

REGRESSION OF MULTIPLE INOPERABLE CARDIAC RHABDOMYOMAS IN A NEONATE AFTER EVEROLIMUS TREATMENT

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INTRODUCTION

Primary cardiac tumors rare during childhood. Rhabdomyoma is the most common cardiac tumour seen in infants and children. It usually occurs in association with tuberous sclerosis complex (TSC). The clinical manifestations of TSC include hamartomatous lesions of the brain, skin, heart, lungs and kidneys.

Cardiac rhabdomyoma (CR) often shows spontaneous regression, but the tumor mass may cause ventricular inflow/outflow obstruction, arrhythmias or congestive heart failure. There are only few previous reports about rhabdomyoma cases that showed regression under everolimus treatment.

As a result of evidence we used everolimus in a neonate with multiple inoperable cardiac rhabdomyomas causing right ventricular outflow-tract obstruction.

Case Report

We report a case of a term male newborn who was born at 39 weeks of gestation. He was resuscitated after birth and then he was placed on mechanical ventilation. Physical examination indicated a grade 2/6 systolic murmur at the apex position and a few hypopigmented skin lesions on the trunk. The postnatal echocardiography (ECHO) revealed multiple rhabdomyomas in right atrium and right ventricle with evidence of right ventricle outflow-tract (RVOT) continuous Doppler (CW Doppler) measured 20 mmHg gradient between right ventricle and pulmonary artery (Figure 2). The mass located in the RVOT was very large (Figure 1). There was a mild mitral regurgitation (Figure 3). There were also smaller masses in right atrium (RA) that originated from interatrial septum and in left ventricle that originated from interventricular septum (Figure 4). He was not tolerate weaning from the mechanical ventilation. Postnatal cranial MRI revealed multiple hamartomas along the periventricular zone. Postnatal cardiac MRI revealed multiple CR (Figures 5-6). Everolimus 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 (Figure 7). Tumor regression continued and at 2 months of age significant reduction in the rhabdomyomas was observed (Figure 8). Mitral regurgitation decreased and the RVOT tumor was hardly visible without significant gradient across the RVOT. During the therapy no side effects were observed. At the time of writing the child was continued everolimus therapy.



Figure 1. RVOT CW Doppler



Figure 2. RV large tumor mass



Figure 3. Mild mitral regurgitation



Figure 4 LVOT obstruction

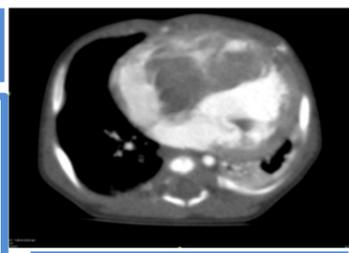


Figure 5. Cardiac MR Axial

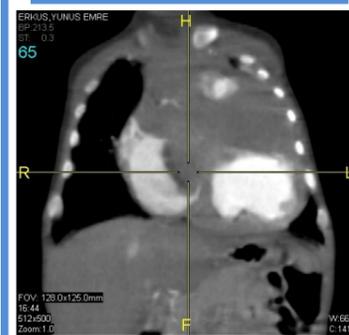


Figure 6. Cardiac MR Cranial



Figure 7. RVOT tumor regression

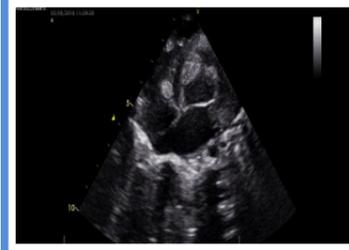


Figure 8. RV tumor regression

CONCLUSION

More than 60% of primary tumours of the heart are rhabdomyomas. Most of the cases are asymptomatic. However cardiac rhabdomyomas may cause ventricular inflow/outflow obstruction, arrhythmias or congestive heart failure depending upon the anatomical location of the tumour. Our patient had a very large mass that infiltrated most of the RVOT region intramurally and caused obstructions. A study conducted by Tiberio et al. in 2011 reported complete resolution of a large left ventricular mass in a seven-year-old patient with TSC treated by everolimus for subependymal giant-cell astrocytomas. Demir et al reported multifocal inoperable cardiac rhabdomyomas that were responsive to everolimus treatment. In our case rhabdomyoma showed rapid regression after everolimus treatment. In conclusion, symptomatic cardiac rhabdomyomas could be managed with everolimus treatment. However further studies needed effectiveness of everolimus and its side effects in children.