Prenatally diagnosis and outcome of fetuses with rhabdomyoma - single centre experience

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Background Cardiac rhabdomyomas (CRs) are the most common primary tumors of the heart in neonates and infants/children. Usually are multiple and, based on the location, can cause haemodynamic disturbance, dysrhythmias or heart failure during the fetal and early postnatal period. CRs have a natural history of spontaneous regression and are closely associated with tuberous sclerosis complex (TSC). It has an association with tuberous sclerosis (TS) and in those cases the tumor may regress and disappear completely, or remain consistent in size.

Objective We aimed to evaluate the prenatal diagnosis, clinical presentation and outcome of CRs and their association with TSC in a single centre. The median follow-up period was three years (range: 6 months - 5 years).

Methods We reviewed medical records of all foetuses diagnosed prenatally with cardiac rhabdomyomas, covering the period from January 2010 to December 2016, which had undergone detailed ultrasound evaluation at a tertiary level centre with limited technical resources.

Results Twelve fetuses were included in the study; all with multiple tumors and, a total of 53 tumors were identified in all patients - the maximum was one fetus with 12 tumors. All patients were diagnosed prenatally by fetal echocardiography. In two foetuses haemodynamic disturbances during the fetal period were noted and pregnancies have been terminated. After long consultation, termination of pregnancy was chosen by the parents in totally 8 cases. In four continuing pregnancies, during the first year of life tumors regressed. TSC was diagnosed in all patients during the follow-up.

Conclusions Cardiac rhabdomyomas are benign from the cardiovascular standpoint in most affected fetuses. An early prenatal diagnosis may help for an adequate planning of perinatal monitoring and treatment with involvement of a multidisciplinary team. Large tumor size, number of tumors and localization may cause hydrops and they are significantly associated with poor neonatal outcome.

Keywords: heart tumor, cardiac rhabdomyoma, fetal echocardiography, tuberous sclerosis complex