OPIS PRZYPADKU CASE REPORT

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Mitral valve infective endocarditis complicating hypertrophic obstructive cardiomyopathy – still a current problem

Infekcyjne zapalenie wsierdzia na zastawce mitralnej jako powikłanie w przebiegu kardiomiopatii przerostowej zawężającej – wciąż aktualny problem

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Key words

hypertrophic obstructive cardiomyopathy, infective endocarditis, dental caries, children

Słowa kluczowe

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Summary

Infective endocarditis in the course of hypertrophic cardiomyopathy is rare. Literature reports mainly contain descriptions of clinical cases, and therefore the risk assessment of infective endocarditis in patients with hypertrophic cardiomyopathy, both in the adults and children, is difficult. We present a case of infective endocarditis in a teenage boy with obstructive hypertrophic cardiomyopathy, in which the inflammatory process has damaged mitral valve, what required an urgent cardiac surgery and mitral valve replacement with a mechanical valve.

Streszczenie

Infekcyjne zapalenie wsierdzia w przebiegu kardiomiopatii przerostowej występuje rzadko. Doniesienia piśmiennictwa zawierają głównie opisy przypadków klinicznych, dlatego też ocena stopnia ryzyka rozwoju infekcyjnego zapalenia wsierdzia u chorych z kardiomiopatią przerostową zarówno w populacji osób dorosłych, jak i u dzieci jest trudna. Przedstawiamy przypadek infekcyjnego zapalenia wsierdzia w przebiegu kardiomiopatii przerostowej ze zwężeniem drogi odpływu z lewej komory u 17-letniego chłopca, u którego proces zapalny wsierdzia doprowadził do uszkodzenia zastawki mitralnej, co wymagało pilnego leczenia kardiochirurgicznego i jej wymiany na zastawkę mechaniczną.

INTRODUCTION

Infective endocarditis (IE) in the course of hypertrophic cardiomyopathy (HCM) is a rare complication, however, it is a serious therapeutic problem (1-4). According to some authors, the probability of IE in patients with HCM within 10 years is about 5% (2). The risk of endocarditis is particularly high in patients with obstruction of the left ventricular outflow tract (LVOTO) and the high value of the systolic pressure gradient in the left ventricular outflow tract (1-6). In the echocardiography bacterial vegetations are usually located on the anterior mitral valve leaflet, on the aortic valve, or both. The inflammatory process can damage the heart valves, cause of their significant regurgitation, what requires urgent cardiac surgery (1-3, 5-7).

CASE REPORT

Currently, 17 year old boy, in whom hypertrophic obstructive cardiomyopathy was diagnosed at the age of 5 years. Since the age of 10 year, he was taken care of a pediatric cardiologist in the district center. From November 2010 (at the age of 15) the patient reported feeling worse, increasing fatigability. In December 2010, he was hospitalized in pediatric cardiology referral center, where the elevated values of inflammatory markers, thickening of the mitral valve leaflets and third degree mitral regurgitation were found. In the chest X-ray pneumonia was diagnosed, and intravenous antibiotic therapy (cefuroxime) has been applied. In January 2011 the boy was urgently admitted to the paediatric cardiology departament with fever 38 degrees, with marked dyspnoea at rest. Tachycardia 137/min,

systolic murmur 3/6 scale of Levin along the left sternal border and at the apex of the heart and liver enlargement were present. In the chest X-ray bilateral parenchymal density, features of chronic congestion in the pulmonary circulation, enlarged cardiac silhouette (cardio-thoracic ratio 0.56) were found. The values of the inflammatory markers were increased, while the blood cultures obtained were negative. The antibiotics (clarithromycin and cefuroxime) have been taken for 11 days to give a slight reduction of the inflammatory markers and the alleviation of fever. In echocardiography features of hypertrophic obstructive cardiomyopathy were present: the thickness of the interventricular septum was 17.9 mm (normal values indexed to the body surface area 4.9-8.5 mm), the thickness of the left ventricle posterior wall was 11 mm (normal values corrected to the body surface area 4.3-9.5 mm), obstruction of the left ventricular outflow tract with the maximum systolic pressure gradient 100 mmHg, systolic motion of the anterior mitral leaflet and its adherence to the hypertrophied interventricular septum and the mitral valve regurgitation third degree also were found (fig. 1, 2).

Moreover, the presence of three bacterial vegetation on the anterior mitral valve leaflet were visualised (one with a diameter of 4.5 mm moving into the left atrium, the second with a diameter of 6.2 mm at the end of the leaflet, the third of the diameter of 8mm in the middle of the leaflet) (fig. 3).

Antibiotic therapy was changed to ampicillin with sulbactam, which was continued for a further 14 days. Due to extensive dental caries the patient was treated urgently. The patient's general condition gradually was getting better, but C-reactive protein still remained elevated and obtained blood cultures were still negative. Subsequent echocardiography examination showed perforation of the anterior mitral valve leaflet and massive mitral regurgitation. The patient was qualified for urgent surgery and antibiotic therapy was converted to vancomycin and netromycin. Surgery was performed in March 2011. The mitral valve was replaced on mechanical valve St. Jude Medical 27MJ-501 and septal myectomy was performed. Of the intraoperative specimen of cardiac muscle and valve bacterial growth was not obtained. The postoperative course was uncomplicated and anticoagulant therapy with acenocoumarol was enabled in a dose-dependent INR values. Postoperative echocardiography showed normal disks' movement of mechanical valve in the mitral position, without perivalvular leak. There was no outflow tract obstruction, the maximum systolic pressure gradient was 4 mmHg (fig. 4, 5).

The patient was discharged in the 58th day of hospitalization. During follow-up, one year after operation, the boy didn't report disturbing symptoms and didn't use to get tired during daily activities. On physical examination there were no symptoms of heart failure, he had regular heart rate 80 bpm, without a murmur over the heart and audible click of mechanical valve

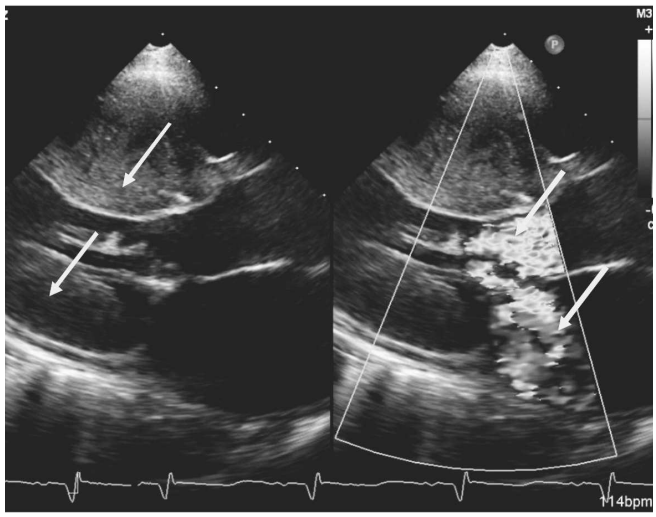

Fig. 1. Septal and left ventricular posterior wall hypertrophy, obstruction of the left ventricular outflow tract, a significant mitral regurgitation (arrows). Echocardiographic parasternal long axis view.

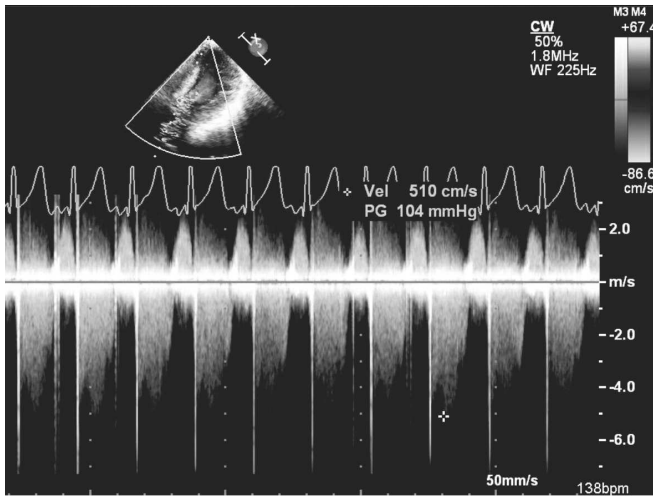

Fig. 2. Hypertrophic obstructive cardiomyopathy. Narrowing the left ventricular outflow tract with maximum systolic gradient of 104 mmHg (maximum velocity in the LVOT 510 cm/sec). Continuous wave Doppler echocardiography in the apical 5-chamber projection.

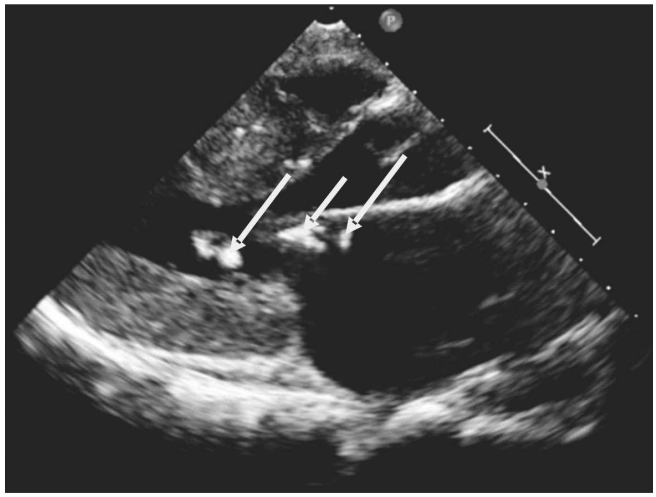

Fig. 3. Hypertrophic obstructive cardiomyopathy. Bacterial vegetations on the anterior leaflet of the mitral valve (arrow). Echocardiographic parasternal long axis view.

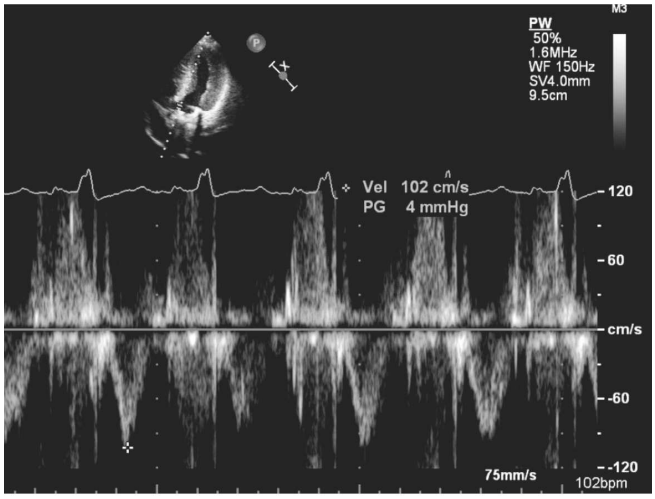

Fig. 4. Hypertrophic obustructive cardiomyopathy after myectomy without obstruction in the left ventricular outflow tract (maximum systolic gradient of 4 mmHg and maximum velocity in the LVOT 102 cm/sec). Continuous wave Doppler echocardiography in the apical 5-chamber projection.

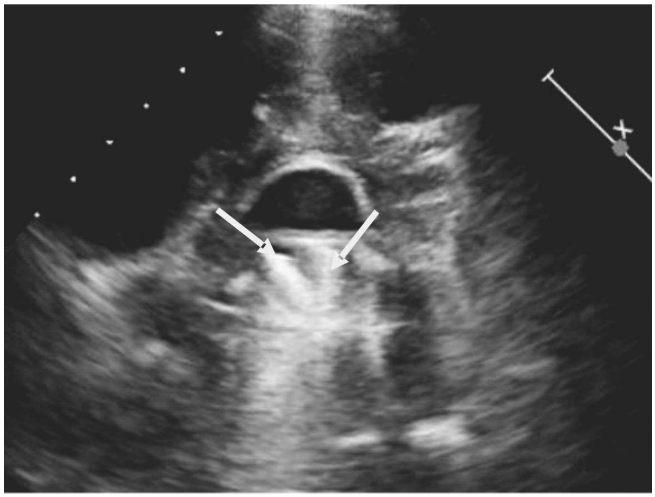

Fig. 5. Hypertrophic obstructive cardiomyopathy after replacing the mitral valve into mechanical. Visible disks a mechanical valve in the mitral position (arrows). Echocardiographic parasternal short axis view at the level of the mitral valve.

was present. Echocardiography showed normal disks' movement of mechanical valve, without perivalvular leak, there was no LVOTO. The patient remains under permanent cardiac care.

DISCUSSION

IE in the course of hypertrophic cardiomyopathy is rare, however, obstruction of the LVOT in patients with HCM is a predisposing factor for the development of IE (1-6). The most common symptoms of endocarditis in patients with HCM are fever, elevated inflammatory markers in laboratory tests and heart failure (shortness of breath, exertional dyspnoea, features of congestion in the pulmonary circulation), systolic cardiac murmur, swelling, hepatomegaly which were present in our patient. In the literature, the clinical cases were described in which the etiological factors of IE (streptococcus sanguisgordonii, staphylococcus aureus, streptococcus viridans, enterococcus) in patients with HCM were identified. Some authors emphasize the role of antibiotic prophylaxis during medical procedures that predispose to IE in patients with HCM (1, 2, 4, 5, 7). Antibiotic prophylaxis of IE in patients with HCM, particularly in the case of obstruction of the LVOT remains a matter of many debates and controversy. According to the experts of the European Society of Cardiology (8) the use of antibiotic prophylaxis in patients with HCM before procedures associated with high risk for IE (dental treatment causing damage to apical area of the tooth) is not recommended, however, some authors suggest the need for its use, because there are no randomized studies in this group of patients, and the risk of complications of IE in patients with HCM is quite high (1, 9). Because IE in patients with HCM is rare, described only a single clinical cases, and therefore it is difficult to determine the true incidence of IE in patients with HCM (1, 9). Perhaps in the future recommendations for endocarditis prevention concerning patients with HCM will be published, but it is necessary to describe some new clinical cases of IE in these patients (1, 9). In our patient, the etiological factor of IE was not detected, the results repeatedly collected blood cultures were negative, which probably was associated with the occurrence of IE with negative blood cultures what is described in the literature in 5% to 31% of all cases of endocarditis (8). The intravenous antibiotics therapy (clarithromycin, cefuroxime, ampicillin + sulbactam) was applied for 25 days with improvement of the patient's clinical condition and decreasing of the value of the inflammatory markers but serious complications of the IE occurred and immediate surgery was needed. An important risk factor for the IE in our patient was undoubtedly an extensive dental caries. Actually more attention is paid not only to the significant impact of dental caries on the development of IE, but also the negative impact of bacteremia occurring during daily activities (i.e. chewing, brushing teeth, using dental floss and toothpicks) in patients with foci of infection in the oral cavity. It is emphasized that the probability of bacteremia increases with rising number of potentially pathogenic bacteria in the oral cavity (10). Therefore, the main emphasis in dental prophylaxis of IE should be placed on maintaining an optimal hygiene and oral cavity health in patients with cardiovascular pathology. In patients with HCM an important risk factor for IE is an obstruction of the LVOT with high systolic pressure gradient and enlargement of the left atrium (1, 3, 5, 6). Bacterial vegetations are usually located on the anterior mitral valve leaflet, but it also concerns the aortic valve, or both causing significant regurgitation and valve perforation. In the literature, clinical cases are described where after the antibiotic therapy bacterial vegetations regression was achieved without damaging the valve (1-3, 5), but in most cases cardiac surgery is necessary, as in our patient (3, 5-7). In described patient, during

surgery the mitral valve was replaced for mechanical valve (St. Jude Medical 27MJ-501) and septal myectomy was efficiently performed. Control cardiological examinations performed one year after surgery showed good hemodynamic effect of the operation and the patient feels good, does not report disturbing cardiovascular symptoms.

CONCLUSIONS

Children with hypertrophic cardiomyopathy and obstruction in the left ventricular outflow tract are a group of patients with an increased risk of endocarditis and development of its complications. In patients with obstructive hypertrophic cardiomyopathy and infective endocarditis, there are indications for early cardiac surgery.

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