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Refractory bleeding from a giant de-differentiated liposarcoma of the chest wall: An indication for neoadjuvant chemotherapy and palliative resection? – A case report

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ABSTRACT

INTRODUCTION: Dedifferentiated liposarcoma (DDLPS) is a heterogenous neoplasm of variable histological grade. DDLPS uncommonly arises from the chest wall. There are limited data available about the tumor's response to chemotherapy and accessible reports indicate minimal benefits. Surgery is thus the cornerstone of management. Here, we demonstrate an uncommon situation where chemotherapy was used to arrest bleeding from a giant DDLPS that was refractory to all available hemostatic agents. This case also presents an uncommon indication for palliative chest wall resection and reconstruction (CWRR).

PRESENTATION OF CASE: A 55-year old woman presented with refractory bleeding from an ulcerated and foul-smelling mass on the anterior chest wall, confirmed histologically to be DDLPS. Chemotherapy with Doxorubicin and Ifosfamide was used to control the bleeding. She subsequently had CWRR to improve her quality of life. The patient made an uneventful recovery but later died from pulmonary embolism.

DISCUSSION: The dedifferentiated component of DDLPS is vascular and may account for why we were able to exhibit a hemostatic response to chemotherapy. CWRR was then employed to improve the quality of life in an advanced, ulcerated and infected tumor of the chest wall.

CONCLUSION: We were able to demonstrate a hemostatic response of DDLPS to neoadjuvant chemotherapy and anticipate that this report may serve as a reference for further studies. Furthermore, we believe that palliative resection may be carried out to improve a patient's quality of life even in the face of advanced disease.

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1. Introduction

Liposarcoma commonly originates from the retroperitoneum and limbs but the chest wall is an infrequent site of affection [1]. De-differentiated liposarcoma (DDLPS) is one of its histologic subtypes. DDLPS is a heterogenous neoplasm with one component being well-differentiated and the other a non-lipogenic sarcoma of variable histological grade. Limited data regarding the sensitivity of DDLPS to chemotherapy are available, with minimal reported ben-

efit [2]. These reports are, however, mostly about retroperitoneal DDLPS. Reports on chest wall DDLPS are scanty. Surgery is thus the cornerstone of treatment of chest wall liposarcoma. Refractory bleeding with severe anemia, malodorous discharge, depression and interference with activities of daily living may sometimes complicate a giant ulcerated DDLPS of the chest wall, reducing the quality of life of the patient. Therefore, neoadjuvant chemotherapy and palliative resection may become clinically relevant, not only to improve survival but to better the quality of life of such patients.

Here, we report the case of a middle-aged woman who had neoadjuvant chemotherapy for recalcitrant bleeding that resisted all available hemostatic agents and subsequently had palliative chest wall resection and reconstruction (CWRR) for an advanced giant DDLPS of the chest wall.

This work has been reported in line with the SCARE criteria [3].

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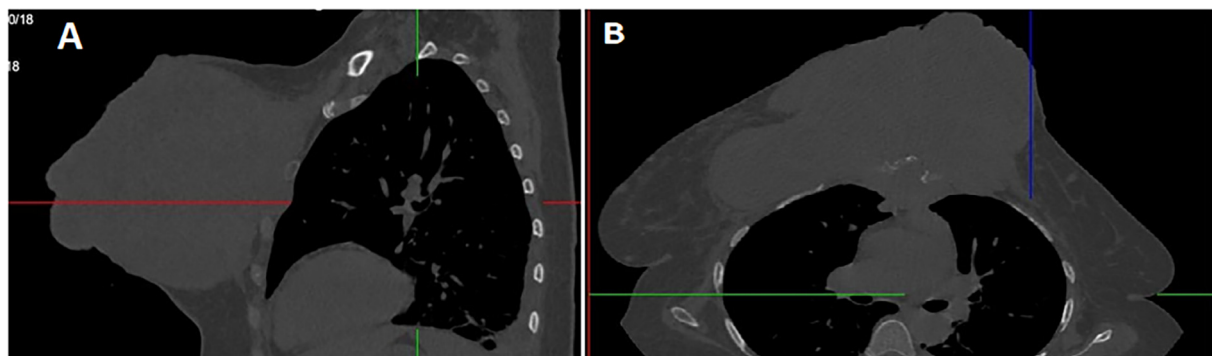


Fig. 1. (A) Coronal Reformat of chest CT scan show a huge hypodense soft tissue mass with irregular margins attached to the muscular layer of the chest wall (B) Axial section of the chest CT scan showing a huge hypodense soft tissue mass within the muscular layer of the anterior chest wall with underlying sternal invasion.



Fig. 2. Immediate preoperative period – Tumor with peripheral areas of necrosis and central yellowish area.

2. Case report

A 55-year old woman presented with a 16-year history of a mid-sternal, pea-sized mass which had maintained its proportions until 3 years prior to presentation when it started to increase in size and subsequently became painful. The mass was about 15 cm × 15 cm × 10 cm, firm, and attached to the underlying structures. There were no discernible peripheral lymph nodes. A tru-cut biopsy of the mass revealed pleomorphic sarcoma. She was subsequently counselled for a CWRR and chemotherapy but she refused and defaulted from the clinic. Two months later, however, she was admitted on account of bleeding from the ulcerated, fetid tumor with severe anemia. The packed cell volume (PCV) was 12%. Other hematologic parameters were within normal limits. She had 8 units of packed cells transfused and was also commenced on intravenous antibiotics and analgesics. Appropriate wound care was established and she was offered TED stockings for deep venous thrombosis (DVT) prophylaxis. A chest CT scan showed a huge hypodense soft tissue mass within the muscular layer of the anterior chest wall with underlying sternal invasion (Fig. 1). Bleeding from the tumor site was unresponsive to available hemostatic methods which necessitated the commencement of a trial neoadjuvant chemotherapy with Doxorubicin and Ifosfamide. Although this successfully stopped the bleeding, it was complicated by anemia and leucopenia, warranting additional units of blood transfusion and Filgrastim administration. She later went into depression associated with a reduction in her quality of life as a result of the malodorous wound

discharge and the compressive weight of the tumor on the chest wall.

She was counselled for CWRR and had enbloc excision of the tumor (Fig. 2), part of the pectoralis major muscles, inferior half of the manubrium sterni, sternal body, xiphoid and adjacent costal cartilages (Fig. 3A & B) with a 4 cm tumor-free skin margin. The pericardium and lungs were free of macroscopic tumor. The excised tumor weighed 3.59 kg and was 25 cm × 21 cm × 14 cm in dimension. The bony defect was reconstructed with methylmethacrylate sandwiched between two Prolene meshes (Fig. 4A). Primary closure of soft tissue was achieved by local flap (Fig. 4B). The histology revealed sheets of malignant lipoblasts with areas of heterologous differentiation predominantly myogenic as well as areas of pleomorphic sarcomatous pattern (Fig. 5).

Recovery was uneventful in the intensive care unit. She however suffered a massive pulmonary embolism about thirty-six hours after the surgery and attempts to resuscitate her proved abortive.

3. Discussion

Liposarcoma usually originates in the extremities and the retroperitoneum but rarely in the thorax [1,4]. It accounts for ≤20% of primary chest wall soft tissue sarcomas [5,6]. There is no gender difference and it most commonly affects people between 40 and 70 years [1]. Histologic variants of liposarcoma include well-differentiated (WDLPS), myxoid, pleomorphic (PLPS); liposarcoma not otherwise specified and dedifferentiated liposarcoma (DDLPS).

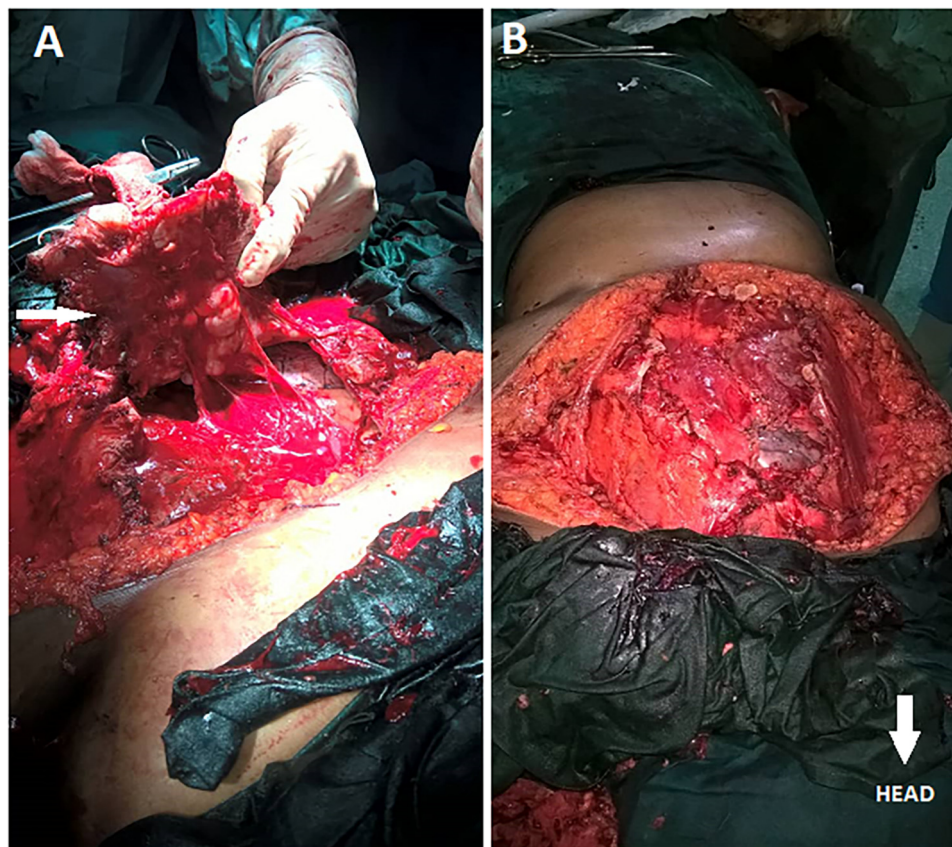


Fig. 3. (A) Resection of the manubrium sterni and (B) Chest wall defect after resection.

DDLPS has been defined as a “nonlipogenic sarcoma” arising in association with well-differentiated liposarcoma/atypical lipomatous tumor. About 90% of DDLPS arise “de novo,” i.e. combined with a well-differentiated tumor in the primary lesion while 10% occur in recurrences of WDLPS. Histopathological characterization is important because dedifferentiation confers the tumor with metastatic capability [7,8] and increased risk of local recurrence following inadequate resection margins [9]. On the other hand, molecular characterization of these tumors has opened up novel treatment targets. The commonest pattern of dedifferentiation is a high-grade sarcoma resembling an undifferentiated pleomorphic sarcoma [10]. In about 5–10% of cases, the dedifferentiated component shows heterologous differentiation containing myogenic elements [11], as in this case presentation. The definition of DDLPS has recently been modified to include a subset of cases that demonstrate lipoblastic differentiation within the dedifferentiated component referred to as homologous lipoplastic differentiation. This has to be differentiated from PLPS because PLPS has a considerably worse prognosis [10].

Percutaneous biopsy may not correctly diagnose or may even misdiagnose DDLPS [7] because the ‘dedifferentiated areas dominate and the well differentiated lipoma-like areas can only be found after careful and generous sampling’ [10]. This may account for why our trucut biopsy showed pleomorphic sarcoma.

Most of the data about the response of DDLPS to chemotherapy are derived from retroperitoneal DDLPS because of the paucity of cases of chest wall DDLPS. These data suggest that the response rate is minimal [2] and as such systemic therapy is not frequently utilized in the primary setting [9]. Notwithstanding the minimal effect of chemotherapy in improving overall survival, this case presentation was able to demonstrate another distinctive

role for chemotherapy in securing hemostasis. The dedifferentiated component of the tumor is the more vascular part [12] and we speculate that ulceration into this part may account for the refractory bleeding from the tumor. Decreased tumor vascularity has been correlated with response to chemotherapy [12] and may explain why chemotherapy had a hemostatic effect in our patient. Anthracycline-based combination chemotherapy reportedly gives the highest objective response rates in patients with DDLPS [2,12]. Combination chemotherapy regimens nevertheless produce this response at the expense of increased toxic effects [13] like in the index case. Even so, the hemostatic effect provided by chemotherapy allowed optimization of the patient and subsequent palliative CWRR.

The inability to do a post-chemotherapy CT scan limited our ability to comprehensively assess response based on the response evaluation criteria in solid tumors (RECIST). Livingston et al. [12] have, however, proposed some vascular criteria to be used to assess post-chemotherapy response in DDLPS. These include necrosis, hyalinization, cytological changes, or any combination of these in the pathology specimen. The post-resection pathology slides of our patient showed extensive areas of necrosis and decreased cellularity (Fig. 5C).

Surgical resection remains the definitive management for operable DDLPS. Treatment often is intended for curative purposes, however, palliative resection in the face of advanced disease is needed to improve the quality of life. The skin ulceration, malodorous wound discharge and weight of the tumor on the chest evidently made CWRR necessary in this case. The principles of oncologic resection [14] were adhered to so as to increase disease-free survival.

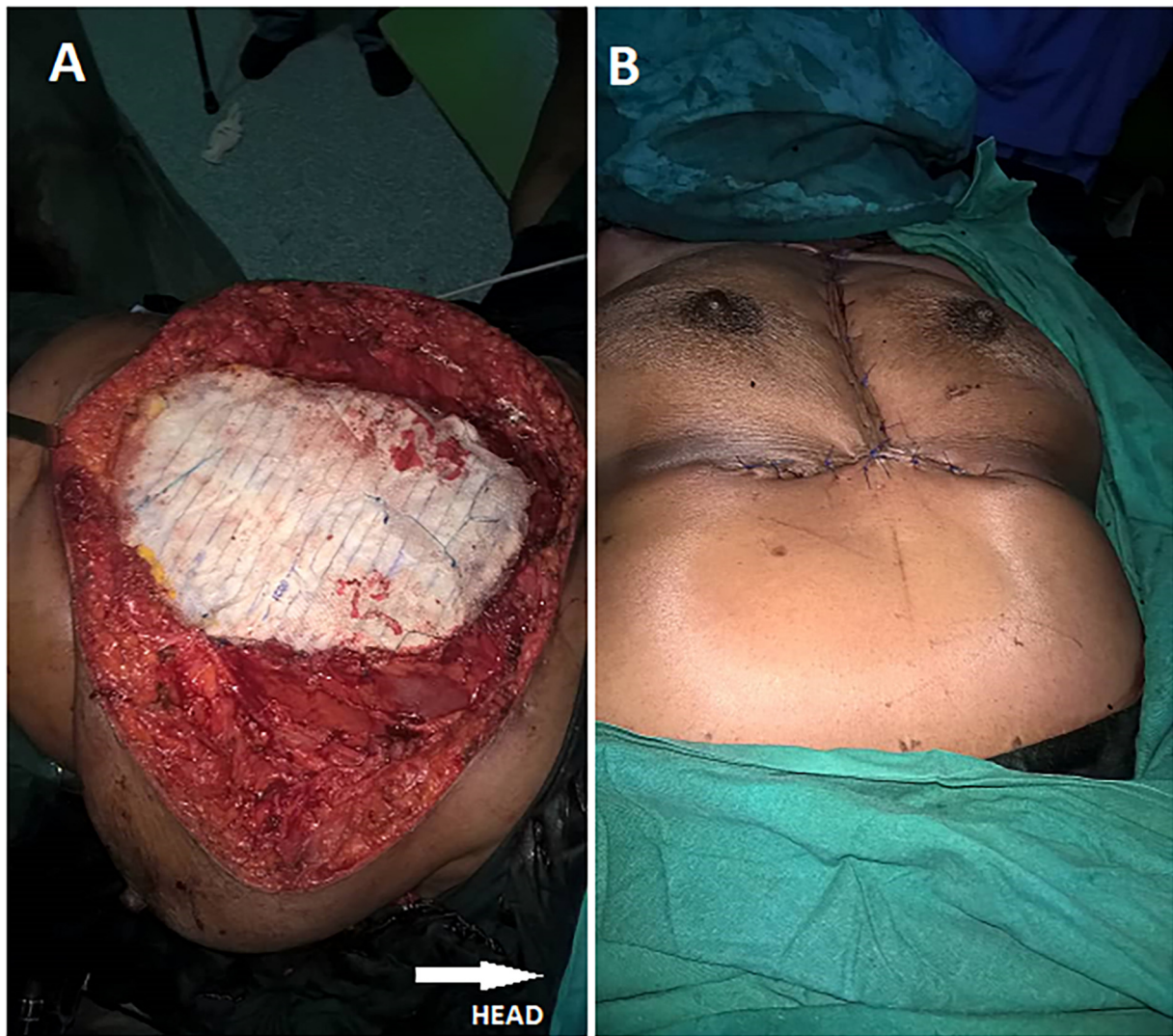


Fig. 4. (A) Chest wall defect reconstructed with methylmethacrylate. (B) After soft tissue reconstruction.

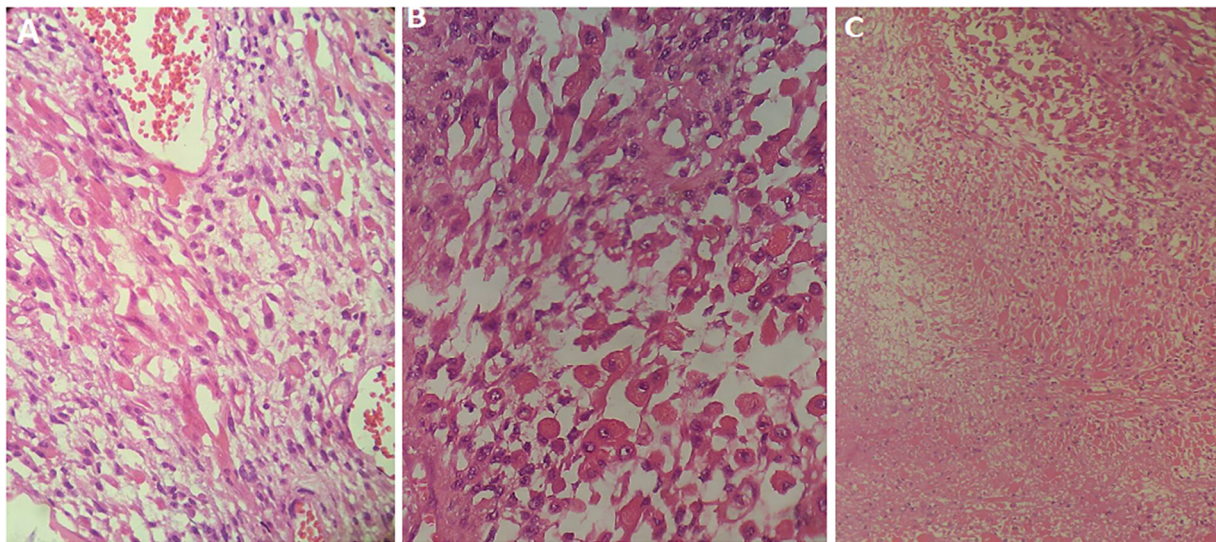


Fig. 5. (A) Sheets of malignant lipoblasts showing myogenic differentiation in highly vascularized stroma. (B) Areas of myogenic differentiation. H&E x 20. (C) Preserved differentiated area around a blood vessel while others show extensive necrosis.

4. Conclusion

This case report demonstrated a unique role of neoadjuvant chemotherapy as a hemostatic agent in DDLPS. However, further studies need to be carried out to authenticate the usefulness of chemotherapy as a hemostatic agent in this subtype of liposarcoma. It also demonstrated that palliative CWRR may be an option in the management of advanced giant, bleeding chest wall tumor in order to improve quality of life of such patients.

Conflict of interest

No conflict of interest declared.

Funding

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Ethical Approval

The study was exempted from ethical approval by the ethics committee, Aminu Kano Teaching Hospital, Kano since it was a case report.

Consent

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

Author contribution

Tunde Nureini Oyebanji – wrote the initial draft.

Literature review – All authors.

Review of the initial draft – Ganiyu Oyediran Oseni, Ismail Mohammed Inuwa, Jameel Ismail Ahmad, Sadiq Garba.

Final approval of manuscript: All authors.

Registration of Research Studies

Not required.

Guarantor

Tunde Nureini Oyebanji.

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