Case Report

Aortic valve replacement in a patient with systemic lupus erythematosus

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Abstract

Valvular heart disease in systemic lupus erythematosus (SLE) is associated with substantial morbidity and mortality. Current therapy includes symptomatic measures and valve replacement. SLE can present major challenges because of accrued organ damage, coagulation defects and complex management regimes. The peri-operative goals are to maintain strict asepsis, avoid use of nephrotoxic drugs and thereby renal insult, and to promote early ambulation post-operatively.

Key words: Anesthesia, aortic valve replacement, systemic lupus erythematosus

Introduction

Systemic lupus erythematosus (SLE) is a multi-systemic autoimmune disorder characterized by a broad range of manifestations alongwith presence of antibodies in the blood directed against one or more components of cell nuclei. Patients with SLE produce abnormal antibodies in their blood that target tissues within their own body rather than foreign infectious agents. The prevalence of SLE ranges from 7.4 to 159.4 per 100,000 of population with female pre-dominance (9:1), and peak age of onset between 15 years and 40 years.[1]

The valvular heart disease in SLE is associated with substantial morbidity and mortality. Current therapy includes symptomatic measures and valve replacement. SLE can present major challenges because of accrued organ damage, coagulation defects and complex management regimes. We report the case of a patient with SLE and glomerulonephritis with severe aortic regurgitation scheduled for aortic valve replacement (AVR).

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Case Report

A 43-year-old hypertensive woman, diagnosed with SLE in 2005, was admitted with complaints of increasing shortness of breath (New York Heart Association (NYHA) class III/IV) and exertional pain in left side of chest. Past history included cerebrovascular accident (right hemiplegia and right facial palsy) with complete recovery. She also had history of renal failure requiring dialysis. Renal biopsy done in 2007 was suggestive of lupus nephritis class V (membranous nephritis). Previous cardiac history included an evidence of endocarditis in 2007. Physical examination was unremarkable except for the presence of malar rash, and obesity (body mass index 38.87 kg/m^2). Pre-operative hemoglobin, platelet count and liver function tests were normal. Activated partial thromboplastin time (aPTT) was prolonged (51.6 s, normal 28-43 s) while fibrin D-dimer level (0.33 μg/ml, normal <0.4 μg/ml) and renal parameters (blood urea nitrogen and serum creatinine) were normal. Routine and microscopic evaluation of urine revealed protein 3+. Urinary protein was high (90 mg.dl^{-1}, normal <15 mg.dl^{-1}) and urine protein/creatinine ratio was also high (1.74, normal 0-0.2). Current medications included oral prednisolone 10 mg once daily, and mycophenolate mofetil 1 g twice daily.

Transthoracic echocardiography showed calcific, non-coapting aortic valve leaflets with severe aortic regurgitation, no aortic stenosis, mild mitral regurgitation, dilated cardiac chambers, mild global hypokinesia of left ventricle with an ejection fraction of 45%, with no intracardiac clot, vegetation, or pericardial effusion. Angiography revealed normal coronary and renal arteries.

Patient was scheduled for AVR and an informed consent was obtained. Though no airway abnormality was detected, however a "difficult intubation kit" consisting of small size endotracheal tubes, McCoy laryngoscope, intubating laryngeal mask airway, and bronchoscope, was kept ready. She was pre-medicated with lorazepam 2 mg orally and pantoprazole 40 mg orally. Standard cardiac monitoring was instituted including invasive blood pressure, cardiac output (CO) by thermodilution method, and trans-esophageal echocardiography (TEE). Cardiac index (CI) and systemic vascular resistance (SVR) and pulmonary vascular resistance (PVR) were calculated using standard formulae. Her pre-operative hemodynamic parameters were: Heart rate 92 beats/min, blood pressure 176/54 (94) mmHg, pulmonary artery pressure 42/18 (25) mmHg, CO = 4.4 L/min, CI = 2.3 L/min/m^2, SVR 1648 dynes.sec/cm^5, and PVR 196 dynes.sec/cm^5.

Cefuroxime 1500 mg and linezolid 600 mg were administered intravenous (IV) as antibiotic prophylaxis. Anesthesia was induced with midazolam 2 mg, fentanyl 3 mcg/kg and thiopentone 150 mg IV. Endotracheal intubation was facilitated with 0.5 mg/kg of atracurium. Anesthesia was maintained with isoflurane in oxygen and air, intermittent doses of fentanyl, and atracurium infusion (0.5 mg/kg/min). After heparinization (4 mg/kg) and achieving an activated clotting time (ACT) of 821 s by Celite method (Hemochron 401, International Technidyne Corporation, Edison, New Jersey, USA), cardiopulmonary bypass (CPB) was instituted by aorto-caval cannulation. During CPB, the hematocrit was maintained between 18% and 25%, perfusion flow between 2.4 L/min/m^2 and 2.8 L/min/m^2, mean arterial pressure between 50 mmHg and 70 mmHg, systemic temperature between 28°C and 30°C and ACT between 700 s and 800 s. Methyl prednisolone 1 g was administered during CPB. A 19 mm mechanical prosthetic valve (St Jude Medical Inc. Minnesota, USA) was implanted at aortic position. The CPB duration was 93 min and aortic cross clamp time was 66 min. Patient was weaned off CPB with inotropic support of epinephrine 0.1 mcg/kg/min and dobutamine 5 mcg/kg/min. Post CPB TEE showed normal functioning of prosthetic aortic valve with no paravalvular leak. Heparin anti-coagulation was reversed with half dose of protamine (total dose given 220 mg, post-protamine ACT 168 s). Patient was shifted to the intensive-care unit in hemodynamically stable condition and mechanically ventilated for 12 h. IV heparin infusion was started on first post-operative day to keep twice the control aPTT. After removal of chest drains, patient was ambulated on post-operative day 1. Heparin infusion was overlapped with acenocoumarol on third post-operative day to achieve an international normalized ratio (INR) of 2.5-3. Patient was discharged from the hospital on tenth post-operative day. Pathological examination of the excised aortic valve showed fibrosis, focal calcium deposits, and infiltration with inflammatory cells. A follow-up at 3 months showed normal function of the valve, and no signs of sepsis, thrombosis or uremia.

Discussion

Patients with SLE can have a wide variety of symptoms and organ involvement of multiple systems. 11 criteria were established by the American Rheumatism Association to establish the diagnosis of SLE.[2] When a person has four or more of these criteria, the diagnosis of SLE is strongly suspected. Our patient had malar rash, photosensitivity, arthritis, proteinuria, and positive antiphospholipid and antinuclear antibodies.

Libman and Sacks[3] first described valvular heart disease caused by SLE in 1924. All SLE patients will have involvement of the heart at some stage during their illness. The prevalence of significant valve dysfunction in patients with SLE is variable. Left ventricular dysfunction in patients with SLE could be secondary to valvular heart disease, myocardial ischemia/infarction, or auto-immunological involvement of the myocardium.

Pre-operative consultation with the patient's rheumatologist will provide accurate information on disease flares, organ damage, and drug history. It would be prudent to delay non-urgent surgery until after recovery from disease flares. Coronary angiography is mandatory in patients with SLE, irrespective of age, with emphasis not only to overt lesions in the major coronary arteries but also to a possible rarefaction of the end-branches as sign of recurrent microemboli.[4] Pre-operative assessment should particularly address the need for peri-operative continuation of immunosuppressants and steroid replacement. In view of increased risk of infection and sepsis, strict aseptic precautions and use of broad-spectrum antibiotic are warranted in the perioperative period. High rate of infection in SLE patients appears related to intrinsic susceptibility and treatment-related immunosuppression, and is an important risk factor for morbidity and mortality in patients with SLE.[5] Ascertaining the presence of anti-phospholipid antibodies is warranted to identify whether there is an increased risk of thrombosis.

Choice of anesthetic technique should account for the potential drug interactions with immunosuppresants, an unexpected difficult airway with subglottic stenosis or laryngeal edema, unrecognized myocardial ischemia, and thrombotic risk. Laryngeal involvement occurs in 0.3-30% patients, and includes mild inflammation,

vocal cord paralysis, subglottic stenosis and laryngeal edema with acute obstruction.[6] All possible precautions should be taken to secure airway in such a situation. The hemodynamic goals are mild tachycardia, a positive inotropic state and a controlled reduction in SVR. The use of pulmonary artery catheter allows determination of basal filling pressures and CO. Concurrent preload augmentation may be crucial to optimize CO when afterload is pharmacologically manipulated. Rhythm disorders such as bradycardia, ventricular fibrillation, tachycardia, and rapid supraventricular rhythm that compromise organized mechanical activity can lead to ventricular distension and compromised coronary perfusion, hence, should be avoided. Use of Intra-operative TEE is helpful not only for evaluation of prosthetic valve function, but also for assessment of biventricular function, titration of inotropes, and fluid responsiveness to optimize hemodynamic goals.[7]

Strategies for renal protection (maintaining urine output, avoiding hypoperfusion and hypotensive states, avoiding nephrotoxic drugs) are advisable even in the absence of overt kidney impairment. Longer acting muscle relaxants such as pancuronium, that depend on renal excretion, should be avoided and atracurium used instead.[8] Pharmacological interactions between anesthetic agents and immunosuppressants should warrant consideration. Azathioprine and cyclophosphamide may interact with muscle relaxants. Non-steroidal anti-inflammatory drugs and methotrexate may cause acute renal failure and pancytopenia. For pain relief, systemic analgesics should be considered. Regional techniques may be helpful provided neuropathies, myelitis and coagulopathies are excluded.[8]

Anti-coagulation management, during and following valve replacement, is important in patients with SLE. CPB activates immunological and coagulation responses, which makes the management of these patients challenging. Using heparin and protamine in routine fashion is one of the options. Other available options are (1) to use extra heparin to double the baseline ACT, (2) use of bivalirudin as an anticoagulant, (3) achieving a heparin concentration of >3 units/ml in blood, (4) plotting heparin/ACT titration curves, or (5) administration of less protamine, e.g., by giving half of the calculated dose or no protamine at all.[9] If the facility for measurement of heparin concentration is not available, more heparin than routine should be administered to prolong the baseline ACT. Celite ACT is recommended for monitoring of anticoagulation, because kaolin ACT is affected by antiphospholipid antibodies. Protamine should be administered only in a low-dose, and slowly (50 mg every 15 min) until the bleeding tendency slows down to an acceptable level. Thromboelastography may be a useful adjunct in this situation, but is yet to be validated to enable coagulation-related decisions in patients receiving heparin.[10] Antifibrinolytic agents such as epsilon aminocaproic acid, tranexamic acid, and aprotinin should be avoided because of increased risk of thrombosis in such patients. Monitoring fibrin D-dimer levels may aid in the detection of subclinical thrombosis. It is advisable to commence anticoagulation early in the post-operative period to prevent thromboembolic episodes. In addition, early ambulation also helps in reducing the incidence of thromboembolic episodes. Due to embolic events in patients with lupus and the pro-coagulant effect of the lupus anti-coagulant, many patients with lupus are placed on warfarin, a vitamin K-antagonist. The target INR should be rather high, at about 2.5-3.5, but no clear cut data support this recommendation. If thrombotic events occur despite "adequate" anticoagulation, the dose of warfarin and target INR can be increased and/or a platelet inhibitor added.[11] Atrial fibrillation frequently affects patients with severe valve disease, so anticoagulation has to be optimum in such patients.

The major anaesthetic challenges in patients with SLE undergoing AVR include accrued organ damage, coagulation defects, drug interactions, potential airway difficulty, increased risk of infection, and renal impairment. The goals of anesthetic management are directed towards maintaining asepsis, avoiding use of nephrotoxic drugs, and promoting early ambulation.

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