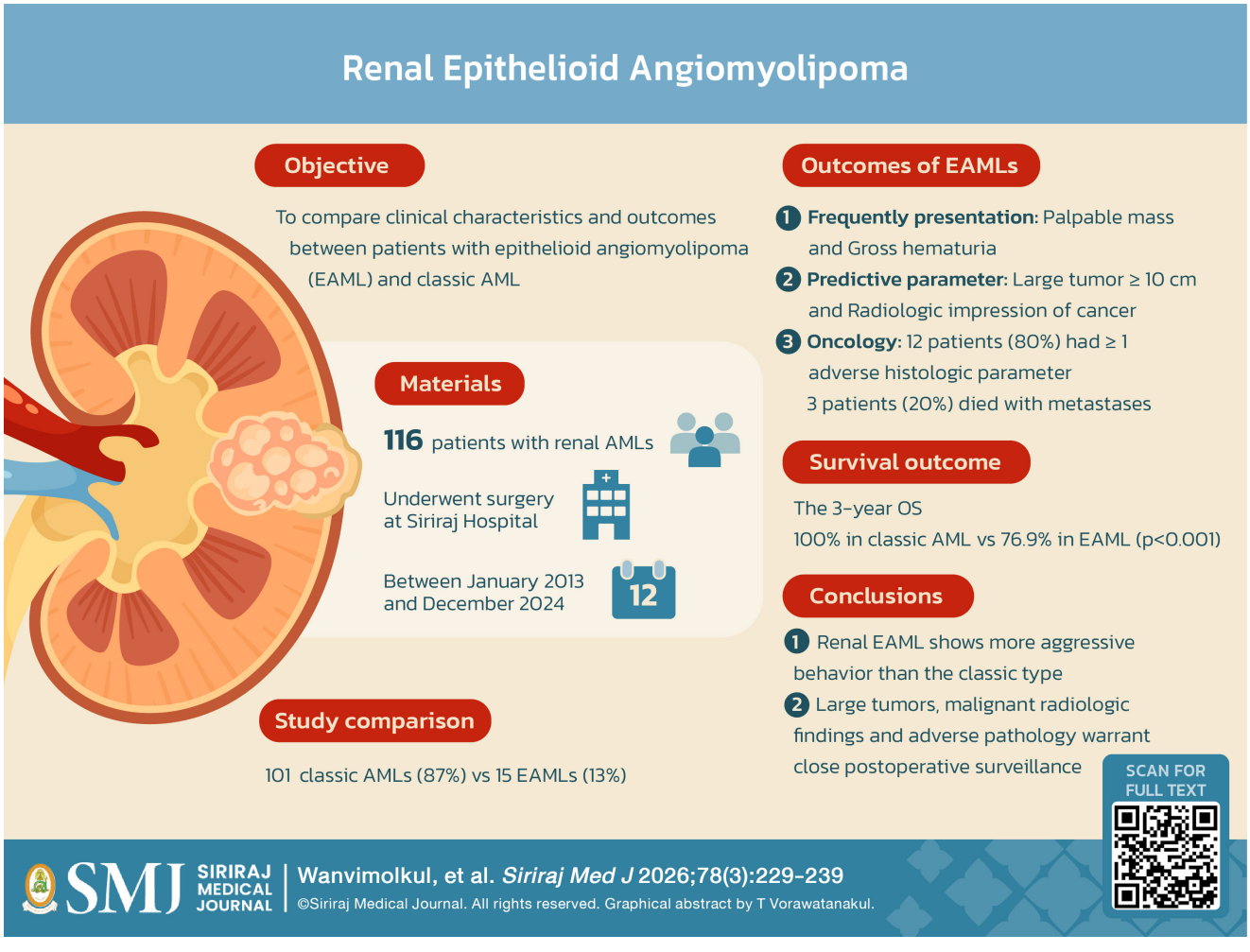
Clinical Characteristics and Surgical Outcomes of Renal Epithelioid Angiomyolipoma: A Comparison with the Classic Type

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ABSTRACT

Objective: To compare clinical characteristics and surgical outcomes between patients with epithelioid angiomyolipoma (EAML) and classic angiomyolipoma (AML), and to identify factors associated with EAML diagnosis.

Materials and Methods: All patients with renal AML who underwent surgery at Siriraj Hospital between January 2013 and December 2024 were reviewed. Clinical features and surgical outcomes were compared between patients with classic AML and those with EAML, and predictors of EAML were evaluated using multivariable analyses.

Results: Among 116 eligible patients, 101 had classic AML and 15 had EAML (12.9%). Most patients were female and were diagnosed in their fifth decade. Demographics, tumor laterality, prevalence of tuberous sclerosis complex gene mutation, and comorbidities did not differ between the 2 groups. Palpable mass (26.7%) and hematuria (13.3%) were more frequent in patients with EAML than in those with classic AML. Most patients with EAML underwent radical or partial nephrectomy due to suspected malignancy. In multivariable analysis, tumor size ≥ 10 cm (odds ratio 15.44; $P = 0.003$) and a radiologic impression of cancer (odds ratio 46.98; $P < 0.001$) independently predicted EAML. Four patients with EAML had adverse pathologic features and experienced poor survival; 3 patients died with metastases. The 3-year overall survival was 100% in classic AML and 76.9% in EAML ($P < 0.001$).

Conclusions: Patients with EAML had less favorable surgical outcomes than those with classic AML. Larger tumor size and a preoperative radiologic impression of malignancy were associated with an EAML diagnosis. Adverse pathologic features in EAML suggest malignant potential.

Keywords: Angiomyolipoma; epithelioid; renal; surgery; classic (Siriraj Med J 2026;78(3):229-239)

INTRODUCTION

Angiomyolipoma (AML) is the most common benign renal tumor. Histologically, AML is categorized as classic AML or epithelioid angiomyolipoma (EAML). Classic renal AML is typically benign and comprises a triphasic mixture of smooth muscle, mature adipose tissue, and thick-walled blood vessels. The 2004 World Health Organization classification of renal neoplasms defines EAML as a potentially malignant mesenchymal neoplasm characterized by predominantly epithelioid-cell proliferation.[1] Most AMLs are discovered incidentally on imaging, although flank pain, gross hematuria, and severe retroperitoneal hemorrhage may occur.[2] AML is generally diagnosed on imaging as a fat-containing renal mass; however, reduced internal fat in EAML makes radiologic differentiation from renal cell carcinoma (RCC) challenging. Surgical management depends on symptoms, tumor size and number, growth pattern, and malignant potential.[3] Definitive diagnosis and subtyping rely on histopathologic examination and immunohistochemical staining.

Although the global incidence of EAML is low, it displays distinct biology from classic AML and carries malignant potential; approximately 20% of patients present with local invasion or metastasis.[4,5] Accurate distinction between classic AML and EAML is therefore critical for treatment selection and prognostication. Several studies have associated EAML diagnosis with younger age, male sex, reduced intratumoral fat, and larger tumor size.[6,7] In contrast, Tsai et al reported no significant associations of age or tumor size between noninvasive and invasive EAML.[8] Based on their analysis of 41 cases and review of 66 published cases, Nese et al proposed that carcinoma-like growth pattern and extrarenal extension or renal vein involvement predict outcomes.[9] More recently, a literature review summarized 46 EAML cases with distant metastasis and recommended long-term follow-up.[10] However, most research comprises case reports or small series, and comprehensive data on clinical and prognostic features remain limited.

We therefore compared clinical characteristics and surgical outcomes between EAML and classic AML in 116 consecutive patients who underwent surgery over 10 years at our center. We also evaluated factors associated with EAML diagnosis. Additionally, we identified EAML cases with malignant behavior and explored predictors of poor outcomes in this subgroup. This is the first EAML cohort study from the Southeast Asian population and may inform future management strategies.

MATERIALS AND METHODS

Study design and ethics

The study protocol was approved by the Siriraj Institutional Review Board (SIRB) (COA no. Si 605/2025). We conducted a retrospective review of 116 patients with renal AML who underwent surgery at the hospital between January 2013 and December 2024.

Cohort and variables

All patients underwent partial or radical nephrectomy, with pathology confirming classic AML or EAML. Collected patient characteristics included sex, age, body mass index, symptoms, and past medical history. Diagnostic variables included radiologic impression, tumor laterality, size, bilaterality, and presence of metastasis. Indications for treatment, management type, and reasons for radical nephrectomy were recorded.

Pathologic definitions and risk stratification

EAML was diagnosed when specimens contained ≥ 80% epithelioid cells, with characteristic positivity for melanocytic markers such as HMB-45 and melan-A and variable positivity for smooth muscle markers (Fig 1). Adverse histologic parameters followed the criteria proposed by Nese et al. since 2011.[9] These included presence of tuberous sclerosis and/or concurrent classic AML, tumor size ≥ 7 cm, carcinoma-like morphology, perinephric fat and/or renal vein involvement, and necrosis. Cases were stratified as low risk (0–1 parameter), intermediate risk (2–3 parameters), or high risk (4–5 parameters).

Outcomes and follow-up

Follow-up data included tumor progression, time to recurrence or metastasis, and patient status at last follow-up. Adverse outcomes were defined as local recurrence, metastasis, or cancer-specific mortality.

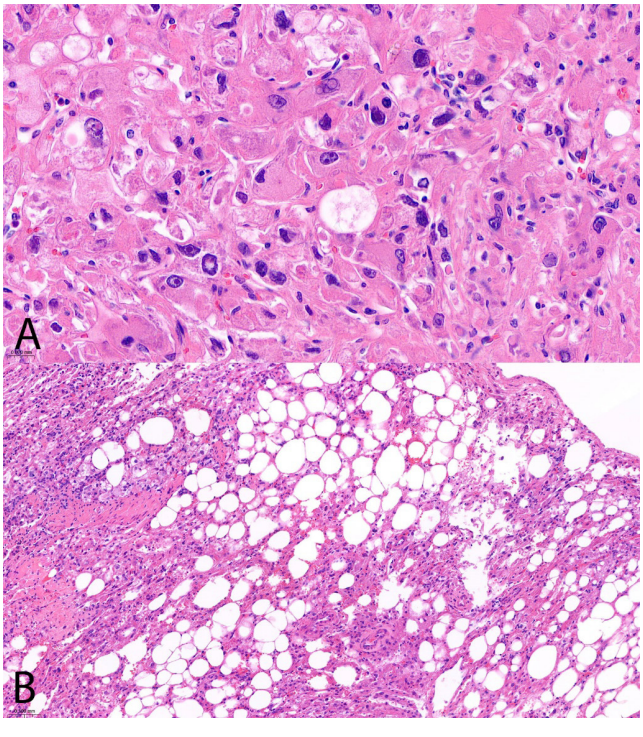


Fig 1. Epithelioid angiomyolipoma of the kidney (H&E stain). **(A)** The tumor comprises predominantly pleomorphic epithelioid cells with eosinophilic cytoplasm. **(B)** Epithelioid AML with concurrent classic AML.

Statistical analysis

Clinical characteristics and surgical outcomes were compared between classic AML and EAML. Continuous variables were summarized as means with standard deviations or medians with interquartile ranges. Between-group differences in categorical variables were tested using the chi-square test or Fisher's exact test, as appropriate. Overall survival was estimated by the Kaplan–Meier method and compared using the log-rank test. Factors associated with EAML diagnosis were identified using univariable and multivariable logistic regression. All tests were 2-sided, with $P < 0.05$ considered statistically significant. Analyses were performed using PASW Statistics version 18 (SPSS Inc, Chicago, IL, USA).

RESULTS

Cohort characteristics

A total of 116 patients were identified: 101 with classic AML (87%) and 15 with EAML (13%). Median age did not differ between the 2 groups (55 vs 53 years; $P = 0.23$), and females predominated in both. Baseline features—including tumor laterality, bilaterality, tuberous sclerosis complex, prior tumor rupture, and comorbidities—were comparable. Clinical characteristics and surgical outcomes are summarized in Table 1.

Presentation and imaging

Patients with classic AML had higher body mass index (mean body mass index 24.0 vs 20.6; $P = 0.024$) and were more often asymptomatic (59.4% vs 26.7%; $P = 0.024$). By contrast, patients with EAML more frequently presented with gross hematuria (13.3% vs 1.0%; $P = 0.044$) and a palpable mass (26.7% vs 5.0%; $P = 0.016$). On imaging, a fat-containing mass consistent with AML was identified more often in classic AML (66.3% vs 13.3%; $P < 0.001$). An impression of RCC was more common in EAML (73.3% vs 23.8%; $P < 0.001$). Representative Computed Tomography (CT scan) image of epithelioid AML from our cohort was presented in Fig 2.

Treatment indications and surgical approach

Indications for surgery in classic AML included suspected malignancy (34.6%), increasing tumor size or size ≥ 4 cm (34.7%), and symptoms or prior rupture (27.7%). The distribution of indications differed significantly between groups: EAML surgeries were driven by suspected malignancy in 13 cases (86.6%) and by symptoms or prior rupture in 2 cases (13.4%). Surgical approach

TABLE 1. Baseline clinical characteristics, imaging impressions, and surgical management in classic AML vs EAML (January 2013–December 2024).

Parameter	Classic AML *n* = 101	EAML *n* = 15	*P* value
Age, median (IQR), y	55.0 (20.0)	53.0 (24.0)	0.238
Female sex, n, %	85 (84.16)	11 (73.33)	0.289
Right side, n, %	45 (44.6)	8 (53.5)	0.524
BMI, median (IQR), y	24.0 (5.6)	20.6 (4.9)	0.024
Co-morbidities, n, %			
Hypertension	32 (31.7)	3 (20)	0.548
Diabetes mellitus	14 (13.9)	2 (13.3)	1.000
CVD	2 (2.0)	1 (6.67)	0.342
Presentations, n, %			
Asymptomatic	60 (59.4)	4 (26.7)	0.024
Abdominal pain	35 (34.7)	5 (33.3)	0.920
Gross hematuria	1 (1.0)	2 (13.3)	0.044
Palpable mass	5 (5.0)	4 (26.7)	0.016
History of ruptured tumor	6 (5.9)	2 (13.3)	0.276
Bilateral AML	4 (4.0)	1 (6.7)	0.506
TSC	1 (1.0)	1 (6.7)	0.243
Tumor size, median (IQR), cm	5.6 (6.5)	10.0 (17.8)	0.828
≥ 4 cm	75 (74.3)	11 (73.3)	1.000
≥ 10 cm	26 (25.7)	9 (60)	0.013
Report of CT/MRI, n, %			
AML	67 (66.3)	2 (13.3)	< 0.001
RCC	24 (23.8)	11 (73.3)	< 0.001
Undetermined	10 (9.9)	2 (13.3)	0.653
Indication for treatment, n, %			
Symptomatic	23 (22.8)	1 (6.7)	0.191
Suspicion of malignancy	35 (34.7)	13 (86.7)	< 0.001
History of ruptured tumor	5 (5)	1 (6.7)	0.573
Pseudoaneurysm	3 (3)	0 (0)	1.000
Increasing size	13 (12.9)	0 (0)	0.213
Size ≥ 4 cm	22 (21.8)	0 (0)	0.071
Type of management, n, %			
Partial nephrectomy	94 (93.1)	8 (53.3)	< 0.001
Radical nephrectomy	7 (6.9)	7 (46.7)	< 0.001
Reason for radical nephrectomy			
Suspicion of malignancy	3 (42.9)	6 (85.7)	0.266
Complex mass	4 (57.1)	1 (14.3)	0.266
Follow-up time, median (IQR), mo	33 (62)	28 (44)	0.091

Data are reported as median (IQR) or n (%), as shown in the table. Imaging impressions derive from CT/MRI reports. Tumor size refers to maximal diameter. Follow-up is reported in months. *P* values are 2-sided.

Abbreviations: AML, angiomyolipoma; BMI, body mass index; CT, computed tomography; CVD, cardiovascular disease; EAML, epithelioid angiomyolipoma; IQR, interquartile range; mo, months; MRI, magnetic resonance imaging; RCC, renal cell carcinoma; TSC, tuberous sclerosis complex; y, years

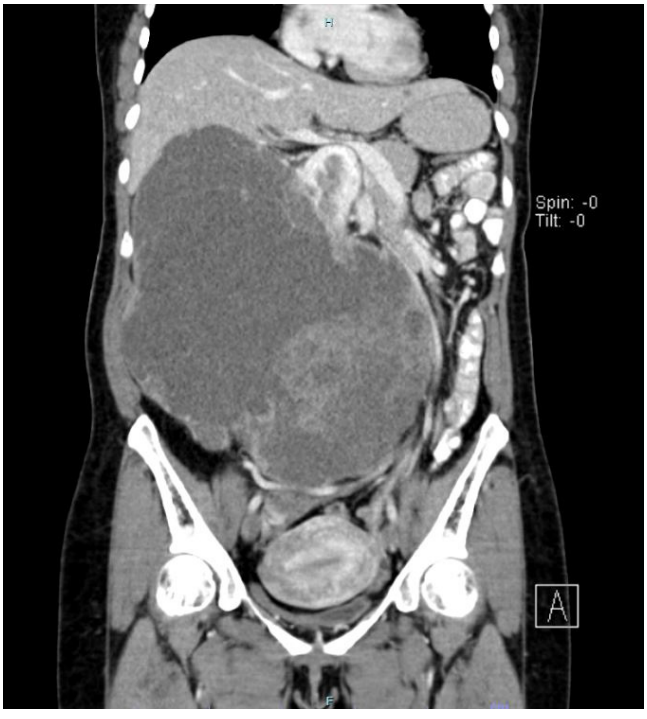


Fig 2. Imaging of epithelioid angiomyolipoma from our cohort. CT scan reveals a 24.4 × 15.0 × 24.0 cm irregular, encapsulated cystic mass with internal irregular nodules and septations, originating from the right kidney.

also differed, with partial nephrectomy more common in classic AML ($P < 0.001$) and radical nephrectomy predominating in EAML ($P < 0.001$).

Follow-up and survival

The median follow-up was 33 months in classic AML and 28 months in EAML. All patients with classic AML remained tumor-free at last follow-up. Four EAML patients (26.7%) experienced tumor progression during follow-up, with mortality of 20% in this group. The 3-year overall survival was 100% in classic AML and 76.9% in EAML ($P < 0.001$; Fig 3).

Predictors of EAML

Univariable and multivariable analyses are summarized in Table 2. Higher body mass index was a protective factor in univariable analysis ($P = 0.024$) but lost significance in multivariable models. Increasing tumor size independently predicted EAML. A tumor size ≥ 10 cm was associated with higher odds of EAML (odds ratio 4.32; 95% CI 1.40–13.30). A radiologic impression of RCC on computed tomography/magnetic resonance imaging also independently predicted EAML (odds ratio 15.35 compared with AML; 95% CI 3.17–74.33). Both associations remained significant in multivariable analysis.

Clinicopathologic features and outcomes of EAML

Clinicopathologic features of patients with EAML are shown in Table 3. Of the 15 patients with EAML, 12 (80%) had ≥ 1 adverse histologic parameter. Localized and locally advanced T stage occurred in a 3:2 ratio. Nearly half of the patients with EAML were high risk for malignant behavior (7/15). Four patients (26.7%) experienced malignant progression with local recurrence, metastasis, or cancer-related death. One patient with positive margin and nodal metastasis (R+/N+) developed liver metastasis at 3 months postoperatively. Another patient with a positive margin without nodal metastasis (R+/N0) developed local recurrence at 9 months postoperatively. Two patients with negative margins and nodes (R0/N0) had longer progression-free survival at 29 and 36 months. No patients with distant metastases received adjuvant systemic therapy. Mortality among the 4 patients with malignant EAML was 75% (3/4). Clinicopathologic features and surgical outcomes of malignant EAML are summarized in Table 4.

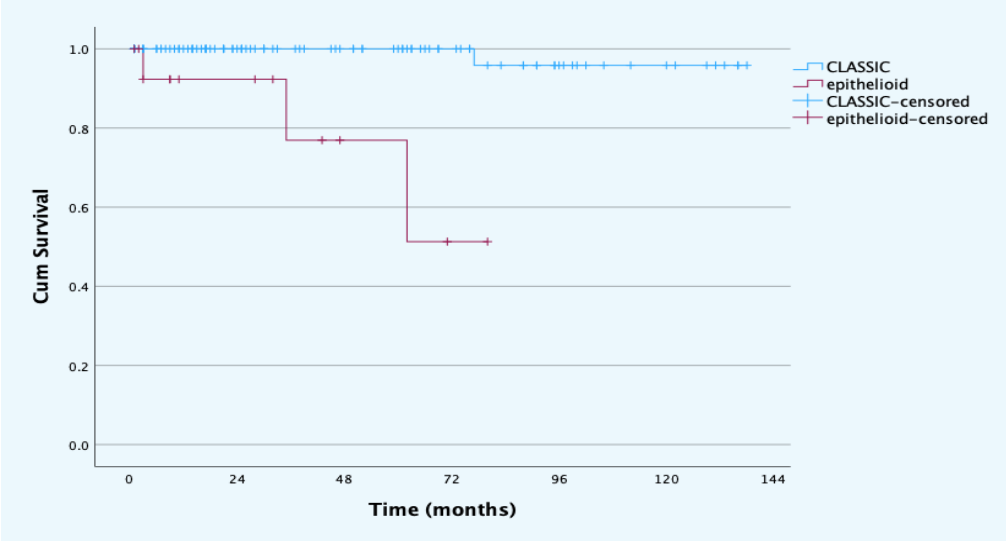


Fig 3. Overall survival after surgery for classic AML vs EAML (Kaplan–Meier). Curves were compared with the log-rank test ($P < 0.001$). Sample sizes were n = 101 for classic AML and $n = 15$ for EAML, with median follow-up of 33 and 28 months, respectively, as per Table 1. **Abbreviations:** AML, angiomyolipoma; EAML, epithelioid angiomyolipoma

TABLE 2. Univariable and multivariable logistic regression for factors associated with EAML diagnosis.

	Univariable			Multivariable		
Variable	**OR**	**95% CI**	***P* value**	**OR**	**95% CI**	***P* value**
Age	0.972	0.933–1.013	0.175			
BMI	0.819	0.688–0.974	0.024	0.816	0.656–1.016	0.069
Male sex	0.518	0.149–1.830	0.307			
Bilateral AML	1.732	0.180–16.629	0.634			
TSC	7.143	0.423–120.757	0.173			
Presentations						
Asymptomatic	0.248	0.074–0.834	0.024			
Abdominal pain	0.943	0.299–2.975	0.920			
Gross hematuria	15.385	1.303–181.707	0.030	15.689	0.063–3923.067	0.328
Palpable mass	6.982	1.629–29.922	0.009			
Report of CT/MRI						
AML			0.003			
RCC	15.35	3.172–74.330	< 0.001	46.989	5.890–374.848	< 0.001
Undetermined	6.70	0.846–53.071	0.072	10.116	0.631–162.160	0.102
Tumor size	1.091	1.016–1.171	0.017			
Tumor size ≥ 4 cm	0.868	0.265–2.844	0.815			
Tumor size ≥ 7 cm	2.385	0.787–7.221	0.124			
Tumor size ≥ 10 cm	4.327	1.404–13.330	0.011	15.443	2.593–91.983	0.003
History of ruptured tumor	2.436	0.444–13.361	0.305			

Odds ratios (ORs) are shown with 95% confidence intervals. For continuous tumor size, the OR corresponds to a 1-unit increase (cm). For "Report of CT/MRI," AML is the reference category. *P* values are 2-sided, with $P < 0.05$ considered significant.

Abbreviations: AML, angiomyolipoma; BMI, body mass index; CI, confidence interval; CT, computed tomography; EAML, epithelioid angiomyolipoma; MRI, magnetic resonance imaging; OR, odds ratio; RCC, renal cell carcinoma; TSC, tuberous sclerosis complex

DISCUSSION

First described by Mai et al, EAML is a rare AML variant with malignant potential.[11] Diagnosis is challenging: specific clinical manifestations are lacking, radiographic distinction is hampered by reduced intratumoral fat, and histologic differentiation from classic AML may be difficult. Surgical management of AML is guided by suspected malignancy, symptoms, and radiologic size; nevertheless, some patients with EAML develop local recurrence or distant metastasis after resection. We therefore retrospectively compared clinical characteristics and outcomes between EAML and classic AML in 116 consecutive patients who underwent surgery for AML.

Consistent with prior studies, middle-aged patients and women predominated in both the EAML and classic AML groups in our cohort.[7,12] However, Lee et al reported that males were 3.3 times more likely than females to have EAML.[6] Female sex hormones have been proposed to explain women's higher AML prevalence, but whether epithelioid differentiation affects sex distribution remains uncertain.[13] In our cohort, tumor laterality, bilaterality, tuberous sclerosis complex, prior rupture, and comorbidities

TABLE 3. Clinicopathologic features and malignancy-risk stratification of EAML.

Clinicopathologic features	EAML *n* = 15
Adverse features	12 (80)
LVI	6 (40)
TSC	1 (6.67)
Tumor size > 7 cm	9 (60)
Concurrent classic AML	7 (46.7)
Carcinoma-like morphological pattern	9 (60)
Involvement of perinephric fat/renal vein	9 (60)
Necrosis	9 (60)
Risk	
Low	4 (26.7)
Intermediate	4 (26.7)
High	7 (46.7)
T staging	
T1	5 (33.3)
T2	4 (26.7)
T3a	5 (33.3)
T4	1 (6.7)
Positive margin	4 (26.67)
Recurrence*	2 (13.3)
Metastasis*	3 (20)
Dead	3 (20)

Risk categories follow Nese et al [9]: low risk 0–1 parameter, intermediate risk 2–3, and high risk 4–5, as defined in **Methods**. Pathologic T stage is shown as recorded. One case experienced both local recurrence and metastasis, as indicated by the asterisk in the table.
Abbreviations: AML, angiomyolipoma; EAML, epithelioid angiomyolipoma; LVI, lymphovascular invasion; TSC, tuberous sclerosis complex

did not differ between the groups. In a series of 587 patients, Lee et al found no difference in hemorrhage incidence between sporadic AML and EAML.[12] Aydin et al reported that EAML is strongly associated with tuberous sclerosis complex and adverse pathologic features.[14] Correspondingly, 1 EAML patient with the complex in our series exhibited malignant behavior. Our findings add to the clinical characterization of AML; however, the biological distinctions between classic AML and EAML require further investigation.

Patients with classic AML had a higher rate of asymptomatic presentation than those with EAML (59.4% vs 26.7%). This finding aligns with a large surgical series that reported 48% of triphasic AMLs were incidental findings in 209 patients.[2] The proportion of asymptomatic patients may be even higher under active surveillance; a Swedish cohort reported 61% asymptomatic at presentation.[15] These data suggest that asymptomatic presentation reflects the benign behavior of classic AML. Conversely, our patients with EAML more often presented with symptomatic

TABLE 4. Clinicopathologic features and surgical outcomes of malignant EAML cases.

Case	Age/ Sex	Presenting symptoms	Surgery type	Tumor size (cm)	Tumor margin (R)/Node status (N)	LVI	Presence of TSC	Concurrent classic AML	Other adverse features*	Risk	Local recurrence/ distant metastasis	Adjuvant treatments	Time to disease progression (mo)	Follow -up time (mo)	Survival status
1	31/M	Hematuria	RN	14.5	R0/N0	N	Y	Y	Y	High	Local/lung	N	29	36	Dead (progression of disease)
2	24/F	Hematuria	RN	14	R0/N0	Y	N	N	Y	High	Bone, lung, liver, brain	Adjuvant RT at bone	36	39	Dead (progression of disease)
3	40/F	Palpable mass	RN	26	R+/N+	Y	N	Y	Y	High	Liver	N	3	3	Dead (progression of disease)
4	66/F	Abdominal pain	PN	10	R+/N0	Y	N	Y	Y	High	Local	Adjuvant RT at tumor base	9	12	Alive

R denotes margin status (R0, negative; R+, positive). N denotes nodal status (N0, negative; N+, positive). "Other adverse features" include carcinoma-like growth, perinephric fat or renal vein involvement, necrosis, and tumor size ≥ 7 cm. Time intervals are in months.

Abbreviations: AML, angiomyolipoma; EAML, epithelioid angiomyolipoma; LVI, lymphovascular invasion; mo, months; N0/N+, nodal status; PN, partial nephrectomy; R0/R+, resection margin status; RN, radical nephrectomy; RT, radiotherapy; TSC, tuberous sclerosis complex.

disease, notably palpable masses and gross hematuria, which likely relates to larger tumors in EAML than classic AML (10 vs 5.6 cm). This pattern may also indicate more invasive EAML biology.

Imaging features differed significantly between the 2 groups. Fat-containing masses consistent with AML were reported more often in patients with classic AML than in those with EAML (66.3% vs 13.3%; $P < 0.001$). Because EAML exhibits variable proportions of adipose tissue, smooth muscle, and blood vessels, its radiologic appearance is heterogeneous and overlaps with other renal tumors.[6,16] Accordingly, radiologists frequently suggested RCC (73.3%) or reported an indeterminate diagnosis (13.3%) in our EAML cases. Thus, radiologic diagnosis of EAML remains challenging, though future work may identify imaging indicators that raise preoperative suspicion.

Indications for surgery differed significantly between groups. In classic AML, the main drivers were risk of bleeding or increasing tumor size (34.7%) and symptoms including prior rupture (27.7%). Nevertheless, one third of classic AML cases underwent surgery due to suspected malignancy. In EAML, suspected malignancy prompted surgery in 86.7% of patients.

The operative approach reflected these indications. Radical nephrectomy was performed significantly more often in cases of EAML, whereas nephron-sparing surgery was favored in classic AML. This pattern suggests potential overtreatment when managing presumed cancer, particularly the use of radical nephrectomy for lesions that are ultimately proven to be AML. In a surgical series of 219 patients, Lane et al reported that 55% had radiographic features prompting surgery for suspected RCC.[2] Given that EAML cannot be reliably diagnosed without immunohistochemistry, percutaneous renal mass biopsy may be necessary for treatment planning in selected indeterminate cases.

In multivariable analysis, larger tumor size (≥ 10 cm) and a radiologic impression of RCC independently predicted EAML. Tumor size ≥ 10 cm conferred higher odds of EAML (odds ratio 15.4, 95% CI 2.59–91.98; $P = 0.003$). A radiographic report suggestive of malignancy increased the odds of EAML diagnosis by approximately 47-fold. In a retrospective comparison of classic AML ($n = 204$) and EAML ($n = 27$), younger age, male sex, and larger tumors predicted EAML.[6] That study also reported that 70.4% of EAML cases were interpreted as RCC on computed tomography.[6] Another series of 9 EAML suggested that a lipid-poor mass without calcification should raise clinical suspicion.[17] Thus, large tumor size and radiologic concern for malignancy should prompt consideration of EAML; however, preoperative tissue diagnosis remains essential when distinguishing malignancy from EAML is required. Higher body mass index appeared protective against EAML in univariable analysis ($P = 0.024$) but lost significance in multivariable models ($P = 0.069$). Obesity has been associated with higher renal neoplasm risk in European cohorts[18,19], yet its relationship with angiomyolipoma is unclear. Notably, our cohort represents a Southeast Asian population with generally lower body mass index; further studies are needed to clarify this association.

Surgical excision remains the primary treatment for renal EAML. In 2008, Aydin et al reported a benign course in all EAML cases ($n = 15$) over a median follow-up of 5.1 years.[14] In our cohort, surgical outcomes differed between classic AML and EAML. Classic AML showed no progression with 3-year overall survival of 100%, whereas overall survival was 76.9% for EAML, consistent with multicenter series.[7,20]

To understand these poorer outcomes, previous studies identified adverse clinicopathologic parameters. Brimo et al analyzed 40 EAML and proposed 4 features: ≥ 70% atypical epithelioid cells; ≥ 2 mitotic figures per 10 high-power fields; atypical mitotic figures; and necrosis. The presence of 3 or 4 features strongly predicted malignant behavior.[21] Similarly, Nese et al proposed 5 parameters for risk stratification: tuberous sclerosis complex or concurrent classic AML, necrosis, tumor size ≥ 7 cm, extrarenal invasion and/or renal vein involvement, and carcinoma-like growth pattern. Low-, intermediate-, and high-risk categories corresponded to 0–1, 2–3, and 4–5 parameters, respectively.[9] In 2015, Lei et al reported that tumor size ≥ 9 cm, venous tumor thrombus, epithelioid cells ≥ 70% or atypical cells ≥ 60%, and necrosis were associated with progression.[20] In our 15 EAML cases, 12 patients (80%) had at least 1 adverse parameter under the Nese scheme.[9] All low-risk patients remained recurrence-free at last follow-up. Seven of 15 patients (46.7%) were high risk, and 4 of these (57.1%) progressed. Prognostic factor analysis for malignant behavior was not feasible given the small number of events. Due to the rarity of EAML, no consensus criteria for predicting malignancy have been established.

Because evidence remains limited, treatment guidelines for EAML are not well established. Complete surgical resection remains the preferred option; however, some patients experience tumor progression after surgery. Reported recurrence and metastasis rates reach 17% and 49%, respectively, and mortality can reach 33%.[9] In our series, 4 patients (26.7%) developed progression and were therefore classified as having malignant EAML; all

had adverse parameters per Nese et al.[9] One patient with positive margins and nodal involvement developed liver metastasis 3 months postoperatively. Another patient with positive margins but no nodal involvement had local recurrence at 9 months.

In contrast, 2 patients with negative surgical margins had longer progression-free survival at 29 and 36 months. These findings underscore that complete tumor removal remains crucial for survival. In our cohort, none of the 3 patients with metastatic EAML received systemic therapy because of poor performance status, which led to rapid progression and tumor-related death; overall mortality was 20%. In 2023, Zhang et al summarized 46 cases of metastatic EAML, reporting poor prognosis and low 5-year survival.[10] Recent reports suggest benefit from mechanistic target of rapamycin inhibitors, tyrosine kinase inhibitors, and immunotherapy in malignant EAML.[5] Based on our data, patients with high-risk adverse parameters warrant close monitoring and may benefit from intensive adjuvant management to improve outcomes.

Our study has limitations. First, its retrospective design restricted analyses to available clinical data. Second, excluding patients managed with angioembolization or active surveillance may have underestimated the true EAML population.[22] Third, radiologic metrics of intralesional fat (Hounsfield units, chemical shift imaging) were unavailable due to incomplete data. Fourth, the number of EAML cases was small because this was a single-center study. Larger multicenter cohorts with standardized radiologic characterization and comprehensive data collection are needed for more robust conclusions.

CONCLUSIONS

EAML presents distinct clinical challenges and is associated with less favorable surgical outcomes than classic AML. Larger tumor size and a preoperative radiologic impression of malignancy were significantly associated with an EAML diagnosis. Adverse pathologic features in EAML indicate malignant potential. Active postoperative management and long-term surveillance are therefore essential to optimize patient outcomes.

DECLARATIONS

Grants and Funding Information

None.

Conflict of Interests

The authors declare no conflict of interest.

Registration number of clinical trial

None.

Author Contributions

Conceptualization and methodology: N.W., T.T., T.H.; Investigation: N.W., V.W.; Formal analysis: N.W., P.L., K.J.; Visualization and writing—original draft: N.W., T.T.; Writing-review and editing: T.T., E.C., S.J., N.T.; Supervision: T.T.

Ethics Statement

The study protocol was approved by the Siriraj Institutional Review Board (SIRB) (COA no. Si 605/2025).

Use of Artificial Intelligence

No artificial intelligence tools or technologies were used in the writing and analysis.

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