

Regression of multiple cardiac rhabdomyomas in an infant after sirolimus therapy

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Abstract

BACKGROUND: Primary cardiac tumors are rare in children, with rhabdomyoma being the most common. It is rarely an isolated finding, and has a strong association with tuberous sclerosis complex (TSC). The clinical manifestations may vary, but most patients are asymptomatic. The complications of cardiac rhabdomyoma (CR) depend on the tumor's location, size, number and degree of invasion of cardiac tissues. Dysrhythmias may occur, the most common of which is supraventricular tachycardia (SVT), especially in those with multiple masses. Although benign and often associated with spontaneous regression, in some circumstances, surgical intervention is recommended when there is sufficient mass effect to adversely affect cardiac output.

OBJECTIVE: The objective of this case report is to observe the efficacy mammalian target of rapamycin (mTOR) inhibitors such as sirolimus to enhance regression of tumors in infants with significant CR.

CLINICAL CASE: This case reports an infant, prenatally diagnosed with multiple cardiac rhabdomyomas by fetal echocardiogram, which was confirmed after delivery. He had multiple episodes of SVT in the first 48 hours of life, and was treated with antiarrhythmics. However, there were several more recurrences of SVT. Because of multiple masses, surgical removal was not an option. Sirolimus was given which successfully reduced the size of the tumors. The patient had no more episodes of SVT. Serial monitoring of sirolimus trough level, blood count, liver function test, lipid profile and bilirubin levels were normal. No other side effects were noted.

CONCLUSION: This report adds to the body of evidence that sirolimus can cause regression of cardiac rhabdomyoma, providing a reasonable alternative to surgical excision. Further studies are needed to standardize treatment protocol using sirolimus for cardiac rhabdomyoma.

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Biography

Dr. Jeremiah Claudine Calacal obtained her medical degree at De La Salle Health Sciences Institute Philippines at the age of

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