Surgical Debulking of Large Ventricular Fibromas in Children

Objective:
The optimal surgical resection strategy for ventricular fibromas is uncertain. We describe the outcomes of tumor debulking, ventricular cavity reconstruction, and atrioventricular valve repair if needed.

Methods:
Single-center, retrospective cohort study of patients undergoing ventricular fibroma resection between 2000 and 2023. Over the course of the study period, the approach has evolved from complete resection to a strategy of tumor debulking.

Results
Among 52 patients, median age at surgery was 2.0 years (IQR 9.2 months-4.6 years) and median weight was 11.5 (IQR 8.5-18.9) kilogram. Median tumor volume index was 69 (IQR 49-169) milliliters/m2. Tumor distorted the AV valve/subvalvar apparatus in 30 (58%) patients, caused outflow tract obstruction in 3 (6%) patients, and 5 (10%) patients had moderate AV valve regurgitation. Surgery was indicated for arrhythmia (n=45, 86%), symptoms (n=14, 27%), and/or hemodynamic compromise (n=11, 21%). Tumor was primarily debulked in 34 (65%) patients, including the last 21 patients. The ventricular cavity was entered in 15 (29%) patients. Concomitant AV valvuloplasty was performed in 18 patients (15 mitral and 3 tricuspid), while 3 patients received intraoperative cryoablation. Residual rim of tumor was left around coronary arteries in patients who had signs of extrinsic coronary compression (n=7, 13%), myocardial bridging (n=6, 12%), and/or intratumor coronary artery course (n=4, 8%). Two patients required early reoperation for residual AV valve insufficiency and one patient late reoperation for LVOT obstruction. Median duration of follow-up was 2.5 (IQR 0.8 -6.5) years. There was no mortality or heart transplantation. During the follow-up period, sustained VT occurred in 1 (2%) patient. No other patients had evidence of ventricular arrhythmia on Holter monitoring or EKG. Among 38 patients with post-op v-stim studies, 3 (8%) experienced inducible VT / VF requiring cardioversion / defibrillation. Two of these patients received an ICD. Pre and post-debulking LV EF did not significantly differ (P=.069). No patients had signs of outflow tract obstruction or > moderate AV valve regurgitation on latest imaging.

Conclusions:
Large ventricular fibromas can be resected safely with appropriate surgical planning and an emphasis on debulking rather than complete resection. Most children are cured of life-threatening arrhythmias and maintain LV function.

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