

Case report on aortic valve replacement in adult woman with systemic mastocytosis (RCD code: VIII)

Monika Różewicz-Juraszek, Tomasz Hryniewiecki, Katarzyna Faber, Elżbieta Abramczuk, Ewa Orłowska-Baranowska, Piotr Szymański*

1st Department of Valvular Heart Diseases, The Cardinal Stefan Wyszyński Institute of Cardiology, Warsaw, Poland

Abstract

We present a case of a 50 year-old woman who was referred to our department with severe symptomatic aortic stenosis and systemic mastocytosis. Conventional mechanical aortic valve replacement was performed successfully with the use of perioperative protocol based on dexamethasone, cetirizine and ranitidine. The only postoperative event was an onset of atrial fibrillation which was treated with amiodarone infusion. 8 days after surgery the patient was discharged home. JRC D 2017; 3 (2): 54–55

Key words: rare cardiovascular disease, aortic stenosis, perioperative management, mast cells, antihistamine agents, steroids

Introduction

Mastocytosis is a rare myeloproliferative disease, caused by pathological growth and accumulation of mast cells which can occur in a cutaneous or systemic form [1]. In case of cutaneous form, mast cells gather only in skin tissue causing typical skin lesions known as urticaria pigmentosa [2].

Symptoms of the disease are caused by tissue infiltration by mast cells or by releasing the substances produced by mast cells, mainly histamine. This causes acute or chronic symptoms, that vary in their severity – from mild to even life threatening. In course of mastocytosis patient may suffer from erythema, itch, nausea, diarrhea, dyspepsia, muscle and bone pain, osteoporosis, hypotonia, tachycardia, headaches, fatigue, depression, anxiety or anaphylaxis [3].

Degranulation of mast cells can be caused by many factors. Moreover, patients' reactions are very heterogenic what requires individual evaluation of risk factors leading to histamine release and being a potential trigger for an anaphylactic reaction. The most common anaphylaxis triggers include: drugs (acetylsalicylic acid, amphotericin B, quinine), sedatives and analgesics (lignocaine, tetracaine, procaine, morphine, codeine, etomidate, thiopental, succinylcholine, enflurane, isoflurane), iodine-based contrast media, preservative agents (parabens), alcohol, physical effort, high or low temperatures, ultraviolet radiation, stress, emotions, infections and allergens [2].

Medical procedures including sedation, analgesia or multi-drug pharmacotherapy may be a potential risk for patients with mastocytosis [4]. Data on perioperative management of patients with mastocytosis undergoing cardiac surgery involving cardiopulmonary bypass is limited and comes mainly from cohort studies.

Case report

We present a case of a 50 year-old woman with severe symptomatic aortic stenosis (AS) and systemic mastocytosis, who was referred for cardiologic evaluation and management. Other medical conditions included hypertension, stable coronary artery disease, impaired glucose tolerance, duodenal ulcers, gastro-eosophagal reflux disease and obesity. The patient had a history of relatively mild course of systemic mastocytosis with predominantly non-specific gastro-intestinal symptoms, constitutional complaints and a single episode of anaphylactoid reaction of unknown origin.

Moderate AS had been diagnosed two years before the admission and it was treated conservatively. After 2 years, the patient was reevaluated due to increasing exercise intolerance and repeating episodes of angina. Transthoracic echocardiography revealed severe stenotic bicuspid aortic valve with mean transvalvular pressure gradient of 55,2 mm Hg and valve area of 0,85 cm². Left ventricular ejection fraction and left ventricular systolic volume were within normal limits.

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* Corresponding author: Department of Valvular Heart Diseases, The Cardinal Stefan Wyszyński Institute of Cardiology, ul. Alpejska 42, 04-628 Warsaw, Poland; tel: 0048 22 343 46 10, fax: 0048 22 343 45 48; e-mail: pszymanski@ikard.pl

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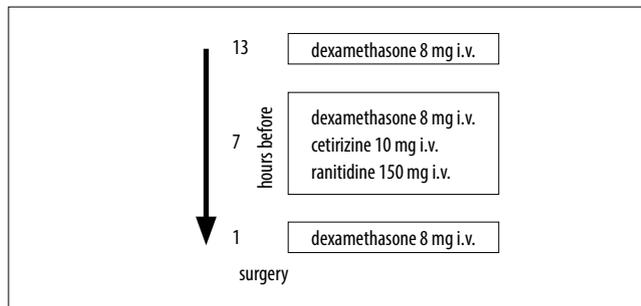


Figure 1. Perioperative medication scheme

Coronary angiography disclosed normal coronary anatomy without stenotic lesions. Due to potential risk of anaphylaxis, no angiotensin converting enzyme inhibitors and beta blockers were introduced.

The patient was referred for surgery. A perioperative protocol based on intravenous infusion of 8 mg of dexamethasone 13, 7 and 1 hour before the surgery with oral administration of 10 mg of cetirizine and 150 mg of ranitidine 7 hours before the surgery was used in order to prevent potential anaphylactic complications (Figure 1).

Conventional aortic valve replacement (AVR) with mechanical St Jude® 21 mm prosthesis implantation was successfully performed via median sternotomy and cardiopulmonary bypass. Propofol and fentanyl were used for sedoanalgesia. The patient was extubated 12 hours after surgery. The only postoperative event was an onset of atrial fibrillation, which was treated with amiodarone infusion. Postoperative echocardiographic assessment revealed normal function of the valve prosthesis and absence of pericardial effusion. The patient was discharged home 8 days after surgery. Since that time, she has remained asymptomatic.

Discussion

Patients with systemic mastocytosis are at higher risk of adverse events during surgery. Many stimuli like emotional or physical stress, drugs can trigger mast cell degranulation. Cardiopulmonary bypass is known to induce systemic inflammatory response [5]. General guidelines for safe surgery in patients with mastocytosis recommend avoiding potentially harmful agents and blocking the cascade of factors released by mast cells. The use of H1 and H2 receptor blockers and steroids perioperatively is advised [4].

This is a case report of a complication-free cardio-surgical procedure with cardiopulmonary bypass in a patient with systemic mastocytosis. There is very limited evidence on this filed available in the literature. Four similar case reports have been published, which included cases of an AVR, valve sparing root and ascending aorta repair, heart transplantation and atrial septal defect closure [6–9]. Wanamaker et al. reported event-free AVR in a 72 year-old female with mastocytosis. They used a combination of H1 and H2 receptor blockers and steroids in the perioperative period [6]. Duggal et al. presented a case report on successful management of diffuse allergic reaction in a patient with systemic mastocytosis undergoing ascending aortic aneurysm repair. Hydrocortisone, diphenhydramine, ranitidine and methylene blue were administered to stabilize the patient [7]. Moro et al. performed heart transplantation in a 56-years-old woman with dilated cardiomyopathy and mastocytosis with no perioperative allergic complications [8]. Also Damo-

dar et al. reported a case of a successful surgical closure of an atrial septal defect in a pediatric patient with systemic mastocytosis [9].

Patient management

Our management included dexamethasone as a steroid, ranitidine as a H1 receptor blocker and cetirizine as a H2 receptor blocker. This strategy is aimed at blocking the cascade of factors released by the mast cell degranulation [4].

No beta blockers or angiotensin receptor inhibitors were used due to reported potential risk of severe anaphylaxis [10–14]. These agents may cause mast cell degranulation. The former can release cellular mediators provoking anaphylaxis through cyclooxygenase mechanism [15], while the latter induce repression of inactivation of bradykinins [12].

Our patient had mild form of systemic mastocytosis. We successfully used a perioperative protocol based on previous reports. It resulted in a complication-free course. It is unknown however, whether this protocol would be suitable for patients with severe forms of systemic mastocytosis.

Conclusions

The use of H1 and H2 receptor blockers and steroids in perioperative protocol in a patient with systemic mastocytosis undergoing surgical AVR showed to be safe and effective in preventing anaphylactic reaction.

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