KIDNEY INVOLVEMENT IN TUBEROUS SCLEROSIS COMPLEX

Sultan Ceren Yıldırım, Gülen Gül Mert, Aysun Karabay Bayazıt*, Şakir Altunbaşak. Çukurova University, Adana, Turkey

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Purpose Tuberous sclerosis complex (TSC) is an inherited neorcutaneous disorder that is characterized by pleomorphic features involving many organ systems. Renal manifestations are the second most significant cause of morbidity and mortality in patients with tuberous sclerosis complex (TSC), and include renal cysts, angiomylipomas (AMLs) and malignant tumors. In this study, we investigated patients with tuberous sclerosis in our clinic for renal involvement.

Methods In our clinic, the renal manifestations of children with TSC between 0–18 years of age were evaluated. Age of the first diagnosis, TSC history of family, renal ultrasonography findings of angiomylipomas and cysts such as size, quantity, location, renal function tests, urinalysis, presence of hypertension, additional organ involvement, and the presence of renal cell carcinoma were assessed.

Results There were 17 (8 male and 9 female) patients with TSC. The mean age of the patients was 11.6 years and first diagnosis time was 2.3 years. Angiomylipoma was the most frequent lesion (15 of 17 patients) and twelve of them were bilateral. At the time of diagnosis 3 patients had angiomylipomas. The sizes of AMLs of the patients were smaller than 5cm. Six patients had also renal cysts and 2 of them with renal cysts had no AML. Additional organ involvement was observed in 3 patients. All of the patients had normal renal function tests and urinalysis.

Conclusion The most common renal lesions in TSC are angiomylipomas and kidney cysts. At the time of TSC diagnosis, all the children must be screened for renal involvement and changing of renal findings in TSC with time should not be forgotten since the new findings can be added to old ones.