Cardiac Myxoma And Wolf-Parkinson-White Syndrome In A Child

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Abstract

A 12-year-old boy is presented with Wolff-Parkinson-White (WPW) syndrome associated with a mass in the left ventricle. Histological examination of the surgically removed tumor revealed the diagnosis of myxoma. A relationship between cardiac myxoma and Wolff-Parkinson-White syndrome seems unlikely.

INTRODUCTION

Cardiac tumors are very rare in children and adults. In a collective series of 731.309 postmortem examinations their prevalence was 0,02% ($_6$). Although the incidence of WPW syndrome is increased in children with congenital heart disease, thus far a relationship between cardiac tumor and WPW syndrome has not been reported. The article describes a child in whom both entities occurred.

CASE REPORT

A 12-year-old boy was referred to our hospital with a history of paroxysmal supraventricular tachycardia (SVT), that was first diagnosed at 4 years of age. The WPW syndrome had been refractory to treatment with verapamil und digoxin. The family history was negative for cardiac disease.

On physical examination the heart rate was 80 per minute and auscultation revealed normal heart sounds with a click after the second heart sound. The blood pressure was 95 / 73 mm Hg. The chest x-ray was normal. The electrocardiogram (ECG) showed a ST-delevation and a negative T-wave in I, II and V3-V8, as well as a delta wave typical of WPW syndrome. In V1 the R-peak was positive. A 24 hour ECG analysis showed a mean heart rate of 159 per minute (minimum 58 per minute, maximum 215 per minute) with 117 single premature ventricular contractations (PVCs), 1500 SVTs and 191 paroxysmal SVTs. Intravenous injection of 50 mg ajmalin resulted in a loss of the delta wave. Transthoracic echocardiography revealed a 1-cm mass in the left ventricle that was in close association with chordae of the septal leaflet of the mitral valve. Because of the potential risk of embolisation of the left ventricular mass an electrophysiologic study (EPS) was not performed.

The patient was operated with the use of cardiopulmonary bypass. The left atrium was opened posterior to the interatrial groove. The mitral valve was normal. Inspection of the left ventricle demonstrated a 1-cm mass of vitreous and gelatinous consistency that originated from the base of a secondary chorda and extended to the anterior mitral leaflet. The tumor including the involved chorda was excised. The mitral valve remained competent and reconstruction was not necessary. Histological investigation of the tumor demonstrated a myxoma.

The postoperative course was uncomplicated. The echocardiographic examination was normal; notably there was absence of mitral regurgitation. On the 16th postoperative day a recurrent WPW syndrome was sucessfully treated with flecainide (3 x 150mg per os daily). The ECG was normal. Three years postoperatively the patient was hospitalized again with recurrent WPW syndrome, which was probably exacerbated by a lack of drug compliance. Treatment with verapamil interrupted the SVT. The patient was discharged with treatment of sotalol p.o. (3 x 80mg daily). Performance of an EPS was declined by the patient.

DISCUSSION

In children myxoma is the most common tumor after rhabdomyoma. The incidence of myxomas increases with age. Whereas in adults approximately 80% of the myxomas are localized in left atrium, in children there is more variation in the localization of the tumor. When present, symptoms consist of dyspnea secondary to mitral valve

obstruction and heart failure. Acute ischemia or cerebral palsy may follow peripheral or cerebral embolisation of the tumor. Pathi and colleagues ([5]) recently reported a left atrial myxoma in a young child and reviewed the literature regarding myxoma in children. Two of 8 children below 6 years of age died because of the aforementioned complications.

An increased incidence of WPW syndrome in children with congenital heart disease is well known. In Ebstein´s anomaly, the associated presence of WPW syndrome is as high as 14% ([1]). Cardiac neoplasms in children may induce ventricular arrythmias ([2,4]). Especially rhabdomyomas are known to be associated with SVT and ventricular tachycardia ([3]). Thus far a relationship between WPW syndrome and myxomas has not been reported. Presence of a symptomatic cardiac tumor is an indication for its surgical removal based on its potential risk of embolisation, myocardial infarction, and sudden death.

Therapy of WPW syndrome follows clinical examination and analysis of ECG and EPS. Emergency drug treament of paroxysmal SVT is by 50mg ajmalin slowly i.v. (alternatively, 70mg propafenon i.v. may be used). Verapamil, as we used in our case, is not indicated for the treatment of SVT because verapamil can transfer an intermittend SVT into atrial fibrillation. In chronic therapy, beta-receptor antagonists and propafenon may be of prophylactic use. Because our patient had been without SVTs for three years postoperatively under drug treatment with flecainide and for the last 6 months with sotalol, we saw no absolute neccessity for performance of an EPS. After consultation, the patient refused an EPS.

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