

Cardiac rhabdomyomas associated with tuberous sclerosis complex in childhood in 11 cases

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Introduction: Cardiac rhabdomyomas (CRs) are the most common pediatric heart tumor and closely associated with tuberous sclerosis complex (TSC).

Methods: We reviewed our patients with CRs who received a diagnosis of TSC previously or during follow-up between June 1996 and January 2012. We evaluated the presentation type, clinical course and outcome of all the cases diagnosed both prenatally and postnatally. All the medical data of the patients obtained from their medical records and computerized database in our hospital, retrospectively.

Results: 11 (% 34) of 32 patients with TSC had a total of 29 CRs. The median follow-up period was 2 years (range: 15 days-15 years). Five patients (% 45) had multiple tumors. Clinical presentation was cyanosis in two patients and cardiac murmur in three patients. Six patients were asymptomatic and CRs detected while cardiac evaluation for TSC. Prenatal screening revealed intracardiac tumors in two patients who received diagnosis of TSC in the follow-up. One of them had single cardiac tumor, epilepsy and supraventricular tachycardia due to Wolff-Parkinson-White syndrome and rhabdomyoma regressed spontaneously in two years. The other patient who had prenatally diagnosed had multiple CRs with significant intracardiac obstruction and ventricular tachycardia. Because he was accepted inoperable, he managed medically (mTOR inhibitor) and obtained rapid response to the treatment initially. Two patients (% 18) underwent cardiac surgery in the infancy period because of hemodynamically significant obstruction and their tumors removed totally. Six of the 29 CRs (% 21) showed complete regression spontaneously in three patients.

Conclusions: CRs are common in the patients with TSC and although they are pathologically benign tumors, sometimes they can cause life threatening events in this population. Thus, cardiac screening of the patients with TSC and fetuses or children of the parents with TSC can reveal significant pathologies in these patient groups.