Regression of Multiple Inoperable Cardiac Rhabdomyomas in a Neonate after Everolimus Treatment

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Introduction: Rhabdomyomas are the most cardiac tumors diagnosed in neonates and infants. We presented a critically sick neonate with multiple inoperable cardiac rhabdomyomas that infiltrated most of the RVOT region intramural and caused obstructions.

Case Report: The term male newborn was born at 39 weeks of gestation and 3550 gr. He was resuscitated after birth and then he was placed on mechanically ventilation. The postnatal echocardiography (ECHO) revealed multiple rhabdomyomas in the left ventricle, right atrium, and right ventricle with evidence of right ventricle outflow-tract (RVOT) obstruction. The mass located in the RVOT was very large and infiltrated most of the RVOT region intramuraly. He was not tolerate weaning from the mechanical ventilation. Postnatal cranial MRI revealed multiple hamartomas along the periventricular zone. Postnatal Renal Ultrasound revealed multiple angiomyolipomas that originate both kidneys. Everolimus (mammalian target of rapamycin (mTOR) inhibitor) therapy was started since the patient was not tolerate extubation and rhabdomyomas were inoperable and causes RVOT obstructions. The patient was tolerated the extubation 8 days following initiation of the medical therapy. Echocardiography at 2 weeks following initiation of everolimus therapy showed regression of the RVOT tumor. Tumor regression continued and at 2 months of age significant reduction in the rhabdomyomas was observed. The RVOT tumor was hardly visible without significant gradient across the RVOT. At the time of writing the child was continued everolimus therapy.

Conclusions: Our patient had a very large mass that infiltrated most of the RVOT region intramuraly and caused obstructions. Our patient is a critically sick neonate, resuscitated after birth secondary to multiple inoperable cardiac rhabdomyomas causing right ventricular outflow-tract obstruction, who was successfully treated with everolimus (mammalian target of rapamycin (mTOR) inhibitor). Symptomatic cardiac rhabdomyomas might be managed with everolimus treatment. However further studies needed for effective dose of everolimus and its side effects in children.