Transarterial embolization of ruptured angiomyolipoma associated with tuberous sclerosis

Shen Yang Lee¹, Li-Jen Wang², Tzung-Hai Yen³ and Chiz-Tzung Chang¹

¹Department of Nephrology and ²Department of Radiology, Chang-Gung Memorial Hospital, Taipei, Taiwan and ³Histopathology Unit, Cancer Research UK, London Research Institute, London, UK

Keywords: angiomyolipoma; transarterial embolization; tuberous sclerosis

Case

A 29-year-old woman, with a past history of tuberous sclerosis diagnosed in her childhood, presented to our emergency department with the acute symptoms of left flank pain. She had no fever and she was fully conscious with stable haemodynamics. Physical examination revealed facial red bumps, and a knocking pain over the left flank. Laboratory investigation showed mild anaemia but normal liver and renal functions. Computed tomography showed a huge ruptured angiomyolipoma of the left lower kidney. Angiography also revealed a hypervascular tumour at the same location (Figure 1). She was treated with transarterial embolization, which was super-selective to the inferior branch of the left renal artery. Post-embolization angiography showed markedly diminished vascularity of the same lesion (Figure 2). The patient developed post-embolization syndrome consisting of fever, flank pain and vomiting after the embolization procedure. These symptoms subsided after acetaminophen treatment. She was then discharged within 2 weeks. Notably, her renal function remained normal at 6 months follow-up.

Discussion

Renal manifestations of tuberous sclerosis comprise angiomyolipomas, cysts and, rarely, renal cell carcinomas. Angiomyolipomas occur in 75% of children with tuberous sclerosis and continue to grow during adulthood. They are usually multiple, bilateral and symptomatic as compared with sporadic angiomyolipomas. An enlarging angiomyolipoma can distort the renal architecture, causing renal failure and even death [1]. Dysmorphic blood vessels in the angiomyolipoma often form microaneurysms, which may rupture and result in renal haemorrhage. Ruptured angiomyolipomas are most commonly seen in lesions >4 cm in diameter [2]. Current management options include observation, embolization and partial or total nephrectomy. Surgery often leads to loss of parenchyma and renal function. Transarterial embolization, due to its relatively lower risk and better preservation of renal function, may be a better option, as demonstrated in this case.

Conflict of interest statement. None declared.

References


Received for publication: 6.12.04
Accepted in revised form: 17.2.05
Fig. 1. Pre-embolization angiography shows marked hypervascularity and aneurysmal dilatation of the angiomyolipoma (arrows) located in the lower pole of the left kidney. There are several smaller angiomyolipomas at the upper pole (arrowheads).

Fig. 2. Left renal arterial angiography after selective embolization of the lower polar angiomyolipoma with the use of a microcatheter shows diminished vascularity of this angiomyolipoma (arrows).