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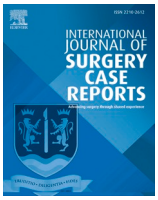

Case series

Case series of high-grade soft tissue sarcoma of the lower limb with delayed diagnosis: Experience at a tertiary hospital in northern Tanzania

Jay Lodhia [a,b,*], Gregory Goodluck [a], Joylene Tendai [a], Ellyagape Urassa [a], Gilbert Nkya [b,c], Alex Mremi [b,c]

[a] Department of General Surgery, Kilimanjaro Christian Medical Centre, P. O Box 3010, Moshi, Tanzania
[b] Faculty of Medicine, Kilimanjaro Christian Medical University College, P. O Box 2240, Moshi, Tanzania
[c] Department of Pathology, Kilimanjaro Christian Medical Centre, P. O Box 3010, Moshi, Tanzania

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ABSTRACT

Introduction and importance: Soft tissue sarcoma is an uncommon form of cancer with poor prognosis. Early diagnosis and treatment are vital for improving the treatment outcomes.
Case presentation: We report a series of high-grade soft tissue sarcomas of the lower extremity with delayed diagnosis to gain insight into the presentation, treatments and outcomes for this rare disease and to determine whether limb-salvage surgery yields reasonable outcomes.
Clinical discussion: Timely health seeking has an impact on the outcome of the treatment of any particular disease. Patient delays are usually socio-economic factors. Soft tissues sarcomas are uncommon malignant tumors that even managed adequately have a poor 5-year survival. Limb salvage becomes questionable especially when patients present late with adverse symptoms.
Conclusion: In this series, we found that patients presented late and this led to unfavorable oncological outcomes, also limb salvage was not an option due to delayed presentation. Thus, early diagnosis is recommended so as to improve treatment outcome.

1. Introduction

An early visit to the health care is sometimes matter of life and death, but the question remains what is late and what is in time. Early health-seeking has an impact on the outcome of treatment related to a particular disease, for example, three months for breast cancer and 2 h for heart attack [1]. Patient's delay is often described as the "length of delay between the onset of signs and symptoms and the patient's first visit to a health care" [1]. Patient's delays are usually associated with socio-demographic factors like age, gender, socioeconomic status or marital status [1]. Herein we present and share our experience from our tertiary centre of three cases of soft tissue sarcoma that presented late hence delayed in diagnosis that impacted limb salvage surgery and unfavorable outcomes.

This work has been reported in line with the PROCESS 2020 criteria [2].

2. Case presentation

2.1. Case 1

A 45-years-old female came in as a referral with the history of mass on the anterior aspect of her the left leg for 7 months with pain and inability to use the limb. This started as a small mass following an animal injury; the mass was excised but recurred 2 months later. The mass was progressive increasing in size associated with bleeding and pus discharge. She reported using herbal medications with no relief. However, on admission she reported of dizziness, awareness of heartbeats and generalized body weakness but denied headaches, fevers, difficulty in breathing or cough. Other systems were unremarkable. No history of chronic illness was reported. She keeps livestock, does not consume alcohol or tobacco.

On examination she was fully alert, moderately pale, afebrile not jaundiced and had inguinal lymphadenopathy. Her axillary temperature was 36.6 °C, blood pressure of 92/85 mmHg, pulse rate of 87 and was

* Corresponding author at: Kilimanjaro Christian Medical Centre, P. O Box 3010, Moshi, Tanzania.
E-mail address: jaylodhia06@gmail.com (J. Lodhia).

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saturating at 98 % in room air. There was an exophytic fungating mass on the anterior aspect of the left leg, measuring approximately 18 by 14 cm. She had precordial hyperactivity with a gallop rhythm. Lab investigations revealed leucocyte of 12.65 × 10^9, hemoglobin of 4.9 g/dl, hematocrit of 17.5 %, ESR of 65 mm/1 h and serum Calcium of 2.07 mmol/L. Her abdominal ultrasound and chest X-ray were normal, X-ray of left tibia and fibula showed proximal boney destruction with cortical reaction (Fig. 1A). She was transfused with blood and she was counseled and scheduled for an emergency transfemoral amputation by the general surgeons (Fig. 2B). She received antibiotics, hematenics, analgesics and physiotherapy. Six days later she was discharged through surgical clinic. Unfortunately she was lost to follow-up. Histopathology of the tumor concluded it to be soft tissue sarcoma, unclassified (Fig. 1C); as Immunohistochemistry tests highlighted positivity for vimentin (Fig. 1D) but negative for Myogenin, Keratin, S100 and melanocytic markers.

2.2. Case 2

A 20-years-old male presented with pain and difficulties in flexing his right knee for 2 years. This started with inability to flex the knee beyond 90°. It was not painful initially but progressively he started feeling severe pain and knee flexion was extremely limited. He denied history of trauma, or a mass or an ulcer formation throughout the course of the illness. Family and social history was unremarkable and past medical history was unremarkable too.

On examination he was clinically stable with stable vitals. Local examination of his right knee had healed traditional marks, non-tender, neuromuscular status was intact. Systemic examination was essentially normal. Histopathology was done which revealed pleomorphic rhabdomyosarcoma (Fig. 2A) with immunopositivity for myoD1 (Fig. 2B). His chest X-ray was unremarkable and X-ray of the femur showed distal femoral cortical thickening with soft tissue extension associated with soft tissue mass (Fig. 2C). CT-scan of the left thigh showed heterogeneous soft tissue mass measuring 2.8 × 7.7 × 10.9 cm in the posterior

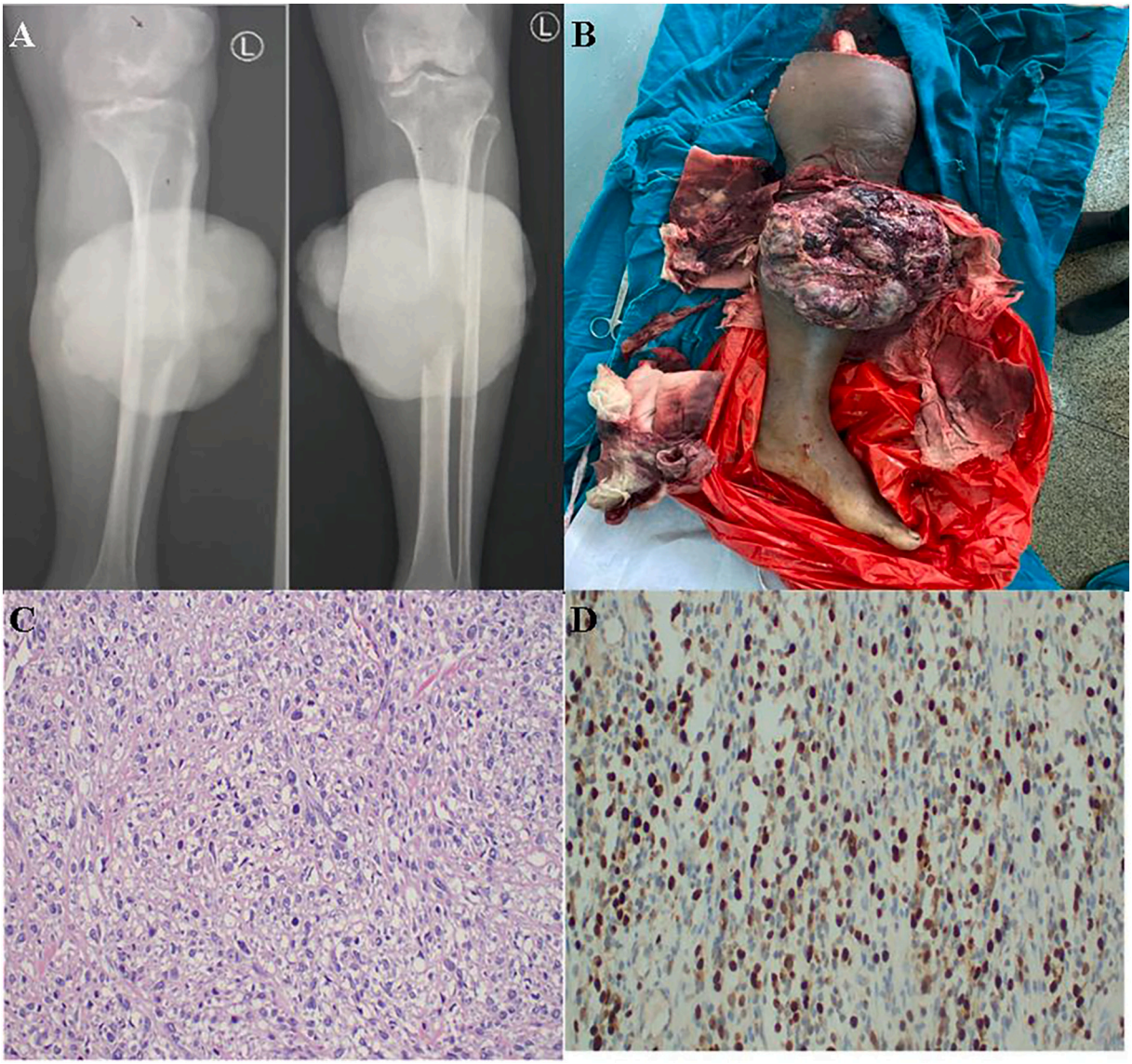

Fig. 1. A. X-ray of left tibia and fibula showing proximal boney destruction with cortical reaction and soft tissue mass.
B. Post transfemoral amputation showing large exophytic mass over the proximal leg.
C. Histopathology of the tumor highlighting diffuse proliferation of large pleomorphic cells ovoid cell with frequent mitoses including bizarre ones; H&E stained sections 100× original magnifications.
D. Immunopositivity of the tumor cells with Vimetin 40× original magnification.

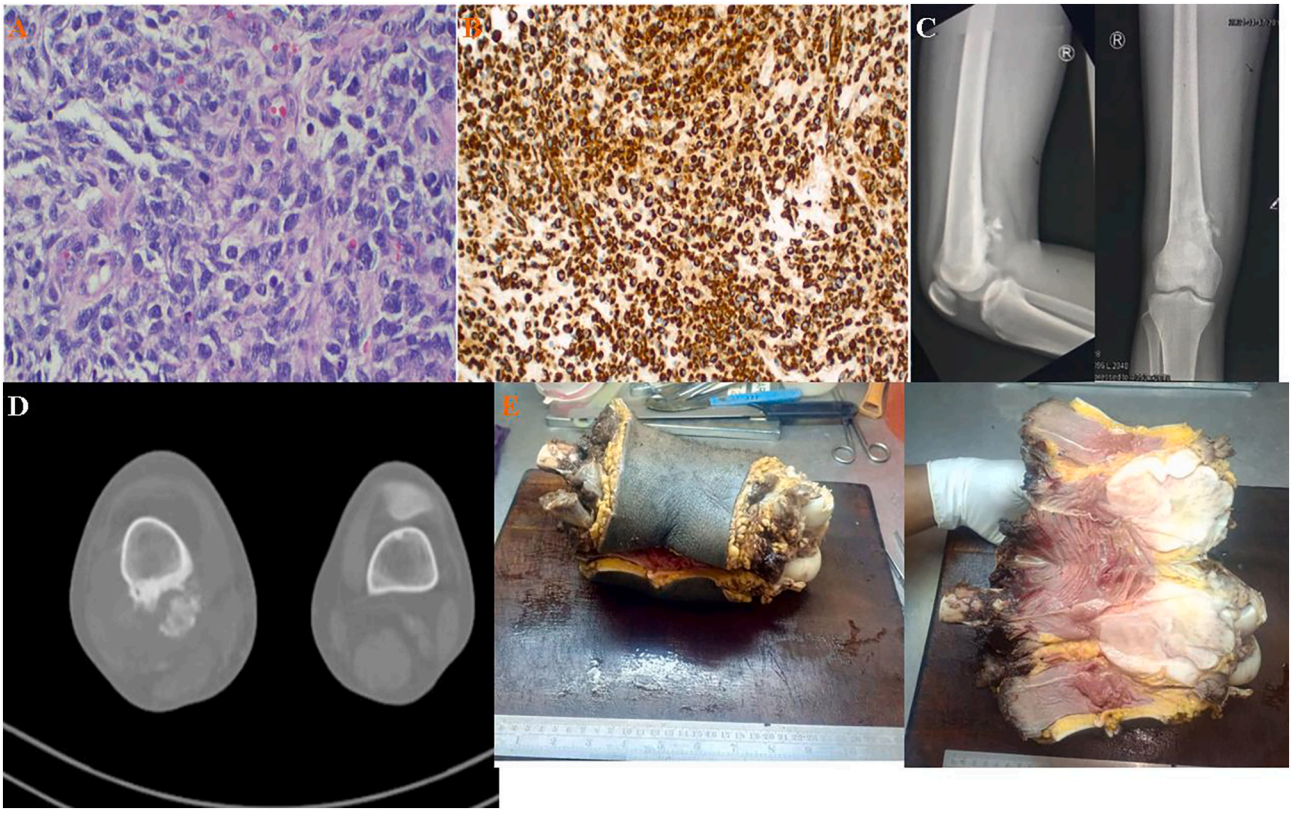

Fig. 2. A. Diffuse proliferation of large pleomorphic cells with rhaboid differentiation; H&E stained sections 200× original magnification.
B. Immunopositivity of the tumor with MyoD1 60× Original magnification.
C. X-ray of the femur showed distal femoral cortical thickening with soft tissue mass.
D. CT-scan showing cortical erosion of the posterior aspect of distal femur and areas of soft tissue calcifications.
E. Photograph showing post transfemoral amputation; lesion on the posterior aspect of distal femur.

distal thigh causing cortical erosion of the posterior aspect of distal femur and areas of soft tissue calcifications (Fig. 2D) and bilateral sub centimeter inguinal lymphadenopathy suggestive of Rhabdomyosarcoma. Transfemoral amputation was performed by the general surgeons (Fig. 2E) and he is currently on adjuvant chemotherapy under oncology care with ongoing occupational therapy.

2.3. Case 3

A 45-year-old female presented to our tertiary facility with a history of progressive left knee swelling for 5 years. Over the course of illness, she had been treated by traditional healers, with the mass being excised three years ago and reportedly recovered but the lesion returned then was treated using herbal medications. The mass continued to increase in size and developed ulcerations discharging foul smelling liquid. This was accompanied with inability to use the limb and carry out her daily activities.

Upon initial examination, she was fully conscious, severely pale, not jaundiced and not cyanotic. On local examination, there was a large cauli-glower like mass on the lateral aspect of the left knee measuring 20 × 15 cm (Fig. 3A). Her systemic examination was unremarkable with no peripheral lymphadenopathy. Lab investigations showed leucocyte count of 16.4 × 10^9/L, severe anemia of 5.1 g/dL, platelet of 319 × 10^9/L, serum creatinine of 43 μmol/L with normal liver enzymes. Abdominal-pelvis ultrasound was normal and left femur X-ray showed soft tissue swelling with no boney involvement (Fig. 3B). She was transfused with six units of whole blood and biopsy of the mass showed atypical oval or spindled shaped cells suggestive of soft tissue undifferentiated sarcoma (Fig. 3C). The tumor was negative for S100.

She consented for transfemoral amputation which was done successfully by general surgeons. Post-operatively she received analgesics antibiotics and wound care then discharged through oncology clinic where she was seen as an outpatient the amputation stump completely healed, there was no evidence of lung metastasis and chest X-ray was clear. She is currently on adjuvant chemotherapy.

3. Discussion

Soft tissue sarcomas are a group of rare malignant tumors and any arise from the limbs of which even managed adequately have a 5-year survival rate of 62–84 % [1]. Many are managed with wide local excision and radiotherapy but a few that are high-grade, proximal, recurrent and involving major neurovascular structures require amputation. Careful assessment and a review at a tertiary centre for exploring treatment options is important before amputation. Nevertheless, amputation is useful palliation procedure for patients with distressing symptoms like pain, bleeding, pain and fungating mass which was in most of the cases in our experience whereby amputation was done for palliation of intractable cancer symptoms [3,4].

For successful treatment of soft tissue sarcoma, early diagnosis is the key [3,4]. Delayed diagnosis of soft tissue sarcoma is common and a number of factors contribute to the delay such as, misdiagnosis as benign tumors because of the presentation. Most of them are painless, mobile, they have slow growth hence both physicians and patients confuse them with benign tumors and appropriate investigations such as biopsies are missed [1]. With regard to our cases we see the patient in cases one and three who had tumor excision without biopsy which was later diagnosed as a soft tissue sarcoma.

Soft tissue sarcomas are best managed by surgical resection with or without radiotherapy. Chemotherapy is mainly reserved for patients

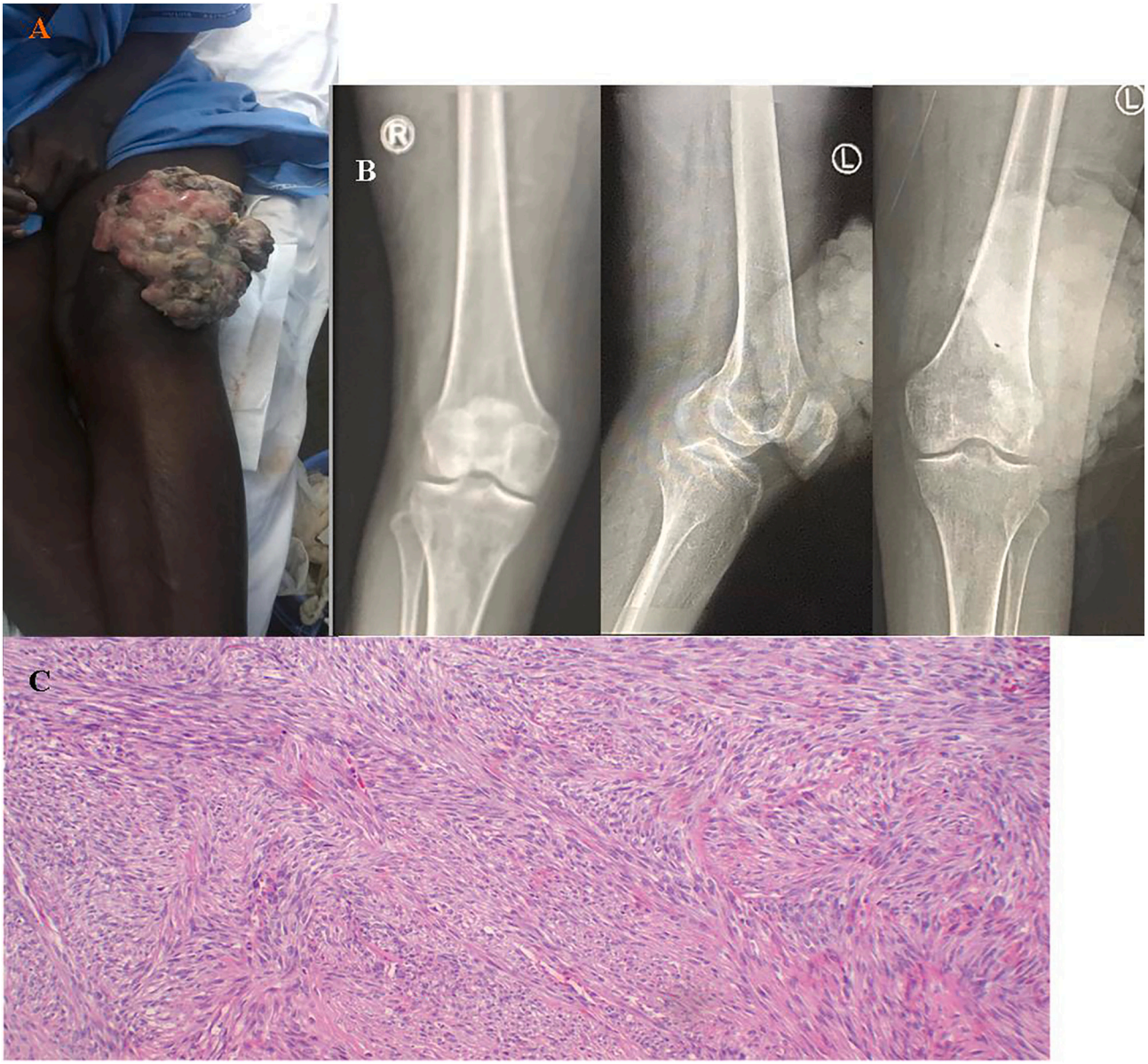

Fig. 3. A. Large cauli-flower like mass over left knee.
B. Distal femur X-ray with soft tissue mass with no boney involvement.
C. Photomicroscopy of the tumor highlighting diffuse tumor made up spindle shaped cells with storiform pattern; H&E 40× original magnification.

who present with metastases or with deep tissue invasion [5,6]. In this series all our patients were managed surgically without radiation therapy and two who showed invasive disease received adjuvant chemotherapy post resection. Radiotherapy has shown to enhance oncologic control of the disease with local recurrence being below 10 % [7].

Limb salvage has been applied as a standard of treatment for most tissue sarcomas for years. Basing on current studies amputations have been as low as 4.1 % [8,9]. For our cases this has been a challenge. For all three cases limb salvage was not successful because of delay in presentation hence making diagnosis. This highlights the importance of educating the general population to improve health-seeking behavior, educate clinicians and importance of early referral for early diagnosis and definitive management. Further cross-sectional study can be done with a larger sample size to establish the factors affecting delayed diagnosis, risk factors of the soft tissue sarcoma and long term outcomes.

4. Conclusion

In the absence of conservative options, major amputations are indicated for the management of advanced tumors with distressing symptoms with effective palliation of debilitating symptoms.

Consent

Written informed consent was obtained from the patients for publication of this case series and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Ethical approval

Ethical approval was obtained from the department of General

surgery, KCMC Hospital.

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Author contribution

- Jay Lodhia – surgeon, conceptualization and writing of the script
- Gregory Goodluck – writing of the script
- Joylene Tendai – Writing of the script and review medical records
- Ellyagape Urassa – reviewed medical records
- Gilbert Nkya – reviewed medical records, Histology analysis
- Alex Mremi – review of medical records, histology slides, editing

Guarantor

Jay Lodhia – Corresponding author.

Research registration

N/A.

Declaration of competing interest

Authors declare no conflict of interests.

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