Medical Expertise

"Development of the European Network in Orphan Cardiovascular Diseases"
„Rozszerzenie Europejskiej Sieci Współpracy ds Sierocych Chorób Kardiologicznych"

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DISCUSSION

Cardiac fibromas are benign primary tumors that typically occur during childhood and may be asymptomatic. The most common cardiac tumor in children is rhabdomyoma, followed by fibroma and teratoma. Fibromas typically involve the free wall or interventricular septum, but they can be located in any cardiac chamber as well as in the pulmonary artery. Echocardiography is the most common initial diagnostic test for the presence of a pathological cardiac mass. Magnetic resonance provides optimal assessment of the location, functional characteristics and tissue features. The diagnosis of fibroma is suggested by the presence of a homogenous tumor, clearly delineated from the surrounding myocardium. The mass shows intermediate signal on T2 weighted images and on T1 weighted black blood images without fat suppression, delayed-contrast hyperenhancement after low 1st-pass perfusion and high signal in an tumor's area on late gadolinium-enhanced images. The natural history of cardiac fibromas is difficult to predict. The size of asymptomatic fibromas can remain constant for over a decade, there is no way to predict whether they will start to grow or affect the heart's performance, for example causing arrhythmias. About one quarter of pediatric patients with cardiac tumors have clinically significant arrhythmias, ventricular tachycardia is the most common type. The major concern with asymptomatic tumors is the potential for sudden death, which can occur without the previous detection of dangerous arrhythmias. Indications for surgical treatment poorly defined and should be determined individually for every patient (along with the extent of operation) regarding all clinical data.

EXPERT'S OPINION

Conservative treatment and close follow-up is recommended at present. Echocardiographic stress test and/or cardiopulmonary exercise test should be considered in order to assess the patient's physical capacity and the tumor's effect on cardiac hemodynamic function.

In case of deterioration of the patient's general condition surgical evaluation is necessary.

CONCLUSION

Conservative treatment and close follow-up is recommended at present.

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