

Bariatric Surgery Impact on Sickle Cell Disease Pain Crisis: A Case Report

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Abstract

Sickle cell disease (SCD) is an autosomal genetic blood disorder resulting in multiple end-organ complications. Malnutrition is a stress factor that can cause a SCD crisis. Bariatric surgery is a weight reduction surgery that involves the binding or removing a part of the stomach or resecting and re-routing the small intestine to a small stomach pouch. It is known to cause malnutrition and stress. Malnutrition affects more than two billion people of all ages worldwide due to different causes. Long-term deficiency of micronutrients leads to reduced immunity, leukopenia, and diseases affecting the psychological, skeletal, and central nervous system. We here present the case of a 20-year-old woman with SCD and class III obesity. She underwent sleeve gastrectomy in 2018 following psychological distress caused by being severely overweight. She had mild SCD pain, but after the bariatric surgery, it became severe, requiring morphine treatment and monthly exchange transfusion beside the NSAID which become not much effective as before bariatric surgery. Our findings show that bariatric surgery, which leads to a stressful condition, can aggravate the SCD pain crisis, thereby highlighting the need for alternative methods of weight reduction in these patients. Controlled studies are required for the proper assessment of bariatric surgery in SCD.

Keywords

Sickle Cell Disease, Bariatric Surgery, Pain Crisis

1. Introduction

Sickle cell disease (SCD) is an autosomal genetic blood disorder leading to severe anemia and impaired tissue perfusion, resulting in multiple end-organ compli-

cations. Pain is the most common complication of SCD. Some SCD patients have mild to moderate pain, while others have severe vaso-occlusive complications, which lead to a pain crisis. Poor hydration increases blood viscosity. Even mild dehydration can have a significant impact on hemoglobin concentration [1]. Stress factors that affect the SCD pain crisis are fever, loss of fluids due to excessive sweating, vomiting, diarrhea, malnutrition, high temperature, and humidity. The renal complications in SCD can impact the urine concentration resulting in higher urine loss and dehydration due to tubular damage [2].

Bariatric surgery (BS) is a weight reduction surgery that involves binding or removing part of the stomach or resecting and re-routing the small intestine to a stomach pouch. The United States National Institutes of Health recommends BS for individuals with a body mass index (BMI) ≥ 40 and those with a BMI of ≥ 35 with comorbidities such as diabetes and other obesity-related severe disorders [3]. All forms of BS reduce food intake and the quality of micronutrients absorption. Following BS, appropriate incorporation of vitamins and minerals can help maintain normal metabolic functions, energy production, hormone synthesis, and decrease the risk of nutrient-related health problems [4]. In bodyweight calculations, a BMI of <18.5 falls within the underweight range, between 18.5 and <25 is the normal range, between 25.0 and <30 is the overweight range, and <30.0 falls within the obese range. Obesity is frequently subdivided into categories: Class 1: BMI of 30 to <35 , Class 2: BMI of 35 to <40 , Class 3: BMI of 40 or higher. Class 3 obesity is sometimes categorized as “extreme” or “severe” obesity [5].

Micronutrient deficiencies cause several diseases while exacerbating many others and affect more than two billion people of all ages. It is associated with 10% of all deaths in children and child welfare. Lack of one or several micronutrients is caused by its inadequate intake, malabsorption, increased requirements, and drug-nutrient interactions. In about 70% of the children and 40% of the adults, these deficiencies cause complications [6]. In many cases, the response to a nutrient deficiency also seems to be genotype-specific. Gene-nutrient interactions are thus a fascinating example of physiological reactions to the environment and diet at the molecular level [7]. Long-term deficiency of micronutrients could cause many disorders, including reduced immunity and leukopenia. Gastrointestinal, central nervous system, psychological, skeletal, and reproductive systems are most affected by these deficiencies [8]. This study aimed to highlight that bariatric surgery is not suitable for all morbidity obese people, and more diseases should be added to the contraindications list of BS.

2. Case Study

The patient was a 20-year-old woman with SCD. She weighed 104 kilograms, was 156 cm tall, and had a BMI of 42. She had neither hypertension nor any metabolic and other chronic disorders. The indication for this surgery was the severe psychological distress from being grade III overweight, and the failure to

reduce weight with many other measures. Although she had SCD, she was never under follow up with a hematologist as she only had mild chronic SCD pain, which was controlled with nonsteroidal anti-inflammatory drugs (NSAIDs). She underwent lap sleeve gastrectomy in 2018, after she signed a consent form. On two weeks post the BS, she was admitted with vomiting and abdominal pain; subsequently, she required several admissions because of dehydration and severe body pain. The SCD pain crisis became severe, requiring morphine to control the pain. She was given intravenous fluids on several occasions. Lately, she has been on a monthly exchange transfusion to control the SCD pain and to reduce the morphine requirement. Her lab findings before and after the BS are as follows:

Before the BS, complete blood count (CBC) on 29-08-2017: White blood cells (WBCs): $17.5 (4 - 10) \times 10^9/L$, red blood cells (RBC): $3.89 (3.8 - 4.8) \times 10^{12}/L$, hemoglobin: 111 (120 - 150) g/L, hematocrit: 0.384 (0.36 - 0.46) L/L, platelet count: $523 (150 - 410) \times 10^9/L$, MCV: 90 (83 - 101) fl, MCH: 29 (27 - 32) pg, MCHC: 348 (315 - 345) g/L, and reticulocytes: 11.6 (0.5 - 2.5)%. Coagulation profile: INR: 1.35 and APTT: 35.2 (27 - 38.3) sec. Serum ferritin: 95.1 (11 - 306) ng/L. Hb electrophoresis: Hb A = 0%, Hb F = 26.1% Hb A2 = 1.3%, and Hb S = 72.6%. G6PD: normal. Liver function test: Total protein: 73 (61 - 79) g/L, albumin: 40 (35 - 48) g/L, total bilirubin: 82 (3 - 25) $\mu\text{mol}/L$, direct bilirubin: 26.2 (1.7 - 8.6) $\mu\text{mol}/L$, LDH: 528 IU/L, ALT: 79 (10 - 60) IU/L, AST: 85 (10 - 42) IU/L, glucose: 5.0 (3.9 - 6.1) mmol/L, and Ca: 2.17 (2.1 - 2.6) mmol/L. Renal function test: BUN: 0.9 (2.5 - 7.1) mmol/L, creatinine: 56 (26 - 77) $\mu\text{mol}/L$, Na: 138 (134 - 144) mmol/L, K: 4.03 (3.6 - 5.1) mmol/L, alkaline phosphatase: 142 (42 - 98) IU/L, corrected Ca: 2.17 mmol/L.

Abdominal ultrasound before the BS was not available.

Eighteen months after the BS, her body weight, height and BMI were 65 kg, 155 cm, and 26.7, respectively. Other lab findings were as follows:

WBC: $12.2 (3.8 - 4.8) \times 10^{12}/L$, RBC: $3.08 \times 10^{12}/L$, hemoglobin: 93 (120 - 150) g/L, hematocrit: 0.271 (0.36 - 0.46) L/L, platelet count: $322 (150 - 410) \times 10^9/L$, MCV: 88 (83 - 101) fl, MCH: 30 (27 - 32) pg, MCHC: 343 (315 - 345) g/L, and reticulocytes: 6.67 (0.5 - 2.5)%. Coagulation profile: INR: 1.22 and APTT: 30.7 (27 - 38.3) sec. Serum ferritin: 269.5 ng/L (11 - 306) ng/L.

Liver function test: Total protein: 58 (61 - 79) g/L, albumin: 33 (35 - 48) g/L, total bilirubin: 91 (3 - 25) $\mu\text{mol}/L$, direct bilirubin: 10.6 (1.7 - 8.6) $\mu\text{mol}/L$, ALT: 17 (10 - 60) IU/L, AST: 36 (10 - 42) IU/L, glucose: 4.6 (3.9 - 6.1) mmol/L, and Ca: 2.08 (2.1 - 2.6) mmol/L.

Renal function test: BUN: $<0.4 (2.5 - 7.1) \text{ mmol}/L$, creatinine: 48 (26 - 77) $\mu\text{mol}/L$, Na: 142 (134 - 144) mmol/L, K: 3.35 (3.6 - 5.1) mmol/L, alkaline phosphatase: 67 (42 - 98) IU/L, and corrected Ca: 2.22 mmol/L.

An abdominal ultrasound after the BS revealed mild hepatomegaly, mild splenomegaly, and cholecystectomy. The enlarged spleen indicated suspected old splenic infarctions due to SCD. There were no indications of intra-abdominal abscess or collections (**Figure 1**).

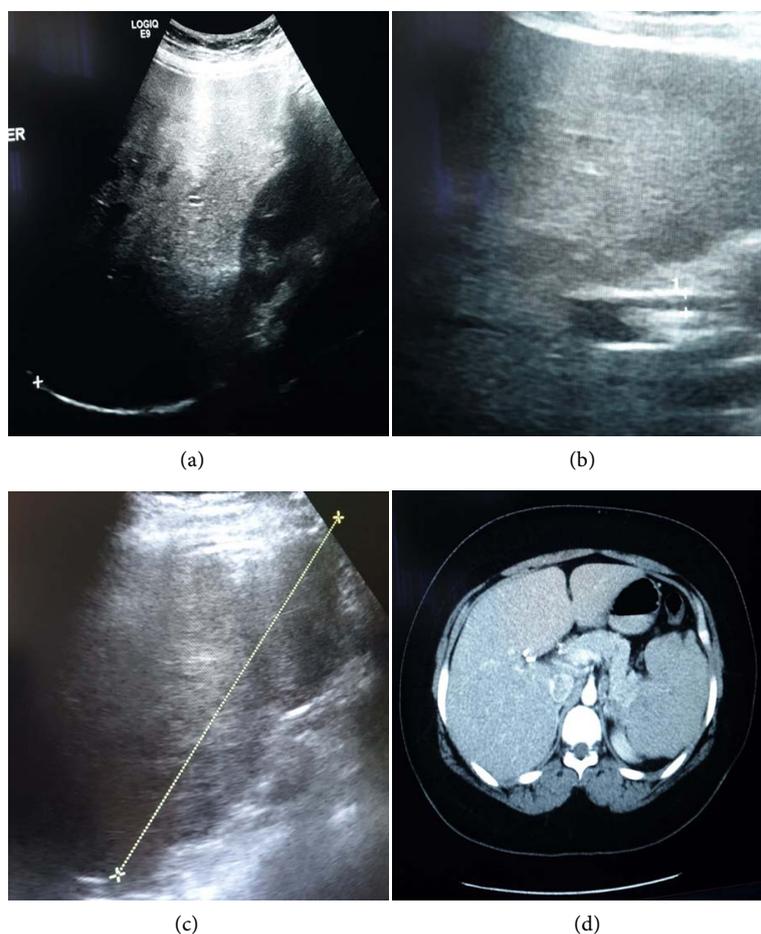


Figure 1. Abdominal ultrasound & CT. (a) Mildly enlarged bright liver measuring about 20 cm in the mid-clavicular craniocaudal dimension; (b) Removed gall bladder with normal diameter of CBD measuring about 5 mm; (c) Enlarged spleen measuring about 14 cm in craniocaudal dimension with irregular outline due to old infarcts; (d) Axial CT image showing hepatosplenomegaly surgical clips of cholecystectomy irregular outline of the.

The patient currently weighs 65 kg, but she has frequent episodes of severe SCD pain, which require morphine and monthly exchange transfusion.

3. Discussion

A SCD patient is a fragile person who needs to stay well hydrated and away from any stress, including psychological, to avoid a painful vaso-occlusive crisis. Common complaints following BS include lethargy, easy fatigue, headaches, and muscle wasting. Reduced food intake is a stressor and contributes to SCD pain and other SCD-related complications [9]. BS deprives the body of water, which increases the blood viscosity resulting in more frequent and severe episodes of SCD crisis.

The aberrant migration of potassium and chloride ions across the RBC membrane causes RBC dehydration, which consequently increases the tendency of hemoglobin to polymerize and sickle [10]. After BS, patients usually develop

dumping syndrome, characterized by abdominal pain, nausea, vomiting, diarrhea, and diaphoresis [11]. A few months or few years after BS, the patients become deficient in micronutrients and trace elements including water, fat-soluble elements such as iron, calcium, phosphorus, copper, magnesium, selenium, chromium, and zinc [12], as well as vitamins such as thiamine, riboflavin, niacin, pantothenic acid, pyridoxine, biotin, folate, cobalamin and vitamins B, D, C, E, K. These nutrients and elements work as enzymatic cofactors in several metabolic processes and biochemical reactions. Therefore, BS can cause a wide range of nutritional, systemic, and psychiatric complications [13]. Arginine and L-glutamine, which are essential micronutrients in SCD, can get severely depleted and influence both the pain intensity and general health of the SCD patients [14]. Proteins are key regulators of metabolism and DNA replication. They also aid in digestion as well as combating infections by strengthening the immune system [15]. The specific risk for developing micronutrient and protein deficiencies cannot be predicted as there is no consensus on the amount of vitamin and mineral supplementation required after BS. However, it is clear that micronutrient deficiencies are relatively common in patients after all types of BS, and so, it is important to screen them before and periodically after the surgery for micronutrients and trace elements [12]. For any type of surgery, SCD patients need special hematological recommendations for maintaining adequate blood level, oxygen saturation, and proper hydration before and after the surgery [16]. Surgical clearance is required from a hematologist before the surgery to avoid any unexpected complications. Pre-operative assessment must include a careful review of the patient's known crisis triggers, baseline hematological profile, transfusion requirements, pre-existing organ dysfunctions, pain degree, and opioid consumption.

4. Conclusion

Compared to healthy individuals, those with SCD have a much lower tolerance for stress. Bariatric surgery puts these patients in a severely stressful condition. This case is significant for both areas of hematology and bariatric surgery. In this study, the SCD pain crisis worsened following the sleeve gastrectomy surgery, indicating that SCD could be a contraindication for such surgery. This case emphasized that bariatric surgery is not suitable for all obese overweight people. Controlled studies are required to assess and define the guideline to improve the outcomes of bariatric surgery, which is now a standard procedure in many countries.

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Conflicts of Interest

The authors have no conflict of interest in this study.

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