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Transfemoral embolization of a large symptomatic renal angiomyolipoma in a horseshoe kidney: a case report and literature review

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Abstract

Renal angiomyolipomas (AML) are the commonest benign renal neoplasms. They are composed of blood vessels, adipose tissue, and smooth muscle in varying amounts**.** It is quite rare to find AML in a horseshoe kidney, although there is at least one such reported case discovered in pregnancy and managed by resection. Spontaneous hemorrhage which could be fatal is the most feared sequelae of AML. The first-line of management for AMLs was previously surgical excision. However, advances in minimally invasive techniques that have a lower risk of complications have broadened treatment options for reducing tumour size and preventing hemorrhage. One of such effective techniques is selective arterial embolization (SAE) of renal AMLs > 4 cm. Patients not requiring any immediate intervention may be followed up with active surveillance. In this case report, we present a case of AML in a horseshoe kidney that was successfully treated with transfemoral embolization as well as a review of the literature.

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Keywords*: Arterial embolization, angiomyolipoma, horseshoe kidney*

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INTRODUCTION

Renal angiomyolipomas (AML) are benign renal neoplasms composed of blood vessels, adipose tissue and smooth muscle in varying amounts. They are commoner in females, especially in the 4th - 5th decade and may occur spontaneously in association with Tuberous sclerosis or rarely with Lymphangioleiomyomatosis [1,2]. They constitute the most common benign renal neoplasia with a reported incidence of about 0.3 - 3% [3,4]. The blood vessels in AMLs lack internal elastic lamina with the replacement of smooth muscle by fibrous tissue. These deficiencies render the vessels fragile and prone to fatal aneurysmal formation and rupture [5,6]. Though surgical excision was previously the first-line management for AMLs, advances in minimally-invasive techniques have allowed for treatment options with very low risk of complications. Selective arterial embolization (SAE) is one of such techniques and is proven to be effective in reducing tumour size as well as preventing haemorrhage of renal AMLs > 4 cm [5-9]. The commonest congenital renal fusion anomaly is the horseshoe kidney, and it is characterized by malrotation and variable blood supply [10]. There have been reports of AML in normal kidneys, and also of other renal tumours in horseshoe kidneys, however, AML in horseshoe kidney are rare. However, AMLs in horseshoe kidneys are rare. As at the time of writing this case report, the authors are not aware of any prior reports of transfemoral embolization of AML in a patient with horseshoe kidney. Here, we present a case of transfemoral embolization of renal AML in a horseshoe kidney.

CASE PRESENTATION

A 58-yr. old woman reported to the hospital with a 2-wk history of right-sided abdominal pain and two episodes of haematuria which had resolved spontaneously. The patient was not known to have any chronic medical condition. She, however, had a previous history of myomectomy and

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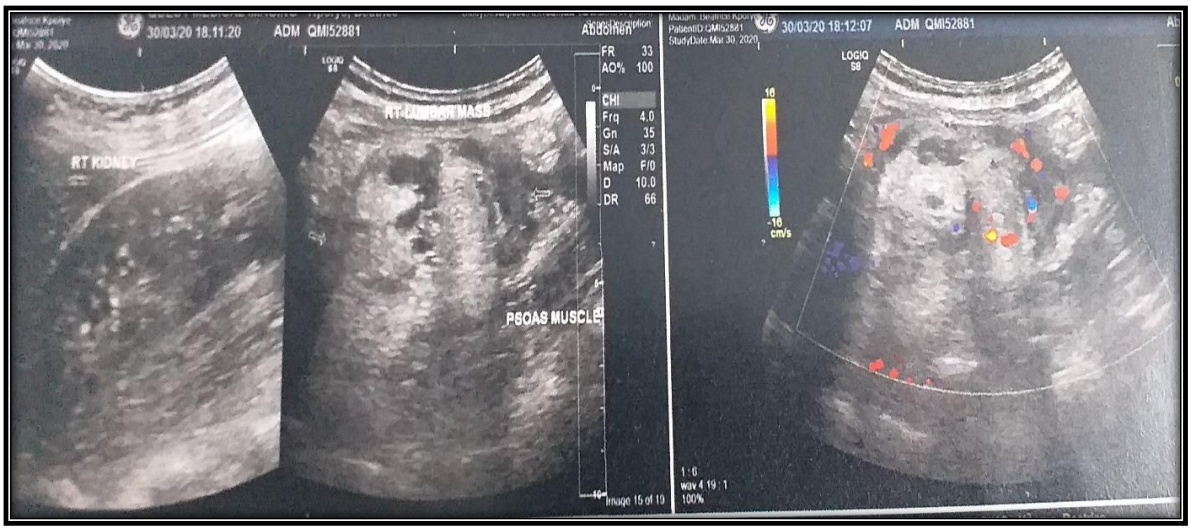
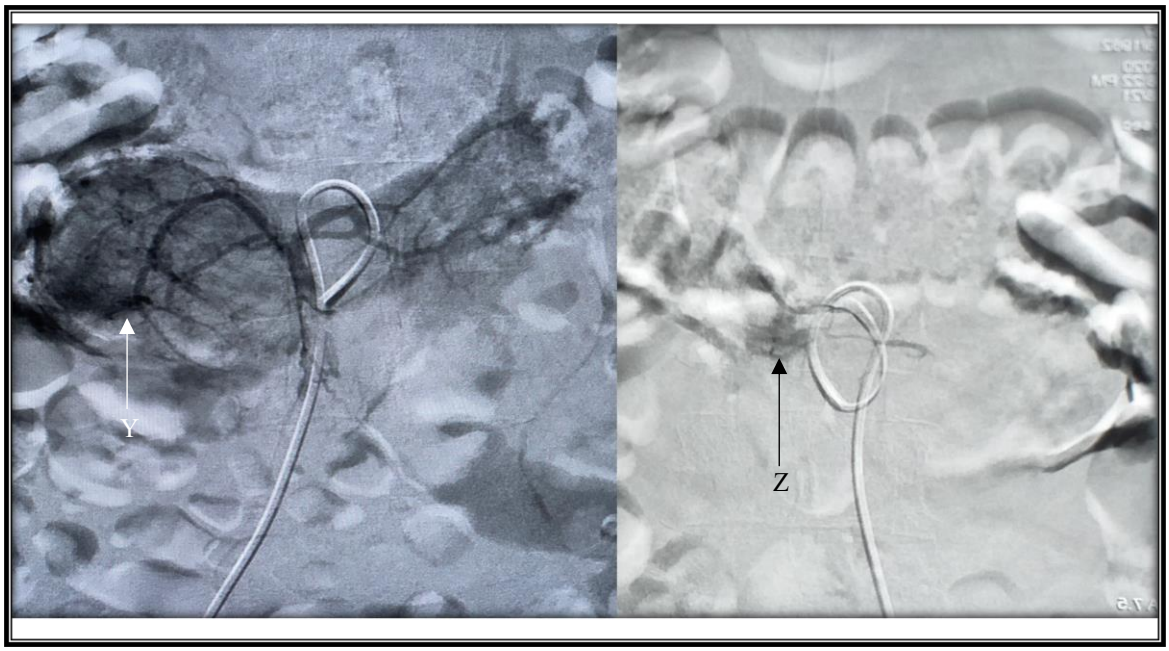


Plate 1: Grayscale ultrasound scan of the horseshoe kidney showing a mass of mixed echogenicity with areas of vascularity in the right lumbar region anterior to the psoas muscle and closely related to the right kidney

lumpectomy on account of uterine fibroids and a benign breast lump, respectively. On examination, the patient was generally stable but mildly pale (haemoglobin level = 9.53 g/dL) and had mild right iliac fossa tenderness. Her blood pressure was 115/65 mmHg and pre-operative blood urea electrolytes, and serum creatinine were normal. An abdominopelvic ultrasound scan done showed a mass of mixed echogenicity with areas of vascularity in the right lumbar region anterior to the psoas muscle and closely related to the right kidney (Plate 1). A contrast-enhanced Computed Tomography (CT) scan revealed normally-positioned kidneys with both kidneys connected at their lower pole by an isthmus of functioning renal parenchyma, measuring approximately 1.83 cm in anteroposterior diameter and lying anterior to the abdominal aorta and inferior vena cava.

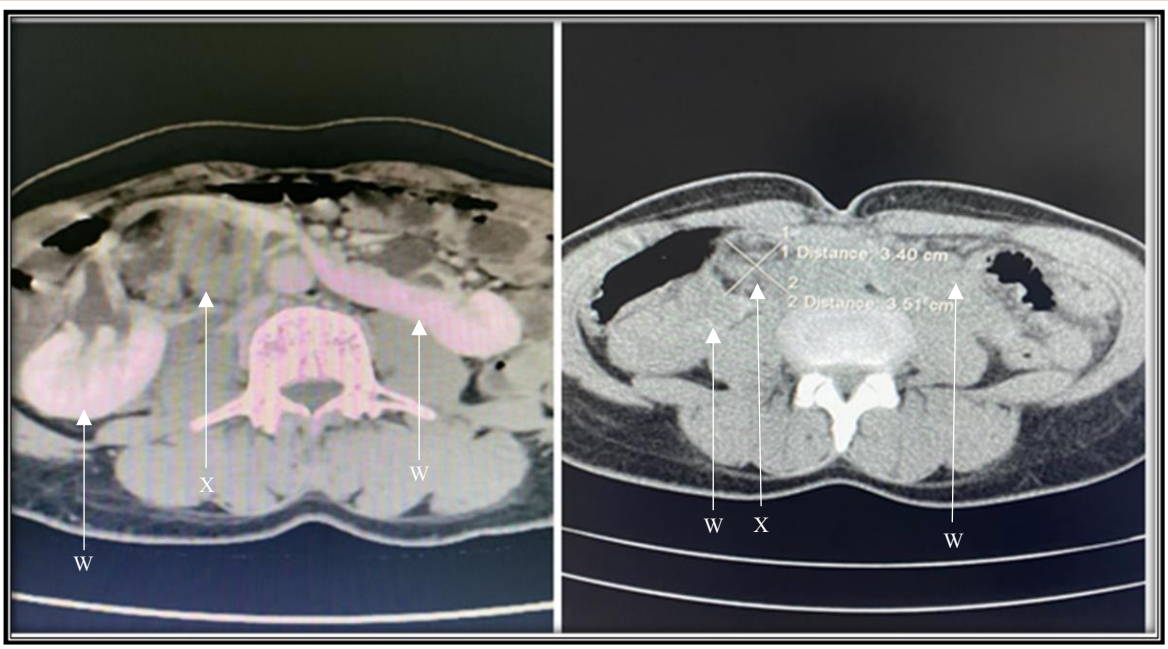
A heterogeneously enhancing mass with few areas of fat attenuation, necrosis, as well as high attenuation blood products were seen in the mid to lower polar region of the right kidney. It measured approximately 5.60 cm transverse by 5.20 cm anteroposterior and caused extrinsic compression of the inferior vena cava. The differential diagnoses given were renal cell carcinoma and renal AML. This meant that the patient required a percutaneous biopsy to confirm the diagnosis. An ultrasound-guided tru-cut biopsy was done which confirmed renal AML. She was counselled on the risk of repeated haemorrhage from the mass due to size (> 4 cm) and embolization was recommended and performed for the patient. In anticipation of the vascular and urological anomalies prone to occur with fused kidneys, the pre-procedure contrast-enhanced CT scan done was assessed for variations in vascular supply and drainage. In our patient, multiple bilateral renal arterial supply was a significant finding. It was necessary to determine which one was the dominant vascular supply. A pre-embolization arteriogram later done was helpful in the identification and super-selective catheterization of the dominant vascular supply. Under ultrasound guidance and using the Seldingher technique, the right common femoral artery was accessed and a 5F femoral sheath was placed. 2000 IU heparin was administered intra-arterially. An aortogram was obtained using a 5F pigtail catheter. The patient was found to have a total of 7 renal arteries (2 main renal arteries, 5 accessory renal arteries) supplying both kidneys. Subsequently, a 5F S1 catheter and a 0.035' hydrophilic guidewire was manipulated under fluoroscopic guidance into the abdominal aorta and the main and accessory renal arteries were selected and arteriograms performed. The dominant vascular supply to the mass was from one of the lower renal arteries (below the inferior mesenteric artery) which demonstrated a well-defined hypervascular mass with significant tumour blush (Plate 2a). This accessory vessel was selected and embolized till there was stasis (Plate 2b) using 350 to 500 µ of contour embolization particles (Boston Scientific, USA).

Post-embolization angiogram showed satisfactory devascularization of the tumour with the disappearance of the tumour blush. Although both kidneys were connected by renal parenchyma, achieving super-selection of the dominant vascular supply and embolization of the tumour to stasis reduced the risk of complication to the normal kidney. Final arteriograms confirmed that there was a good flow to both kidneys. All intravascular devices were removed, and hemostasis achieved using manual compression for 20 min. The patient was asked to remain supine with strict bed rest for 4 h. The patient left the interventional radiology recovery in a stable condition after an overnight stay. No immediate complications were encountered. A CT scan that was done 4 wk post-procedure revealed a reduction in the size of renal AML from 5.60 cm x 5.20 cm to 3.51 x 3.40 cm (Plate 3).



(a) (b)

Plate 2: (a) Arteriogram through the dominant vascular supply to the mass demonstrating a well-defined hypervascular mass 'Y' with significant tumour blush (b) Post-embolization arteriogram showing significant tumour devascularization 'Z'



(a) (b)

Plate 3: (a) Pre-embolization contrast-enhanced axial CT scan demonstrating heterogeneous renal angiomyolipoma 'X' with areas of macroscopic fat. 'W' shows the horseshoe kidney (b) Post-embolization unenhanced axial CT scan shows AML has significantly decreased in size from (5.60 x 5.20) cm to (3.51x 3.40) cm. 'W' shows the horseshoe kidney

DISCUSSION

Although AMLs are not uncommon, it is quite rare to find them in a horseshoe kidney. In 2010, Dave et al. reported of a case of an asymptomatic AML in a horseshoe kidney in pregnancy which was successfully resected during laparotomy, following an elective caesarean section at 39 wk and confirmed by histopathology [11]. Most patients with renal AMLs are asymptomatic when diagnosed since the majority are incidental findings on imaging [7,12]. Symptomatic presentation is more common with renal AMLs associated with Tuberous Sclerosis Complex [7]. Spontaneous retroperitoneal haemorrhage is the most common symptomatic presentation. Patients may also present with a palpable mass, flank pain, haematuria, hypertension, anaemia, urinary tract infection, or renal failure [7,12,13]. Demonstration of macroscopic fat within the lesion is a distinctive feature on all imaging modalities. However, not all AMLs can be diagnosed solely with imaging due to variable quantities of fat. Image-guided percutaneous renal biopsy is useful in these instances in addition to differentiating between benign and malignant renal lesions. Differential diagnoses for renal AML include renal cell carcinoma, peri-renal liposarcoma and renal oncocytoma [7]. The main indications for treatment include the presence of symptoms, presence in women of childbearing age or pregnant women, suspicion of malignancy, and a size larger than 4 cm. Patients not requiring any immediate interventions may be followed up with active surveillance (AS) [6,7].

Earlier studies have suggested 4 cm as the limit above which intervention should be considered. One of such is a popular review published in 1986 by Oesterling et al. where it was reported that 82% of patients with renal AMLs larger than 4 cm experienced symptoms. Furthermore, over 50% of those with tumour size > 4cm in that review had active retroperitoneal haemorrhage with a third presenting in shock [14]. Other studies also revealed that tumours with sizes of more than 4 cm had interval growth and needed treatment often [15-17]. However, a large series on AMLs found no difference in growth rates between AMLs > 4 and ≤ 4 cm during AS and thus concluded that lesions > 4 cm do not require early intervention based on size alone. These findings suggest an initial AS for large (> 4 cm) asymptomatic lesions is a safe management option [16].Treatment options include partial nephrectomy, radical nephrectomy, selective renal artery embolization, cryo- and radiofrequency ablation, as well as treatment with mammalian target of rapamycin inhibitors [6,7]. Traditionally, the first-line treatment for AMLs was surgical, in the form of either radical nephrectomy or partial nephrectomy. This was mainly because malignancy could not be ruled out. This however has changed since AMLs can now be accurately diagnosed. The main drawback of surgery is the risk of surgery-related complications, notably the potential deterioration of renal function in radical nephrectomy, especially in a patient with variant vascular anatomy [6,7]. Due to these drawbacks with surgical treatment and coupled with the increasing evidence for the use of less invasive techniques, many clinicians now opt for embolization as first-line treatment [6]. Embolization has been validated by previous studies to be efficacious in causing tumour shrinkage as well as prevention and management of tumour rupture [6,8,9,18]. A study by Hocquelet et al. showed a significant reduction of tumour burden, with a mean volume reduction of 72% after a median follow-up time of 28 mos. Another important finding was that high-fat content lesions were less amenable to embolization than low-fat content lesions [9].

Before tumour embolization, pre-procedure contrast-enhanced computed tomography (CT) or magnetic resonance imaging (MRI) are reviewed to assess tumour burden as well as delineate vascular anatomy and variations that may negatively impact on its success [6]. Also, pre-procedure computed tomography angiography is useful in identifying dominant vascular supply and may demonstrate arteriovenous shunting if present [6]. Arterial access is traditionally obtained via the femoral artery although the trans-radial route, is an equally effective alternative route. The procedure is performed under local anaesthesia or moderate sedation. Pre-embolization arteriograms are done for intra-procedural confirmation of vascular anatomy as well as to aid in the super-selective catheterization of artery most likely supplying the tumour. A co-axial microcatheter may be helpful to achieve the latter [6,19]. The appropriately determined embolic agent, which may be particles or a mixture of ethanol and ethiodized oil is administered into the feeding artery which is then embolized till stasis is achieved [6,20]. Ethanol and lipiodol mixtures or particles such as spheres or polyvinyl alcohol cause complete occlusion of small vessels feeding tumours, thus are the preferred choice for embolization [6]. Post-embolization arteriography is performed for confirmation of tumour devascularization and preservation of vascularity in the remainder of the kidney [6,20]. Complications after embolization include post-embolization syndrome which occurs due to an inflammatory response to necrotic tissue. This occurs within 3 - 7 days after the procedure with symptoms of abdominal pain, fever, nausea, and leukocytosis [6,7,20]. This condition is self-limiting and resolves with standard supportive care including antipyretics, anti-emetics and analgesia [12,17,20]. Other uncommon complications include non-target embolization of normal parenchyma, renal infarction with abscess formation and arterial access site complications [6,7,13].

Conclusion

Renal AML is the commonest benign renal tumour with the potential to result in life-threatening sequelae if proper management is not executed. Diagnostic accuracy is fundamental to initiating appropriate treatment. Although several treatment options exist, symptomatic patients with tumours > 4 cm should be considered for arterial embolization which is an effective, evidenced-based and safe minimally invasive technique for reducing the size of tumours as well as preventing tumour rupture.

DECLARATIONS

Ethical considerations
The authors obtained informed consent from the patient. All personal identifiers were removed to ensure patient confidentiality and prevent traceability of publication to the patient. Euracare Advanced Diagnostics and Heart Centre Ghana Limited, Accra, Ghana, approved the use of patient data for publication.

Consent to publish
All authors consented to the publication of the manuscript.

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Competing Interests
No conflict of interest was reported by the authors.

Author contributions
BDS, DAA and BBJ contributed to the management of the case, drafting of the report, and final review of the report.

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Availability of data
All relevant data are provided in the manuscript. The published data is available from the corresponding author on a reasonable request.

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