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# Commonest modes of presentation of bone and soft tissue tumors at sarcoma services of A Tertiary Care Hospital of KPK.

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**ABSTRACT… Objectives:** The aim of this study is to assess the commonest mode of presentation at our sarcoma unit and the duration from onset of symptoms till presentation. **Study Design:** Retrospectively study. **Setting:** Sarcoma Unit of Hayatabad Medical Complex, Peshawar. **Period:** June 2016 to December 2019. **Material & Methods:** Two hundred and eighty two patients were included in this study of which 172 patients were male and 110 were female. Sample size was calculated through non-probability consecutive sampling technique. **Results:** Average age of patients presenting to us was 30.2years (±18.5) ranging from 3years to 83years. Highest numbers of patients fell into the age group category of 11-20years i.e. 94 (33.33%). The average duration of symptoms among our patients was 19.6 months (±26.09) ranging from 0.1months (3 days) to 168 months (14years). Highest number of patients fell into the 7months-12months category i.e. 86 patients (30.5%). The commonest mode of presentation among our patients was painful lump i.e. 128(45.3%) while patients presenting with painless lump were 57(20.2%) and those presenting with only pain were 28(9.9%). **Conclusion:** The study concluded that there is a need of greater awareness among general population and health care professionals regarding red flag signs and symptoms in order to avoid the delays in diagnosis which may lead to poor prognosis.

**Key words:** Bone Sarcoma, Delayed Diagnosis, Early Detection, Sarcoma, Soft Tissue Sarcoma, Sign and Symptoms.

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## INTRODUCTION

Bone and soft tissue sarcomas are a heterogeneous group of tumors originating from the mesenchymal germ layer.[1] As they are a rare group of tumors their early recognition is very important as they are often missed.[2] For early detection, clinicians should have background knowledge about the worrying features associated with these tumors. Alarming symptoms for bone tumors include unexplained bone pains/tenderness and/or a limp. Some patients may even present with a pathological fracture.[3] As we do not have a sarcoma registry in Pakistan, it is not possible at this stage to mention the incidence of sarcomas in Pakistan. However, in the UK, almost 2700 new cases are diagnosed every year accounting for 1% of all the cancers.[3]

It is well documented that the key to better outcomes is early diagnosis and prompt treatment by sarcoma surgeons.[4] However, patients present late due to numerous reasons including lack of awareness both in public and medical professionals and the absence of dedicated sarcoma units. According to the National Institute for Health and Clinical Excellence (NICE) guideline (2005) urgent referral should be sought after for a patient having a soft tissue mass with any of the following features; size >5cm, lump increasing in size, deep to deep fascia, painful, recurrent lesion after previous excision.[5]

Health care providers having a strong suspicion of bone sarcoma with or without radiological evidence should also refer their patients to a specialist center. The Department of Health further suggests that any suspected case of cancer should be referred to a respective cancer

specialist within two weeks.[6] Many studies suggest that these guidelines are not being followed and unfortunately patients present very late to a concerned health care provider.[7] A study done in 2010 states that only 15% of the cancer patients are referred within the two weeks of first presentation.[8] A large number of patients are still presenting late to a cancer specialist, with the average delay of almost 14months.[9]

This delay has significant effects on the outcome of management. In particular, the size of the tumor has a direct relationship with the outcome of these patients including resection and long-term survivorship.[10] This delay can also have disastrous effects on the mental well-being of the patient. It has been reported in a recent study in US that the patients with bone and soft tissue sarcoma have higher incidence of suicide among them, especially those that have sarcoma of the vertebral column and pelvis.[11]

The main purpose of our study was to determine the commonest symptoms our patient presents with to our institute and the average time these patients have been suffering with their symptoms before they presented to a concerned health-care provider.

## MATERIAL & METHODS

Data was collected retrospectively from a prospectively held database which has been in place since June 2016 in the primary institute.

After collecting the demographic data each patient's data was analyzed for commonest modes of presentations and duration of symptoms before presentation. The diagnosis was confirmed by histopathology after local and systemic staging.

The data was further examined using excel sheets to find out the commonest symptoms with which patients presented to the primary institute and the duration it took for them to present to us.

### Inclusion Criteria

All patients diagnosed with bone or soft tissue sarcoma.

### Exclusion Criteria

Tumors that are ectodermal in origin.

## RESULTS

Two hundred and eighty two patients were included in this study at the time of data capture of which males were 172 (60.9%) and 110 (39.2%) were female. Average age of patients presenting to us was 30.2years (±18.5) ranging from 3years to 83years. On sub-group analysis, the highest numbers of patients fell into the age group category of 11-20years i.e. 94 (33.33%) and 31-50 years i.e. 68 (24%) (Figure-1).

Out of collected data, 111 (39.3%) were reported to have a soft tissue sarcoma and 171 (60.6%) had a sarcoma related to bony origin.

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The average duration of symptoms among our patients was 19.6 months (±26.09) ranging from 0.1months (3 days) to 168 months (14years). On sub-group analysis, the highest number of patients fell into the 7 months-12 months category i.e.86 patients (30.5%) (Figure-2).

The commonest mode of presentation among our patients was a painful lump i.e. 128 (45.3%) while patients presenting with painless lump were 57(20.2%) and those presenting with only pain were 58(20.5%). Patients presenting with pathological fracture were 28 (9.9%). Patients who presented to us with a fungating lesion were 11(4%) (Figure-3).

To further analyze the duration of symptoms before presentation to our unit, we categorized our results according to years from 2016 to 2019. According to our results the average duration of symptoms of the patients presented in 2016 was 25.3 Months. After that we noted a steady decline over the next years (Figure-4). In 2019, the average duration of symptoms came down to 10.09 Months (*p*-value 0.01), which was statistically significant difference.

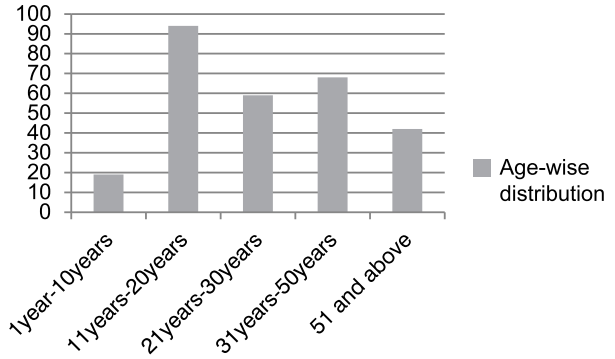


**Figure 1. Age-wise distribution**

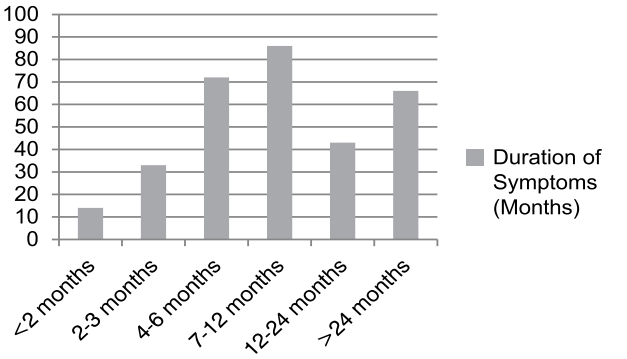


**Figure-2. Duration of symptoms (Months)**

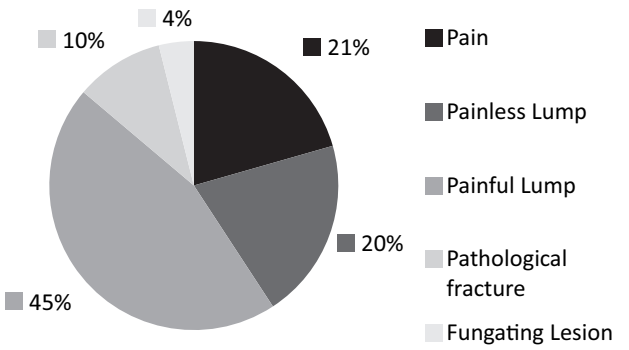


**Figure-3. Modes of presentation**

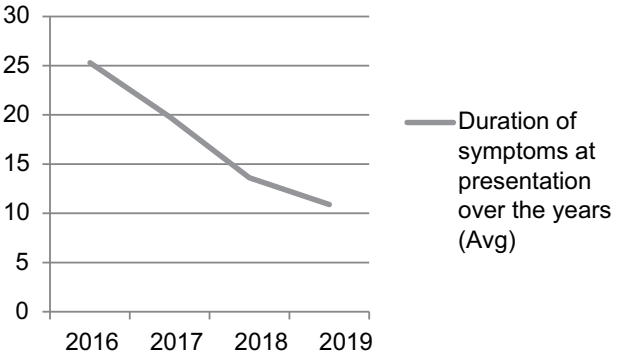


**Figure-4. Duration of symptoms at presentation**

## DISCUSSION

Bone and soft tissue tumors are rare and therefore their early detection can be missed by health care providers.[2] This study describes the pattern of presentations of bone and soft tissue tumors at sarcoma services of a tertiary care hospital in KPK province of Pakistan. Although a painless lump is more prone to be missed and managed conservatively, our study reported a large number of patients presenting with a painful lump i.e. 161 (57%). This may be due to the very large size of tumors that we see in our practice which causes pain due to pressure effects. Deep pain is an alarming feature and if it's worsening it should be explored.[3] According to our research, 74 (26.2%) patients presented with only pain, which after investigating turned out to be a bone sarcoma. Thus we recommend that any deep lasting pain and worsening, in particular, should be thoroughly investigated. Night pain, although, could have multiple causes but a study reported 20% of their bone sarcoma cases presenting with only night pain.[12] A small percentage of our cases presented with a pathological fracture i.e. 25 (6.5%), similar to previous studies suggesting between 5% and 10%.[13,14] This again signifies a high grade aggressive tumor which may have caused enough local destruction leading to fracture or suggesting delay in presentation after onset of symptoms.

Our results point to the fact that most of these patients present very late and this directly affects the therapeutic outcome of their disease course. Johnson *et al*[4] reported a delay of almost 5.7months for soft tissue sarcomas. While Ashwood *et al*[15] reported a delay of almost 7.5months for bone and soft tissue sarcomas. However, our results show the average duration of symptoms reported to be 19.6 months. Out of 282 of our cases 236(83.6%) patients presented to us with delay of as late as 30.1 months. This directly influences their clinical outcome and puts the patients at risk for amputation as compared

to limb salvage surgery and also leading to poor survivorship.

Tumor size is one of the most important prognostic factors for sarcoma outcome.[10] According to the guidelines it has been advised that any swelling bigger than a golf ball (42mm) must be referred for further evaluation.[16] For every centimeter increase at the time of definitive diagnosis, there is an estimated 3-5% reduction in overall survival for STS.[5] In our study, the average size of STS was 13cm. Therefore, it is recommended that any lump having the size of >4cm should be evaluated.[17] For other lumps we recommend that they should be monitored and an increase in size should prompt a referral to a sarcoma service.

Patient and health care professionals' education is of vital importance regarding these rare types of tumors. As our results indicate, our patients are presenting with shorter duration of symptoms now as compared to earlier. We believe that this positive trend is due to patient and clinician awareness and availability of a dedicated sarcoma unit leading to early referrals, early diagnosis and prompt treatment. Referral to a dedicated sarcoma unit would also save the patient from an unplanned excision known as whoops procedure. This happens where the surgeon performs an excision without prior staging, biopsy, diagnosis and without respect to resection margins and then later on finds out that the tumor was a sarcoma.[18] Hence, the term "whoops" procedure.

## CONCLUSION

- We emphasize on educating public and health care providers about potential worrying lesions, particularly those inexperienced surgeons who are likely to carry out a biopsy and or excision of a presumed benign lump(whoops procedure).[18]
- We recommend that urgent referral should be sought for a patient having a soft tissue mass with any of the following features; Size >5cm, increasing in size, deep to deep fascia, painful, recurrent after previous excision.
- Development of dedicated sarcoma centers and national sarcoma registry.
- If a health care provider has any suspicion of a bony or a soft tissue sarcoma, such cases should be referred without further delay to a sarcoma unit under supervision of dedicated teams.



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## AUTHORSHIP AND CONTRIBUTION DECLARATION

| Sr. # | Author(s) Full Name | Contribution to the paper | Author(s) Signature |
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| 2 | Zeeshan Khan | Concept, Data collection, Manuscript. | |
| 3 | Muhammad Saeed | Data collection, Manuscript. | |
| 4 | Israr Ahmad | Data collection, Manuscript. | |
| 5 | Salik Kashif | Data collection, Manuscript. | |
| 6 | Arif Khan | Data collection, Manuscript. | |