**CASE REPORT**

Combined minimal invasive methods for renal angiomyolipomas treatment

**Abstract**

Angiomyolipomas are composed of variable amounts of three components: blood vessels (angioid), smooth muscle (myoid) and mature fat (lipoid) components and consists the most common benign non cystic renal lesion. Most of AML cases are found incidentally when the kidneys are imaged for other reasons. However they do have the risk of rupture with bleeding or secondary damage of surrounding structures as they grow. The risk of bleeding and surrounding tissue damage is proportional to the size of the lesion (>4 cm diameter). Other symptoms and signs include palpable mass, flank pain, urinary tract infections, haematuria, renal failure, or hypertension. AMLs found incidentally are usually small and so require no therapy. Lesions that present with retroperitoneal haemorrhage often require emergency embolization as a life saving measure. Preventing treatment for larger AMLs, or those that have been symptomatic, include tumor resection or partial nephrectomy or selective arterial embolization. Although it is considered effective in preventing hemorrhage, the last seems less efficient in reducing AML regrowth risk especially in patients with multiple or large AMLs. Here we discuss the combination of selective arterial embolism and radiofrequency ablation in the treatment of giant renal angiomyolipomas.

**Key words**

- embolism;
- radiofrequency ablation;
- angiomyolipoma;
- solitary kidney

**Introduction**

Angiomyolipomas (AMLs) have been classified among the perivascular epithelioid cells tumour group (PEComas). They are composed of variable amounts of three components: blood vessels (angioid), smooth muscle (myoid) and mature fat (lipoid) components. AMLs consists the most common benign non cystic renal lesion. The liver is the second most frequent site. Two types have been described: the sporadic and multiple. The first occurs as a single tumour in one kidney. It accounts for 80% of renal AMLs and it is typically identified in adults, with a strong female predilection. The second occurs as larger tumour and/or multiple tumours in both kidneys and accounts for 20% of renal AMLs. It affects both sexes at a younger age than sporadic AML. It is seen in association with neuro-ocular-cutaneous disorders such as tuberous sclerosis, Von Hippel-Lindau syndrome and neurofibromatosis type 1. AMLs are benign and usually asymptomatic. In fact, most of AML cases are found incidentally when the kidneys are imaged for other reasons however they do have the risk of rupture with bleeding or

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The risk of bleeding and surrounding tissue damage is proportional to the size of the lesion (>4 cm diameter). Other symptoms and signs include palpable mass, flank pain, urinary tract infections, haematuria, renal failure, or hypertension. AMLs found incidentally are usually small and so require no therapy. Lesions that present with retroperitoneal haemorrhage often require emergency embolization as a life saving measure. Preventing treatment for larger AMLs, or those that have been symptomatic, include tumor resection or partial nephrectomy or selective arterial embolization. Although it is considered effective in preventing hemorrhage, the last seems less efficient in reducing AML regrowth risk especially in patients with multiple or large AMLs.

**Case Presentation**

A 78 y.o. patient with known AMLs on the left kidney, history of right nephrectomy due to mass renal haemorrhage caused by spontaneous rupture of a large pelvic AML and mild renal and cardiac insufficiency, was referred to the emergency department for abdominal pain, weakness, light-headedness and shortness of breath. Clinical symptoms and decreased haematocrit were suggestive for internal haemorrhage. In the

**Figure 1.** Axial CT images prior to intervention. (A) Unenhanced image shows the typical appearance of a large angiomyolipoma with fat and soft tissue (*) densities. The mass compresses the inferior vena cava (arrow). (B) Contrast-enhanced image (venous phase) shows a thrombus causing an enhancement defect (arrow) at the lowest part of the inferior vena cava

**Figure 2.** Axial CT images post embolization and ablation. (A) Unenhanced image shows that the angiomyolipoma is smaller, with relative shrinkage of the soft tissue component (*) in favor of the fat. The compression of the IVC is now less striking. (B) Contrast-enhanced image (venous phase) shows complete resolution of the caval thrombus (arrow).
ultrasound, fluid accumulation in the perirenal space and a 5 cm nodular hyperechoic lesion in the left kidney suggested a heterogeneous AML and probable haemorrhage. Radiological investigation confirmed the above findings and showed that haemorrhage was limited to the perirenal space. Despite transfusion and intravenous fluid replacement blood loss wasn’t stopped and therefore patient underwent selective arterial embolism. Patient was haemodynamically stabilised and none of the usual complications of arterial embolism occurred except a slight increase of body temperature. Upon follow up visit, no tumor regression was detected. Moreover, the tumor was found in close proximity to inferior vena cava where a thrombus of 1.5 cm in diameter was developed. The caval thrombus was treated with anticoagulant (warfarin in an average maintenance dose of 5 mg on the first and second days for an INR value in the range of 2.0 to 3.0). Since anticoagulant treatment had proved ineffective ten months later, it was decided to definitely manage the patient. A combined interventional treatment (consisted of a second selective arterial embolism session along with radiofrequency ablation of the AML plus conservative management of inferior vena cava thrombus) was decided. Transarterial embolization was performed first. Superselective approach was achieved and embolization was performed with tightly calibrated microspheres (Embozene, Celonova) with diameters of 250 and 400 micrometers. Post-embolization angiogram showed devascularisation of the AML and no signs of renal infarction. Radiofrequency ablation (with the use of radiofrequency electrode Jet-Tip, RF Medical Co) was applied 20 days later. The procedure was monitored ultrasonographically, and no complications were observed. The patient received intravenous hydration and antibiotics and was discharged the following day. Anticoagulation treatment was for continued for 2 more months. Upon follow up evaluation, AML size was reduced with relative shrinkage of the soft-tissue component in favor of the fat. The compression of the inferior vena cava was less striking while the caval thrombus was completely resolved.

**Discussion**

Management of symptomatic AMLs can be problematic in patients not suitable for surgery. Moreover, nephron preservation is essential for patients with impaired renal function and remain a key treatment consideration in many other patients (eg those with tuberose sclerosis complex and those who can have multiple, bilateral and very large AMLs). In these cases interventional radiology techniques can provide an alternative approach. In our case treatment intentions were both to decrease the risk of haemorrhage recurrence and decrease tumor size -and thus to facilitate the resolution of IVC thrombus- with the minimal effect on the renal function. Given the general health condition our patient and the fact that the thrombus was not actually resulted from vascular spread from the AML to the inferior vena cava, interventional radiology treatment was preferred instead of nephron-sparing surgery plus caval thrombectomy.

Selective arterial embolization of renal AMLs is currently uniformly performed to prevent hemorrhage in patients with AMLs larger than 4 cm. Although several studies have shown a low incidence of recurrence after embolization, this is true only for isolated renal AMLs. In fact, patients with multiple AMLs and patients with tuberosclerosis and Von Hippel-Lindau syndrome as well, suffer of recurrences after embolotherapy. Moreover, while regular selective embolotherapy can reduce the tumor size, there is a danger of non-target embolization. On the other hand, superselective embolization guarantees minimal tissue loss; however it may be ineffective both in the prevention of recurrences and tumor size reduction. Of note, the true long term recurrence rate is currently unknown and depends of the number of coexisting AMLs.

Radiofrequency ablation therapy (RFA) was proposed as an alternative to angio-embolization and nephron-sparing surgery for AML treatment. In fact, focused RFA is a nephron-sparing treatment option since destroys only the solid and vascular elements of the tumor, without encroaching on any normal renal tissue. The efficacy of RFA against AML was proven in a small number of cases and mainly for small, growing AMLs. Castle et al., treated successfully 15 cases of small renal AMLs. They report a low complication rate (13.3%) and no radiographic recurrences at a mean follow-up of 21 months. It should be mentioned that tumors -especially large- could not be significantly reduced after RFA and thus treatment may be misinterpreted as failed. Actually even when no change in overall tumor volume on follow-up radiological im-
aging occur, in most of the cases the tumors became fattier with involution of the soft-tissue elements (decrease in mean soft tissue-to-total tumor ratio). Further evidence of treatment effect is the development of a visible capsule around the ablation zone\textsuperscript{12}. Grego- ry et al., treated four large AMLs (maximal axis 6.1–32.4 cm). They report no complications and significant decrease in mean soft tissue-to-total tumor ratio during a minimum 48-month period. Prevoo et al., however, report a decrease in tumor size from 4.5 cm to 2.9 cm at 12 months after RFA of a sporadic AML in a patient with a solitary kidney. No complications occurred and no AML recurrence was observed during the 12-month follow-up\textsuperscript{13}.

Little information exists regarding the efficacy of the combination of selective arterial embolism and radiofrequency ablation in AML treatment: Sooriakumaran et al. treated two large sporadic AMLs (tumor size greater than 9cm) who had received selective arterial embolization before RFA and found a reduction in tumour mass of 20\% with evidence of significantly reduced vascularity after RFA and minimal enhancement of the treated areas on CT or MRI in all two cases after a median follow-up of 7.5 months\textsuperscript{5}. The above findings are comparable with that of our case.

Conclusions

Both selective arterial embolization and RFA are effective for AML of < 4 cm however their efficacy in the treatment of larger tumours is questionable. For this reason complementary radiofrequency ablation therapy to super-selective arterial embolization of renal AMLs is a reasonable alternative approach. Current data suggests that the above combined interventional treatment appears to be effective in the treatment of large AMLs but is insufficient to provide conclusive evidence. Large randomized prospective studies would be needed to establish the efficacy of this treatment.

Τα αγγειομυολιπώματα αποτελούνται από ποικίλες ποσοτή- τες τριών συστατικών: αιμοφόρα αγγεία, λείες μυικές ίνες και λίπος και αποτελούν την πιο συνηθισμένη καλοήθη μη κυστική νεφρική βλάβη. Στις περισσότερες περιπτώσεις βρίσκονται τυ- χαία, όταν οι νεφροί απεικονίζονται για άλλους λόγους. Όμως ενέχουν τον κίν- δυνο ρήξης με αιμορραγία ή δευτεροπαρα- θές βλάβης των γύρω δομών λόγω της ανάπτυξής τους και της συνεπακόλουθης πίεσης. Ο κίνδυνος αιμορραγίας και βλά- βης των γύρω ιστών είναι ανάλογη με το μέγεθος της βλάβης (διάμετρος> 4 cm). Άλλα συμπτώματα και σημεία περιλαμβάνουν ψηλαφητή μάζα, πόνο στα πλευρά, λοιμώξεις του ουροποιητικού συ- στήματος, νεφρική ανέπαρκεια ή υπέρταση. Τα αγγειομυολιπώματα που βρίσκονται παραπεμπτότως είναι συνήθως μικρά και έτσι δεν απαιτούν θεραπεία. Αντίθετα οι βλάβες που παρουσιάζονται με οπιοθεραπευτική αιμορ- ραγία συχνά απαιτούν επείγουσα αντιμετώπιση -συνήθως με εμβολισμό- για τη διάσωση του αρρώστου. Η προληπτι- κή θεραπεία για τα μεγαλύτερα αγγειομυο- λιπώματα, ή εκείνων που συνδέονται με τα συμπτώματα που αναφέρθηκαν παρα- πάνω περιλαμβάνει την εκτομή του όγκου ή τη μερική νεφρεκτομή ή τον επιλεκτι- κό αρτηριακό εμβολισμό. Παρόλο που o τελευταίος θεωρείται αποτελεσματικός στην πρόληψη της αιμορραγίας, φαίνε- ται πως είναι λιγότερο αποτελεσματικός στη μείωση του κινδύνου αναγέννησης του όγκου ιδιαίτερα σε ασθενείς με πολλαπλά ή μεγάλα αγ- γειομυολιπώματα. Στο άρθρο αυτό συζητάμε τον συνδυασμό του επιλεκτικού αρτηριακού εμβολισμού με την καυτηρίαση με ραδιοσυχνότητες στην αντιμετώπιση γιγάντιων νεφρικών αγγειομυολιπώματων.

**Lέξεις ευρετηριασμού**

εμβολισμός, καυτηρίαση με ραδιοσυχνότητες, αγγειομυολίπωμα, μονήρης νεφρός
References