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Spindle Cell Leiomyosarcoma of Gluteal Muscle- A Case Report

Marwa Al Sharji[1*], Hazem Elgohary[2], Yasser Abbas[3] and Maimuna Alsaadi[4]

[1]General foundation program trainee, Oman medical specialty board. Muscat, Oman

[2]Department of general surgery, Khawlah Hospital, Muscat, Oman

[3]Department of general surgery, Khawlah Hospital, Muscat, Oman

[4]Department of histopathology, Khawlah Hospital, Muscat, Oman

[*]Corresponding author:

Marwa Al Sharji,
General foundation program trainee, Oman medical specialty board. Muscat, Oman,
E-mail: m.alsharji-112@hotmail.com

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1. Abstract

44 years old male patient, presented with left gluteal swelling representing a collection, the patient had incision and drainage done followed by excision and biopsy, final histopathology report showed spindle cell leiomyosarcoma.

This case report is of spindle cell sarcoma, it is a rare type of sarcomas. This patient had an atypical presentation of the tumour initially presenting as a collection mimicking gluteal abscess, initially requiring incision and drainage.

2. Background and Case Report

2.1. Introduction

Soft tissue sarcomas are derivatives of mesenchymal cells precursors that develop into different tissues such as (muscles, connective tissues and adipose). Leiomyosarcomas are smooth muscle tumours, it the most common type of soft tissue sarcomas with an incidence of 11%. Local recurrence and distant metastasis are both possible. Tumour size and location are the most prominent prognostic factors. Soft tissue leiomyosarcoma is mostly found in retroperitoneum and large blood vessels. While the most commonly affected non-retroperitoneum is in the lower extremity accounting for 10-15% of all limbs soft tissue sarcomas [1]. Typically, almost all leiomyosarcomas present with a painful mass, pain is moderate in intensity, other clinical features depend on the tumour site. Histologically, fibrous and myxoid changes are seen, spindle cells arranged in spirals. Large tumours usually have hypocellular areas with coagulative necrosis [2].

3. Case Report

This is a case report of 44 years old male, with no significant medical or surgical background who initially presented to the emergency department with a 2-week history of painful swelling with pain radiating to the left leg. On examination, the patient had a mildly tender, indurated swelling, 10 by 10 centimetres in size over the left gluteal region. laboratory workup was done for the patient, inflammatory markers were within normal. Soft tissue ultrasound was done and was reported as a large well defined cystic lesion with thick internal fibrous septations at the left gluteal area measuring 18*15cm of turbid content, No internal vascularity, no surrounding oedema, no calcification. The impression was of haematoma/abscess. The patient was taken for incision and drainage, intra-operatively, 2 litres of liquified hematoma were drained. Samples taken and sent for cytology and biopsy and culture. Fluid microscopy showed 440(cells) /uL of total white blood cells (80% was of neutrophils and 20% of lymphocytes, 143,500 cells/uL of total red blood cells. The fluid culture showed no growth. Fluid cytology showed heavily blood-stained smears containing scattered multi-nucleated giant cells, foamy histocytes and inflammatory cells. Few cells noted with irregular hyperchromatic nuclei and vacuolated cytoplasm occasionally binucleated. The cell block was acellular.

The patient was initially managed with daily dressing, after 11 days from the Incision and drainage patient came to the emergency again with pus discharge from the wound. When examining the patient, he was afebrile, other vitals within normal, there was minimal slough over the wound. Aspiration attempted and around 100ml collected and sent for laboratory workup. Fluid culture reported afterwards as moderate growth of Enterobacter cloacae complex and staphylococcus aureus. The patient was offered ad-

mission afterwards again for surgery as the patient had discharge draining. The patient's inflammatory markers remained slightly high. Magnetic resonance imaging done. The patient was taken for revision of the previously done incision and drainage, intra-operative findings were of a large intramuscular hematoma with necrosis of the gluteus maximus, sample for biopsy was taken for histopathology. Microscopic findings showed extensively ulcerated fragments with underlying granulation tissue, the granulation tissue was replaced by malignant neoplasm composed of atypical spindle cell with irregular, large pleomorphic, hyperchromatic nuclei and abundant pale to eosinophilic cytoplasm, scattered bi- and multinucleated and bizarre forms are noted. There was brisk mitotic activity with atypical neoplasm. The tumour cells are strongly positive for vimentin and SMA stain, desmin and CD 99. Negative staining of AE1/AE3, MNF116, CD34,S100, p63. The picture was consistent with malignant spindle cells neoplasm with morphology and immune profile of leiomyosarcoma. The patient was then scheduled for excision of the leiomyosarcoma.

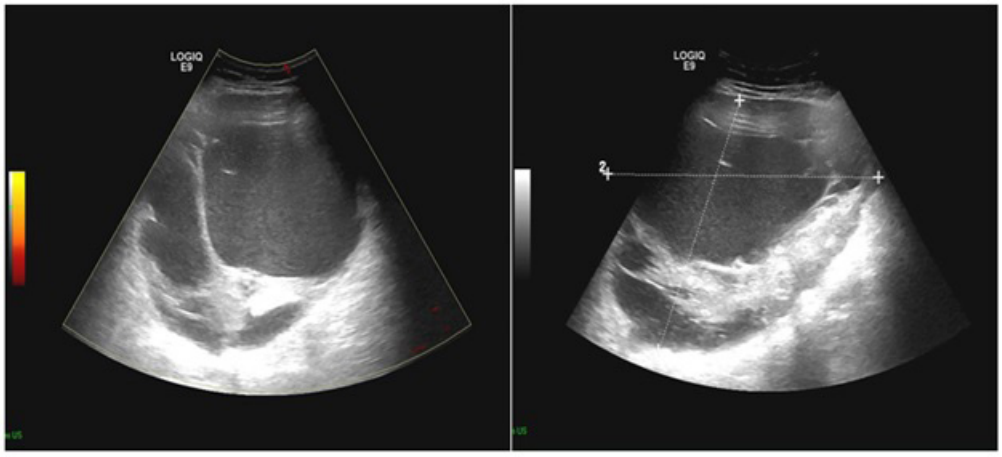
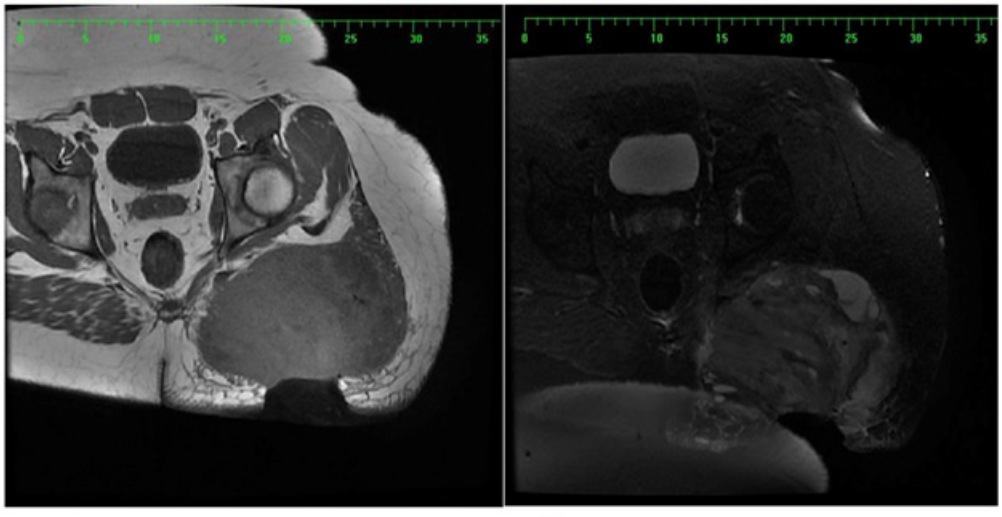
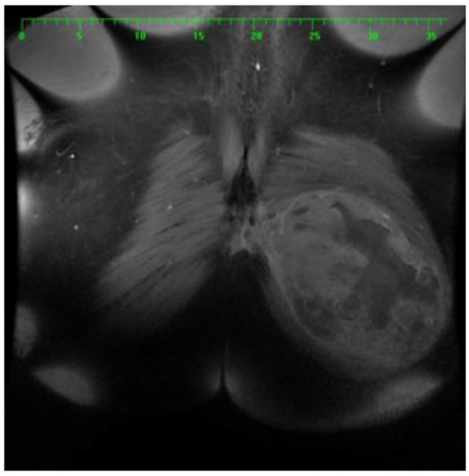


Figure (1): (A), (B) Ultrasound showing a large cystic lesion measuring 18*15 cm. Internal septation with solid component is noted at the left gluteal area.





C

Figure 2: MRI images taken after initial incision and drainage showing soft tissue mass lesion over left gluteal muscle with areas of necrosis.

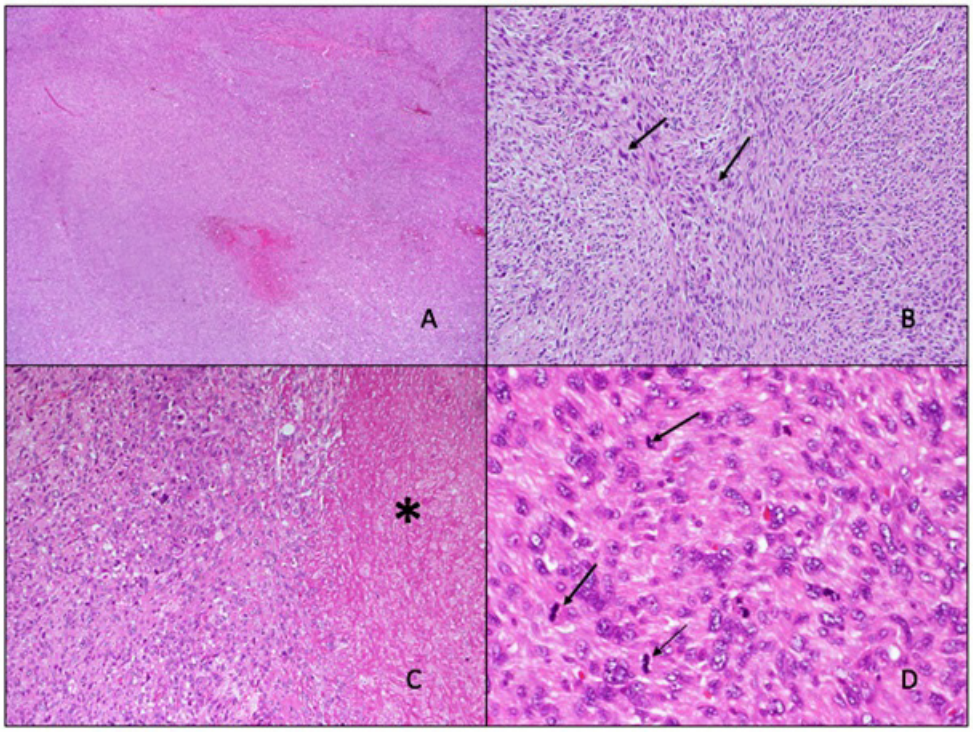


Figure 3: (A). Low power (2x) showing a cellular lesion, **(B).** Interlacing fascicles of spindle cells with atypia (arrows) (10x), **(C)** Tumor on the left with necrosis on the right (*) (10x), **(D).** High power (40x) showing cells with atypical nuclei and frequent mitosis (arrows).

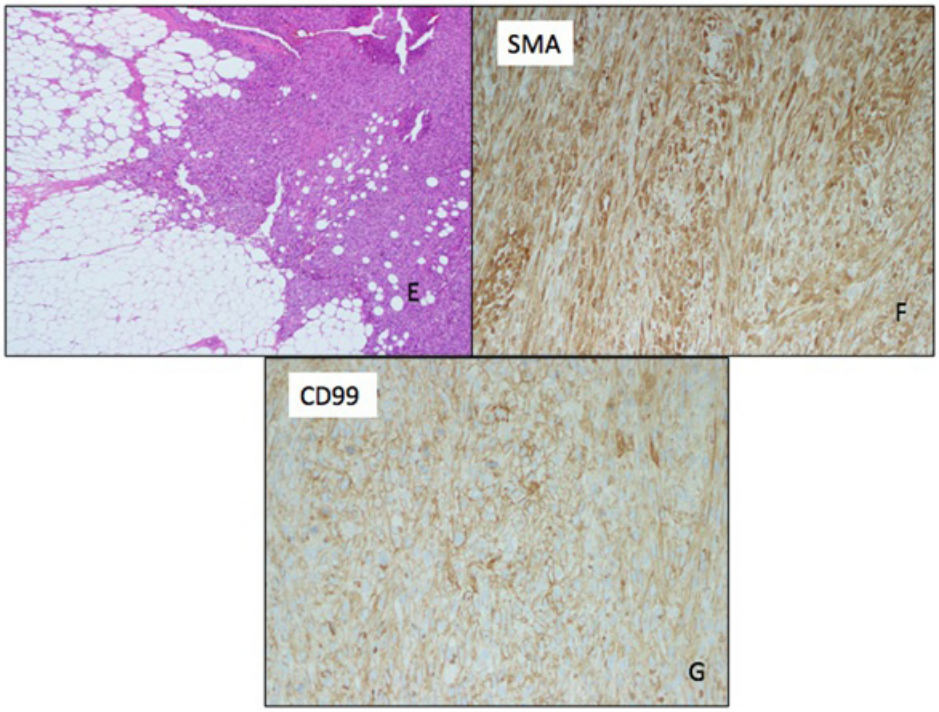


Figure 4: (E). Tumor infiltrating into surrounding adipose tissue (4x), **(F).** Tumor cells show diffuse positivity for SMA immuno-stain (20x), **(G).** Tumor cells showing CD99 positivity (20x).

4. Discussion

Spindle cell Sarcoma is an unusual, atypical soft tissue sarcoma. Other variants of sarcomas are pleomorphic and round cells. Sarcomas generally account for less than 1% of all malignant cancer cases. Undifferentiated sarcomas may rise in different locations, including head, neck, upper/lower limbs and retroperitoneum. A delay in establishing a diagnosis is possible given that sarcomas are a rare type of tumours. It was also reported in the literature that inappropriate resection of the masses is a common finding before the final histopathology report. The latter may increase the chance of metastasis due to disruption of the tumour. Thus, it is recommended to fully investigate the patient before attempting surgical resections. In our case, as the patient initially presented with a picture of collection/hematoma as per ultrasound, incision and drainage were attempted and then a revision surgery was attempted to excise the full mass.

Soft tissue sarcomas metastasize primarily to the lungs, Computed Tomography of the chest should be done regularly for evaluation. However, to investigate the primary lesion, it is recommended to use MRI for masses found in the trunk, peripheries and head and neck. While CT is recommended for intra-abdominal masses [3].

Spindle cell leiomyosarcomas diagnosis should be established in an equipped medical centre, it is identified by the presence of typical spindle cells for smooth muscle differentiation which is typically found in fascicles and meet and perpendicular angles. Moreover, immunohistochemically, the presence of smooth muscle actin(SMA) and other markers like ad desmin and h-caldesmon. In our case, a biopsy was positive for desmin and SMA [4]. Other stains used in diagnosing leiomyosarcoma are vimentin, Pan-K, laminin, CAM-52, PC10, hematoxylin, phosphatungstic acid, Masson trichrome and periodic acid-Schiff stain [5]. Surgery is the most reliable mode of treatment for leiomyosarcoma, the margin of excision should be at least 1 cm in all directions. However, if surgery and radiation are used together, the excision margin should be of 0.5 cm approximately [5]. For patients with advanced disease, cytotoxic chemotherapy with agents like doxorubicin and ifosfamide to decrease the size of the mass to make it operable [6].

5. Conclusion

Spindle cell sarcoma is a very rare variant of sarcomas. Suspicious enlarging masses should alert clinicians and warrant them for further investigations and imaging and if possible biopsy before surgical excision. Clinical staging and surgical margins are important factors in determining a patient's outcome. Implementing the role of a multidisciplinary team in managing and integrating these tumours is important.

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