

Case Report – Malignant Renal Epithelioid Angiomyolipoma

Authors: Dr Stephanie Zhang and Dr Jeanne Boissiere

Background

Renal epithelioid angiomyolipomas (EAML) are mesenchymal tumours consisting of adipose tissue, smooth muscle cells, abnormal thick-walled blood vessels (1). Malignant renal EAML is a rare entity with a challenging diagnosis, prognostication and management (2). It is also unclear if and how histological appearances, immunohistochemistry and next-generation sequencing can be used to predict the behaviour and clinical outcome of malignant EAMLs (1) (3) (4). We are presenting this case of a malignant renal EAML to improve understanding of this rare condition.

Case Presentation

- Retired 78-year-old gentleman presented with worsening shortness of breath on exertion
- Had an exudative left sided pleural effusion with predominantly lymphoid cells
- CT thorax and abdomen – 9.2 cm solid, complex mass in left kidney concerning for primary renal cell neoplasm with no local invasion or metastases
- Had a left radical nephrectomy and recovered well
- Initial Histopathology report – 92 mm Grade 4 clear cell renal cell carcinoma with extensive rhabdoid and sarcomatoid differentiation
- Subsequent immunohistochemistry showed strong and diffuse staining for Melan-A and HMB45, weak staining for CD68, and negative staining for PAX8
- Revised Histopathology report after multi-disciplinary discussion – malignant renal EAML, pT3a
- Management – high-risk surveillance, repeat CT scan in 3 months and follow up

Histopathological Findings

Figure 1



Figure 2

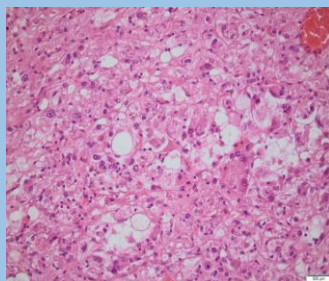


Figure 3

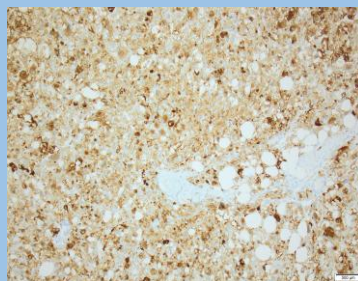


Figure 1: Macroscopic image depicting tumour's atypical features – colour and location

Figure 2: Microscopic image demonstrating high-grade cytological and atypical features, including dilated and distorted vascular spaces, and mixed adipocytic background

Figure 3: Immunohistochemistry result showing strong staining for Melan-A

Discussion

Renal EAMLs are often misdiagnosed as high-grade clear cell renal carcinomas (RCCs) as they show similar cytological features and RCCs are far more common (4)(5). Both EAMLs and clear cell RCCs exhibit haemorrhagic, necrotic and cystic features; this further contributes to confusion between the two diagnoses (5). Treatment options are different between the two diagnoses, including either active surveillance, partial or radical resection, and other targeted therapies, and so making the correct diagnosis is important (5)(6)(7). Furthermore, given the rare nature of renal EAMLs, it can be difficult to predict its malignant potential.

On review of existing literature, we have identified the following techniques to help with these diagnostic challenges:

- 1) Clear cell RCCs have a clear capillary network while renal EAMLs have discohesive sheet-like patterns (6).
- 2) Clear cell RCCs positively stain with PAX8 and PAX2 while renal EAMLs are negative for PAX8, expression of cathepsin K and PNL2 have high sensitivity for diagnosing renal EAMLs.
- 3) Mutations to the TSC and TFE3 genes are associated with renal EAMLs (3)(7).
- 4) The Clear Cell Likelihood Score, a scoring system based on radiological findings, can help distinguish between renal EAML and clear cell RCC (5). For example, EAMLs result in hypointensity while clear cell RCCs result in iso/hyperintensity (5).
- 5) The following features are significantly linked to metastatic disease: $\geq 70\%$ atypical epithelial cells, ≥ 2 mitoses per 10 high-power fields, atypical mitoses, necrosis (3).
- 6) Negative smooth muscle actin (SMA) expression and $\geq 10\%$ expression of Ki-67 was significantly associated with malignancy, and poorer prognoses (8).

Conclusion

This case provides insight in to how these rarer tumours can present atypically, highlights the complexities in diagnosing and managing renal EAMLs, and draws attention to ongoing gaps in our knowledge for further research. This case also illustrates the importance of multi-disciplinary discussion and review of the latest literature when faced with challenging and rarer cases.

References

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