CASE REPORT

Pregnancy in a Patient With Severe Aortic Stenosis Requiring Unusual Management

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IN DEVELOPED COUNTRIES, congenital heart disease is the most common cardiovascular disease reported during pregnancy, with the majority of the cases of aortic stenosis in pregnant women being caused by congenital bicuspid aortic valve.[1,2] While patients with asymptomatic mild, moderate, or even severe aortic stenosis usually can continue pregnancy to term, all symptomatic patients with severe aortic stenosis or asymptomatic patients with impaired left ventricular function are at high risk for cardiac complications.[1] Aortic valve replacement is indicated before pregnancy in these women.[1] Percutaneous aortic valvuloplasty also can be considered during pregnancy.[3–5] More invasive techniques, such as aortic valve replacement under normothermic cardiopulmonary bypass during pregnancy, are associated with a high risk of mortality and morbidity for both the mother and the fetus[6–8] The authors report here a case in which the global management of a pregnant patient with severe aortic stenosis was reconsidered in light of an unexpected echocardiographic report of calcified valves with a high risk of emboli.

CASE REPORT

A 41-year-old woman (163 cm, 92 kg) with a second pregnancy arrived in the emergency department of a primary center during the 26th week of gestation with the onset of dyspnea on exertion, worsening gradually during pregnancy and interfering with regular daily activities (NYHA class III). She did not have any notable history apart from two miscarriages and a previous normal pregnancy 10 years ago that was well tolerated and did not require any medical attention, resulting in a child who weighed 1,800 g and was delivered vaginally at term with no further complications.

All obstetric echocardiographies performed during the current pregnancy revealed a normal fetus with a normal developing status and a cranial presentation. There was no sign of fetal distress.

On admission, functional status was evaluated as NYHA class II. Auscultation revealed a grade II/VI systolic murmur in the aortic area and a diminished second heart sound. The electrocardiogram revealed normal sinus rhythm with evidence of left ventricular hypertrophy and subsequent repolarization abnormalities. Transthoracic echocardiography showed evidence of left ventricular hypertrophy with a septal wall of 16 mm, no dilatation of any fraction, and normal right ventricle function. The aortic valve had 3 leaflets and was calcified, with restricted motion. Mean gradient was estimated at 18 mmHg and valve area at 0.9 cm². There was neither other valve disease nor dilatation of the ascending aorta. No estimation of the cardiac output was made at that point. No therapeutic measures were taken, and the patient was discharged from the hospital with the indication for further cardiac follow-up. The patient was instructed to visit a cardiologist every 4 weeks for a complete cardiac evaluation and to present at the emergency room at any moment if her status deteriorated, but no formal follow-up actually was planned.

During the 32nd week of gestation, she was readmitted urgently to the emergency room of the same center following 2 syncopal episodes and was referred to the authors' cardiac referral center. Following transfer, a clinical examination revealed a functional status evaluated at NYHA class III, and BP was 142/87 mmHg with a regular pulse of 102 bpm and no signs of congestive heart failure. A grade III/VI systolic murmur in the aortic area and absence of a second heart sound with normal pulmonary auscultation were detected. The electrocardiogram had a similar appearance to that performed at the time of first admission, with normal sinus rhythm, evidence of left ventricular hypertrophy, and subsequent repolarization abnormalities. The laboratory workup revealed mild microcytic anemia (11.2 g/dL hemoglobin), NTproBNP of 1,004 pg/mL and hs-cTnT (highly sensitive troponin T assay) of 48 pg/mL. Echocardiography showed a calcified congenital bicuspid aortic valve with severe stenosis, an aortic area of 0.7 cm^2, an indexed aortic valve area of 0.36 cm^2/m^2, a mean transvalvular gradient of 124 mmHg, and a maximal transvalvular gradient at 210 mmHg from a right parasternal window using a Pedoff probe. Left ventricular stroke volume was 77 mL, the cardiac output was 8.09 L/min, and the indexed cardiac output was 4.11 $L/min/m^2$. A mobile calcified nodule was observed at the extremity of the posterior cusp prolapsing inside the left ventricular outlet, measuring a diameter of 0.4 cm. The left ventricle had normal systolic function, concentric hypertrophy with septal predominance of 16 mm, impaired relaxation with E/A = 0.77, and high filling pressures with E/E' = 15. There was no evidence of left ventricular dilatation, the left ventricular telediastolic diameter being 5.44 cm and the left ventricular telesystolic diameter 3.78 cm. The left ventricle ejection fraction was calculated at 58%. There was no evidence of aortic regurgitation or ascending aorta dilatation, but minimal signs of mitral and tricuspid regurgitation were identified, with no hemodynamic significance. Right ventricular function and pulmonary artery pressures were normal. Obstetric examination revealed a normal course of pregnancy with a normally developed viable fetus.

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Although percutaneous aortic valvuloplasty initially was discussed, the risk of arterial embolization of calcified fragments was considered to be excessive because of the extreme mobility and dimensions of the calcified nodule, and this procedure was not performed. After multidisciplinary discussion involving cardiologists, obstetricians, pediatricians, and anesthesiologist, the planned strategy was to accelerate pulmonary fetal maturation by steroid therapy until the 35th week of gestation and perform C-section immediately followed by aortic valve replacement. It was proposed to delay aortic valve replacement by 48 hours in order to optimize hemodynamic parameters. However, the risk of major complications in the case of any hemodynamic instability or other complication following C-section, including coagulation disorders and prolonged bleeding, was considered to be excessive given the severity of aortic stenosis and its poor functional tolerance. Thus, the decision was made to perform a combined procedure, starting with a C-section and immediately followed by aortic valve replacement.

The patient was hospitalized in the cardiac intensive care unit. Since she remained asymptomatic at rest and there was no sign of congestive heart failure, no medical treatment or hemodynamic optimization was needed except a correction of the iron-deficiency anemia with oral iron and gastric protection using rabeprazole.

Surgery was planned 14 days after admission and was performed in the cardiac operating room. Because of the risk of hemorrhage and possible hemodynamic instability related to uterine bleeding after C-section during systemic heparinization for extracorporeal circulation, the multidisciplinary decision to perform total hysterectomy was made in case of difficult bleeding control. The patient and her relatives were informed about and accepted the risks and possible complications.

Thirty minutes prior to induction of anesthesia, 150 mg of effervescent ranitidine were administered as prophylaxis for acid aspiration syndrome. A 16-G cannula for fluid management was inserted in a peripheral vein on the right hand. An invasive arterial catheter in the left radial artery and a central venous catheter in the right internal jugular vein were placed under local anesthesia prior to induction. A bispectral index monitor was used to evaluate depth of anesthesia. Before induction, BP was 160/95 mmHg and pulse was 120 bpm.

A rapid-sequence induction was performed using 40 mg of etomidate (0.4 mg/kg) and remifentanil titrated for an effect-site concentration of 2.5 μg/mL using the Minto model. A dose of 100 mg of succinylcholine (1.08 mg/kg) was used for muscle relaxation and tracheal intubation. Hemodynamic stability was obtained and, after induction, systolic BP varied between 125-145 mmHg, and pulse varied between 98-110 bpm. A fractional inspired oxygen concentration (F_IO_2) of 1.0 was used before extraction, and the patient was maintained on desflurane, titrated in order to obtain a BIS value between 40-60.

A female baby weighing 2,200 g (ranked in the 10th percentile), with a length of 42.5 cm (30th percentile) and a head circumference of 32 cm (30th percentile) was delivered. The APGAR score was 4/5/7/9 at 1/2/5/10 minutes. The baby cried immediately after birth, her oropharynx was suctioned, and she was assisted using supplemental oxygen by facial mask for mild respiratory distress, probably due to incomplete pulmonary maturation and the opioid used during anesthesia. She had a pulse of 140 bpm and SpO_2 of 70%, which rapidly increased to 97%, with pH 7.22 and serum lactate of 3.3 mmol/L. After initial stabilization, the baby was transferred to the neonatal intensive care unit. Induction-delivery, abdominal skin incision-delivery, and uterine incision-delivery times were 17 min, 14 min, and 9 min respectively.

Full heparinization for cardiopulmonary bypass was performed immediately after delivery with 300 IU/kg of unfractionated heparin. This decision to perform full heparinization before uterine closure was make in order to ensure perfect surgical hemostasis and minimize the risk of uterine bleeding during cardiopulmonary bypass. Uterine muscle tone was stimulated simultaneously with uterine massage and rapid infusion of 15 units of oxytocin followed by continuous infusion of 1 unit/h. After complete surgical hemostasis and closure of the abdominal wall, the total bleeding volume was 950 mL, and moderate uterine muscle tonus was obtained.

Transesophageal echocardiography was performed 15 minutes after delivery under general anesthesia and showed a mean aortic transvalvular gradient of 42 mmHg and high left ventricular filling pressures. The anesthetic technique was switched to total intravenous anesthesia using propofol, titrated for an effect-site concentration of 3 mg/mL, and sufentanil, titrated for an effect-site concentration of 0.4 ng/mL, using the Schnider model, according to the local cardiac surgery protocol, ensuring hemodynamic stability and a BIS value between 40-60. As the patient had a history of a possible penicillin allergy, vancomycin, 15 mg/kg, and gentamicin, 5 mg/kg, were chosen for antibiotic prophylaxis Aortic valve replacement was performed under normothermic cardiopulmonary bypass with non-pulsatile flow and anterograde blood cardioplegia. Mean arterial pressure was maintained between 50 mmHg and 60 mmHg with a pump flow rate of 2.4 L/min/m^2 (4.8 L/min), and aortic valve replacement was performed with a 21-mm CE Magna Ease biologic valve. There was no need for hemodynamic drug support after weaning off cardiopulmonary bypass, and the patient immediately recovered normal sinus rhythm. The cardiopulmonary bypass time was 47 min, and the aortic cross-clamping time was 36 min. Protamine was administered to antagonize the initial heparin dose, the thorax was closed after surgical hemostasis, and 390 mL of blood autotransfusion were given.

The patient was transferred to the cardiac surgical intensive care unit. The postoperative course rapidly was favorable, and the patient was extubated 6 hours after admission to the ICU with no need for hemodynamic support. There was no evidence of coagulopathy or abnormal bleeding in the pericardial drains, and only 1 unit of packed red blood cells had to be transfused during the ICU stay. Standard anticoagulation with unfractionated heparin was introduced 6 hours after surgery and was switched to enoxaparin 2 days after surgery. Oxytocin was administered at a dose of 20 IU/day for 2 days after surgery, and normal uterine tone was obtained. The patient was discharged from the ICU 4 days after surgery after full recovery. Her baby also had a favorable course and was discharged from the neonatal intensive care unit 7 days after birth.

DISCUSSION

At the present time, 0.2% to 4% of all pregnancies in western industrialized countries are complicated by cardiovascular diseases, and an increasing number of patients develop cardiac diseases during pregnancy.[1] Hypertension is still the most common event, occurring in 6% to 8% of all pregnancies. In developed countries, congenital heart disease is the most common cardiovascular disease reported during pregnancy (75%-82%). In other countries, the most common cardiovascular disease is rheumatic valvular disease (56%-89%). Other diseases such as cardiomyopathies are rare but can induce severe complications.[1]

Marked anatomic and physiologic changes occur in women during pregnancy to meet the increased demands of the mother and fetus, including increased plasma volume and cardiac output and decreased venous return and systemic vascular resistances, with changes in cardiac preload and afterload. Systolic function initially increases but then decreases in the last trimester due to modifications of ventricular volumes. Hemostatic changes also are documented, with increased concentrations of coagulation factors and fibrinogen and platelet adhesiveness leading to an increased risk of