

CASE REPORT

Rare Metastatic Patterns in Rhabdomyosarcoma: A Report of Two Cases with Soft Tissue-to-Soft Tissue Spread

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ABSTRAK

Rabdomiosarkoma ialah sejenis sarkoma tisu lembut malignan yang jarang berlaku dan lazimnya menyerang kanak-kanak. Walaupun metastasis biasanya melibatkan paru-paru, tulang dan otak, metastasis dari tisu lembut ke tisu lembut adalah sangat jarang. Laporan kes ini membentangkan dua kes luar biasa rabdomiosarkoma yang menunjukkan metastasis antara tisu lembut. Kes pertama melibatkan seorang lelaki berusia 34 tahun dengan rabdomiosarkoma alveolar yang berasal dari tangan kanan dan merebak ke lengan atas, pelvis dan saraf tunjang. Kes kedua melibatkan seorang remaja perempuan berusia 14 tahun dengan rabdomiosarkoma paravertebra yang merebak ke saluran tulang belakang dan metastasis ke lengan bawah kiri. Kedua-dua pesakit menunjukkan perkembangan penyakit yang pesat walaupun telah menerima rawatan awal, menunjukkan sifat agresif rabdomiosarkoma serta corak metastasis yang luar biasa. Kes-kes ini menekankan keperluan bagi pengamal perubatan untuk mempertimbangkan corak metastasis yang jarang berlaku dalam rabdomiosarkoma, dengan penekanan kepada kepentingan pengimejan diagnostik yang menyeluruh dan pengurusan multidisiplin untuk penjagaan yang optimum. **Kata kunci:** Corak metastasis atipikal; metastasis tisu lembut; rabdomiosarkoma; sarkoma tisu lembut; subjenis alveolar

ABSTRACT

Rhabdomyosarcoma is a rare malignant soft tissue sarcoma, primarily affecting children. While common metastatic sites include the lungs, bones and brain, soft tissue-to-soft tissue metastasis is highly unusual. This report presents two rare cases of rhabdomyosarcoma demonstrating soft tissue-to-soft tissue metastasis. The first case involves a 34-year-old man with alveolar rhabdomyosarcoma originating in the right hand and metastasising to the upper arm, pelvis and spinal cord. The second case is a 14-year-old female with paravertebral rhabdomyosarcoma extending to the spinal canal and metastasising to the left forearm. Both patients exhibited rapid disease progression despite initial treatment, highlighting the aggressive nature of rhabdomyosarcoma and its atypical metastatic behaviour. These cases highlight the need for clinicians to consider rare metastatic patterns in rhabdomyosarcoma, emphasising the importance of comprehensive diagnostic imaging and multidisciplinary management for optimal care.

Keywords: Alveolar subtype; atypical metastatic pattern; rhabdomyosarcoma; soft tissue sarcoma; soft tissue metastasis

INTRODUCTION

Rhabdomyosarcoma is the most common malignant soft tissue sarcoma in children, constituting 5-8% of paediatric cancers and 50% of all childhood soft tissue sarcomas (Dondapati et al. 2021; Shouman et al. 2005). It originates from mesenchymal cells, typically affecting children under 10 years old (Shouman et al. 2005), with a male-to-female ratio of 1.5:1. Although primarily a paediatric malignancy, rhabdomyosarcoma is rare in adults (Dondapati et al. 2021). The two primary histological subtypes of this sarcoma are the embryonal and alveolar types (Dondapati et al. 2021; Shouman et al. 2005), while other less common variants include pleomorphic, botryoid and nonspecific subtypes (Shouman et al. 2005). Embryonal rhabdomyosarcoma predominantly occurs in the orbit, whereas alveolar rhabdomyosarcoma, often found in extremities, demonstrates the highest rates of recurrence and metastasis (Shouman et al. 2005). Common metastatic sites include the lungs, bones, bone marrow, liver, distant lymph nodes and brain (Dondapati et al. 2021; Shouman et al. 2005). However, soft tissue-to-soft tissue metastasis is exceedingly rare. This report details two unusual cases of rhabdomyosarcoma with such metastasis, illustrating the challenges in diagnosis and treatment.

CASE REPORT

Case 1

A 34-year-old male teacher presented with a painless swelling on his right palm, which had been progressively enlarging over three months before he sought medical attention. The swelling subsequently ulcerated, bled and became painful, with the pain described as sharp and localised, partially relieved by analgesics (Figure 1A). He denied any history of hand infections, systemic symptoms, such as fever, night sweats, or chronic cough, and had no known exposure to tuberculosis. One month later, he developed bilateral lower limb swelling and heaviness, accompanied by left foot numbness. He also

experienced intermittent urinary incontinence and constipation requiring manual stool evacuation, though he could still ambulate at a slower pace without significant limb pain.

On examination, the patient appeared alert and non-cachectic, with stable vital signs. Lung examination revealed normal breath sounds without crepitations, and the abdomen was soft, non-tender, with no organomegaly. There were no palpable lymph nodes in the cervical, axillary or inguinal regions. Local examination of the right hand revealed a 6 x 5 cm ulcerated, firm mass over the hypothenar region, with no signs of infection and intact neurovascular status. Bilateral lower limb examination showed gross swelling, more pronounced on the left side, without erythema, tenderness or pitting oedema. While the right lower limb demonstrated normal motor and sensory function, the left lower limb had reduced sensation at the L3 dermatome and significantly diminished motor strength (grade 4 for L2 and L3, and grade 0 for L4, L5, and S1). Reflexes were decreased at the left knee and absent at the ankle, with equivocal Babinski signs bilaterally. Sensation over the perianal region (S2–S4) was diminished, and anal tone was lax.

Laboratory investigations revealed anaemia (Hb 9.4 g/dL), elevated alkaline phosphatase (ALP) (288 IU/L) and lactate dehydrogenase (LDH) levels (1500 U/L). Other tests were unremarkable. Ultrasound of the right hand initially suggested a hematoma, prompting incision and drainage, but the wound deteriorated postoperatively. A subsequent magnetic resonance imaging (MRI) of the hand later revealed a lobulated, heterogenous soft tissue lesion measuring 6.3 x 5.1 x 4.5 cm, isointense on T1 and hyperintense on T2, with heterogeneous gadolinium enhancement, suggestive of a large soft tissue tumour (Figure 1B). A trucut biopsy confirmed a diagnosis of high-grade alveolar rhabdomyosarcoma.

The patient underwent four cycles of chemotherapy, but the tumour progressed, spreading to the right arm. Mid-cycle MRI revealed a multilobulated soft tissue mass in the right arm with extension to the shoulder joint, right posterior pleura, intercostal muscles and

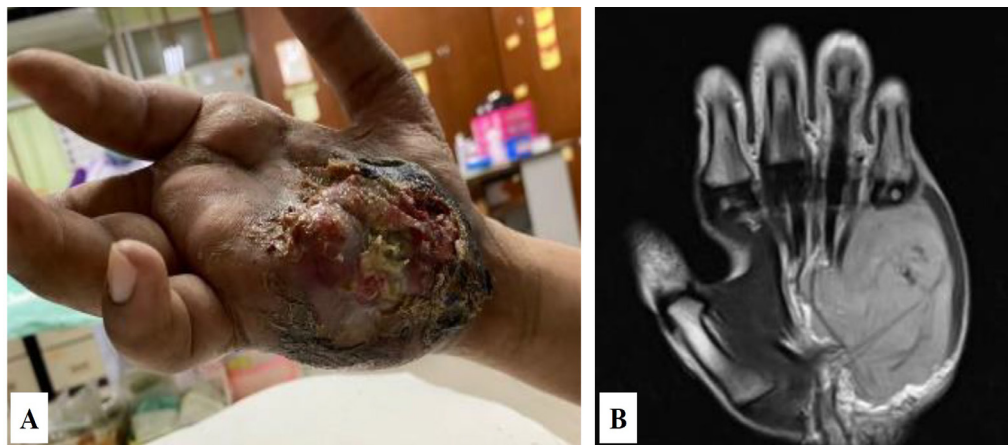


FIGURE 1: Case 1 (A) A fungating and ulcerated soft tissue mass over the hypothenar region of the right hand; (B) Coronal T1-weighted MR image showing a lobulated, homogeneous lesion involving the hypothenar muscles.

multiple lytic bone lesions from the humeral head to the proximal humerus. Following five additional chemotherapy cycles, repeat computed tomography (CT) imaging showed metastases in the left sacral ala with extension into the pelvic cavity and spinal canal. Lumbosacral MRI performed when the patient developed acute neurological symptoms in the lower limbs revealed an enhancing left sacral alar mass with similar extensions. Due to rapid disease progression, palliative care was initiated. The patient ultimately succumbed to disease progression, with widespread metastases, including to the brain and lungs.

Case 2

A 14-year-old right-handed female presented with a one-week history of urinary retention and worsening right lower limb weakness that had significantly deteriorated on the day of admission. She reported experiencing mild weakness in the right lower limb two weeks earlier but had remained ambulatory and attended school during that time. On admission, she had complete loss of bladder and bowel control. She denied any history of trauma, falls or fever, and there was no family history of malignancy or tuberculosis. Additionally, she reported a painless swelling

on her left forearm that had been progressively enlarging over the past month, accompanied by unintentional weight loss and reduced appetite.

Clinical examination revealed intact upper limb neurology but tenderness over the mid-thoracic spine. Bilateral lower limb power was grade 0 from L2 to S1, with diminished sensation below the T6 level. Reflexes were absent in both lower limbs, Babinski's reflex was upgoing on the left, and anal tone was lax with reduced perianal sensation. Examination of the left forearm showed a firm, non-tender mass measuring 10 x 7 cm over the medial aspect of the proximal forearm. The swelling was hard, not attached to the skin and had well-defined margins. Peripheral nerve assessments for the median, ulnar and radial nerves were intact. There were no palpable axillary lymph nodes and a breast examination was unremarkable. Blood investigations showed no abnormalities.

Plain radiograph of the left forearm showed soft tissue shadow. MRI studies revealed a lobulated soft tissue lesion in the right paravertebral region at the T2-T5 vertebral levels, measuring 5.5 x 4.6 x 4.3 cm, with extension into the spinal canal. The lesion appeared isointense on T1, hyperintense on T2, not suppressed in STIR sequence and heterogeneously enhanced with post-contrast imaging (Figure 2). A similar lesion

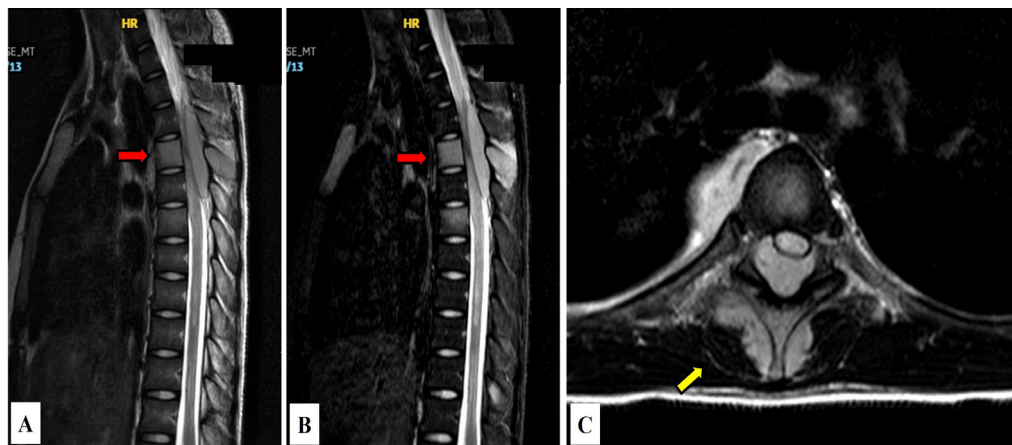


FIGURE 2: Case 2. MR images of the thoracic spine: (A) Sagittal T1-weighted (T1WI); (B) sagittal STIR sequence; and (C) axial T2-weighted at the T4 vertebral level. T4 was marked with a red arrow in images (A) and (B). A well-defined, lobulated right paravertebral soft tissue lesion at the T2–T5 vertebral levels was highlighted with a yellow arrow in image (C). The lesion appeared iso-hyperintense to muscle on T1WI (A), hyperintense on T2WI (C), not suppressed on FLAIR and STIR sequences (B), and demonstrated heterogeneous enhancement on post-contrast imaging. The lesion extended posteriorly, invaded the erector spinae muscles bilaterally and compressed the posterior thecal sac through the spinal canal.

was identified in the right paravertebral region at the T9-T10 levels. MRI of the left forearm showed a heterogeneous lobulated mass in the volar compartment, involving the flexor digitorum profundus and flexor pollicis longus muscles, extending from the common flexor tendon to the mid-forearm (Figure 3). The neurovascular bundle was compressed but remained intact proximally and distally. Additional smaller lesions were identified in the medial arm and forearm.

The patient underwent a T3-T6 posterior laminectomy with tumour debulking; however, her neurological status remained unchanged postoperatively. Histopathological examination of the spinal tumour confirmed rhabdomyosarcoma. Her condition rapidly deteriorated, with sudden-onset dyspnoea and severe abdominal distension due to ascites. Biopsy of the left forearm mass confirmed metastatic rhabdomyosarcoma. Secondary complications, including urosepsis and grade 3 sacral sores, further compromised her general condition. Despite medical interventions, she succumbed to her illness.

DISCUSSION

The first case involved a 34-year-old male diagnosed with alveolar rhabdomyosarcoma originating in the right hand, which exhibited an atypical metastatic pattern, spreading to the soft tissues of the arm, pelvic cavity and spinal cord. The second case described a 14-year-old female with an extradural, extramedullary rhabdomyosarcoma originating in the right paravertebral region at T4-T5, extending into the spinal cord and metastasising to the soft tissue of the left forearm. A key feature of these cases is the unusual occurrence of soft tissue-to-soft tissue metastasis, a rare phenomenon in rhabdomyosarcoma.

Soft tissue sarcomas are rare malignancies, comprising 1-2% of adult cancers (Younger et al. 2020). According to a study by Younger et al. (2020), about one-third of adolescents and young adults diagnosed with soft tissue sarcomas presented with synchronous metastases, while the remainder developed metastases after an average interval of 16 months. In both cases discussed here, the primary tumour and

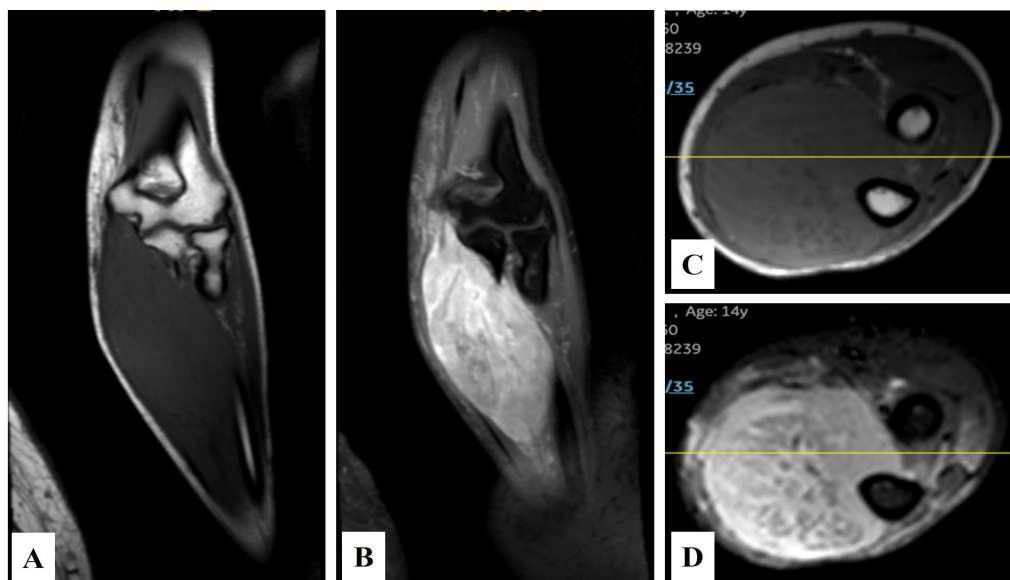


FIGURE 3: Case 2. MR images of the left forearm: (A) Coronal T1-weighted (T1WI); (B) coronal T1WI post-gadolinium; (C) axial T1WI, and (D) axial STIR. The images revealed a heterogeneous, lobulated mass in the volar compartment extending from the common flexor tendon to the mid-forearm. The lesion appeared iso-hypointense on T1WI (A and C), hyperintense on T2-weighted imaging, heterogeneously enhanced post-contrast (B), and not suppressed on STIR (D).

metastatic lesions were present at the time of initial diagnosis. The first patient presented with a hand mass and concurrent cauda equina syndrome due to spinal cord compression, while the second patient exhibited acute bilateral lower limb paralysis alongside a forearm mass, both indicative of synchronous metastatic disease.

Histological subtypes of soft tissue sarcomas, as identified in the Younger et al. (2020) study, include leiomyosarcoma, synovial sarcoma, Ewing sarcoma, rhabdomyosarcoma, pleomorphic sarcoma, liposarcoma and malignant peripheral nerve sheath tumours (Younger et al. 2020). These subtypes influence the differential diagnosis for patients presenting with soft tissue masses. Primary tumour sites in the study were most commonly extremities (42%), followed by retroperitoneum (15%), intra-abdominal and pelvic regions (9%), gynaecological sites (8%), intrathoracic locations (8%) and head and neck (7%). Consistent with this distribution, the primary tumour in the first

case was in the extremity, while the second case involved an intrathoracic origin.

Rhabdomyosarcoma predominantly affects children, particularly in its embryonal subtype. In adolescents and young adults, the alveolar subtype is more common and is associated with poorer outcomes due to its aggressive nature and higher likelihood of lymph node involvement and metastasis. The alveolar subtype accounts for 63% of cases in this age group and is often linked to the expression of the PAX-FOXO1 fusion gene, a marker of poor prognosis (Younger et al. 2020). In this series, the first patient succumbed to the disease within 15 months, while the second, younger patient passed away within just three months, underscoring the aggressive course of this malignancy in both cases.

Rhabdomyosarcoma typically spreads via the haematogenous route, with common metastatic sites including the lungs, bones, lymph nodes, liver and brain (Chen et al. 2022). Staging investigations such as chest CT scans, bone scans

and positron emission tomography imaging are crucial for detecting locoregional and distant metastases, as well as identifying unusual patterns of spread (de la Monte et al. 1986). The metastasis pattern observed in these cases, soft tissue-to-soft tissue, highlights a unique and rare progression pathway for rhabdomyosarcoma, challenging conventional understanding of its metastatic behaviour and emphasising the need for comprehensive diagnostic imaging and tailored therapeutic approaches.

CONCLUSION

These cases emphasise the rare phenomenon of rhabdomyosarcoma metastasising to soft tissue, challenging conventional metastatic patterns. They underscore the importance of early detection, comprehensive imaging and multidisciplinary management to address the aggressive nature of such cases and improve clinical outcomes.

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Conflict of interest: The authors declare no conflicts of interest.

Ethics statement: This case report was prepared in accordance with the ethical principles outlined in the Declaration of Helsinki. Approval for the study was granted by the institutional ethical board (Ref: USMKK/PPP/JEPeM/21080582). As it was a retrospective review of existing medical records, the requirement for informed consent was waived by the approving ethics committees. However, consent was obtained from patients

whose photographs were taken and included in the publication. All patient data were anonymised and de-identified to ensure confidentiality and privacy throughout the research process.

REFERENCES

- Chen, J., Liu, X., Lan, J., Li, T., She, C., Zhang, Q., Yang, W. 2022. Rhabdomyosarcoma in adults: case series and literature review. *Int J Womens Health* **14**: 405-14. <https://doi.org/10.2147/IJWH.S352143>
- de la Monte, S.M., Hutchins, G.M., Moore, G.W. 1986. Metastatic behavior of rhabdomyosarcoma. *Pathol Res Pract* **181**(2): 148-52. [https://doi.org/10.1016/s0344-0338\(86\)80003-6](https://doi.org/10.1016/s0344-0338(86)80003-6)
- Dondapati, M., Reyes, J.V.M., Ahmad, S., Stern, A.S., Lieber, J.J. 2021. Rare adult subtype of rhabdomyosarcoma, a common childhood soft tissue carcinoma. *J Investig Med High Impact Case Rep* **9**: 23247096211042236. <https://doi.org/10.1177/23247096211042236>
- Shouman, T., El-Kest, I., Zaza, K., Ezzat, M., William, H., Ezzat, I. 2005. Rhabdomyosarcoma in childhood: A retrospective analysis of 190 patients treated at a single institution. *J Egypt Natl Canc Inst* **17**(2): 67-75.
- Younger, E., Husson, O., Asare, B., Benson, C., Judson, I., Miah, A., Zaidi, S., Dunlop, A., Al-Muderis, O., van Houdt, W.J., Jones, R.L., van der Graaf, W.T.A. 2020. Metastatic soft tissue sarcomas in adolescents and young adults: a specialist center experience. *J Adolesc Young Adult Oncol* **9**(6): 628-38. <https://doi.org/10.1089/jayao.2020.0010>