Case Report

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Double Valve Replacement Surgery in a Rare Case of Systemic Sclerosis with Restricted Mouth Opening

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Abstract

Systemic sclerosis is rare autoimmune connective tissue disorder often localized to the skin. Some occasion; it spread to heart, gastrointestinal system, lung and kidney. The cardiac structures like valve, coronary artery and conduction systems are mostly affected. Systemic sclerosis patients have associated pulmonary hypertension, restrictive pulmonary function, thick skin with ulcers and dysrhythmias. We discuss the management of a 39 year female diagnosed case of systemic sclerosis presented for double valve replacement. She had previous mitral valve commisurotomy 25 yrs back, past episode of stroke, restricted mouth opening resulting in difficult tracheal intubation and restrictive lung disease. The case report described the meticulous medical and surgical management for double valve replacement andsuccessful intubation with fibreoptic bronchoscope.

Keywords: Systemic Sclerosis; Restrictive Mouth Opening; Cardiac Surgery; Double Valve Replacement.

Introduction

Systemic sclerosis (SSc) is an autoimmune connective tissue disease leading to hardening of skin due to increased fibroblastic activity [1]. Skin, mucosa, vessel wall and connective tissue became thicken, fibrosis and fused leading to restricted function of the organ [2].

The thick skin produces difficulty in accessing the vein; vasculitis and thrombosis in vein. Attrition of tip of fingers and toes are commonly found in SSc. The cardiac involvements are coronary artery disease, myocarditis and valvular stenosis [3]. Systemic sclerosis affects the conduction tissue in heart and manifest as heart block and life threatening dysrhythmias. Renal involvement produces vasculitis leading to acute renal failure [4]. The gastro-intestinal (GI) system manifests with dilation of gut, ulcerations and dyspepsia. The facial, oral and dental tissues are also affected by SSc and creates restricted mouth opening resulting in difficult tracheal intubation during general anaesthesia [5].

We report the rare case of SSc with mitral valve stenosis, aortic valve stenosis, perioperative arrhythmia, GI dysplasia, restrictive lung disease and restricted mouth opening producing difficult intubation. Literature search in English language do not find any such publication. Hence we describe the perioperative management of the case undergoing double valve replacement surgery.

Case History

A 39 year lady with body weight of 48kg presented to the hospital with complains of palpitation and dyspnea on exertion.She was a diagnosed case of systemic sclerosis.She had previous intervention of percutaneous trans-venous mitral commisurotomy (PTMC)for treatment of mitral stenosis 24 years back. The patient had a past episode of cerebro-vascular accident by thrombus embolus migration. There were associated gastrointestinal problems like gastritis and dyspepsia.

The preoperative examination detectedsclerosed skin, attrition of finger tips with few ulcers. Culture

sensitivity of the fluid from ulcer detected sterile. The mouth opening was small and the inter-incisor passage was allowing one and half fingers measuring approximately 2cm, with perioral fibrosis.This was giving the anticipation of difficult intubation. The CT scan study of larynx, trachea and bronchus detected no stricture or compression. Auscultation of chest detected mid-diastolic murmur at mitral area and ejection systolic murmur at aortic area. The air entry to both lungs was equal, reduced and without any added sounds. Echocardiography detected thick mitral valve leaflets, restricted movement, subvalvular apparatus diseasewith fused chordae, and mid-diastolic gradient of 15mmHg, valve area of 0.7cm^2 suggestive of severe mitral stenosis and no regurgitation. The aortic valve leaflets were thick, restricted movement and peak gradient of 90mmHg suggestive of severe aortic stenosis. Tricuspid valve closure was showing mild regurgitation in color Doppler. The left ventricular ejection fraction was 50%. The pulmonary function test detected mild restrictive lung disease with a predicted functional vital capacity of 75%. Laboratory examination of complete hemogram, liver and renal functions detected normal values except hemoglobin of 9.6gm%, creatinine 1.6mg% and ESR of 55mm in 1sthour.

She was planned for double valve replacement surgery under cardiopulmonary bypass. Premedication in the form of alprazolam 0.25mg at bedtime and morphine 4mg, promethazine 20mg 30min before shifting to operating room(OR)were administered to the patient to alley anxiety and produce preemptive analgesia. Peripheral venous access was difficult because of thick skin and thin thrombosed veins. Two small caliber (20G) peripheral venous cannulas were established with great difficulty. Invasive arterial line was established in left radial artery. Methylprednisolone 1g and ceftazidime 1g were injected slowly. Pulse oximetry couldn't be applied in the fingers due to the attrition, fibrosis and ulcers, but was applied on the ear lobe with reliable reading. Anaesthesia was induced with fentanyl 500µg, midazolam 1mg and etomidate 15mg after preoxygenation for 5min with 100% oxygen. Bag and mask ventilation was confirmed. Due to anticipated difficult intubation fibreoptic bronchoscope was used to intubate the trachea with a 7mm cuffed endotracheal tube. Central venous catheterization via right internal jugular vein, trans-esophageal echocardiography (TEE) and bispectral index were used to monitor intraoperative cardiac function and level of sedation. Anaesthesia was maintained with O2, air, sevoflurane, fentanyl, midazolam and atracurium; and patient was mechanically ventilated.

Intraoperative TEE confirmed severe mitral and aortic stenosis (Figure 1). Systemic anticoagulation was achieved with 400IU/ kg of heparin. After aorto-bicaval cannulation CPB was commenced.Under moderate hypothermia, 32^{0}C mitral and aortic valve were replaced with 27mm and 21mm size mechanical prosthetic valve from St Jude Medical, St. Paul, Minnesota 55117 USA. Ultrafiltration was continued during and after CPB to reduce the inflammatory marker and hemo-concentrate the blood. Patient was weaned from bypass with dobutamine 5µg/kg/min, nitroglycerine 0.5µg/kg/min, sinus rhythm and mechanical ventilation of lungs. Heparin was reversed with protamine. She was shifted to ICU and ventilated overnight in lieu of difficult intubation. The postoperative blood loss was 150mL. The postoperative pulmonary care was adjusted with bronchodilator, humidification, incentive-spirometry and chest physiotherapy. She was shifted to ward on 3rdpostoperative day. She developed episodes of ventricular ectopic and atrial fibrillation that was controlled with amiodarone therapy. The subsequent period was uneventful and she was discharged from hospital on 12th postoperative day.

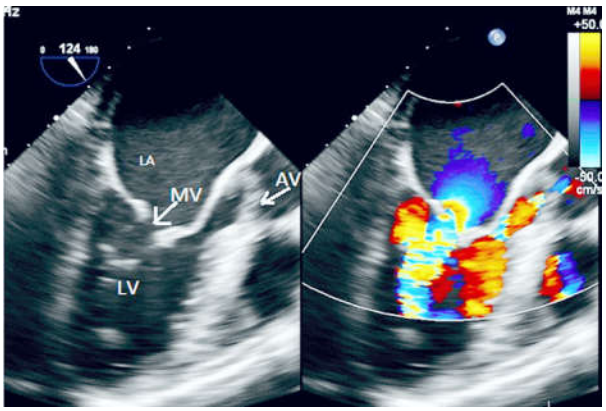

Fig. 1: Transesophageal echocardiography midesophageal aortic long axis view color compare showing both aortic valve (AV) and mitral valve (MV) stenosis. LA- left atrium, LV- left ventricle

Discussion

Systemic sclerosis (SSc) is an autoimmune disease predominately involve the skin [4].

Systemic sclerosis more commonly found in female (male: female 1:3 [2,6]. The localized form called scleroderma, where the skin is thick and fibrosed resulting difficulty in peripheral venous cannulation. The etiology of SSc is exactly unknown. Initially vascular inflammation and cellular

apoptosis are the common happening. Subsequently collagen deposition occurs in skin and many organs [4]. There may be ulcers at fingertips or skin due to necrosis of the area. The systemic form involves lung, kidney, gastro intestinal (GI) system and heart. The CREST syndrome is a manifestation of SSc where the combination of Calcinosis, Raynaud's phenomenon, Esophagealdysmotility, Sclerodactyly and Telangiectasia [6]. Systemic sclerosis also involve facial, head and neck tissue, where there will be perioral fibrosis leading to restricted mouth opening, temporo mandibular joint fibrosis and ankyloses are detected because offibrosis of soft tissue and ligaments [7]. This condition creates problem for anaesthetists during management of airway for cardiac surgery or intervention in cardiac catheterization laboratory.

Cardiac manifestations are coronary artery disease, conduction abnormality, aortic stenosis, mitral stenosis. Conduction problem mayassociate with heart black atrio- ventricular dysrhythmia. The patient may require pacemaker or automated implantable cardiovertr defibrillator (AICD) placement. Valve involvement with associate renal and pulmonary hypertension is better managed with catheterization laboratory intervention like trans catheter aortic valve implantation (TAVI), PTMC and balloon valvotomy [8].

Lung pathology associated with SSc is detected as pulmonary hypertension (PHT)[2] and restrictive lung disease. Many patient die due to severe PHT. Stress Dopppler echocardiography is successful in 95.2% cases to detect PHT [6]. The use of sildenaphil, busentan, chest physiotherapy, bronchodilator and steroids are effective to manage the PHT and restrictive lung disease. The requirement of lung & heart transplant reported in some cases. Esophageal dysmotility, reflux gastritis and stricture may be observed in the advanced stage of SSc. Balloon dilatation and bougienage are performed in patients developing narrowing of esophagus. Esophagectomy in Scleroderma has been reported by Yekelera E et al in a case of esophageal stricture in a 20yr patient [7]. Kidney is affected in 60-80% of cases. The manifestations are systemic sclerosis renal crisis (SRC), interstitial fibrosis and glomerulonephritis [5]. Systemic sclerosis renal crisis requires prompt diagnosis and treatment with angiotensin converting enzyme inhibitors. Renal biopsy confirms diagnosis and provides critical insight into the pathogenesis [5]. Endothelin is a logical potential target for therapy in renal scleroderma.

The present case was unique with multiple organ and system derangement due to SSc. In our case the patient had thick skin, ulcers, attrition of distal extremities, restricted mouth opening, anemia, mild derangement of renal function, mild restrictive lung disease, acute inflammation and most importantly the cardiac involvement. She had mitral stenosis 25 yr back, that was managed with PTMC. The present time showed restenosis of mitral valve and severe aortic valve stenosis with mild tricuspid regurgitation. This situation produced surgical challenge. Multi-organ derangements need modification of drugs that are less dependent on renal metabolism. The use of ultrafiltration and steroid helped us to reduce the tissue inflammation especially in a situation like SSc. The restrictive lung function needed perioperative bronchodilator, steroid, respiratory exercise and chest physiotherapy. Restricted mouth opening was a challenge for tracheal intubation; that was managed with fibreoptic bronchoscope intubation. Postoperative arrhythmia was controlled with amiodarone.

In summary, systemic sclerosis is a connective tissue disorder that can affect major organ like heart, lungs, kidney, GI system, skin and airway. Thorough screening, assessment and investigation will help in identifying the risks before cardiac surgery. Perioperative optimization of pulmonary, renal gastrointestinal and immune function will achieve a successful outcome after double valve replacement or any other form of cardiacsurgery.

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