The Egyptian Cardiothoracic Surgeon

In Press

Original Article

Bentall Procedure for an Adolescent with Sickle Cell Disease, Hodgkin's Lymphoma, and old Cerebrovascular Accident

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Abstract

Cardiopulmonary bypass (CPB) in patients with sickle cell anemia can trigger lethal vaso-occlusive crises, especially in cases of hypoxia, hypothermia, acidosis, or low-flow states. We described a patient with sickle cell anemia who had bicuspid aortic valve stenosis and aneurysmal dilatation of the ascending aorta complicated with infective endocarditis. The patient had a history of stroke. During routine workup, Hodgkin's Lymphoma was diagnosed. The patient underwent exchange transfusion preoperatively and immediately before the initiation of CPB. We performed a Bentall procedure, and the patient was discharged in a stable condition. Sickle Cell Disease can be very challenging during CPB, and special precautions are required to prevent vaso-occlusive crises.

KEYWORDS

Sickle Cell Disease; Hodgkin's Lymphoma; Aortic valve replacement; Cardiopulmonary

Introduction

Sickle cell anemia is an autosomal recessive inherited disorder that is prevalent among people of African descent [1,2]. Sickle Cell Disease (SCD) is characterized by its recurrent vaso-occlusive crises, which may lead to multi-organ damage and a reduction in survival rate [3]. Sickle cell disease patients are at higher risk for developing malignancies, especially hematological malignancy. Hodgkin's lymphoma is common in those patients [4], which may affect the management. With the improvements in diagnostic and surgical techniques along with anesthetic management, the number of patients with SCD who undergo cardiac surgery has increased [2].

Hypoxia, hypothermia, acidosis, and low flow states can trigger a lethal vaso-occlusive crisis. Cardiopulmonary bypass (CPB) provokes these factors and can lead to fatal vaso-occlusive crises during cardiac surgery. Stabilization of the hemoglobin (Hb) level preoperatively and the use of exchange transfusion along with careful monitoring for the triggering factors can decrease the risk of sickling crisis [1].

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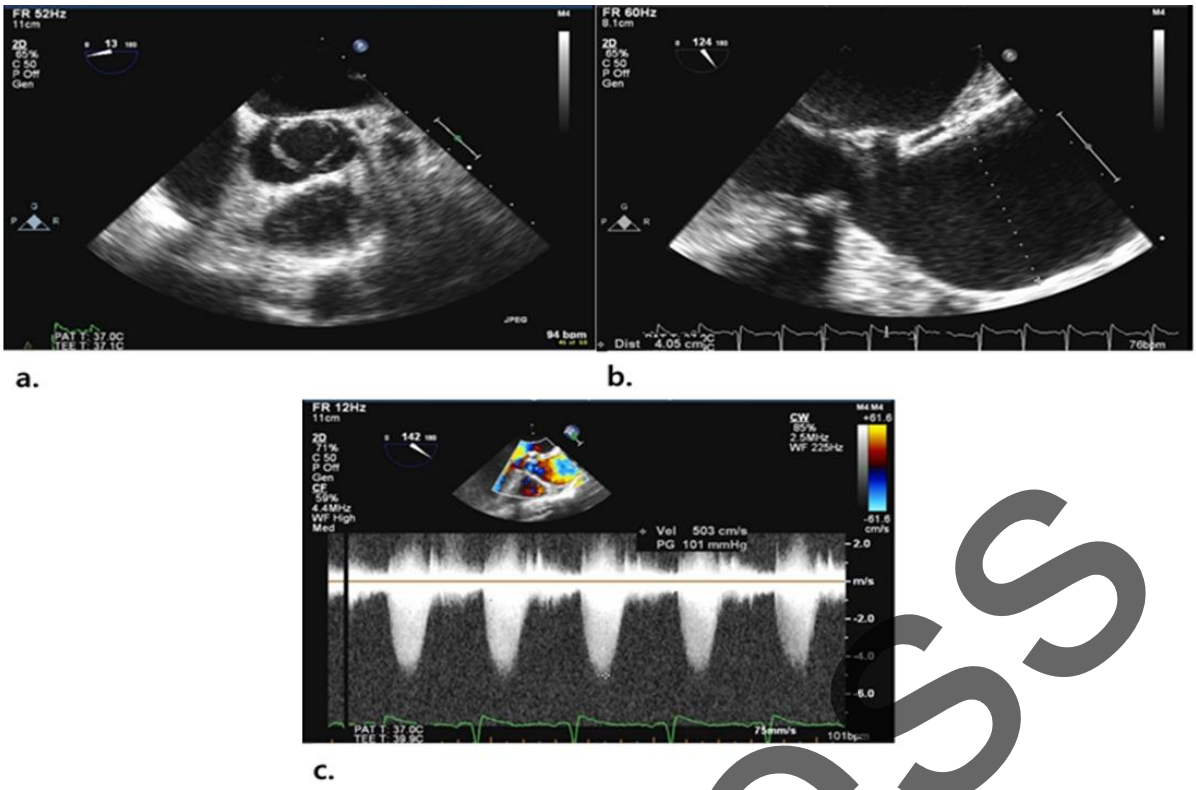

Figure 1: Transesophageal echocardiogram demonstrating. (a) Bicuspid aortic valve. (b) The aortic root. (c) Color Doppler demonstrating a severe aortic stenosis

Case Presentation:

A male patient aged 20-year old, presented with fever. The patient had a sickle cell anemia and a history of stroke with right-side weakness. The patient was referred to our center as a case of acute infective endocarditis, bicuspid aortic valve stenosis, and post stenotic aortic root dilatation. On examination, the patient was conscious, oriented, and hemodynamically stable. The chest was clear on auscultation, with normal oxygen saturation on room air. The patient was febrile with a temperature of 38C.

Precordial auscultation showed a harsh ejection systolic murmur with crescendo-decrescendo configuration, medium pitch with grade III intensity, best heard over the aortic area and radiated to the carotid.

The hemoglobin (Hb) was 7.4 g/dl, HbS 92.2%, and white blood cells (WBC) 21000 per microlitre. Three blood cultures from different sites were negative and respiratory, and urine cultures were also negative. Q-fever, Brucella, and Dengue fever workup were negative. The patient was started on empirical intravenous ceftriaxone 2 gm/24 hours and vancomycin 600/8 hour.

Transthoracic echocardiography showed severe concentric left ventricular (LV) hypertrophy, and the LV ejection fraction was 60%. Aortic valve (AV) was bicuspid with no evidence of vegetation, and the aortic root was 4.1 cm. The transesophageal echocardiography showed thickened and restricted aortic valve leaflets. There was severe aortic valve stenosis with peak and mean gradients of 160 and 90 mmHg, respectively. There was no vegetation or aortic root abscesses to support the infective endocarditis diagnosis. (Figure 1 a, b, c).

Computed tomography (CT) scan of the chest and abdomen showed generalized lymphadenopathy in the axilla, abdomen, and inguinal area. An excisional biopsy of the left inguinal lymph node (LN) showed Hodgkin lymphoma (Figure 2).

Brain CT angiogram demonstrated multiple old infractions, mainly in the occipital lobe, posterior limb of the right internal capsule, and the corona radiata with encephalomalacia (Figure 3). Magnetic resonance imaging (MRI) confirmed the CT findings.

Positron emission tomography (PET) scan showed multiple FDG avid enlarged lymph nodes seen in the left inguinal, obturator and external iliac lymph node chain. The largest measured around 3.2 cm (SUV max 15) (Figure 4).

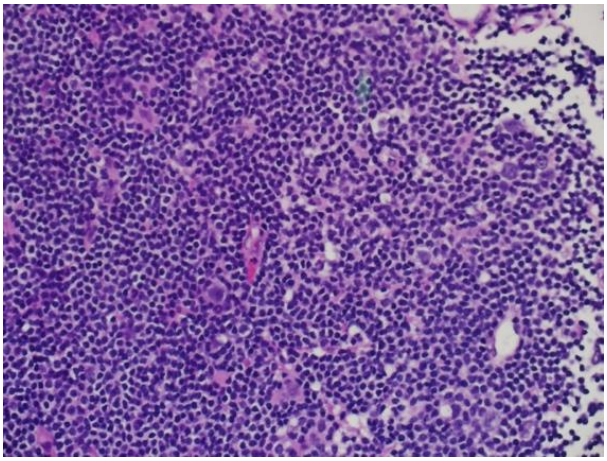

Figure 2: Microscopic view demonstrating scattered large atypical cells in the background of small lymphocytes, H&E 100x

The patient was scheduled for aortic valve and ascending aorta replacement. We performed an exchange transfusion using packed red blood cells (PRBCs) in the preoperative period and immediately before the initiation of CPB.

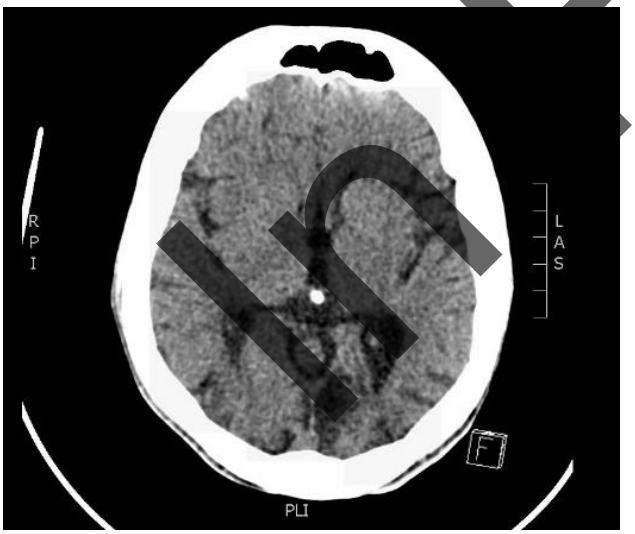

Figure 3: CT scan demonstrates multiple old infractions mainly seen in the occipital lobe, posterior limb of the right internal capsule, and the corona radiata with encephalomalacia changes

The circuit was adapted for the exchange transfusion by adding a 1/4-3/8-3/8" Y connector within the venous line. The two 3/8" ends of the Y were connected to the standard venous line of the circuit. The 1/4" end of the Y was connected to 1/4" tubing and attached to a separate cardiotomy reservoir (cell saver).

The circuit was primed with 900 ml of crystalloid fluid. After that, the crystalloid fluid was replaced with 250 ml of 5% albumen. After the circuit was primed, the excess crystalloid fluid was drained from the circuit, and four units of PRBCs were added to the venous reservoir.

Once the chest was opened, and the heart was exposed, the patient was systemically heparinized. An aortic cannula was placed in the ascending aorta, and venous drainage was accomplished through a straight cannula placed in the right atrium. Once an activated clotting time of 371 seconds was achieved, the exchange transfusion was initiated. With the venous line clamped distal to the Y connector, the line to the separate cardiotomy reservoir was opened, allowing the patient's blood to drain. At the same time, blood from the CPB machine was transfused to the patient to maintain the patient's hemodynamics. After an exchange transfusion of 1500 mL and reducing HbS down to 3.6 g/dl, the patient was placed on CPB.

Normal hypothermia was used, and the patient was cooled to a bladder temperature of 34°C. A mechanical aortic valve size 21-mm was implanted, and ascending aorta was replaced with size 26mm vascular graft. The CPB flows were maintained at a minimum cardiac index of 2.4 L/min. Venous saturation was kept >70%, and no acidosis occurred during the CPB period. Two units of PRBCs were given during the bypass. Hemoglobin level was 10.6g/L. The patient was re-warmed to a bladder temperature of 36°C. The aortic cross-clamp time was 134 minutes, and the total CPB time was 163 minutes. There was surgical oozing with no obvious site of bleeding, blood products were given, and a decision was made to leave the chest open and transfer to the intensive care unit (ICU).

In the ICU, the patient required surgical re-exploration for hemostasis with no detected site for bleeding, and the chest was closed on the second postoperative day [6]. The pathology

report of the aortic valve leaflets showed signs of endocarditis.

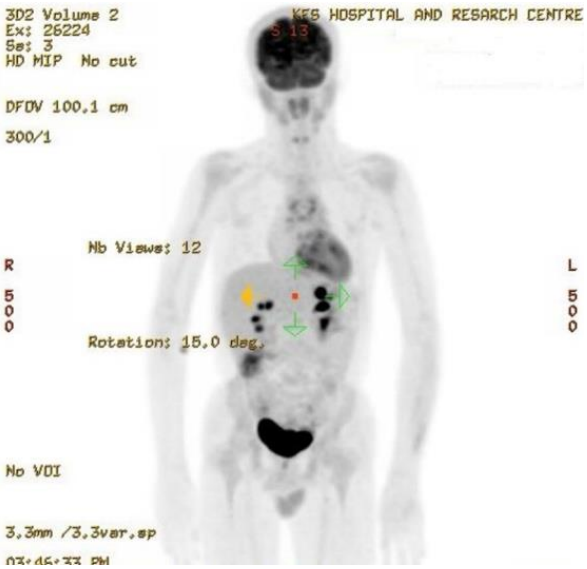

Figure 4: There are multiple FDG avid enlarged lymph nodes seen in the left inguinal, obturator, and external iliac lymph node chain. The largest measures around 3.2 cm (SUV max 15).

The patient was discharged in a stable condition on warfarin with a targeted INR (international normalized ratio) of 2-3. Follow up appointments with oncology and hematology outpatients' clinics were given for the management of lymphoma.

Seven months after the surgery, the patient was admitted with acute right occipital ischemic stroke. Regarding the cardiac point of view, there was no complaint, and he showed a significant improvement in the symptoms. There were no major surgical complications. The admission duration was two weeks, after which he was discharged in a stable condition.

Discussion

This case presents several challenges that we had to tackle, one of which was a vaso-occlusive crisis during aortic root replacement, which is considered one of the major surgeries. A similar case report had the same challenge but managed with a different approach. We performed exchange transfusion, which can aid in two ways, one is by reducing HbS, and the other is by increasing the preoperative hematocrit level. Thus, facilitating better oxygen delivery to the tissues [2,7]. This measure was taken preoperatively; additionally, perioperative exchange transfusion was done to reduce the HbS level and decrease the probability of the vaso-occlusive crises. Simple sequential transfusions had also been reported to reduce the HbS level in a similar case with no intraoperative complications [1].

Mild hypothermia (> 32°C) is traditionally used for myocardial preservation by decreasing the metabolic rate and reducing oxygen demand. However, hypothermia in patients with sickle cell anemia is still controversial, with almost half of the published reports, patients were maintained on normothermia during CPB, the rest underwent mild hypothermia (> 32°C) [1].

In our case, we performed mild hypothermia of 34°C during the surgery, CPB flows were maintained at a minimum cardiac index of 2.4 L/min, venous saturation was kept >70%, and no acidosis occurred during the CPB period. There was no major vaso-occlusive crisis that occurred throughout the surgery. On the other hand, in a similar case report, they opted for normothermia that resulted in a good outcome [1].

There is no consensus regarding the superiority of either cold or warm cardioplegia during cardiopulmonary bypass, even in patients with SCD [5]. Warm cardioplegia aids in removing HbS from the coronary circulation, thus reducing the possibility of a sickling crisis, yet it is associated with less myocardial protection [5]. On the other hand, cold cardioplegia has a longer duration [5], and this was the reason we used cold cardioplegia for this case.

Another challenge is the use of mechanical vs. biological valves. Even though mechanical valves are superior in durability, they are linked with an increased risk of thrombosis, hemolysis, sickle cell crisis, and lifelong anticoagulation use, which can predispose the patient to hemorrhagic complications [1, 5]. In our case and because of his young age, and the durability of the mechanical valves, we prioritized the use of a mechanical valve. In contrast, in the similar case report, they chose a biological valve prosthesis based upon several factors one of which is the that SCD patient

has reduced life expectancy compared to the general population, another factor is the potential risk that the mechanical valves could develop complications as mentioned before [1].

A multidisciplinary team approach that involved pain management, hematology, and oncology was essential in this case. Pain management is of a major consideration due to chronic pain from a recurrent vaso-occlusive crisis [5,8]. Respiratory splinting as a result of the surgery may affect the patients' breathing and may result in hypoventilation and subsequent acidosis, resulting in a sickling crisis [8]. On the other hand, higher doses of opioids may suppress respiration leading to acidosis [8]. For these reasons, optimal pain management is an essential element in the recovery phase [5,8].

Conclusion

Sickle Cell Disease can be very challenging during cardiopulmonary bypass, which can trigger vaso-occlusive crises. Exchange transfusion can reduce HbS and increase the hematocrit level and mild hypothermia may be beneficial in such cases.

Conflict of interest: Authors declare no conflict of interest.

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