

# Metastatic Epithelioid Sarcoma- A Case Report

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## Introduction

Epithelioid sarcoma (ES) is a rare soft tissue sarcoma distinguished by epithelioid cell nodules that are immunoreactive to CD34, cytokeratins (CKs), and epithelial membrane antigen. (1) It is commonly found in young adults with a mean age of incidence of 35 years and manifests as a result of SMARCB1/INI1 deficiency or other proteins of the SWI/SNF complex. (2)

## Background

Commonly misdiagnosed as infected warts, ulcers, or abscesses, (3) it represents less than 1% of all soft tissue sarcomas, (4) with a prevalence of 0.4 per 1 million population. (5) Despite vigorous multimodal therapies, it has a fatal outcome in ~50% of cases. (6)

## Case

A 23 year old adult male diagnosed with epithelioid sarcoma presented with a several weeks' history of worsening pain and limited movement in the left lower limb, general weakness and decreased working ability. Inspection revealed a necrotic area measuring 10-7-7 cm on the left ankle. Tumors were palpated in the left popliteal fossa and the left groin. Imaging results of PET-CT scan of the whole body, CT scans of the pelvis and lower limb revealed regional metastatic disease. An above the knee amputation, along with inguinal lymph node dissection was decided as the definitive course of treatment.

## Discussion

For further evaluations and support of clinical decision-making, scientific publications from the last 10 years were reviewed using the keyword 'Epithelioid sarcoma' on PubMed by applying appropriate filters. It has two distinct variants: classic ES and proximal ES. The former is usually observed in distal extremities manifesting as a subcutaneous or deep dermal mass, with uniform epithelioid cells having central necrosis, while the latter usually favors proximal limbs, limb girdles and midline of the trunk with more atypical cells of variable rhabdoid morphology. (7) In addition to having a high recurrence rate and localized lymph node metastases, ES is renowned for its aggressive behavior, leading to an expected survival of about a year. Wide surgical resection is accepted as a reliable treatment of localized ES. Alternative approaches include administration of doxorubicin, tazemetostat and immunotherapy (8)

## Conclusion

Treatment selection depends on characteristics of the disease, staging upon presentation, regional lymph node involvement, presence of comorbidities, and the patient's performance status. (9) The likelihood of a misdiagnosis can be reduced, resulting in prompt and efficient management of epithelioid sarcoma, by identifying common misinterpretations of ES and putting strategies like immunohistochemical staining, multidisciplinary collaboration, and raising awareness among healthcare professionals into practice.