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# An unusual cause of hepatic venous outflow obstruction: Epithelioid angiomyolipoma of the kidney

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**Authors:**
Muhammad Ayob[1,2]
Muhammed Vally[1,2]
Mohammed M. Ismail[1,2]
Vikash Lala[1,2]
Adam Mahomed[1,2]

**Affiliations:**
[1]Division of Medical Gastroenterology, Department of Internal Medicine, Faculty of Health Sciences, University of the Witwatersrand, Johannesburg, South Africa

[2]Division of Medical Gastroenterology, Department of Internal Medicine, Wits University Donald Gordon Medical Centre, Johannesburg, South Africa

**Corresponding author:**
Muhammad Ayob,
mayob97@gmail.com

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Epithelioid angiomyolipoma (EAML) is a rare, aggressive perivascular epithelioid cell tumour (PEComa) with malignant potential. A 36-year-old man presented with a large hypervascular left renal mass. Computed tomography showed a 12.5 cm heterogeneous tumour with thrombus extending from the renal vein into the inferior vena cava and right atrium, complicated by Budd–Chiari syndrome. Biopsy confirmed Melan-A–positive epithelioid cells. Epithelioid angiomyolipoma can mimic advanced renal cell carcinoma. Management included selective transarterial angioembolisation to reduce haemorrhage risk.

**Contribution:** This report documents a rare EAML presentation complicated by Budd–Chiari syndrome, highlighting the utility of pre-procedural angioembolisation for managing aggressive, hypervascular variants with extensive venous involvement.

**Keywords:** epithelioid angiomyolipoma; renal mass; tumour thrombus; Budd–Chiari syndrome; selective transarterial angioembolisation; inferior vena cava.

## Introduction

Renal epithelioid angiomyolipoma (EAML) is a rare mesenchymal neoplasm belonging to the perivascular epithelioid cell (PEComa) family.[1] Although EAML accounts for approximately 4.6% – 7.7% of all renal angiomyolipomas (AMLs),[2] it is clinically significant because of its recognised malignant potential. In contrast to the typically indolent course of classic AML, EAML has been associated with aggressive features, including local invasion, vascular extension, recurrence and distant metastasis, most commonly to the lungs, liver and regional lymph nodes.[1,3] Its variable behaviour and overlap with renal cell carcinoma (RCC) on imaging further complicate diagnosis and management.

Morphologically, EAML is defined by a predominant population of at least 80% epithelioid cells, characterised by large polygonal cells with eosinophilic cytoplasm, marked nuclear polymorphism, prominent nucleoli and minimal adipose content.[4,5] This 'fat-poor' profile creates a formidable diagnostic dilemma, as radiological features such as a lack of macroscopic fat and heterogeneous enhancement frequently overlap with RCC.[6]

Definitive diagnosis requires immunohistochemical confirmation, with EAML typically expressing melanocytic markers such as human melanoma black-45 (HMB-45) and melanoma antigen recognised by T cells 1 (Melan-A) while lacking the epithelial immunophenotype characteristic of RCC.5 Assessment of malignant potential relies on validated risk stratification systems, most notably the Brimo criteria.[7]

The combination of extensive tumour thrombus reaching the right atrium and secondary Budd–Chiari syndrome illustrates both the malignant potential of EAML and the importance of strategic preoperative management, including selective angioembolisation, in mitigating haemorrhagic risk. Here, we report a rare case of EAML with extensive venous invasion, illustrating the diagnostic challenges and clinical implications of high-risk disease. Although EAML is recognised to possess malignant potential, extensive proximal venous extension is uncommon.[8,9] Furthermore, Budd–Chiari syndrome occurring secondary to EAML-associated tumour thrombus is exceptionally rare and remains poorly characterised in the literature.[8,9]

Therefore, the purpose of this report is to highlight the diagnostic challenges posed by this aggressive EAML variant, discuss the role of multimodal imaging and histopathological confirmation in establishing the diagnosis and demonstrate the value of preoperative angioembolisation in facilitating definitive surgical management.

## Case presentation

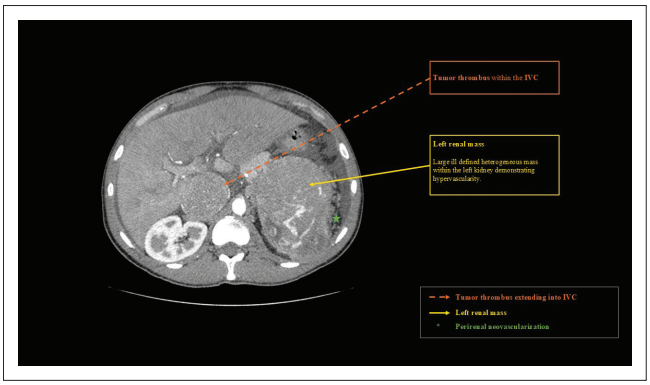
A 36-year-old man with no significant medical or surgical history presented with a 1-month history of progressive left-sided flank pain and abdominal distension. Physical examination revealed a large, ballotable left flank mass with moderate ascites and no splenomegaly. Initial laboratory evaluation demonstrated normocytic anaemia consistent with anaemia of chronic disease and mild renal impairment. Inflammatory markers showed a markedly elevated C-reactive protein with low procalcitonin. Liver function tests revealed hepatocellular injury with a preserved INR. Human immunodeficiency virus and hepatitis screening were negative (Table 1).

An urgent multi-phase computed tomography (CT) scan of the abdomen and pelvis was performed, which identified a 12.5 × 11.4 × 10.6 cm heterogeneous, hypervascular left renal mass with areas of central necrosis (Figure 1). Magnetic resonance imaging (MRI) was subsequently performed to better define the extent of venous involvement, confirming tumour thrombus extension to the atriocaval junction and associated Budd–Chiari physiology. Subsequent T2-weighted MRI sequences corroborated the CT findings, demonstrating a large heterogeneous renal mass with areas of high T2 signal intensity corresponding to cystic or necrotic components (Figure 2 and Figure 3). The lesion demonstrated an aggressive, infiltrative growth pattern, with a tumour thrombus extending from the left renal vein into the inferior vena cava (IVC), reaching the atriocaval junction. Associated findings included hepatic venous outflow obstruction with mottled hepatic enhancement, in keeping with Budd–Chiari physiology, as well as an incidental right lower lobe segmental pulmonary

**TABLE 1:** Baseline laboratory findings on admission.

| Category | Test | Result | Reference range |
|---|---|---|---|
| Haematology | Haemoglobin | 10.5 g/dL | 13.8–18.8 |
| | MCV | 87.9 fL | 79–100 |
| | MCH | 25.8 pg | 27–35 |
| | Platelets | 360 × $10^9$/L | 150–450 |
| | WCC | 8.08 × $10^9$/L | 4.0–12.0 |
| Iron profile | Serum iron | 2.4 µmol/L | 11.6–31.3 |
| | Transferrin | 19.1 µmol/L | 20.2–44.8 |
| | % Saturation | 6.30% | 20–55 |
| | Ferritin | 631 ng/mL | 20–300 |
| Renal function | Creatinine | 127 µmol/L | 80–115 |
| | eGFR (CKD-EPI) | 62 mL/min | > 90 |
| | Urea | 4.7 mmol/L | 2.1–7.1 |
| Inflammatory markers | CRP | 54 mg/L | < 5 |
| | Procalcitonin | 0.09 ng/mL | < 0.05 |
| Liver function | ALT | 130 IU/L | < 50 |
| | AST | 74 IU/L | < 50 |
| | GGT | 270 IU/L | < 64 |
| | ALP | 206 IU/L | 53–128 |
| | Total bilirubin | 10 µmol/L | 3–26 |
| | Conjugated bilirubin | 7 µmol/L | 2–7 |
| | Albumin | 29 g/L | 35–50 |
| Coagulation | INR | 1.22 | 1.00–1.25 |
| | aPTT | 35.7 s | 24.6–31.2 |
| Genetics and thrombophilia screen | Factor V Leiden | Not detected | - |
| | Prothrombin G20210A | Not detected | - |
| | JAK2 V617F | Not detected | - |
| Viral screen | HIV (4th Gen) | Negative | - |
| | Hepatitis B surface Ag | Negative | - |
| | Hepatitis C Ab | Negative | - |

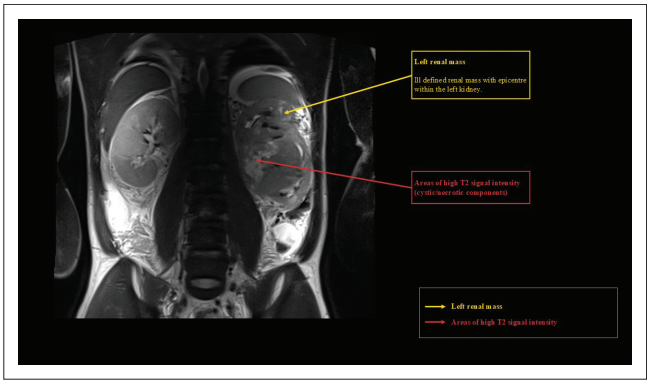
MCV, mean corpuscular volume; MCH, mean corpuscular haemoglobin; WCC, white cell count; eGFR, estimated glomerular filtration rate; CKD-EPI, Chronic Kidney Disease Epidemiology Collaboration; CRP, C-reactive protein; ALT, alanine aminotransferase; AST, aspartate aminotransferase; GGT, gamma-glutamyl transferase; ALP, alkaline phosphatase; INR, international normalised ratio; aPTT, activated partial thromboplastin time; HIV, human immunodeficiency virus.




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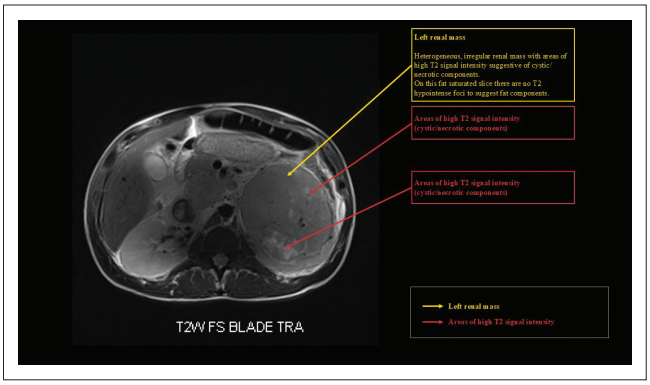
IVC, inferior vena cava.

**FIGURE 1:** Axial contrast-enhanced computed tomography showing a hypervascular left renal mass with inferior vena cava tumour thrombus.




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**FIGURE 2:** Coronal T2-weighted MRI showing a large heterogeneous left renal mass with cystic or necrotic components.




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**FIGURE 3:** Axial T2-weighted fat-suppressed MRI showing a heterogeneous left renal mass with cystic or necrotic components.

embolism. A subsequent thrombophilia screen was negative (Table 1).

The absence of macroscopic fat and the presence of infiltrative features suggested a non-classic, potentially malignant neoplasm, prompting a diagnostic core biopsy. Histological examination demonstrated a neoplasm composed of greater than 80% epithelioid cells arranged in nests and sheets, with abundant eosinophilic to granular cytoplasm, prominent nucleoli and occasional intranuclear pseudoinclusions. Definite tumour necrosis was not identified within the sampled biopsy cores. Immunohistochemically, the tumour cells showed strong positivity for HMB-45, Melan-A and smooth muscle actin (SMA), while staining was negative for AE1/AE3, PAX8, CD10, CK7, synaptophysin, inhibin, CD117 and S100. This immunophenotypic profile, characterised by co-expression of melanocytic and smooth muscle markers in the absence of epithelial or adrenal markers, established the diagnosis of EAML.

Following the definitive histological confirmation of EAML, the management strategy prioritised tumour devascularisation. Given the lesion's extreme hypervascularity and large size, selective transarterial angioembolisation was performed as a strategic necessity to mitigate the risk of life-threatening haemorrhage and facilitate subsequent surgical intervention. Staging evaluation, including CT imaging of the chest, abdomen, pelvis and brain, demonstrated no evidence of distant metastatic disease. Following successful embolisation, the patient was planned for radical nephrectomy with curative intent, as there was no evidence of metastatic disease and definitive resection offered the best opportunity for local disease control. However, the patient demised before definitive surgical intervention could be performed. Consequently, nephrectomy was not undertaken, and no resected specimen was available for pathological assessment.

## Discussion

The clinical presentation of a large renal mass accompanied by venous tumour thrombus extending into the IVC represents a formidable diagnostic and therapeutic challenge. In our case, the additional development of a Budd–Chiari-like clinical picture, characterised by hepatic venous outflow obstruction, immediately necessitated consideration of more aggressive pathological entities rather than the typically indolent renal masses commonly encountered.

In contrast to classic angiomyolipomas, which generally follow a benign course, EAML demonstrates a distinctly more aggressive phenotype.[1,2] The presence of venous extension signifies progression towards the more malignant end of the PEComa spectrum and raises concern for invasive biological behaviour. Such vascular involvement shifts the clinical approach towards a more comprehensive diagnostic evaluation, detailed cross-sectional imaging assessment and carefully planned multidisciplinary management.

Differentiating EAML from RCC remains a critical diagnostic hurdle, particularly as EAML often presents as a 'fat-poor' mass lacking the macroscopic adipose tissue typical of classic angiomyolipomas.[6] On non-contrast CT, these tumours usually appear heterogeneous, and many demonstrate a fast-in and slow-out enhancement pattern with arterial phase uptake and delayed retention.[6] Larger or high-risk lesions (≥5 cm) may instead show persistent enhancement and irregular intralesional vascularity.[6] Because T2-weighted MRI characteristics overlap substantially with clear cell RCC, imaging alone is frequently insufficient for definitive diagnosis.[6]

This diagnostic overlap necessitates tissue confirmation. Although percutaneous renal mass biopsy is generally safe and diagnostically useful, nondiagnostic results and an imperfect negative predictive value remain limitations, and bleeding-related complications such as haematoma, haematuria and haemorrhage have been reported.[10] Consequently, definitive classification depends on immunohistochemistry, with EAML typically expressing melanocytic markers including HMB-45, Melan-A and microphthalmia-associated transcription factor (MiTF), often accompanied by Cathepsin K expression and variable positivity for smooth muscle markers such as SMA and desmin.[5]

The differentiation of indolent from potentially malignant EAML is central to clinical decision-making. The Brimo criteria provide a histopathology-driven framework based on four adverse microscopic features: (1) ≥70% atypical epithelioid cells, (2) ≥2 mitoses per 10 high-power fields, (3) atypical mitotic figures, and (4) tumour necrosis, with high-risk behaviour predicted when ≥3 features are present.[7] In contrast, the Nese criteria incorporate five adverse parameters: (1) tumour necrosis, (2) carcinoma-like growth pattern, (3) extrarenal extension and/or renal vein involvement, (4) tumour size >7 cm and (5) association with tuberous sclerosis complex, stratifying cases into low, intermediate and high-risk groups according to cumulative features.[11] While Brimo emphasises cytologic atypia and mitotic activity, Nese integrates tumour size and anatomical extension, offering a broader risk model.

In the present case, the renal mass with central necrosis, as well as both renal vein and IVC involvement fulfills multiple adverse parameters under the Nese model, supporting high-risk classification. Preoperative application of the Brimo criteria is inherently limited, as core biopsy sampling may not reliably capture mitotic burden or atypical mitoses across a heterogeneous tumour. Thus, although biopsy findings demonstrated marked epithelioid atypia, definitive Brimo stratification requires comprehensive histopathologic assessment, whereas the Nese criteria are more readily applicable in the preoperative setting. This limitation was particularly relevant in our patient, as death before planned nephrectomy precluded examination of the entire tumour specimen and prevented definitive post-operative histopathological risk stratification.

The high-risk features present in our patient are particularly noteworthy because extensive venous involvement remains an uncommon manifestation of EAML.[2,3,11] While renal vein invasion is recognised within contemporary risk stratification models and reflects aggressive biological behaviour,[11] tumour thrombus extending into the IVC and cavoatrial junction has only rarely been described.[8,9] Furthermore, hepatic venous outflow obstruction resulting in Budd–Chiari syndrome is exceptionally uncommon, with the literature largely limited to isolated case reports.[9]

In resource-limited settings, management must therefore remain pragmatic and grounded in established surgical principles. While nephron-sparing surgery is reserved for smaller, low-risk lesions (<4 cm), infiltrative EAML with venous extension necessitates radical nephrectomy to achieve local control.[2,11,12] In our patient, the extensive tumour burden and cavoatrial extension further justified an aggressive surgical approach following preoperative angioembolisation. Additionally, post-operative surveillance remains central to monitoring for local recurrence and metastatic progression.

This case therefore adds to the limited body of evidence describing aggressive EAML with extensive venous extension and highlights important learning points for clinicians, radiologists and pathologists. Clinicians should recognise that EAML may present with life-threatening vascular complications despite its classification within the angiomyolipoma spectrum. Radiologists should consider EAML in the differential diagnosis of a hypervascular fat-poor renal mass associated with venous tumour thrombus. For pathologists, immunohistochemical confirmation remains essential to distinguish EAML from RCC and guide appropriate management.

# Conclusion

Failure to recognise the malignant potential of EAML has significant clinical consequences. Its capacity for venous extension and Budd–Chiari physiology demonstrates that EAML can behave aggressively and mimic advanced RCC. Accurate histopathological diagnosis is therefore essential to guide appropriate surgical management and prognostic counselling.

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## Competing interests

The authors, Muhammad Ayob; Muhammed Vally; Mohammed M. Ismail; Vikash Lala and Adam Mahomed, declare that they have no financial or personal relationships that may have inappropriately influenced them in writing this article.

## CRediT authorship contribution

Muhammad Ayob: Conceptualisation, Writing – original draft, Writing – review & editing. Muhammed Vally: Conceptualisation, Writing – original draft, Writing – review & editing. Mohammed M. Ismail: Conceptualisation, Writing – original draft, Writing – review & editing. Vikash Lala: Conceptualisation, Writing – original draft, Writing – review & editing. Adam Mahomed: Conceptualisation, Writing – original draft, Writing – review & editing. All authors reviewed the article, contributed to the discussion of results, approved the final version for submission and publication and take responsibility for the integrity of its findings.

## Ethical considerations

Written informed consent was obtained from all individual participants involved in the study.

An application for full ethical approval was submitted to the Wits Human Research Ethics Committee (Medical) on 08 January 2026. Ethical clearance to conduct this study was obtained from the University of the Witwatersrand Human Research Ethics Committee (Medical) (No. M260245).

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## Data availability

Data sharing is not applicable to this article as no new data were created or analysed in this study.

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