Malays J Pathol 2025; 47(2) : 315 – 318

CASE REPORT

A case of large renal angiomyolipoma resulted in hydronephrosis

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Abstract

Introduction: Renal angiomyolipoma is one of the members of the perivascular epithelioid cell (PEC) tumour family. It has a characteristic triphasic morphology featuring varying proportions of dysmorphic blood vessels, smooth muscle cells, and mature adipose tissue. Large angiomyolipomas pose a risk of haemorrhagic complications. *Case report:* A 47-year-old woman presented with right abdominal pain for 2 months, and radiological investigation revealed a 14.2 cm renal mass causing hydronephrosis. The symptoms persisted after arterial embolisation and hence, nephrectomy was performed. The excised renal tumour weighed 1,442 grams. There was a well-circumscribed yellow mass confined within the Gerota's fascia. Histological examination revealed a classic triphasic morphology comprising mature adipose tissue, dysmorphic blood vessels, and smooth muscle. Notably, there were post-embolisation histological changes in extensive fat necrosis and foreign body giant cell reaction. Immunohistochemically, it expressed HMB45, MelanA, and SMA, while it was negative for PAX8 and pan-cytokeratin. *Discussion:* We described a case of renal angiomyolipoma with classic gross appearance of adipose-rich tumour and the triphasic histological features and discussed the post-embolisation histological changes.

Keywords: Angiomyolipoma, embolisation, kidney, PEComa, prognosis

INTRODUCTION

Renal angiomyolipoma (AML) stands as the most common benign mesenchymal neoplasm affecting the kidney, comprising approximately 1% of all renal tumours and affecting up to 0.6% of the general population.[1,2,3] These tumours are classified under the perivascular epithelioid cell tumour (PEComa) family. It is classically characterised by a triphasic histology consisting of 3 components, including dysmorphic thick-walled blood vessels, spindle and epithelioid smooth muscle cells, and mature adipose tissue.[1,4,5] This is a case of large AML in a 47-year-old woman causing obstructive hydronephrosis. She was treated with arterial embolisation, followed by nephrectomy due to persistent symptoms post-embolisation. We describe the classic gross and microscopic appearances of a case of AML and highlight the importance of recognising the histological changes secondary to post-embolisation treatment.

CASE REPORT

A 47-year-old woman presented to our hospital with a 2 months history of increasing right flank pain and a lower abdominal swelling. Her full blood count and renal functions were unremarkable. Subsequently, an abdominal and pelvic CT scan revealed a well-circumscribed mass measuring approximately 15.0 × 11.7 × 9.9 cm, located at the right kidney. The mass compressed the renal pelvis and proximal ureter resulted in right hydronephrosis. She underwent preoperative arterial embolisation of the tumour. However, she continued to experience pain. Consequently, she underwent a right radical nephrectomy.

Gross findings

The nephrectomy specimen weighed 1,442 grams. Serial sectioning showed a large, well-circumscribed yellow mass measuring 14.2 × 11.5 × 7.5 cm. The tumour replaced almost the

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entire renal parenchyma and was confined within the Gerota's fascia, without extrarenal extension. The cut section revealed a yellow adipose tissue-like appearance with cystic spaces filled with yellowish oily fluid. (Fig. 1).

Microscopic findings

Histologically, the tumour composed of predominantly mature adipose tissue, comprised 80-90% of the tumour volume, admixed with interlacing fascicles of spindle-shaped smooth muscle cells and thick-walled blood vessels (Fig. 2). A diagnosis of classic triphasic renal angiomyolipoma was made. Notably, post-embolisation histological changes were seen throughout the specimen, including fat necrosis and multiple groups of foreign body-type multinucleated giant cells (Fig. 2). Immunohistochemically, the spindle cells and some adipocytes were positive for the melanocytic marker HMB-45. The adipose tissue showed focal positivity for Melan-A, and there was diffuse positivity for smooth muscle actin (SMA). The tumour cells were negative for S100, PAX8, and cytokeratin.

DISCUSSION

The majority of AML are sporadic and primarily affect middle-aged women ranging from 40 to 60 years.[6,7] A proportion of cases are associated with tuberous sclerosis complex, in younger individuals which often leading to larger and bilateral tumours.[2,4] The pathogenesis of AML is found to be linked to genetic mutations in *TSC1* or *TSC2* tumour-suppressor genes, resulting in the hyperactivation of the mammalian target of rapamycin (mTOR) pathway. This dysregulation ultimately leads to unchecked cellular growth and proliferation.

The average size of AML is about 6 cm and large tumours may be associated with haemorrhage. Hence, larger tumours may require prophylactic treatment to avoid spontaneous, life-threatening bleeding.[1] Bhatt *et al*. (2016) in a cohort of 582 AMLs showed about 70% of tumours > 4 cm were asymptomatic, and only 0.4% had urgent bleeding.[8] Current guidelines from the urological associations recommend active surveillance for asymptomatic tumours of any size, with intervention reserved for tumours that grow rapidly, patient becomes symptomatic or are linked to a high-risk pregnancy.[9]

The utility of arterial embolisation could complicated histopathological evaluation. The possible histological changes in these samples include coagulative necrosis, fat necrosis, haemorrhage, extensive inflammatory infiltrates and multinucleated cells with foreign-body giant cell reactions.[1,4,5,7] Some of these histological changes may resemble tumour necrosis, and may lead to misdiagnosis of a more aggressive tumour.[3,10] Hence, awareness of these features

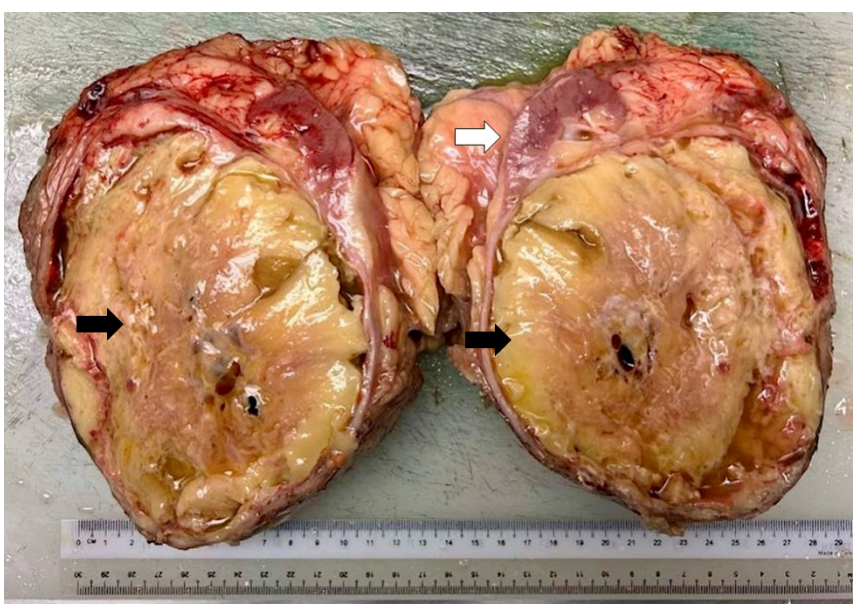

Fig. 1. Gross appearance of angiomyolipoma. The nephrectomy specimen demonstrating a large, well-circumscribed tumour with a yellow-tan cut surface (black arrow). A few cystic spaces were observed. The residual renal parenchyma was pushed to the periphery (white arrow).

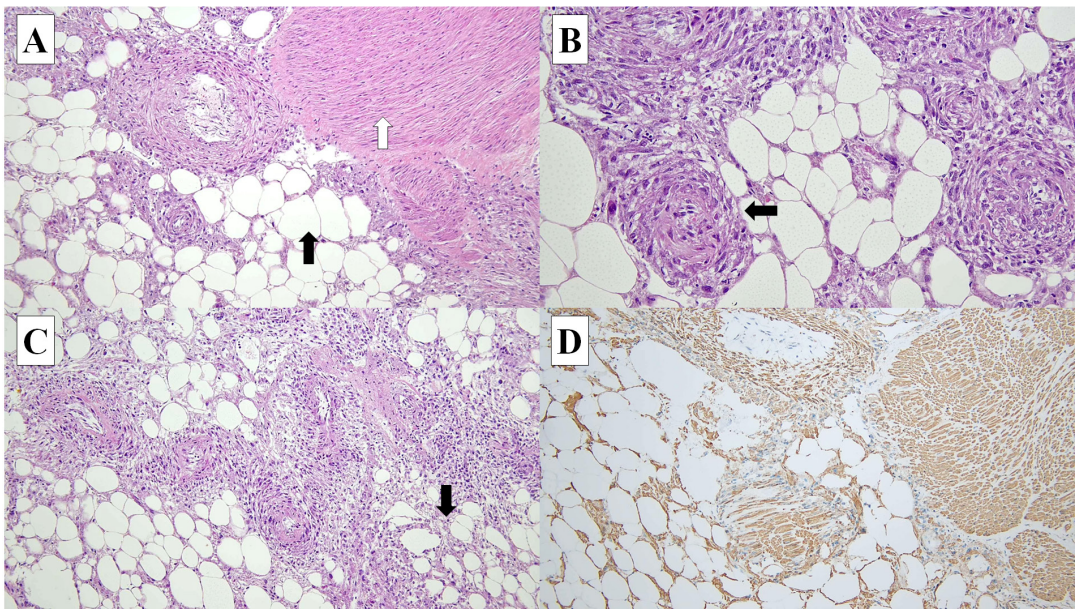

Fig. 2. Microscopic appearance of angiomyolipoma. (A) Tumour composed of triphasic morphology of mature adipose tissue (black arrow), smooth muscle (white arrow) and (B) thick-walled blood vessels (arrow) (H&E, A×100, B×200). (C) Post-embolisation changes – fat necrosis (black arrow) (H&E, ×100). (D) The tumour showed diffuse smooth muscle actin (SMA) expression (SMA, ×100).

is essential. In a study on post-embolisation of uterine artery in 10 cases of leiomyomas, they described the characteristic histologic features of post-embolisation such as massive necrosis, dystrophic calcification, vascular thrombosis, intravascular foreign material that elicited a histiocytic and foreign-body giant cell reaction, and in occasional cases, there were foci of myometrial necrosis and microabscess formation.[11] In another study on meningioma, post-embolisation changes included necrosis, intravascular particles and fibrinoid necrosis of the vascular wall. They cautioned against misdiagnosis the meningioma as a higher-grade tumour due to the presence of necrosis.[12]

In conclusion, we described a case of large AML with mass effect causing symptomatic hydronephrosis which was successfully treated with arterial embolisation followed by nephrectomy. This report also highlighted the importance of recognising the histological changes associated with post-embolisation by pathologists to avoid misinterpretation.

Acknowledgement: We thank the patient for the verbal consent to publish this report.

Author Contributions: All authors contributed equally to the development of this manuscript.

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

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