19th World Congress in Fetal Medicine

Cardiac Rhabdomyoma – a case series assessing the implication of prenatal findings on long term postnatal outcomes

Jain A, Acharya V, Shettikeri A, Sahana R, Radhakrishnan P
Bangalore Fetal Medicine Centre, Bangalore, India

Objective

Prenatal detection of cardiac rhabdomyoma is rare, even though it remains the commonest fetal cardiac tumour. They are associated with Tuberous Sclerosis which is an autosomal dominant genetic condition with cardiac and extra-cardiac involvement particularly, that of the brain and kidneys. The objective of this study was to assess the antenatal cardiac and extra-cardiac findings and their implication on long term follow up amongst the survivors.

Methods

This is a retrospective study of prospectively collected data from a tertiary care fetal centre of fetuses diagnosed with cardiac rhabdomyoma from 20-36 weeks. The study period was from January 2007 to December 2014. Advanced fetal echocardiography was performed by FMF operators for all with suspected cardiac defects and recorded on Astraia fetal database software. The prenatal records were examined for clinical history and scan findings with cardiac and extra-cardiac defects. All parents were offered genetic consultation, counselling with a paediatric cardiologist and fetal surveillance. Long term pregnancy outcomes were obtained by telephonic/electronic communication with the parents.

Results

In total, there were 10 fetuses detected during the study period. Antenatal diagnosis was made in all cases, with 4 being detected in the second trimester and 6 in the third trimester. They comprised of 9 singletons and 1 twin in a monochorionic diamniotic pregnancy. There was a significant family history in 4 cases with Tuberous Sclerosis present in one of the parents. There were 6 cases with solitary tumour and 4 with multiple tumours. 1/6 (16.6%) with solitary tumour and 2/4 (50%) with multiple tumours (75%) had extra-cardiac involvement, respectively. All 3 cases with extra-cardiac findings had intracranial tubers. In addition, one case had echogenic kidneys along with intracranial tubers and multiple rhabdomyomas. 1 case with multiple rhabdomyomas had cardiac bradyarrhythmia. 9/10 (90%) pregnancies resulted in live births and 1/10 (10%) pregnancy underwent termination. On long term follow up, 5/9 (55.6%) survived, 3/9 (33.3%) had neonatal death (NND) and 1/9 (11.1%) had infant death. Amongst the 5 survivors, 3 (60%) had solitary tumour and 2 (40%) had multiple rhabdomyomas. None of the survivors had extra-cardiac defect. The one with bradyarrhythmia remained stable postnatally, didn't require any pacemaker and remains under the care of a paediatric cardiologist till date. There was no expressed concern for the survivors by the family and they have grown appropriate for age with no functional impairment. Data for postnatal echocardiography was available for 3 cases which showed resolution of the tumour with age. Amongst the 4 deceased, 2/4 (50%) had solitary tumour and 2/4 (50%) had multiple rhabdomyomas. Both the cases with solitary tumours had early NND, but this was secondary to meconium aspiration syndrome and premature termination of the monochorionic pair because of TTTS, respectively. Both the cases with multiple tumours which resulted in NND and infant death, respectively had intra-cranial involvement as well, resulting in seizures.

Conclusion

Our study showed long term survival with minimal morbidity in those with isolated cardiac rhabdomyoma. Extra-cardiac involvement was associated with poor perinatal outcomes. Genetic evaluation of parents is warranted in such cases for the possibility of Tuberous Sclerosis. Successful outcomes require multi-disciplinary approach with fetal medicine specialist, geneticist, neonatologist and paediatric cardiologists. Enhanced fetal surveillance for monitoring the progression of the tumour and development of hydrops and/or bradycardia, should be done to provide optimal outcomes. Postnatally, a detailed evaluation followed by periodic clinical assessment with echocardiography can result in increased overall survival rate.

Figure 1 : Multiple Rhabdomyomas

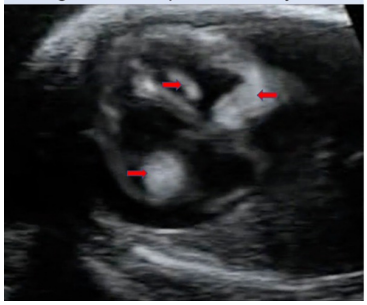

Figure 2 : Intracranial Tubers

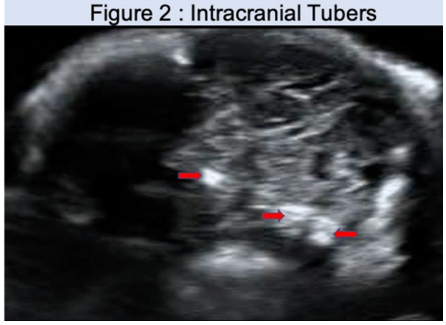

Table 1 : Patient characteristics with outcomes

Patient	Solitary /Multiple	Extracardiac Involvement	Outcome	Long Term follow up
Fetus 1	Solitary	No	NND	NND
Fetus 2	Solitary	No	LB	Live at 14 years
Fetus 3	Multiple	Hydrops, Brain	LB	Infant Death
Fetus 4	Solitary	No	LB	Live at 12 years
Fetus 5	Multiple	No (Cardiac Arrythmia)	LB	Live at 11 years
Fetus 6	Multiple	No	LB	Live at 12 years
Fetus 7	Multiple	Kidneys, Brain	NND	NND
Fetus 8	Solitary	No	NND	NND
Fetus 9	Solitary	Brain	Termination	Termination
Fetus 10	Solitary	No	LB	Live at 8 years