Managing a massive renal angiomyolipoma

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DECLARATIONS

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Renal angiomyolipoma is a benign tumour of the kidney that can lead to long term morbidity irrespective of renal function.

Case report

A giant renal angiomyolipoma (AML) measuring 18 × 13.2 × 22.4 cm was found in the left kidney of this 43-year-old female (with no evidence of tuberous sclerosis [TS]). Her other kidney was atrophic contributing only 3% of total renal function (damaged by chronic pelvi-ureteric junction obstruction). The grossly hypertrophied left kidney was causing displacement of the ureter, pancreas and spleen (Figure 1). There was gross neovascularization within the AML, and the patient suffered three life threatening episodes of haemorrhage requiring interventional radiological procedures which attempted to coil the arteriovenous malformations. On one of these occasions the procedure caused a renal artery aneurysm and a radiological attempt at repair resulted in contrast induced anaphylaxis thereby precluding any further radiological attempts. Owing to the risk of further haemorrhagic episodes, a difficult decision was made to attempt a partial nephrectomy to excise the angiomyolipoma – at this risk of rendering the patient effectively anephric and dialysis dependent if total nephrectomy had to be performed to control bleeding.

Intraoperatively the surgeons were able to successfully resect a segment of AML measuring 14 × 19 cm. Postoperatively there was a prolonged period of hypotension requiring inotropic support and blood product replacement. She underwent three further abdominal operations to investigate and treat ongoing blood loss and abdominal compartment syndrome. This was subsequently exacerbated by intra-abdominal sepis – perihepatic collections necessitating ultrasound guided drainage and episodes of pneumoniae with multiple courses of intravenous antibiotics. During her ITU course she required haemofiltration, ventilation via a tracheostomy and total parenteral nutrition. Upon clinical improvement a right sided homonymous hemianopia was noted, with evidence of a left posterior circulation artery territory ischaemia demonstrated on head MRI scan (Figure 2). The patient made a very slow recovery and was discharged 50 days later, with normal renal function, a large open abdominal wound (left to heal by secondary intention) and a new visual field defect. She has subsequently developed an incisional hernia and the visual defect has failed to resolve.

Discussion

AML was first described by Bourneville (1880), and is considered a benign hamartoma with polyclonal proliferation. AML can be found in multiple forms; classic, microscopic, cystic, epithelioid, oncocytoma-like.[1] They are more common than previously thought and it is now estimated that they affect 13 per 10000 adults.[2] Recently, clonal neoplasm and AML have been reclassified within a family of peri-vascular epithelioid cell tumours (PEComas).[1]

The most frequent form or classic AML; is composed of adipose tissue, spindle cells, epithelioid smooth muscle cells and abnormally thick walled blood vessels.[1] There is an association (estimated 10%) with TS, where the AMLs are more likely – and are frequently bilateral, small and multifocal.[2] However, cases of massive AMLs (measuring up to 45 × 20 × 15 cm) have also

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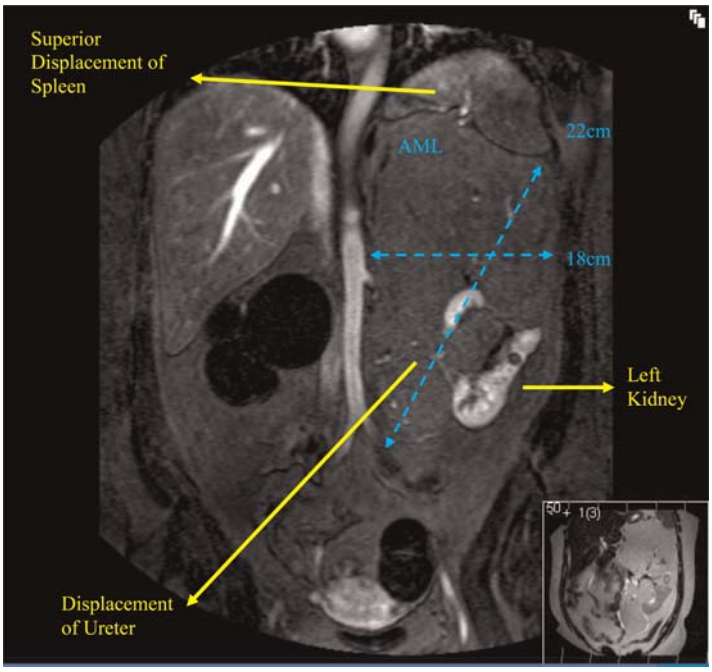
been reported.[3] Sporadic AMLs have a female preponderance, occurring between the ages of 30 and 50 years, and are more often unilateral and larger in size.[1]

Diagnostically AMLs are a challenge as radiological appearances vary, making it difficult to differentiate from a renal cell carcinoma (RCC). Current modalities of radiological analysis are ultrasound (US), computed tomography (CT) and magnetic resonance imaging (MRI). On US, AML is suggested by a strongly hyper-reflective lesion with acoustic shadowing, however they can also be hypo- or isoreflective. Siegel *et al*.[4] demonstrated an overlap in US appearances; such that up to 12% RCCs were sonographically indistinguishable from AML. Uncertainty on US usually leads to investigation with CT, where the classical appearance is a fat containing homogenous mass. MRI scanning reveals high signal intensity from fat on T1-weighted images. Using these techniques, 100% of AMLs were correctly identified in one study.[5]

Lesions less than 4cm are generally managed conservatively, however larger AMLs (>4 cm) are symptomatic in approximately 90% of cases and necessitate intervention.[5] Common problems are abdominal pain, palpable mass causing compressive symptoms and spontaneous aneurysm rupture.[5] Oesterling *et al*. (1986) proposed that patients with symptomatic AML greater than 4 cm should undergo angiogram and transcatheter renal artery embolization, enucleation or partial nephrectomy.[5] In practise this remains difficult to implement as evidenced by a case described by Galanis *et al*.[6] who reported a very large unilateral AML (31 × 16.5 × 8.5 cm) for which a complete nephrectomy was performed.[6] More recent evidence suggests that aneurysm size is more important than tumour size in determining risk of bleeding. A total of 14.3% of patients with lesions studied by Yamakado *et al*., with minimal vascularity (few small stretched vessels), presented with haemorrhage versus 50% of those displaying marked vascularity (multiple, large tortuous vessels).[7]

Novel treatment options investigate the role of vascular endothelial growth factor (VEGF) inhibition. AML cells produce VEGF *in vitro* which subsequently drives angiogenesis thereby further increasing vascularity within the AML. Anti-angiogenic drugs such as cyclooxygenase-2 inhibitors, interferons, thalidomide, retinoids have been poorly studied in benign tumours – however Arbiser *et al*. (2002) hypothesize that these agents may impede growth as they do in malignant tumours.[8] Bissler *et al*. (2008) demonstrated twelve months of treatment with sirolimus caused a reduction in AML volume to 27% of baseline – upon discontinuation of treatment there was subsequent increase in size; multicentred placebo controlled trials are currently underway to explore this further.[9] We have not considered this option in this patient who has residual AML because of possible side effects (sirolimus is associated with impaired wound healing and excess rates of hernia formation[10]) in a lady whose clinical course to date has been very challenging.

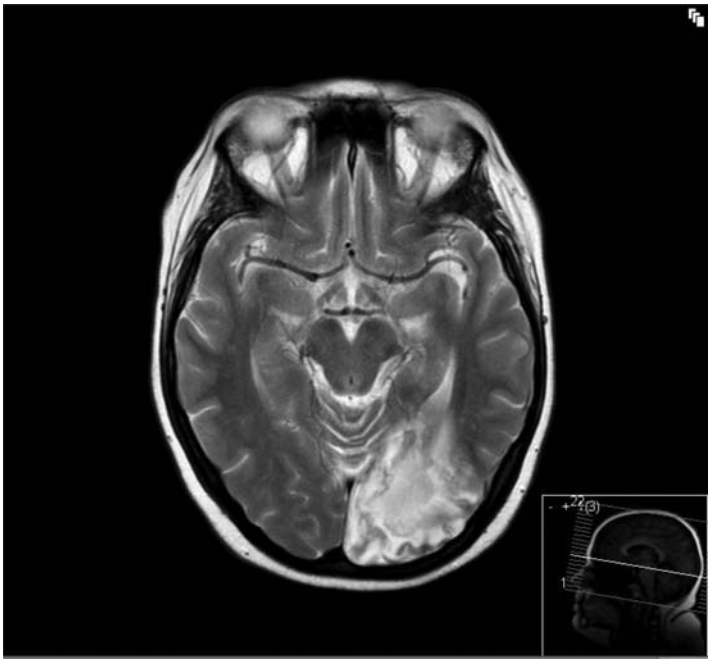
Figure 1
Magnetic resonance angiogram: Massive left sided AML with displacement of spleen and ureter



Summary

A case of allegedly benign giant renal AML is presented here because of its uncommon occurrence in an effectively single kidney and the huge clinical dilemma it presented for her clinicians, as well as the poor overall outcome for the patient despite eventual satisfactory renal outcome.

Figure 2
Axial T2 weighted MRI image: High signal intensity in left occipital lobe demonstrating acute ischaemic stroke



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