Management of Complications Caused By a Massive Left Ventricle Tumor in a Neonate

Running Head: Left Ventricle Tumor in a Neonate

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Abstract

We report a case of a neonate born with a giant fibroma occupying the entirety of her left ventricle. Due to the extensive resection, her postoperative course was complicated by severely diminished left ventricular function and complete heart block necessitating extracorporeal support. Ultimately, cardiac resynchronization therapy was employed, after which the infant's ventricular function gradually improved and she was successfully discharged to home.

Cardiac fibroma is the second most common primary cardiac tumor in the pediatric population [1]. Most commonly, cardiac fibromas are single, firm, non-encapsulated tumors involving the left ventricle (LV) free wall or septum [2]. Management will vary depending on age, weight, and tumor size and location. Complete resection, if feasible, is the preferred approach, but partial resection, heart transplantation, or conservative management have also been applied. We describe a neonate with a massive tumor in her left ventricle (LV) who, due to the extensive resection required, suffered unavoidable and unusual complications that necessitated a relatively novel management strategy that ultimately resulted in a successful discharge to home.

The patient is a female neonate prenatally diagnosed with a massive cardiac tumor occupying her LV. She was born at 38 weeks gestation via planned Caesarian section without complication. Echocardiogram postnatally revealed a massive LV tumor and consequent LV outflow obstruction. A prostaglandin E_1 infusion was initiated immediately after birth to maintain ductal patency. Cardiac MRI confirmed a large, well circumscribed mass occupying the LV without evidence of perfusion (Figure 1). Cardiac catheterization demonstrated coronary compression of left anterior descending artery.

At 4 days of life, she was taken to the operating room for surgical resection.(Figure 2 – artist sketch) Upon direct examination, a poorly vascularized mass burrowing through the LV was observed. A thin layer of epicardium and myocardium that was overlying the mass was dissected. Upon reaching the inferior border, a space between the myocardium and tumor was created and the myocardium was dissected away from the tumor mass until much of the tumor was unroofed.(Figure 3) The tumor base was then dissected away from the septum and the tumor was resected. Frozen biopsy revealed a benign fibroma.

Following resection, a ventricular septal defect (VSD) remained and the left anterior descending coronary artery was “free-floating” until it attached to the distal third of the LV. The thinned-out ventricular septum was closed and the inner half of the myocardium was approximated, both of which were accomplished using running sutures. The epicardium and superficial half of the LV myocardium were then sutured in a simple continuous running fashion, taken from the base of the heart down towards the apex. At this point, the LV was reconstructed to the extent possible.

Postoperative transesophageal echocardiogram (TEE) showed marginal LV function and a small 2-mm residual VSD. Also, complete atrioventricular block was present, for which temporary atrial and ventricular pacing wires were placed. Due to these unavoidable sequelae of the extensive resection, she failed to wean from cardiopulmonary bypass. She was placed on extracorporeal membrane oxygenation (ECMO) via central cannulation and transferred to the cardiovascular intensive care unit.

On postoperative day 2, TEE showed slightly improved LV function with good motion of the posterior wall and apex, but with a hypokinetic and thin-walled outlet septum. More concerning, the size of the residual VSD appeared larger than previously noted, with considerable left-to-right shunting. On the following day, a 5.5-mm pulmonary artery (PA) band was placed at bedside. Over the ensuing several days, LV function appeared to be improving but RV function was diminished, deemed to be related to the PA band. Consequently, the PA band was removed and on postoperative day 7, the VSD was closed with a Dacron patch. After this intervention, the patient tolerated weaning from ECMO support over the next 24 hours. Five days later, to address her persistent complete atrioventricular block, a Medtronic pacemaker pulse generator and new epicardial electrodes were surgically placed.

Over the next several weeks, despite the initial encouraging improvement, she continued to require mechanical ventilation and vasoactive support. Serial echocardiograms demonstrated persistent depressed LV function with paradoxical septal motion, due in large part to her paced rhythm and extensive septal patch. Further, the patient's size and the extensive ventricular septal reconstruction raised concerns regarding the effectiveness of a ventricular assist device. After a thorough discussion with family and the multidisciplinary team, the consensus was to continue ventilator and vasoactive support and list her for heart transplantation.

Unfortunately, she remained on mechanical ventilation for many weeks awaiting heart transplantation. As the risks associated with prolonged mechanical ventilation mounted, the team decided to attempt cardiac resynchronization therapy (CRT) as a means of improving her cardiac function and facilitate extubation. On postoperative day 96, she underwent implantation of a biventricular permanent pacemaker. Intraoperatively, her previous left upper abdominal quadrant pacemaker site was reopened and her pacemaker was temporarily explanted. The site that corresponded to the apex of the heart was then exposed and a Medtronic epicardial surface lead (model 4965-25) was sutured to this site. The leads were then secured to a Medtronic biventricular permanent pacemaker (Viva CRT-P, model C6TR01) and the device was implanted into a subcutaneous pocket. DDD pacing was initiated, with a lower rate of 130 beats per minute and upper tracking rate of 180 beats per minute.

Over the next few weeks, cardiac function gradually improved. On postoperative day 124, she was successfully extubated from mechanical ventilation. She was then slowly transitioned from intravenous vasoactive medications to an oral congestive heart failure regimen and, on postoperative day 179, was discharge home. At 16 months of age, she remains stable as an outpatient with her native heart, steadily gaining weight and progressing developmentally.

Comment

Cardiac fibromas are benign tumors which are typically solitary, circumscribed, firm, non-encapsulated and, in many cases, calcified centrally [3]. Cardiac fibromas can intertwine with the ventricular muscle and detrimentally affect cardiac function by replacing functional cardiac mass. A newborn infant with a LV fibroma can present with left-sided heart failure and mimic ductal dependent systemic blood flow lesions, while right-sided tumors can present with severe cyanosis and right heart failure, mimicking ductal dependent pulmonary blood flow lesions [3]. In older pediatric patients, symptoms can present from incidental findings in an otherwise healthy patient to congestive heart failure, life-threatening arrhythmias, and sudden cardiac death.

Though complete resection is preferred, alternative management approaches include use of the Damus-Kaye-Stanzel procedure, a modified Norwood procedure with partial tumor resection, heart transplantation, or temporary single ventricle palliation as a bridge to transplantation [4,5]. In our patient, complete resection of the mass was chosen, considering the near complete obliteration of her LV cavity, obstruction of left LV outflow, and less than ideal long-term morbidity associated with single ventricle palliation. Unfortunately, the postoperative course was complicated by an unavoidable ventricular septal defect and complete atrioventricular block. Consequent severely hypokinetic ventricular septum and less than optimal atrioventricular synchrony resulting from univentricular pacing then lead to chronic refractory heart failure and dependence on invasive mechanical ventilation.

CRT is an established treatment in adults with heart failure, but efficacy and indication in children is less clear [6]. The outcome of CRT in a neonate with poor LV function and ventricular dysynchrony resulting from the resection of a massive LV tumor was likewise unclear. Fortunately, CRT lead to significant improvement in her cardiac function and successful discharge to home.

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Figure Legends

Figure 1. MRI reveals a large, well circumscribed, non-vascularized left ventricular mass measuring 4.2 cm X 4.1 cm X 3.8 cm.

Figure 2: Two layers closure of left ventriculotomy with endocardial alignment of papillary muscles to provide mitral support and competent.

Figure 3: The left ventricle, which consisted of a thin layer of epicardium and myocardium, was dissected and a space between the myocardium and tumor was created, allowing access into the LV cavity.