Accepted Manuscript

Submission Date: 2023-10-30
Accepted Date: 2024-04-03
Accepted Manuscript online: 2024-04-10

Archives of Plastic Surgery

A Rare Case Of Primary Sarcoma Arising Within Free Muscle Transfer.

ANCHAL JAIN, Bilal A Rafique, Amer Durrani, Ahid Abood.

Affiliations below.

DOI: 10.1055/a-2303-4940

Please cite this article as: JAIN A, Rafique B A, Durrani A et al. A Rare Case Of Primary Sarcoma Arising Within Free Muscle Transfer. Archives of Plastic Surgery 2024. doi: 10.1055/a-2303-4940

Conflict of Interest: The authors declare that they have no conflict of interest.

Abstract:
Soft tissue sarcomas are one of the rarest forms of cancer. We describe a unique case of a 35-year-old patient who sustained an open lower limb fracture requiring an intra-medullary nail and free latissimus dorsi muscle flap reconstruction. He had a complex post-operative course including osteomyelitis, a re-fracture and chronic pain. Eleven years following the injury, he presented with pain and localized swelling around the flap. Histological analysis confirmed a rhabdomyosarcoma (RMS) within the LD muscle and he underwent a trans-femoral amputation. He is now in remission and walks on a prosthesis pain free.

Corresponding Author:
ANCHAL JAIN, Cambridge University Hospitals NHS Foundation Trust, Plastic & Reconstructive Surgery, Cambridge, United Kingdom of Great Britain and Northern Ireland, anchal.jain@nhs.net

Affiliations:
ANCHAL JAIN, Cambridge University Hospitals NHS Foundation Trust, Plastic & Reconstructive Surgery, Cambridge, United Kingdom of Great Britain and Northern Ireland
Bilal A Rafique, Cambridge University Hospitals NHS Foundation Trust, Plastic & Reconstructive Surgery, Cambridge, United Kingdom of Great Britain and Northern Ireland
Amer Durrani, Cambridge University Hospitals NHS Foundation Trust, Plastic & Reconstructive Surgery, Cambridge, United Kingdom of Great Britain and Northern Ireland
Ahid Abood, Cambridge University Hospitals NHS Foundation Trust, Plastic & Reconstructive Surgery, Cambridge, United Kingdom of Great Britain and Northern Ireland

A Rare Case of Primary Sarcoma Arising Within Free Muscle Transfer

Mr. Bilal Rafique[1*], Dr. Anchal Jain[1*], Mr Amer J Durrani[1], Mr Mr. Ahid Abood[1]

[1]Department of Plastic and Reconstructive Surgery, Addenbrooke's Hospital, Cambridge University Hospitals NHS Trust.

* Joint first authorship: These two authors have worked together on this publication and contributed equally to the work

Corresponding address: Dr Anchal Jain, CT1 in Plastic and Reconstructive Surgery, Department of Plastic and Reconstructive Surgery, Addenbrookes Hospital, Cambridge University Hospitals NHS Foundation Trust, Hills Road, Cambridge, CB2 0QQ, UK. Email: anchal.jain@nhs.net

INTRODUCTION

Muscle and fascio-cutaneous flaps are common reconstructive options for patients with traumatic lower limb soft tissue defects. The latissimus dorsi (LD) is one of the most commonly used muscles for coverage of such defects. It lends itself well to this purpose by offering a robust, large and well vascularised muscle with a long pedicle length and the option of a skin paddle. Alternatively, a split thickness skin graft can be applied to resurface the muscle.

Soft tissue sarcomas (STS) are rare forms of cancer with an incidence in England of 7.7 cases per 100,000 persons and approximately 4300 new cases in England each year [1]. RMS is a rare primitive mesenchymal type of STS made up of cells that differentiate into skeletal or striated muscle. It is the most common soft tissue malignancy in children but exceedingly rare in adults [2].

The following case highlights a sinister presentation of a rhabdomyosarcoma following a complex lower limb reconstruction. To our knowledge, only one other case report exists in the literature demonstrating a similar tumour within a muscle flap [3]. Our case report highlights how this rare tumour may present, particularly within the context of a complex postoperative course, other variables and more likely diagnoses. It hence emphasises this crucial but rare differential diagnosis.

CASE PRESENTATION

A 35-year-old male was involved in a high speed road traffic accident thirteen years ago. He sustained a closed fracture of the right femur and an open fracture of the left femur and right tibia and fibula with an overlying soft tissue defect (Gustilo and Anderson IIIb) (Fig 1).

The bilateral femoral fractures were treated with intramedullary (IM) nails (Fig 2 a). The right leg underwent a joint orthoplastic debridement and external fixation on day of admission. On Day 5 the external fixator was replaced with an IM nail and the soft tissue defect was reconstructed using a free latissimus dorsi flap and split skin graft (Fig 2 b, Fig 2 c & Fig 3). He was discharged four weeks following initial admission.

Over the course of the next few years, he had various hospital admissions due to osteomyelitis (OM), a re-fracture following trauma and chronic pain (Fig 4). Three months post operatively he developed OM of the right tibia which was treated within the bone infection unit with removal of IM nail and IV antibiotics. Three years post operatively he sustained trauma to the leg causing a refracture and had an IM nail re-inserted. He continued to have chronic pain in the limb which was maximally treated with analgesics but eventually underwent removal of his second IM nail.

Eleven years following the initial injury, he presented with swelling and pain. This was most likely thought to be a recurrence of osteomyelitis and he was managed with prolonged course of antibiotics. An MRI revealed a large inflammatory phlegmon most in keeping with infection, however a biopsy was advised to rule out neoplastic changes (Fig 5). He progressed to have a PET-CT which showed low-grade uptake in soft tissue of right anterior lower leg and confirmed no evidence of metastatic disease. A biopsy of the muscle flap revealed a spindle cell/sclerosing rhabdomyosarcoma.

He was treated in a regional sarcoma centre in line with UK guidelines on management of soft tissue sarcomas [4]. There was no evidence of distant metastatic disease and he was treated with the standard IVADo chemotherapy regimen (ifosfamide, vincristine, doxorubicin, dactinomycin) followed by a right above knee (transfemoral) amputation. (Fig 6)

Histopathology showed densely cellular tumour comprising sheets of tumour cells with round hyperchromatic nuclei and scant cytoplasm. Tumour cells exhibited a spindled morphology in some areas (Fig 7). On immunohistochemistry, the tumour showed strong diffuse staining for Desmin and Myo-D1 in keeping with the final diagnosis of pT2, N0, M0, Grade 3 sclerosing rhabdomyosarcoma (100 x 40 x 20 mm).

On regular clinic reviews, he continued to show positive progression with phantom limb pain which eventually settled. Surveillance staging scans showed no evidence of recurrence or metastases. He continues to be seen by specialist amputee therapy services and is fully independent walking with a prosthesis and is pain free (Fig 8).

DISCUSSION

Soft tissue sarcomas are rare in adults making up less than 1% of all adult malignancies. Of all STS, RMS accounts for 3% of cases [5]. The latest Cancer Research UK data shows 4300 new cases in England each year with an average 10 year survival rate of 45%. Many of these cases present with haematogenous metastatic disease [1]. The mainstay of treatment in adults with STS is generally adjuvant or neoadjuvant chemotherapy, surgery and radiotherapy.

Our patient interestingly presented with a primary RMS arising within the latissimus dorsi free muscle flap eleven years after it was transferred to his right leg. This was on a background of significant traumatic injury followed by a prolonged course of osteomyelitis, repeat trauma and surgery and chronic pain. One of the key learning points is that this may theoretically occur within any type of soft tissue transfer. The other is within this patient's presentation. Eleven years later when he represented with pain and swelling, the most likely, and possibly concurrent, diagnosis was osteomyelitis as demonstrated by the MRI. On exploration and bone biopsy it is therefore prudent to not only send samples for microbiology but also histopathological analysis. Without this it is likely the diagnosis and potentially prognosis would have been delayed.

It is difficult to pin point the exact cause of this sarcoma and whether or not this may have occurred still if the muscle was not transferred. Most cases of sarcoma arise de novo however risk factors include family or genetic history (RMS may occur in patients with Gorlins Syndrome), exposure to toxins such as vinyl chloride or arsenic, radiotherapy or HIV. Our patient had no history of risk factors. A possible theory in this case may include a sustained inflammatory response due to chronic infection and repeated trauma.

Within the context of lower limb salvage and rehabilitation, this case demonstrates very well that limb amputation should also be considered, in select cases, as a good surgical procedure and not failure. Our patient following this has been able to integrate well back into society, has returned to work and is now pain free walking well on a prosthesis.

CONSENT

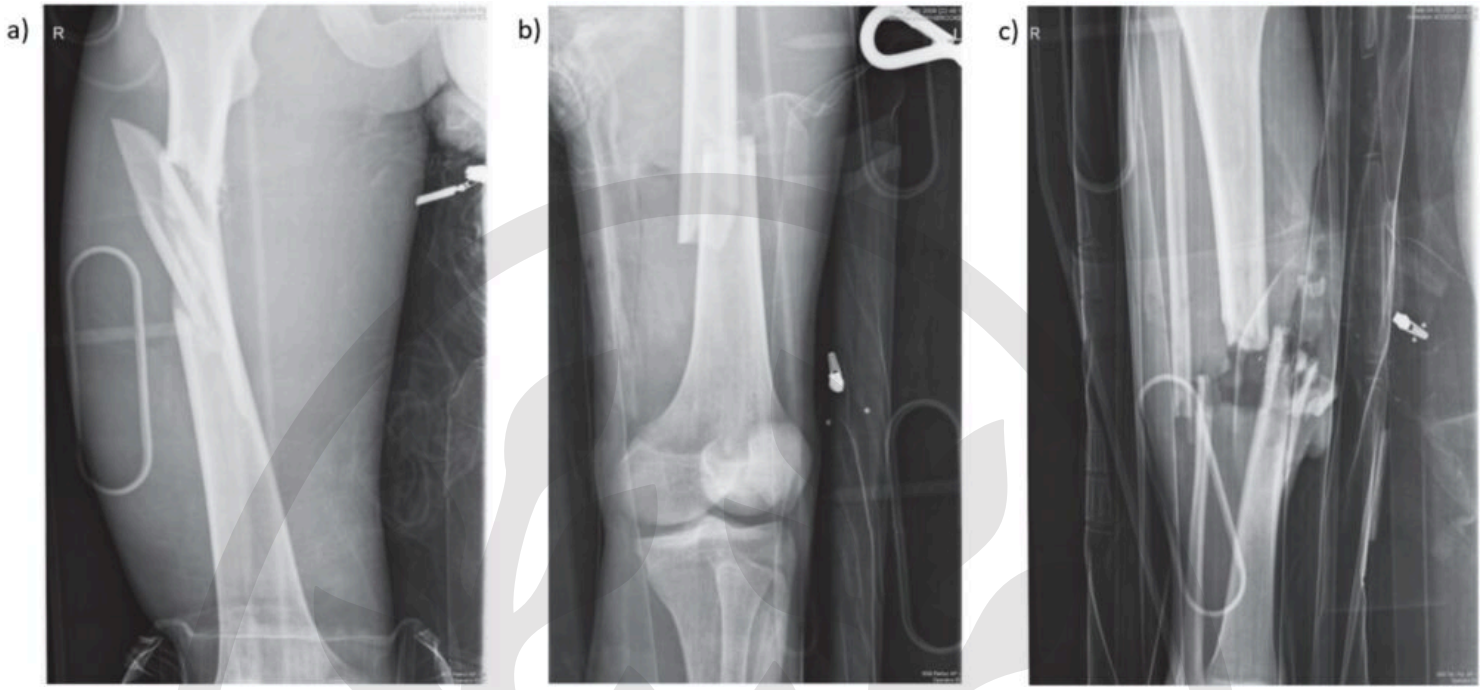
In accordance with ethical guidelines, the authors confirm that informed consent was obtained from the patient for the publication of their case report, documenting the process in their medical records and ensuring voluntary participation with full understanding of the implications involved.

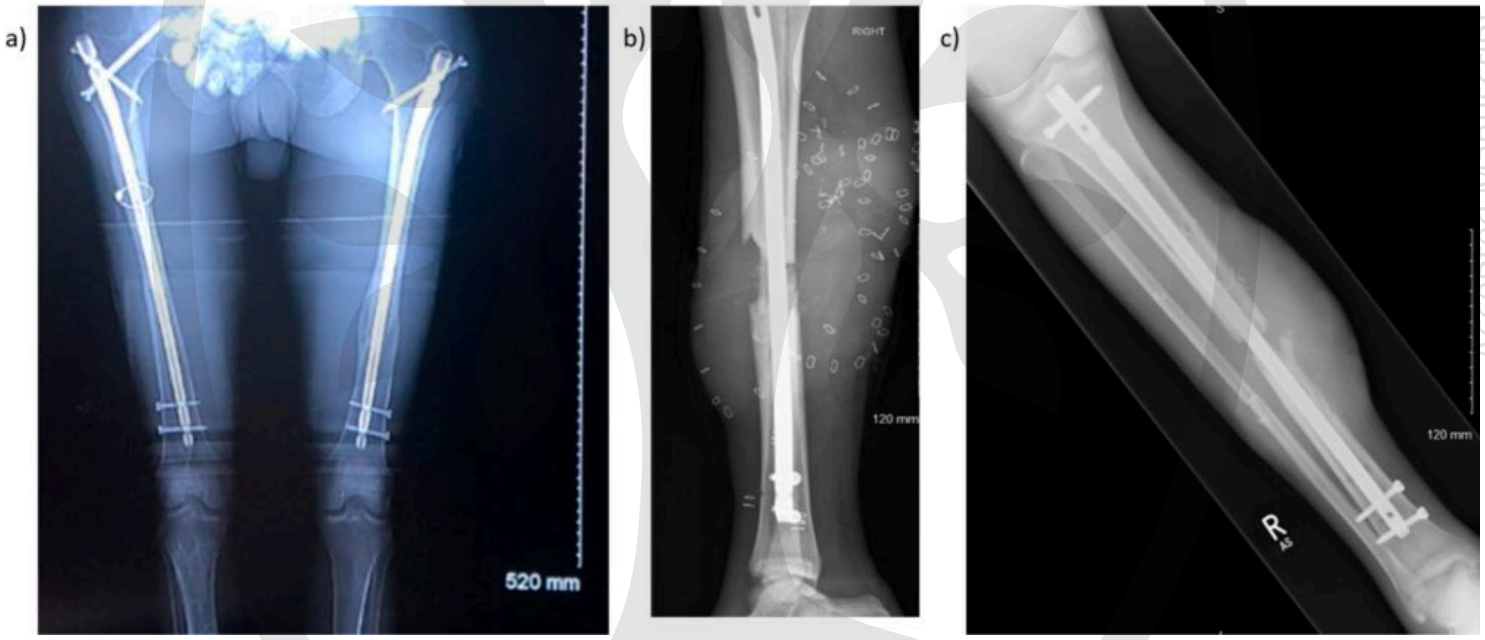
DECLARATION OF INTEREST

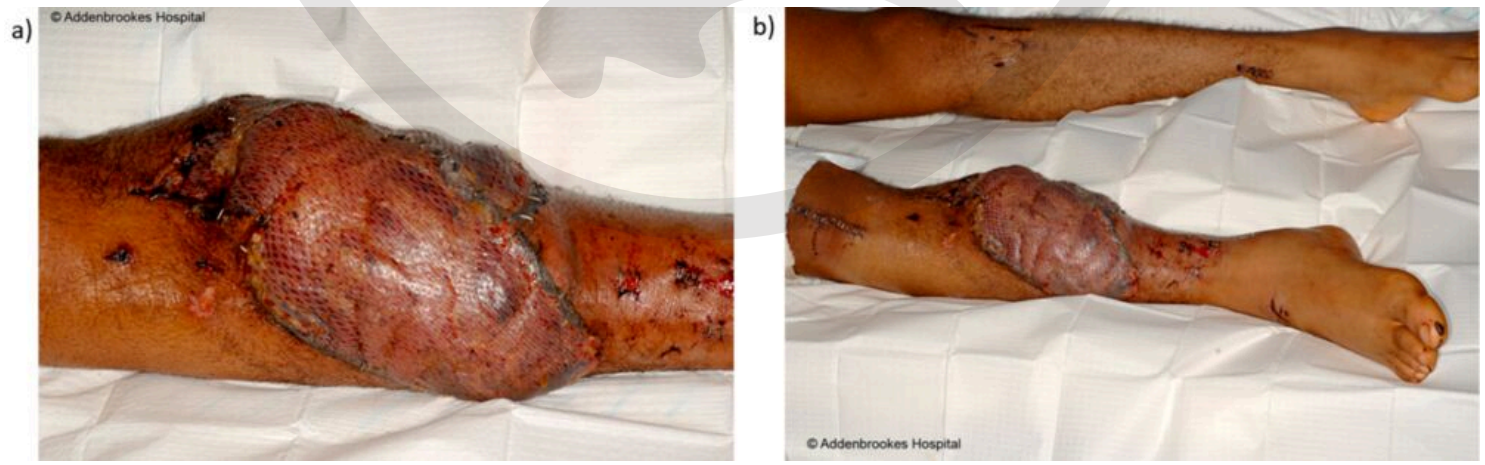
The authors declare no conflict of interest and confirm no funding was required for this case report.

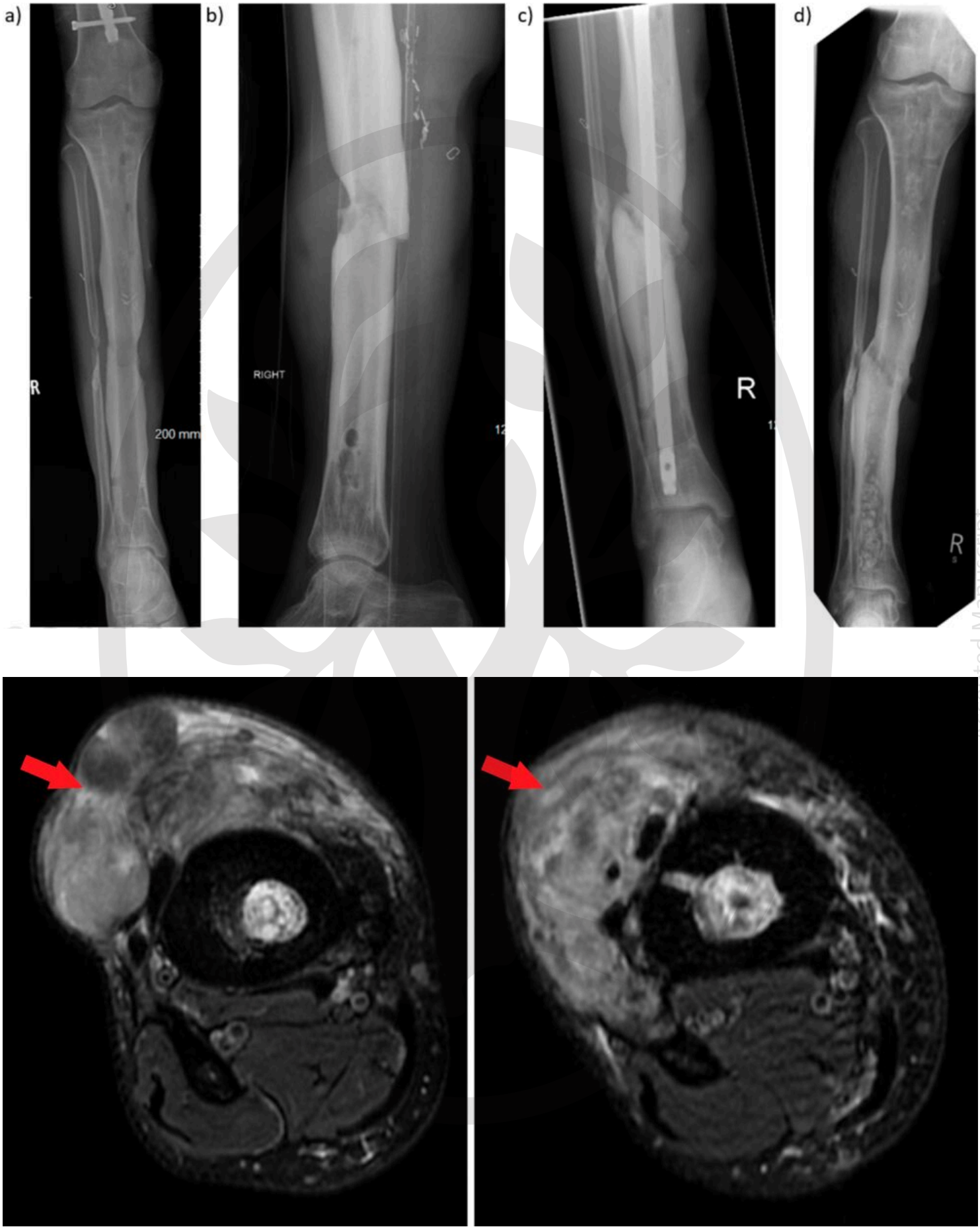
REFERENCES

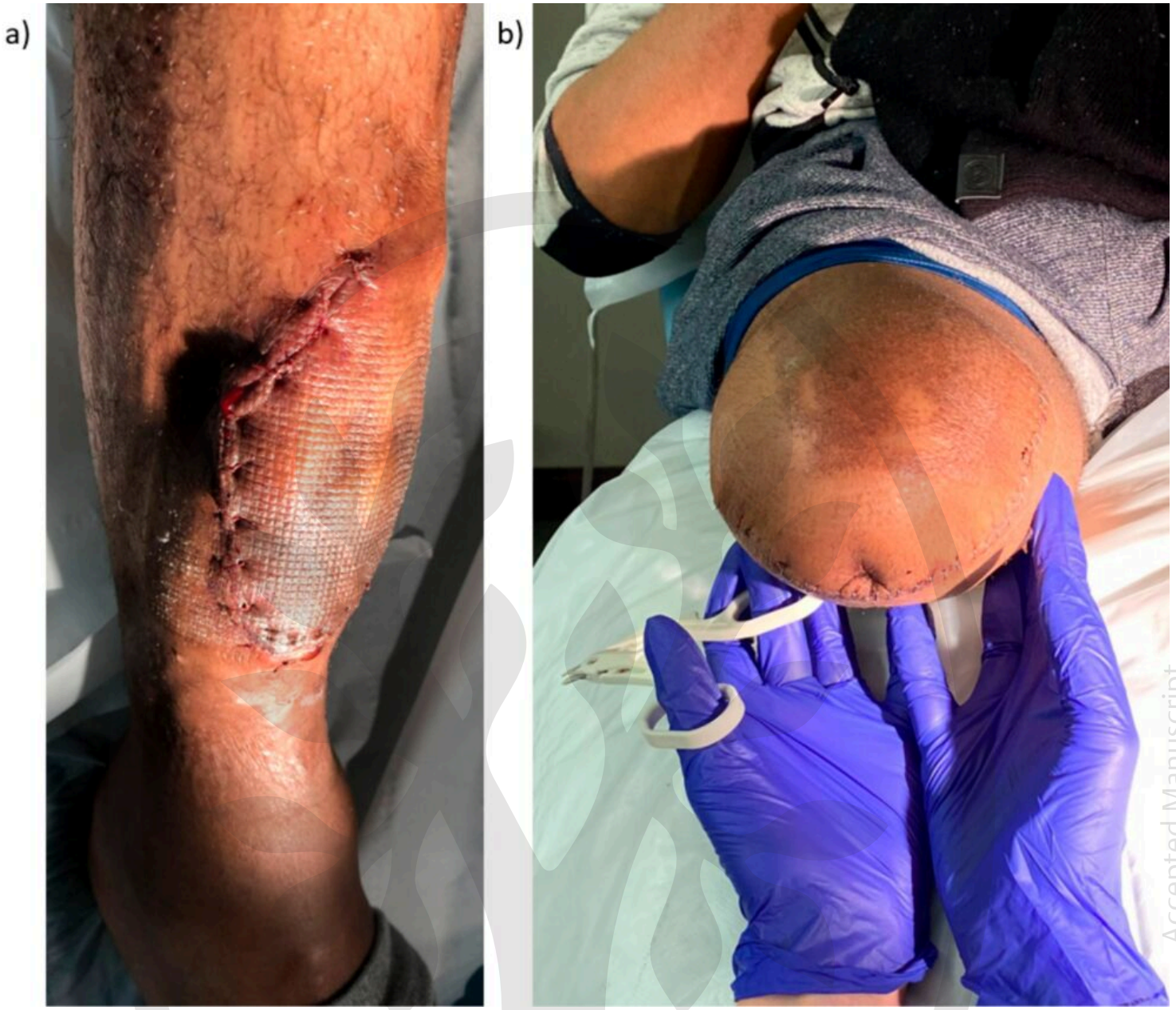
1. Soft tissue sarcoma statistics. Cancer Research UK. Published May 14, 2015. https://www.cancerresearchuk.org/health-professional/cancer-statistics/statistics-by-cancer-type/soft-tissue-sarcoma
2. Chen J, Liu X, Lan J, et al. Rhabdomyosarcoma in Adults: Case Series and Literature Review. *International Journal of Women's Health*. 2022;Volume 14:405-414. doi:https://doi.org/10.2147/ijwh.s352143
3. Fisher MH, Woods J, Bartlett EK, Nelson JA. A rare case of rhabdomyosarcoma identified in a VRAM flap after lower extremity reconstruction. *Journal of surgical case reports*. 2023;2023(3). doi:https://doi.org/10.1093/jscr/rjad083
4. Dangoor A, Seddon B, Gerrand C, Grimer R, Whelan J, Judson I. UK guidelines for the management of soft tissue sarcomas. *Clinical Sarcoma Research*. 2016;6(1). doi:https://doi.org/10.1186/s13569-016-0060-4
5. Ferrari A, Dileo P, Casanova M, et al. Rhabdomyosarcoma in adults. *Cancer*. 2003;98(3):571-580. doi:https://doi.org/10.1002/cncr.11550

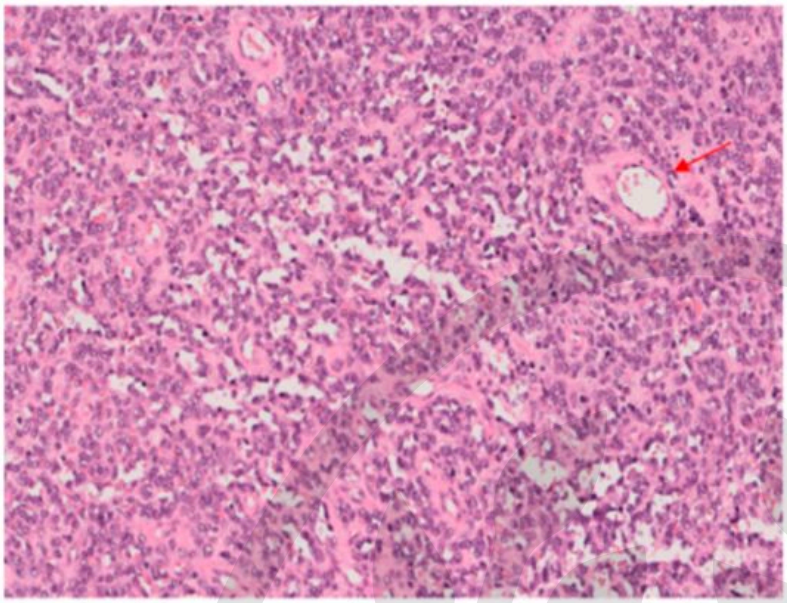




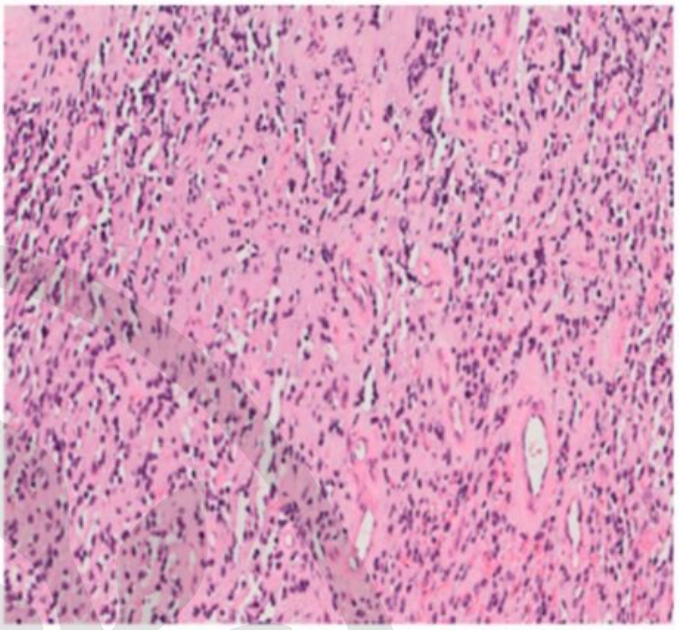




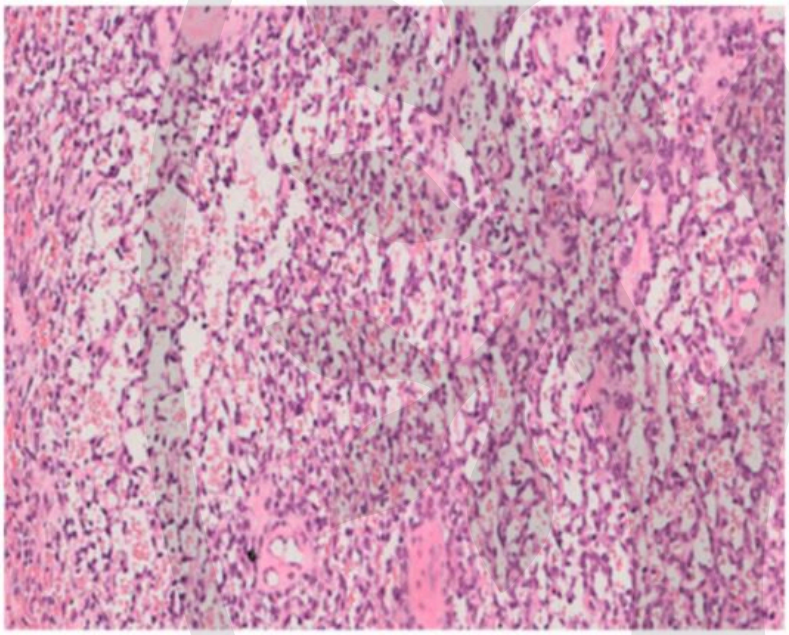




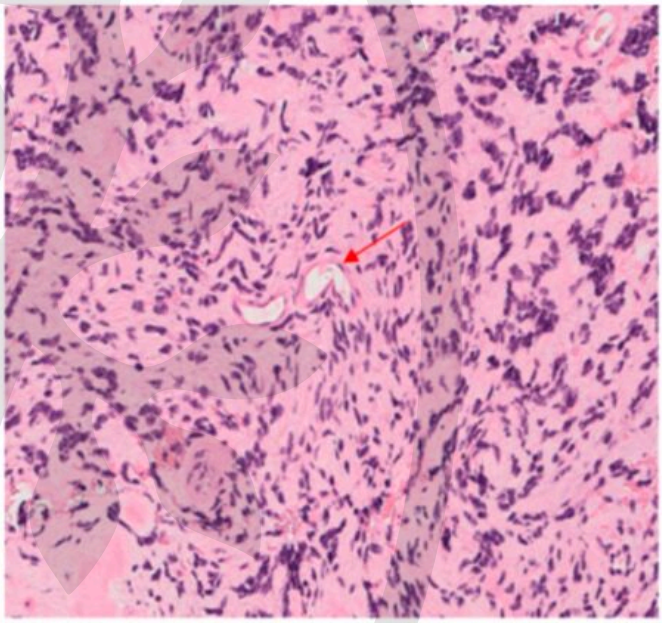
Densely cellular tumour comprising sheets of tumour cells with round hyperchromatic nuclei and scant cytoplasm



Tumour cells separated by a fibrous & hyalinised stroma focally



There is a suggestion of a pseudo- alveolar pattern in places



The tumour cells exhibit a spindled morphology in some areas

©Dr Alopa Malaviya