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Principles of Surgical Management and Tissue Preservation in Cardiac Rhabdomyoma

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Description

As recently discussed, cardiac rhabdomyomas often present management challenges in young infants that require a thoughtful and methodical approach to treatment [1]. These tumors are benign and are mostly associated with tuberous sclerosis type 1 and type 2. Although relatively uncommon, rhabdomyomas represent the most frequent type of cardiac tumor in children, accounting for 45% of all pediatric cardiac tumors, of which 80-90% are associated with tuberous sclerosis [2]. They are hamartomatous, mesenchymal tumors of striated muscle origin which may result from genetic modifications during the development of striated muscle, although no mechanism has been definitively elucidated [2]. The majority of rhabdomyomas are clinically asymptomatic and regress spontaneously with time, thus requiring no intervention other than monitoring [2,3]. They can occur anywhere within the heart and have a wide spectrum of presentations, ranging from complete lack of symptoms to life-threatening hemodynamic instability from obstruction or arrhythmia [2,3]. This significant variability in presentation and the unpredictable natural history that these tumors take in each individual patient necessitate a methodical yet flexible management approach.

Because of their benign nature and high rate of spontaneous regression, it is often challenging to decide exactly when intervention beyond observation is needed. The two most effective forms of intervention are mTOR inhibitor therapy, such as everolimus or sirolimus and cardiac surgical resection [4,5]. The mTOR pathway is overactive in both TSC1 and TSC2, leading to dysregulation of protein synthesis and apoptosis, ultimately resulting in tumor formation [5]. Inhibitors of this pathway can mitigate this process and induce tumor regression in as high as 91% of patients [6]. While mTOR inhibitors are highly effective, they are not without adverse effects and associated risks, including pulmonary hemorrhage, mucositis, rash, and metabolic derangements [7]. Although highly safe, open cardiac surgery is invasive and does carry inherent risk. As such, surgical resection of cardiac rhabdomyomas is typically reserved for patients exhibiting hemodynamically significant arrhythmias or outflow tract obstruction.

When cardiac surgical resection is necessary, there is no "one size fits all" approach. These rhabdomyoma tumors vary widely in size and location within the heart, and, therefore, require an individualized strategy. There are, however, several important principles to keep in my mind when performing these tumor resections. Tissue preservation is especially important in the pediatric heart in order to maintain current cardiac function and allow for cardiac growth with time. This principle in pediatric patients is clearly highlighted in the successful surgical management of infective endocarditis, where some limited residual valvar regurgitation is considered acceptable if all infected material is adequately debrided and valve replacement can be avoided [8]. When applied to cardiac rhabdomyomas, given that these tumors can often be multiple and in different chambers of the heart, only the tumor focus or foci that are causing the arrhythmia or obstructive symptoms should be resected. If complete resection of a symptomatic focus is not possible, cryoablation of residual tumor is safe and may assist with further regression [1].

Overall, management of cardiac rhabdomyoma tumors can pose a challenging clinical scenario due to their variable presentation and known possibility of spontaneous regression. The decision to pursue surgical intervention should be made with the input of a multidisciplinary team, and surgical resection itself should focus on the tumor site or sites that are directly causing symptoms. Furthermore, as demonstrated recently, tissue preservation should be made a priority whenever possible [1].

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