Letter to the Editor

WHAT IS THE PRESENTATION OF RENAL ANGIOMYOLIPOMAS?

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To the Editor

Renal angiomyolipomas (AMLs) are commonly benign tumours formed by fat, blood vessels and non-striated muscle, which tend to be associated with neuroendocrine syndromes, or turn up sporadically (1). They can, depending on their histological features, express cell growth markers and show high proliferation levels, in addition to atypia and an increment of mitosis. In this way, they can be classified as benign and malign tumours (2). Basically, renal AMLs manifest in two forms: incidental finding after imaging tests for another pathology process, or by a spontaneous rupture accompanied by retroperitoneal bleeding that may even be lethal (3). An extensive series of individuals studied by Nelson et al reports that 80% of renal AMLs occur sporadically and without associated syndrome complex, while renal AMLs associated to neuroendocrine syndromes are less frequent (2, 4).

Our experience in renal AMLs in the last 5 years is summarized in 15 cases (14 women and 1 men), with a mean age of 55 years at the time of diagnosis. In 13 cases, renal AMLs manifested sporadically, were unilateral and showed a mean size of 51 mm as maximum diameter. Only in 2 cases the condition had a multiple form and a bilateral presentation, as one of them was associated with Von Hippel-Lindau disease, and the other with tuberous sclerosis. Nine of the aforementioned 15 patients had to undergo surgical treatment either because of a spontaneous retroperitoneal bleeding that lowered haemoglobin levels and led to hypotension, or because of the size of the renal mass with increased risk of spontaneous rupture. The remaining 6 cases, in which the diagnosis was incidental due to ultrasonography or CT-scan, are at present under regular follow-up. Only one case out of the nine that underwent surgery showed a histological result of epithelioid AML, which is a malign variant of perivascular epithelioid cell tumour (PEComas). The remaining 8 tumours after surgery proved to be benign AMLs.

As can be seen from the literature, the main problem with renal AMLs is a spontaneous rupture, also known as Wunderlich syndrome, which can lead to potentially lethal complications. Open and laparoscopic surgery procedures are necessary to arrest haemorrhage, remove the bleeding lesion, and, if necessary, perform a partial or total nephrectomy to save the patient's life (2, 3, 4).

Because of the peculiar presentation of renal AMLs, when a patient complains of pain in the renal fossa or on one side, associated with both arterial hypotension and vegetative syndrome (sweating, dizziness, chills, nausea), it is advised to perform imaging procedures to eliminate the occurrence of a bleeding retroperitoneal renal mass. When the finding of the renal AML is accidental, the follow-up and treatment will depend on the size and localization of the tumour. When the tumour is smaller than 40 mm, our option is to establish a regular follow-up of the mass through ultrasonography or CT-scan. When the size is over 40 mm, we consider either to remove the lesion or to embolise the lesion, given the risk of spontaneous rupture.

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