EARLY DEGENERATED BIOPROSTHETIC MITRAL VALVE

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ABSTRACT

Mitral valve prolapse (MVP) is fairly common in children, often being randomly found in asymptomatic, as well as in symptomatic patients with conjunctive tissue diseases (CTD). The authors report the case of an 11-year-old girl, diagnosed at the age of 6 with MVP and severe mitral regurgitation (MR). She had surgery at the age of seven, when the mitral valve replacement with a biological valve prosthesis was performed. Four years later, the girl was admitted for sudden onset of dyspnoea with orthopnoea, haemoptysis and generalized cyanosis. The diagnosis of acute pulmonary oedema and degenerated biological valvular prosthesis correlated with severe mitral stenosis was evoked by means of clinical and echocardiographic examinations. The evolution was favourable after the replacement of the bioprosthesis with a mechanical prosthesis.

Keywords: bioprosthesis, mitral valve, child

INTRODUCTION

The mitral valve ring is nonplanar and saddle-shaped. Usually, the valvular leaflets are shoved towards each other by the papillary muscles contraction and by the chordae tension during ventricular systolic, which leads to the coaptation of the edges of the valve. The valvular prolapse occurs when one valve slides under this coaptation area. The idiopathic mitral valve prolapse (MVP) can occur congenitally, but it is often diagnosed later during adolescence or adulthood. Some of the complications may include arrhythmias, heart failure due to severe mitral regurgitation (MR) or, sometimes, thromboembolic events. Familial cases are autosomally dominant with variable penetrance and expression. Treatment is directed by the presence or the absence of complications.

Thus, the asymptomatic cases presenting minimal MR do not require medical treatment, but those with ventricular dysfunction and severe MR require surgical treatment.

CASE

We present the case of a female child, aged 11 years old, diagnosed at age of 6 with mitral valve prolapse and severe mitral regurgitation. When she was 7, she underwent the replacement of the mitral valve with tissue valve prosthesis which was performed at the Institute for Cardiovascular Diseases and Transplantation from Târgu Mureş. The procedure was followed by chronic treatment with anticoagulants (Sintrom, Aspenter).

Four years after inserting the prosthetic valve, the girl presented sudden dyspnea, orthopnea, hemoptysis and generalized cyanosis. She was admitted to the pediatric ward of a county hospital and she was treated with antibiotics (Cefort), mucolytics and cortisone preparations for 7 days. Symptoms initially improved but eventually got worse, so that pulmonary embolism was suspected and the patient was transferred to Acute Care Department of 1-st Pediatrics Clinic from Iaşi.

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On admission, physical examination revealed severe general condition, suffering face, pale skin and mucous membranes, postoperative scar on the mid-sternal line, significant dyspnea and orthopnea, respiratory frequency = 56/min, productive cough, hypoxemia – SaO_2 = 85-89% in the atmospheric air. The pulmonary auscultation revealed bilateral crepitation rales, rhythmic heart sounds, tachycardia 130/min, systolic murmur grade III/6 at the apex with posterior radiation, normal liver.

The biological exam highlighted neutrophilic leukocytosis, mild hepatocytolisis, elevated creatinphosphokinase level, elongated prothrombin time and decreased prothrombin activity – due to the anticoagulant treatment. Chest X-ray showed adjoining macro-opacities occupying almost entirely the both pulmonary areas, bulging cord with right lower arch, normal vascular pedicle. ECG showed sinus tachycardia 140/minute, QRS axis of + 90°, PQ = 0.12 sec; ST depression (4 mm) in the PRD, DIII, AVF, V3-V5; biatrial overload; QT = 0.28 sec (normal).

Echocardiography performed by emergency (see Fig. 1) showed hyperechoeic prosthesis in dysfunctional mitral position, limited opening, creating severe stenosis and grade III mitral regurgitation, significant dilatation of the pulmonary artery and right heart, grade III tricuspid regurgitation.

Positive diagnose was established: acute pulmonary oedema, severe mitral stenosis by degenerated bioprosthesis, pulmonary hypertension. The patient was urgently transferred to the Cardiovascular Surgery Clinic of Iasi, where the replacement of bioprosthesis with a mechanical prosthesis was immediately performed. The postoperative evolution was favourable, overall condition improved, yet a complication occured: an externally popliteal sciatic nerve paresis,which was cured through physiotherapy.

DISCUSSIONS

Classic MVP is defined as the displacement of one or both leaflets during systole, exceeding the mitral valve annular plane with 2 mm or more under the mitral ring, having as result leaflet thickening. Non-classic prolapse refers to leaflet displacement without valve thickening. The etiology of MVP is not clear and is probably multifactorial. It can result from excessive leaflet tissue (redundancy), myxomatous proliferation of the spongious layer of the valves, and elongation of the chordal apparatus. These alterations are met in the case of individuals with a wide range of congenital heart malformations as well as in acquired heart disease including collagen vascular disease (Marfan syndrome, Loeys-Dietz syndrome), ischemic heart disease, hypertrophic cardiomyopathy, and pectus excavatum, as well as in the case of thin patients (1). Isolated prolapse can be sporadic or familial, with autosomal dominant and x-linked transmission.

Prevalence rates are 1-2% in children and 5-15% in adolescents and young adults, twice more frequent in females than in males (2,3).

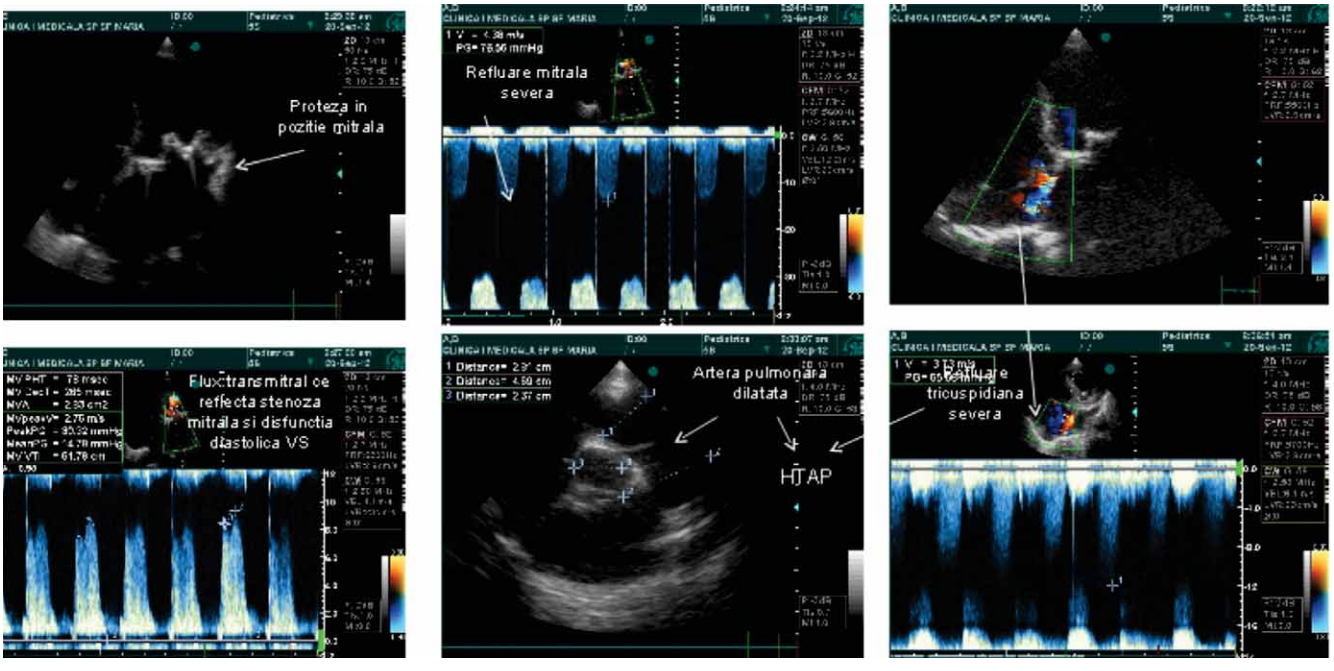

FIGURE 1

MVP is usually diagnosed on the clinical basis of a mid-systolic click of the mitral valve and a late systolic murmur of mitral regurgitation.

Most patients are asymptomatic, and MVP is an incidental auscultatory finding, when a short systolic murmur is discovered in the mitral area. In time, various symptoms can occur: palpitations, fatigability and exertion dyspnea, chest pain, neuropsychiatric symptoms (panic attacks, nervousness, presyncope and syncope). Some patients present skeletal abnormalities that do not fit into any of the recognized connective tissue disorders (height-to-weight ratio greater than normal, scoliosis, arachnodactily, pectus excavatum or pectus carinatum) (4).

Arrhythmias described at rest or during exercise include premature atrial (23.6%) or ventricular (27.3%) contractions, supraventricular tachycardia, and conduction abnormalities (5). Some studies report a prevalence of ventricular arrhythmias over 30% (6). Also, in 8-16% of the patients with refractory ventricular tachycardia, the only cardiac anomaly was MVP. There is a connection between MVP and sudden death (7,8). If the incidence of sudden death in MVP is not well established, the studies suggest that the risk is 5 or 10 times greater in the cases in which there is also severe mitral regurgitation. It is considered that the relation between MVP and sudden death is due to ventricular arrhythmias (9). In children, the mortality rate is very low.

The appearance of mitral regurgitation (MR) and the progression from mild or moderate MR to severe MR are important determinants to morbidity. A study made by Deng showed that the prevalence of MR increased from 29% of patients to 43% of patients during the four years of observation (10). Other possible complications include congestive cardiac failure, rupture of chordae tendineae, infective endocarditis (in 0.1-0.3 cases per 100 patient years), thromboembolic phenomena including cerebrovascular accidents, and sudden death. Cardiac arrhythmias such as ventricular tachycardia and ventricular fibrillation are more common in MVP (2).

In childhood, MVP is not progressive, and the majority of patients do not require specific therapy, but only periodic observation. Asymptomatic patients with isolated mitral systolic clicks need only counseling and reassurance. They are recommended to avoid excessive use of caffeine, cigarettes, alcohol, and prescription or over-the-counter drugs that contain stimulants such as epinephrine or ephedrine to minimize catecholamine and cyclic adenosine monophosphate (AMP) stimulation. The most recent recommendations have limited the indications of subacute bacterial endocarditis antibiotic prophylaxis only to patients with the highest risk, undergoing the highest risk procedures. In this category one can include: patients with a prosthetic valve or a prosthetic material used for cardiac valve repair, patients with previous endocarditis, patients with cyanotic congenital heart disease without surgical repair or with residual defects, palliative shunts or conduits, patients with congenital heart disease with complete repair with prosthetic material whether placed by surgery or by percutaneous technique, up to 6 months after the procedure and patients with a residual defect at the site of implantation of a prosthetic material or device by cardiac surgery or percutaneous technique. The antibiotic prophylaxis is no longer recommended for other valvular diseases or congenital heart diseases (11)

The surgical treatment (plastic surgery of mitral valve or replacement with prosthesis) has precise indications established by the American and European guidelines: patients with moderate-severe MR, symptomatic patients with acute severe MR, symptomatic patients with severe chronic MR with cardiac failure New York Heart Association (NYHA) functional class II-IV symptoms , asymptomatic patients with chronic severe MR and mild-to-moderate LV dysfunction or with preserved LV function, new onset atrial fibrillation, or pulmonary artery hypertension. In the case of children, surgery is indicated only when the medical treatment or the mitral valve repair have failed. In the case of patients younger than one year, the mitral valve replacement must be delayed as much as possible because it is associated with substantially increased risk of morbidity and mortality. The mitral valve replacement has as a result the highest mortality from all the other pediatric valve procedures (10-30%) and it has the worst long-term prognosis – between 5 and10 years the survival is between 50-80% (13). Bioprosthetic xenografts have been found to have limited durability at the mitral position in the case of children, with a mortality between 79%, 75% and 74% at 1, 5 and 10 years respectively. This suggests the idea that the most part of the cases of mortality occur in the immediate postoperative period. (14)

Comparing with mechanic prosthetic valves, bioprosthetic valves have the distinct advantage of not needing lifelong anticoagulation, but only three months after the surgery. The tissue of origin is generally porcine with the exception of a bovine pericardial valve manufactured from fixed bovine

pericardium. Xenografted valves calcify when placed into the human circulation, ultimately leading to their failure. This calcification process appears to occur more rapidly in the case of younger patients. Structural deterioration of bioprosthetic valves is an inevitable consequence of their utilization in the humans. This incidence is nonlinear with deterioration and subsequent "failure" increasing at a greater rate after a certain period of time. In the case of children this time frame is often very short (14). Actual data show that the medium life of a bioprosthesis in the case of adult in15 years (15).

The given case follows the general trend described in literature, that of premature deterioration of the mitral bioprosthesis, in the first four years. The replacement by a mechanical valve saved the life of the patient, with a slow favorable evolution after surgery. Anticoagulant treatment was recommended, with regular INR follow-up, which must be maintained at an optimal value of 3.5-4.5. Another recommendation was the endocarditis antibiotic prophylaxis. A peculiarity of the case is the external sciatic nerve paresis as a postoperative complication, alleviated through physiotherapy.

CONCLUSIONS

The cases with MVP with severe MR, like the presented case, require surgery for mitral repair or replacement. The bioprosthesis offer the advantage of a short course of anticoagulant treatment, but has the major disadvantage of deterioration, as early as the patient is younger.

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