Spindle cell sarcoma: a case report of diagnostic and therapeutic quandary in a low resource setting

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Abstract

Sarcomas can present differently in different parts of the body and showcase varied histopathological features and tend to recur locally and metastasize to distant sites. We discuss a case of a 37-year-old male with local recurrence of spindle cell sarcoma of the paraspinal muscles of size 20 × 20 cm² with overlying ulceration and discharge with possible pulmonary metastasis. The mass was evaluated using magnetic resonance imaging/computed tomography and the histology was confirmed by biopsy. Wide surgical resection of the mass was done and the patient was referred to another center for radiotherapy and further treatment. The large size of the sarcoma and the possible pulmonary metastasis poses a risk of significant morbidity and mortality in this patient. This case showcases the scenario of many patients in developing countries where the patients are lost to follow-up due to various reasons and present later with grave consequences.

CASE REPORT

We present a case of a 37-year-old man with spindle cell sarcoma of the paraspinal muscles, which recurred locally after surgery.
At the time of presentation, the picture shows a mass in the left upper back of size 20 \times 20 \text{ cm}^2 with overlying ulceration and minimal discharge at the time of presentation.

On local examination, he had a 20 \times 20 \text{ cm}^2 tender hard mass at left upper back with overlying ulceration and discharge (Fig. 1). Magnetic resonance imaging (MRI) of the chest with contrast revealed a large lobulated heterogeneously enhancing soft-tissue mass in the subcutaneous plane of the posterior aspect of the chest wall along with a suspicious nodule in the azygous lobe of the right lung, which warranted further investigation (Fig. 2). Biopsy of the mass reported spindle cell sarcoma with possibility of neuronal cell origin. Immunophenotyping was advised for further confirmation but could not be done due to lack of resources.

The tumor was surgically excised by a team of surgeons (Fig. 3). The patient has been sent for radiotherapy of the tumor bed and further evaluation of his pulmonary nodule to the cancer hospital nearly 300-km away. Due to the unavailability of the plastic surgeon for skin grafting, the patient was again referred to another hospital nearly 150-km away.

**DISCUSSION**

The evaluation of a patient with a suspected soft-tissue sarcoma includes history, imaging and biopsy [6]. MRI is the preferred modality for the evaluation of soft-tissue masses of the extremities, trunk, head and neck [7]. All patients diagnosed with sarcoma should also have a computed tomography scan of the chest at the time of diagnosis and in follow-up [6].

Spindle cell sarcomas affect people of almost any age and sex [8]. Two separate studies by Feng et al. and Smith et al. showed the median age at presentation of 57 years [9, 10]. In contrary to these studies, the age at presentation in our case is quite early (37 years). In the same study by Smith et al., the median tumor size found was 9.87 cm [10]. Our patient had a tumor size of 5 \times 4 \text{ cm}^2 at the first presentation while it grew into a massive size of 20 \times 20 \text{ cm}^2 when it recurred later after the surgery. This sort of presentation is quite rare as patients generally tend to present earlier with the mass with a median duration of 20 weeks [10], whereas our patient completely ignored the mass until it grew to a massive size and started hindering his day-to-day activities. This highlights that in poverty and destitution, people ignore medical attention until the very last stage [11].

Due to their rarity, delay or misdiagnosis is common for sarcomas especially in settings with limited facilities. [12, 13]. In our case, the mass was initially identified as liposarcoma. Later, repeat biopsies revealed spindle cell sarcoma. The rarity of the case and the little experience in diagnosing rare cases might have led to this error in histological diagnosis [14].

Two separate studies by Swamsura et al. and Diageler et al. reported local recurrence after surgery within a median duration of 19 months and 15.7 months, respectively [15, 16]. Our patient...
had a recurrence within 3–4 months after surgery, which is quite rare. A frequent follow-up is therefore advised after excision of sarcoma especially in the first 2 years [17].

In our case, the lack of proper communication between involved hospitals and doctors might be one of the reasons for his loss to follow-up [18, 19]. Besides his financial constraints and ignorance, inadequate counselling about the diagnosis may have led to the discontinuity of care, which is very common in developing countries [18, 19]. Due to limited resources in hospitals, it is common in developing countries to refer patients to other centers for further care [20]. Even tertiary care center like ours cannot provide comprehensive care to the patient. This creates a disadvantage in continuity-of-care for many patients. Hence, one of the main focuses regarding this patient would be to ensure adequate follow-up. To avoid such circumstances in the future, it is better if the health care team takes charge and arranges continuity-of-care for the patient so that any future patients do not leave without completing their course of treatment. Use of telemedicine is proven to be highly effective to ensure follow-up in developing countries [21]. It is, of course, necessary to address the main underlying issues like the financial burden for the patient and aim for sustainable treatment strategies.

**CONCLUSION**

The management of soft-tissue sarcomas is best done in a center with appropriate expertise in multiple fields. In absence of appropriate expertise or resources, patients do not get adequate treatments in time. This may lead to grave consequences like metastasis or recurrence causing significant problems to the patients. Regular follow-up after treatment with history, physical examination and chest imaging is of utmost importance.

**CONFLICT OF INTEREST STATEMENT**

None declared.

**FUNDING**

None.

**REFERENCES**

1. Siegel RL, Miller KD, Fuchs HE, Jemal A. Cancer statistics, 2021. CA Cancer J Clin 2021;71:7–33.
2. World Health Organization Classification of Tumours Editorial Board. Soft tissue and bone tumours. In: *International Agency for Research on Cancer*, Vol. 3, 5th edn. Lyon, France: IARC, 2020, ISBN: 9789283245025.
3. Lawrence W Jr, Donegan WL, Natarajan N, Mettlin C, Beart R, Winchester D. Adult soft tissue sarcomas. A pattern of care survey of the American College of Surgeons. *Ann Surg* 1987;205:349.
4. Christie-Large M, James SLJ, Tiessen L, Davies AM, Grimer RJ. Imaging strategy for detecting lung metastases at presentation in patients with soft tissue sarcomas. *Eur J Cancer* 2008;44:1841–5.
5. Doyle LA. Sarcoma classification: an update based on the 2013 World Health Organization classification of soft tissue and bone. *Cancer* 2014;120:1763–74.
6. Dangoor A, Seddon B, Gerrand C, Grimer R, Whelan J, Judson I. UK guidelines for the management of soft tissue sarcomas. *Clin Sarcoma Res* 2016;6:1–26.
7. Panicek DM, Gatsonis C, Rosenthal DI, Seeger LL, Huvos AG, Moore SG, et al. CT and MR imaging in the local staging of primary malignant musculoskeletal neoplasms: report of the radiology diagnostic oncology group. *Radiology* 1997;202:237–46.
8. Borden EC, Baker LH, Bell RS, Bramwell V, Demetri GD, Eisenberg BL, et al. Soft tissue sarcomas of adults: state of the translational science. *Clin Cancer Res* 2003;9:1941–56.
9. Feng L, Wang M, Yibulayin F, Zhang H, Yang YL, Ren F, et al. Spindle cell sarcoma: a SEER population-based analysis. *Sci Rep* 2018;8:1–10.
10. Smith GM, Johnson GD, Grimer RJ, Wilson S. Trends in presentation of bone and soft tissue sarcomas over 25 years: little evidence of earlier diagnosis. *Ann R Coll Surg Engl* 2011;93:542–7.
11. Sapkota T, Houkes I, Bosma H. Viscous cycle of chronic disease and poverty: a qualitative study in present day Nepal. *Int Health* 2021;13:30–8.
12. Noria S, Davis A, Kandel R, Levesque J, O’Sullivan B, Wander J, et al. Residual disease following unplanned excision of a soft-tissue sarcoma of an extremity. *J Bone Joint Surg* 1996;78:650.
13. Kostopoulou O, Delaney BC, Munro CW. Diagnostic difficulty and error in primary care—a systematic review. *Fam Pract* 2008;25:400–13.
14. Arbiser ZK, Folpe AL, Weiss SW. Consultative (expert) second opinions in soft tissue pathology: analysis of problem-prone diagnostic situations. *Am J Clin Pathol* 2001;116:473–6.
15. Sawamura C, Matsumoto S, Shimoji T, Okawa A, Ae K. How long should we follow patients with soft tissue sarcomas? *Clin Orthop Relat Res* 2014;472:842–8.
16. Daigeler A, Zmarsly I, Hirsch T, Goertz O, Steinau HU, Lehnhardt M, et al. Long-term outcome after local recurrence of soft tissue sarcoma: a retrospective analysis of factors predictive of survival in 135 patients with locally recurrent soft tissue sarcoma. *Br J Cancer* 2014;110:1456–64.
17. Rothermundt C, Whelan JS, Dileo P, Strauss SJ, Coleman J, Briggs TW, et al. What is the role of routine follow-up for localised limb soft tissue sarcomas? A retrospective analysis of 174 patients. *Br J Cancer* 2014;110:2420–6.
18. Kattel S. Doctor patient communication in health care service delivery: a case of Tribhuvan University Teaching Hospital, Kathmandu. *Master in public policy and governance program thesis. North South University Bangladesh*, 2013.
19. Tiwary A, Rimal A, Paudyal B, Sigdel KR, Basnyat B. Poor communication by health care professionals may lead to life-threatening complications: examples from two case reports. *Wellcome Open Res* 2019;4:7.
20. Poudel KK, Sims D, Morris D, Neupane FR, Jha AK, Lamichhane N, et al. Cancer cases referral system in Nepal. *Nepal J Epidemiol* 2018;8:748.
21. Combi C, Pozzani G, Pozzi G. Telemedicine for developing countries. A survey and some design issues. *Appl Clin Inform* 2016;7:1025–50.