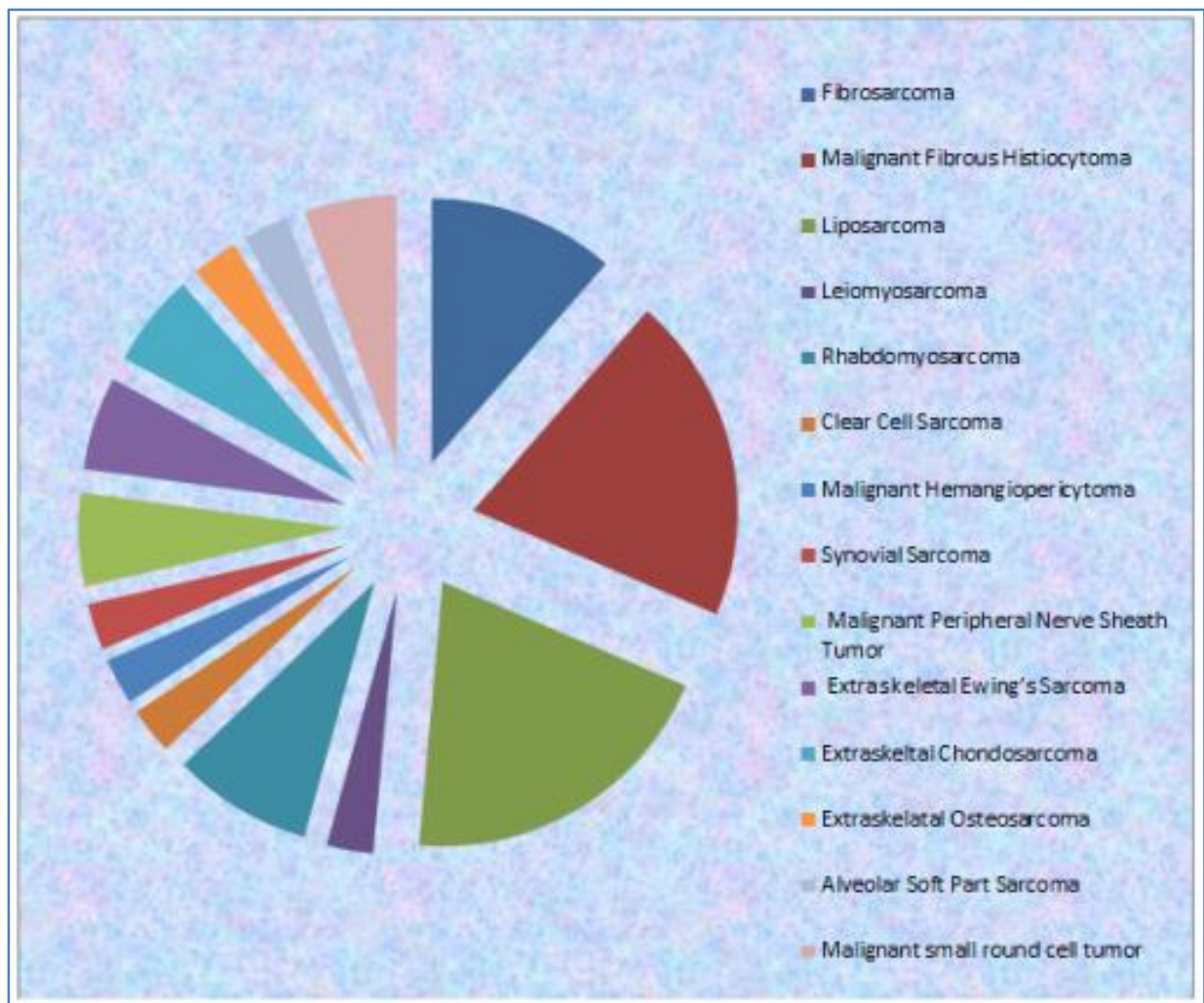
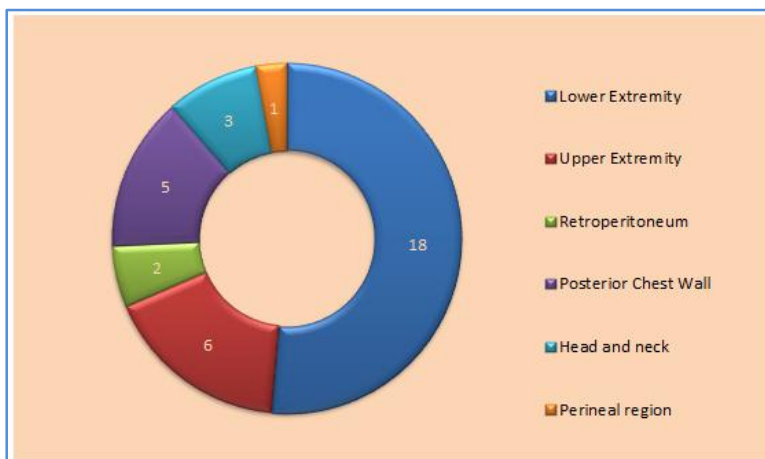
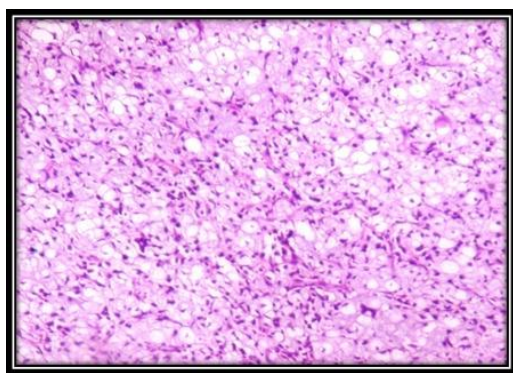


Fig. 1

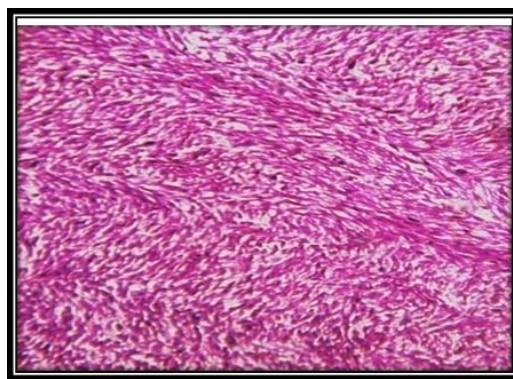
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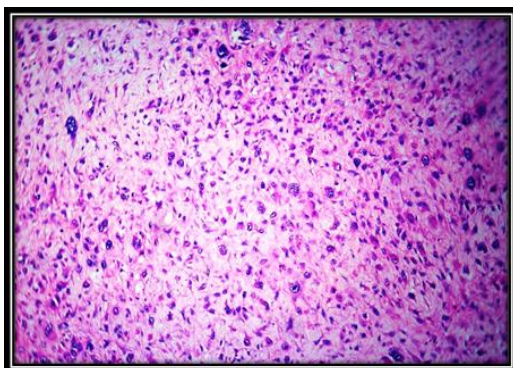
**Fig. 2: Site Distribution**



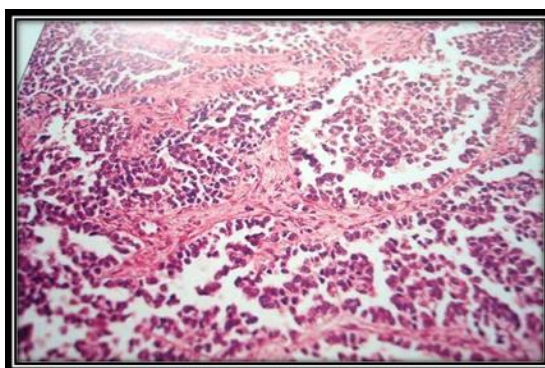
**Fig. 3: Myxoid Liposarcoma showing 'Chicken wire' pattern (H & E, 40 X)**



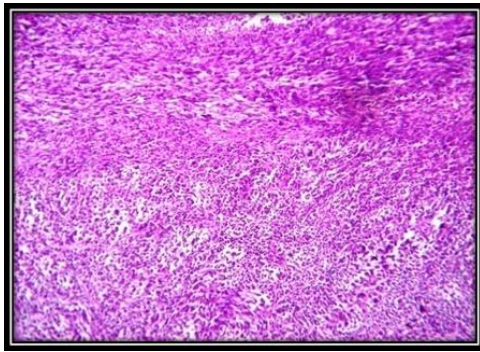
**Fig. 4: Herringbone pattern seen in Conventional Fibrosarcoma (H & E, 40x)**



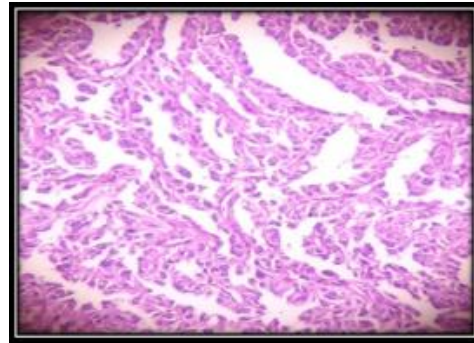
**Fig. 5: Histopathological picture of undifferentiated high grade pleomorphic sarcoma/ Pleomorphic malignant fibrous histiocytoma showing pleomorphic cells.(H&E, 40X)**



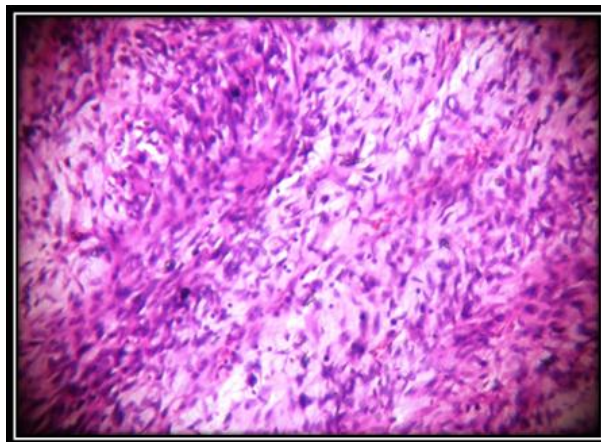
**Fig. 6: Histopathological picture of alveolar rhabdomyosarcoma. (H & E, 60 X)**



**Fig. 7: Histopathological picture of Synovial Sarcoma (Biphasic pattern)**



**Fig. 8: Histopathological picture of alveolar soft part sarcoma (H & E, 60 X)**



**Fig. 9: Histopathological picture of malignant peripheral nerve sheath tumour showing hypocellular as well as hypercellular areas. (H & E, 40 x)**

| Age Group   | Number |
|-------------|--------|
| < 1 year    | 2      |
| 1-10 years  | 0      |
| 11-20 years | 3      |
| 21-30 years | 3      |
| 31-40 years | 6      |
| 41-50 years | 9      |
| 51-60 years | 10     |
| 61-70 years | 2      |

**Table 1: Age Distribution**

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| Clinical Findings | No. of Cases |
|-------------------|--------------|
| Swelling          | 33           |
| Pain              | 11           |
| Trauma            | 3            |
| Recurrence        | 1            |

**Table 2: Incidence of Clinical Findings**

| Author                    | Lower Extremity | Upper Extremity | Head and Neck | Trunk     | Retroperitoneum | Total |
|---------------------------|-----------------|-----------------|---------------|-----------|-----------------|-------|
| Potter et al <sup>4</sup> | 152(49.5%)      | 59(19.2%)       | 12(3.9%)      | 48(15.6%) | 36(11.7%)       | 307   |
| Buhari et al <sup>5</sup> | 80(41.6%)       | 19(9.8%)        | 27(14%)       | 61(31.7%) | 5(2.6%)         | 192   |
| Present study             | 18(51.42%)      | 9(25.71%)       | 1(2.8%)       | 4(11.42%) | 2(5.7%)         | 35    |

**Table 3: Incidence of anatomic sites in soft tissue sarcomas**

| Tumour Characteristics                             | Gabhane et al (n=8) <sup>10</sup>                     | Nthumba et al (n=31) <sup>11</sup>                    | Present Study(n=2)                              |
|--|---|---|---|
| Location in descending order of frequency          | 1.Lower Extremities<br>2.Upper Extremities<br>3.Trunk | 1.Trunk<br>2.Lower Extremities<br>3.Upper Extremities | Lower Extremities                               |
| Clinical Features in descending order of frequency | 1.Mass lesion<br>2.Pain<br>3.Neurological deficit     | 1.Mass lesion<br>2.Pain<br>3.Neurologic deficit       | 1.Mass lesion<br>2.Pain<br>3.Neurologic deficit |
| Association with Neurofibromatosis 1               | 2 cases (25%)   | 15 cases (48.3%)                                      | None  |

**Table 4: Malignant Peripheral Nerve Sheath Tumour as compared with that of Gabhane et al<sup>10</sup> and Nthumba et al<sup>11</sup>**

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