

Complicated Sickle Cell Anemia: A Challenging Case.

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ABSTRACT

Sickle cell anaemia may lead to serious problems during anaesthesia and surgery, due to the nature of the disease and the various Multi-Organ affections and complications that can ensue, SCD patients are at high risk, thus making anaesthesia stressful and challenging even in a state of the art settings. This is a case study of a Saudi patient with complicated SCA who was brought in for an urgent surgery after a fracture of the right neck of the femur following a Road Traffic Accident (RTA). The evidence base oriented approach was taken to decide and stratify patient cardiac condition for prediction of the risk and its management, beside his end stage kidney diseases and his dependency on regular hemodialysis. At the same time the effect of sickle cell anaemia itself and the problem of his raised pulmonary hypertension. This case study emphasizes the proper preoperative assessment and evaluation of SCD patients in addition to the collaboration with other medical specialists, including a hematologist, physician, cardiologist, anesthesiologist, and an orthopaedic surgeon in multi-disciplinary approach.

Keywords:Sickle cell anaemia, Dilated Cardiomyopathy, End stage renal disease, Anaesthetic management.

INTRODUCTION

Sickle cell disease (SCD) is a haematological disorder of the human red blood cell, caused by the inheritance of the sickling gene alone or in combination with another abnormal haemoglobin gene. The commonest genotypes are homozygous sickle cell disease (SS), sickle cell haemoglobin C disease (SC) and sickle cell β -Thalassemia (SI3thal).

The disease has its origins in sub-Saharan Africa and the Middle East, hence it is most prevalent in individuals of African descent as well as in the Caribbean, Middle East, parts of India and the Mediterranean, and South and Central America. SCD is now of increasing importance worldwide and there are increasing numbers of affected individuals in Europe and the USA due to immigration of people from endemic areas. In Kingdom of Saudi Arabia (KSA) Sickle Cell Anemia is found mainly in the eastern and southern provinces; many studies have classified this part of the gulf as an endemic area.

Every year, many patients with sickle cell anaemia undergo anaesthesia for different Major Surgical Procedures that are usually associated with high morbidity and mortality. Although sickle cell anaemia is a hemoglobinopathy disorder.

stroke and other complication associated with repeated blood transfusion.^[1]

CASE REPORT

A 30 years old male weighing 30 kg presented with fracture of the neck of the right femur 2 days prior to his presentation due to a Motor Vehicle Accident (MVA), and was scheduled for InterTAN surgery. He was a known case of complicated sickle cell anaemia (SCA) leading to End Stage Kidney Disease and Dilated Cardiomyopathy. The patient is dependent on hemodialysis with history of many hospitalizations during last 7 years of congestive heart failure and sickling crisis during which he received medical treatment and blood transfusions. Consultation with Haematology, Nephrology, Cardiology and Anaesthesia Department were carried out accordingly.

On Clinical assessment patient looked ill dyspneic, pale, but not jaundiced or cyanosed, the Pulse rate was 115 bpm and regular, Blood pressure was 170/100. Physical examination revealed a pulsatile pericardium with a positive thrill and a dynamic Apex at the 6th Intercostal space. A Pan systolic murmur at mitral and tricuspid areas were identified as features of mitral and tricuspid regurgitation. The second heart sound was also palpable. The Liver was enlarged 2 cm below the costal margin and it was tender, JVP however was not raised. No LL edema but there was sacral edema.

Investigation during the time of presentation revealed the following parameters:

Urea=174 mg/dl, Creatinine = 9.1 mg/dl, Sodium= 125 mmol/l, Potassium = 6.7 mmol/l, albumin = 3.9g/dl, Hb = 6.2 g/dl, Hct = 20.8%, Platelet Count = 266 103/UL, APTT = 38.2 sec, PT = 11.8 sec.

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It can lead to serious multisystem complications, including congestive heart failure, severe pulmonary hypertension, end stage renal diseases,

His ECG showed NSR with a rate of 122bpm high voltage QRS in left chest leads [Figure 1], ECHO = dilated left ventricle with EDD 7.1 CM. depressed LV function with ejection fraction EF of 34% [Figure 2, 3]. Severe mitral and tricuspid regurgitation. Systolic PAP = 65 mmHg.

Blood transfusion was planned immediately, the patient received two units of PRBCs, hemodialysis during the consecutive 2 days before surgery as Hb was 6.7 gm/dl, and CCU bed was arranged.

On the morning of the surgery, his lab parameters were as follows:

Hb = 9.5 gm/dl, Htc = 30%, Sodium = 132 mmol/l, Potassium = 4 mmol/l and Creatinine = 3.7 mg/dl, glucose = 109 gm/dl. High risk consent was obtained as per hospital policy and the patient was reassured. No pre-medications were given.

On the day of surgery, patient heart rate was 80 bpm, oxygen saturation was 94% on room air, and blood pressure was 115/85 mm Hg [Figure 4]. A left radial arterial line was inserted under local anaesthesia prior to the induction of anaesthesia while the patient had already a right subclavian central line. The patient was connected to a standard 5-leads monitor. Anaesthesia was induced with 25 mg xylocart, 100 mcg fentanyl, 65 mg propofol and 25 mg atracurium with 100 % oxygen and 1% sevoflurane. Gentle intubation was done with endotracheal tube and anesthesia was maintained with oxygen-air, with FiO₂ 0.6 and 1-1.5% sevoflurane, increments doses of atracurium and fentanyl.

Duration of surgery was 125 minutes and during this time the vital signs were as follows: Systolic Blood pressure ranged from 115 mmHg to 128 mmHg while diastolic was 80 mmHg to 88 mmHg. His heart rate ranged from 86 to 90 beats/min, while his SPO₂ was 99 – 100%, CVP 7-9 cm H₂O, ETCO₂ 36 – 38 mmHg [Figure 5]. There was one episode of hypotension when blood pressure dropped to 89/76 mmHg after the second dose of 50 mcg of fentanyl one hour after induction of anaesthesia. The approximate total blood loss was 170 ml and total fluids given to the patient are 200 ml PRBC and 60 ml normal saline. No urine output. Patient was given 50 mcg of fentanyl before transfer to CCU without a drop in his arterial blood pressure. At the end of operation patient vital signs was stable and as follow, HR = 90/ min, BP = 127/82. He was transferred to CCU and electively mechanically ventilated for 24 hours with stable vital signs. His CCU stay was uneventful and his 12-lead ECG did not show any new changes after being connected to the ventilator [Figure 6].

DISCUSSION

In order to reduce the problems associated with anaesthesia and surgery in patient with complicated

sickle cell disease (SCD), clinician must have a good knowledge about the disease itself and its complication. SCD have a great and high morbidity and mortalities due to co-morbidities of cardiovascular, respiratory, and renal systems. In every system this may range from mild form of complication up to end stage and life threatening condition.

In-patient with sickle cell disease vaso-occlusion and haemolysis are the main clinical presentation while acute chest syndrome is one of the most serious complications of SCD, with a mortality rate of 10% [2]. Good pain relief not only intraoperatively but also postoperative with thromboembolic prophylaxis, oxygen administration and chest physiotherapy are essential with serial ABG monitoring [2]. Recommendation for elective operation is that patient should be free of crisis and infections to reduce precipitation of crisis [2]. Simple blood transfusion is safer and effective than aggressive blood transfusion or exchange transfusion aiming for preoperative hemoglobin of 10mg/dl [3]. The main stay of managing patient as general include well rehydration, proper oxygenation and avoidance of hypoxia, avoidance of acidosis, hypothermia and vascular stasis with good monitoring of patient [3].

Renal manifestation of SCD range from only renal impairment up to end stage renal diseases with shrunken kidney. Survival time after developing of ESRD in patient with SCD was 4 years leading to death time of 27 years old as the median age of ESRD was 23 years old. [4] ESRD account for 16%-18% of mortality. [5] Management of patient with sickle cell nephropathy aiming for complications prevention and reduction of morbidity. Preoperative preparation and optimization benefit all patient regardless their risk and goal directed therapy is used to improved tissue oxygen delivery for high-risk patient while dialysis is usually scheduled about 12–24 hours prior to surgery. [6] Patient with sickle cell anemia need well rehydration, however this may precipitate heart failure or pulmonary edema in-patient with ESRD. Thus cautious during fluids administration is very important which should be guided by CVP if possible.

Clinical picture of dilated cardiomyopathy may vary from only cardiomegaly to severe congestive heart failure (CHF). Dysfunction of cardiac muscle result in valvular lesion specially aortic and mitral regurgitation while arrhythmias and embolism are (systemic or pulmonary) also common. [7] Preoperative risk stratification of those patient using up-to-date American heart association (AHA) and American college of cardiology (ACC) guidelines is of valuable importance. Mortality is increased when the ejection fraction is less than 40% however, heart failure has a great risk even

with preserved ejection fraction.^[8] During anesthesia for patient with cardiomyopathy the main goals are to void tachycardia, minimize the effects of negative inotropic agents, especially anaesthetic drugs, prevent increase in afterload, and maintain adequate preloading in addition to maintenance of sinus rhythm and ^[9]. Intra and postoperative inotropes can be required including dobutamine, dopamine, phosphodiesterase inhibitors, and levosimendin but dopamine with its positive inotropic, chronotropic and vasoconstrictive effects may be the drug of choice ^[7-9].

While anesthetic management of patient with DCM is difficult, this will be of high morbidity and mortality when combined with severe pulmonary hypertension. Pulmonary hypertension affecting approximately 20-40% of patient with SCD, which necessitate good awareness of anesthesiologist about the cause itself, and severity with avoidance of drugs which may increase hypertension like nitrous oxide and at the same time avoidance of hypoxia and acidosis.^[10]

The case was rather challenging in regards to the patient's Sickle cell anemia and the risk of crisis as well as management of associated dilated cardiomyopathy with its risk of ischemia and heart failure. End stage renal disease was another challenge, especially for fluids management and problem of anuria, which may needs early dialysis. A lot of measures were taken in the CCU for pain management, sedation and mechanical ventilation of the patient during the first 24 hours besides fluids therapy, antibiotic, and anticoagulants protocol.

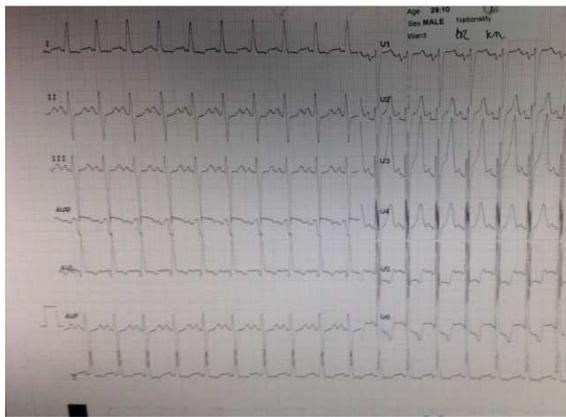


Figure 1: Patient ECG at presentation

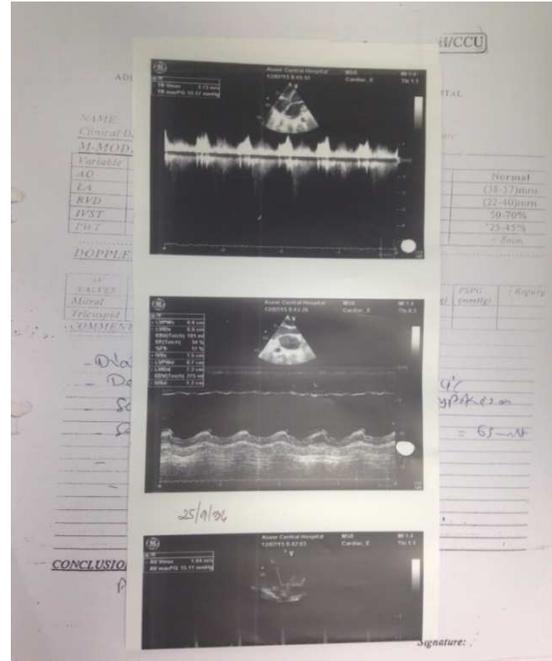


Figure 2: Echo at presentation.

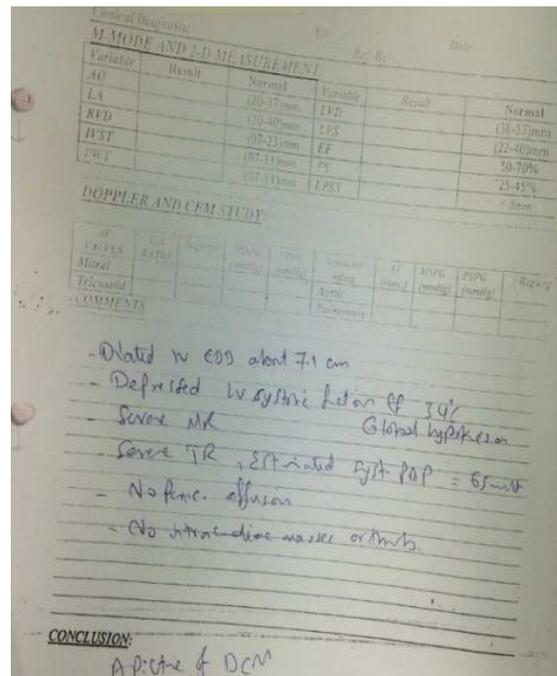


Figure 3: Echo report.

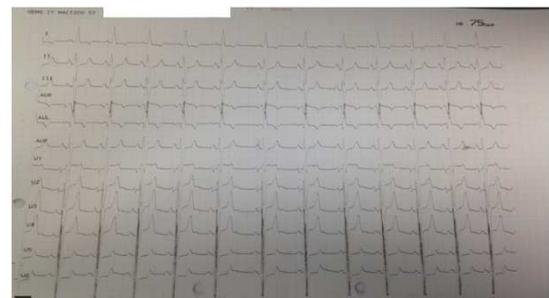


Figure 4: Patient ECG after dialysis and blood transfusion.

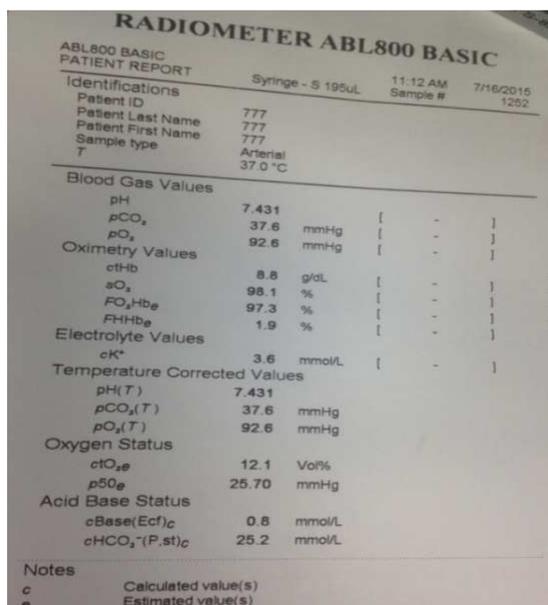


Figure 5: Acid-base analysis report.

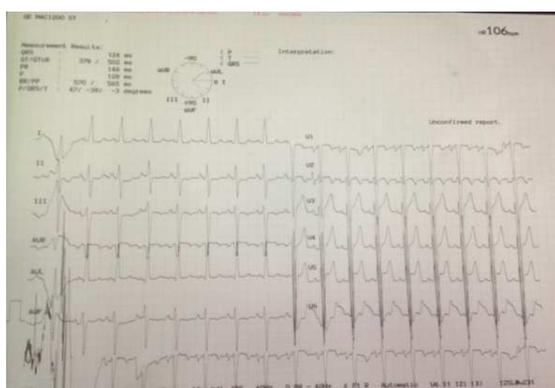


Figure 6: Patient ECG postoperatively and just after initiation of mechanical ventilation in CCU..

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CONCLUSION

Sickle Cell Anemia associated with Dilated Cardiomyopathy, End Stage Kidney Diseases and Pulmonary Hypertension puts the patients at high risk, and possess a great deal of challenge to anesthesia with high risk of morbidities and mortality in the perioperative period. For those patients proper preoperative assessment and evaluation with a Multi-Disciplinary team will lead to optimization of management and facilitate a proper anesthetic plan and will ensure tight detection and early managements of anticipated intra and post anaesthesia complications.

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