Patient Case Observation: Fetal Cardiac Rhabdomyoma

A 19 years old primigravida Bahraini female, of a first degree consanguineous marriage and spontaneous conception, had a regular antenatal follow-up in Bahrain Defence Hospital. She is a known case of bicornuate uterus diagnosed through early pregnancy. Her last menstrual period was on 12 June 2017 with an early scan showed expected date of delivery 18 March 2018. There was no major illness in the past, no history of any congenital anomalies in the family. Has a laser cosmetic therapy for upper lip mole. The patient did an anomaly scan which showed a single fetus with microcephaly and intraventricular rhabdomyoma.

Antenatal fetal echo study confirmed multiple masses related to the ventricular myocardium, the largest of which measured 18x12mm in the left ventricle, the other two masses measured 14x8mm and 15x7mm related to the posterior interventricular septum. There was no obstruction to blood inflow or outflow from the ventricles. The presence of multiple cardiac tumors consistent with rhabdomyoma most likely secondary to tuberous sclerosis. The pregnancy may allow going term and may have normal vaginal delivery. The patient came in early labor at thirty-seven weeks gestation with breech presentation, delivered by emergency lower segment caesarian section; a live male baby of 2.48 kg with an Apgar score of nine and ten at one and five minutes after delivery respectively. Patient’s postnatal course was uneventful and was discharged on third day postpartum.

At day 1 of life, the infant developed tachypnea and desaturation kept on nasal continuous positive airway pressure (CPAP) but still oxygen requirement is high so intubated and kept on PTV mode still not maintaining then shifted to High-frequency osculatory ventilation (HFO). He needed nitric oxide along with a norepinephrine for Persistent pulmonary hypertension (PPHTN), weaned gradually as kept on room air since day nine of life. Fetal US brain done showed both cerebral hemispheres are of normal morphology and echo pattern. No abnormal focal echogenicity. Unremarkable appearances of the ventricular system with no evidence of hydrocephalus. No detectable intracerebral, intraventricular or subarachnoid hemorrhage now.

Tuberous sclerosis was confirmed. Fetal echo has done after birth showed a large rhabdomyoma attached to the intraventricular septum to the left ventricle side measured 18x14mm, additional two large ones at the atrioventricular groove measured 5x4mm and 3.5x5mm and right ventricle apex measured 10x6.9mm. A multiple rhabdomyoma was seen at the left ventricle/right ventricle free wall and intraventricular septum. No vessel obstruction. PFO shunt seen with a right to left flow. Trivial mitral regurgitation with mild tricuspid regurgitation. PDA shunt 2-3mm seen with right to left flow. Normal aortic arch, no coarctation of the aorta. Ejection fraction 71%. Good biventricular systolic function. No pericardial effusion.

The infant upon discharge was in stabled condition. Discharged on day 16 of life with a weight of 2.375 kg, and has been managed as an outpatient in pediatric and pediatric cardiology clinic at our hospital. After delivery, examination by a pediatric cardiologist confirmed that the infant had a cardiac tumor and the cardiac function of the infant was diagnosed as normal.Stayed at hospital for approximately 3 weeks duration and discharged in stable condition.

At 1 month of age, readmitted with an episode of cyanosis and uprolling of eyes lasted for few

seconds. Discharged after 5 days in stable condition. At 2 months of age, readmitted as decreased activity to rule out sepsis. During admission developed supraventricular tachycardia which aborted with adenosine, admitted 1 day in ICU. Discharged after 3 days in stable condition. Repeated echo at age of 3 months; the largest tumor slightly reduced in size while the others almost remain same size.

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