DOI: 10.14260/jemds/2015/2019

ORIGINAL ARTICLE

VARIOUS SURGICAL PRESENTATIONS AND THEIR OUTCOME IN SICKLE CELL DISEASE: A PROSPECTIVE STUDY

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HOW TO CITE THIS ARTICLE:
Manju Singh, Sandeep Chandrakar, Amit Agrawal, Gambhir Singh. "Various Surgical Presentations and their Outcome in Sickle Cell Disease: A Prospective Study". Journal of Evolution of Medical and Dental Sciences 2015; Vol. 4, Issue 81, October 08; Page: 14207-14212, DOI: 10.14260/jemds/2015/2019

ABSTRACT: Sickle cell disease is a common genetic disorder which represents major medical problem in certain parts of world. In India Chhattisgarh is one of the states in which sickle cell disease is common. Purpose of this study was to evaluate the patients of sickle cell disease with various surgical problems and their outcome. A prospective study of 100 patients with diagnosed sickle cell disease was done in the department of General Surgery of Pt. JNM medical college and attached hospital. Maximum patients with surgical presentation were above 40 yrs. of age (48%) with male preponderance. Most common presenting symptom were generalized weakness and abdominal pain (69%). Important clinical sign was pallor 85%, splenomegaly 12%, vasoocclusive crisis in 53%, hemolytic crisis in 4% sequestration crisis in 5% and aplastic crisis seen in 1%. Various surgical presentations were acute appendicitis 14% cholelithiasis 15%, hollow viscous perforation 13% and various other presentations. 63 patients underwent surgical treatment and 37 were managed conservatively. Post operatively delayed wound healing was the common complication 52% due to wound infection 51%. Mortality was 5%. Although causes are multifactorial but patients of sickle cell disease are more prone to develop intra and post-operative complications so it is important for a surgeon to understand the pathophysiology and management of sickle cell disease to reduce the morbidity and mortality of these patients with surgical presentations.

KEYWORDS: Sickle Cell Disease, Vaso Occlusive Crisis, Hemolytic Crisis, Surgical Presentation.

INTRODUCTION: Sickle cell disorder refers to the state in which the red cells become sickle shaped in deoxygenated environment. It is a genetic disorder and occurs in homozygous state of hemoglobin–S gene, hemoglobin-SC, hemoglobin-SD, and hemoglobin–S beta thalassemia states.[1]

The first reported case in the literature was in 1910 by Herrick in a dental student at Chicago whose blood smear contained pear shaped elongated red blood cells.[2]

It is characterized by hemolytic anaemia and vasoocclusive crisis which can lead to wide spread vascular occlusion by sickled red blood cells leading to multiple organ infarction causing various clinical condition such as peptic ulcer, ischemic colitis, appendicitis, cholecystitis, splenic infarct, splenic abscess.[3,4,5,6,7]

The disease prevalence is quite variable but it is estimated that 8% of black people in America and 40% in certain countries of tropical Africa have the sickle cell trait.[8] Sickle cell disease is prevalent in many parts of India where it may range from 9.4% to 22.2% in endemic areas.[9] A high incidence of sickle cell disease is found among schedule caste and tribe population of Chhattisgarh region.[10]

Surgical management in these patients is associated with high morbidity and mortality. Close collaboration between medical and surgical specialties is essential. Precautions to guard against factors predisposing to vasoocclusive crisis will greatly contribute to reduction in morbidity and mortality associated with surgery in this high risk group.

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OBJECTIVE OF STUDY:

1. To study various surgical presentation in sickle cell disease.
2. To study the outcome of surgical patients with sickle cell disease.

MATERIAL AND METHODS: This prospective study was conducted from September 2009 to September 2011 in 100 patients of sickle cell disease diagnosed by history and by investigations, who attended the surgical OPD and admitted in Dr. BRAM Hospital, Pt. JNM Medical College Raipur. Patients of all age group of both sex with sickle cell disease and surgical problem were included in this study. Clinical evaluation as well as related hematological, biochemical and radiological investigations were carried out in all patients. This study was conducted according to the principle established and approved by ethical committee of Pt. JNM Medical College Raipur Chhattisgarh.

OBSERVATION: Out of 100 cases studied 63 were admitted because of complaints of sickle cell disease with surgical problem and 37 were admitted because of surgical condition and were diagnosed sickling positive. In this study 64% patients were male and 36% were female with male: female Ratio of 1.77:1. The most common age group presenting with surgical presentation was above 40yrs (48%).In this study 45% were of Sahu community which is also prevalent community in CG, 15% Kurmi, 7% Gonds, 11% Satnami, 2% Mahars, 9% Yadav, 10% Muslims and Oriya were 1%.

Most common symptoms seen in this study were generalized weakness and pain in abdomen 69% and 67%, as shown in Table–1. Most common clinical sign seen in the study is pallor 85% followed by tenderness over abdomen48%. Table–2.

Vasoocclusive crisis was present in 53% of which abdominal pain was common manifestation, hemolytic crisis was present in 4%, Acute splenic sequestration seen in 5%, it was associated with sudden fall in hemoglobin with increase in spleen size, Aplastic crisis was seen in 1%.

Various surgical presentations of patient with sickle cell disease is shown in Table–3. In this acute appendicitis, hollow viscous perforation and cholelithiasis were the common presentation.

In this study 63 patients underwent surgical intervention and 37 patients managed conservatively. Preoperatively 18% patient received blood transfusion.

Most common postoperative complication in the study was delayed wound healing 82.54% as a result of wound infection 80.95%, chest infection 28.57% and UTI in 19.04%

There were total 5 death in our study. One patient died of abdominal crisis with prepyloric perforation, two patient were of cholelithiasis with cholecystites with septicaemia. One patient was of rectal perforation and one of diabetic foot with septicaemia. 95 patient were discharged after complete recovery from surgical pathology.

Symptoms	No. of Patients	Percentage (%)
Abdominal pain	67	67
Abdominal distension	30	30
Vomiting	44	44
Not Passing Flatus and Motion	20	20
Hematuria	7	7
Fever	58	58
Chest pain	7	7

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Cough	12	12
Breathlessness	3	3
Palpitation	4	4
Discoloration of digits	4	4
Generalized weakness	69	69
Body pain	35	35
Lump in abdomen	17	17
Ulcer over body	15	15

Table 1: Distribution of symptoms

Signs	No. of Patients	Percentage (%)
Pallor	85	85
Icterus	3	3
Edema	12	12
Ascitis	10	10
Signs of dehydration	30	30
Generalized lymphadenopathy	9	9
Abdominal tenderness	48	48
Abdominal Guarding	29	29
Abdominal Rigidity	2	2
Hepatomegaly	8	8
Splenomegaly	12	12
Lung signs	18	18

Table 2: Distribution of clinical signs

Diagnosis of Patient	No. of Patient	Percentage (%)
Acute Appendicitis without lump	8	8
Acute appendicitis with lump	4	4
Acute appendicitis with abscess	1	1
o/c/o Acute appendicitis with fecal fistula	1	1
Cholelithiasis	8	8
Cholelithiasis with Cholicystitis	6	6
Cholelithiasis with cholidocholithiasis	1	1
Peptic ulcer	1	1
Gastric perforation	2	2
Duodinal perforation	1	6
Prepyloric perforation	6	1
Ileal Perforation	3	3
Rectal perforation	1	1
Intestinal obstruction	3	3
Carcinoma stomach	4	4

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Carcinoma Gall- bladder	2	2
Cholangiocarcinoma and Chronic pancreatitis	1	1
EHPVO and Hypersplenism	1	1
Hypersplenism	2	2
Sickle cell trait with splenomegaly	1	1
Hepatic hemangioma, mild splenomegaly, UTI	1	1
Liver abscess	3	3
Cellulitis	8	8
Non- healing ulcer	2	2
B/L Inguinal Hernia	1	1
PVH	3	3
Multiple Renal calculi	1	1
Ureteric calculi	1	1
B/L Hydroureteric nephrosis	1	1
RCC	2	2
Carcinoma of Penis	1	1
Hypospadias	1	1
Gluteal abscess	2	2
Thigh abscess	2	2
Breast abscess	2	2
Psoas abscess	1	1
Buerger's disease	4	4
Fournier's gangrene	1	1
Pyothorax	3	3
Lymphoma	1	1
Hemorrhoids	2	2

Table 3: Various Surgical presentations

DISCUSSION: In our study the most common age group with surgical presentation was above 40 yrs. (48%). Diagnosis of sickle cell disease is usually established in childhood but patients do not develop symptoms until the onset of puberty, pregnancy or early adult life.[11] Male preponderance seen in our study is similar to previous study in our area; this is because males are more prone to known precipitating factor and there is a gender bias in the community. Distribution of maximum number of sickle cell disease was found in sahu community which signifies the distribution of sickle cell gene in various communities and its prevalence throws light on migration and ethnicity. With the advent of DNA finger printing technology the links between various groups could be better established.

This will also point genetic difference in certain ethnic group.[12,13] Pain abdomen is one of the most common complaint as reported by various studies was also the common complaint in our study.[14,15] Hepatomegaly and spleenomegaly as abdominal sign was comparable to other study.[14] Various surgical presentation in study group includes commonly acute appendicitis, hollow viscous perforation, cholelithiasis with cholecystites, and leg ulcer signifies vasoocclusive and hemolytic crisis in sickle cell disease which is responsible for various surgical presentation in this disease.[16,17]

Sickle cell disease is not a single factor responsible for all surgical pathology as all these presentations have multifactorial etiology so further study is needed for any conclusion.

Post-operative complication, particularly delayed wound healing is quite higher in the study as these patients are usually have anaemia, and hypersplenism at the time of presentation. Chest complication are also common in these patient. So these patients require proper anaemia correction, oxygenation and folic acid supplementation before, during and after surgery for better outcome as we have done in our study.[18,19,20]

There were total five deaths in our study which were mainly associated with systemic illness along with abdominal crisis and acute chest infection. Careful preoperative evaluation, intraoperative vigilance and scrupulous postoperative management can reduce morbidity and mortality.

CONCLUSION: In sickle cell disease with surgical presentation it is important for surgeon to understand the pathophysiology and management of this high risk disease. Awareness of various surgical manifestations with adequate precaution to guard against factors predisposing to vasoocclusive crisis will greatly contribute to the reduction in mortality and morbidity associated with surgery in sickle cell disease.

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DOI: 10.14260/jemds/2015/2019

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FINANCIAL OR OTHER COMPETING INTERESTS: None

Date of Submission: 15/09/2015.
Date of Peer Review: 18/09/2015.
Date of Acceptance: 29/09/2015.
Date of Publishing: 08/10/2015.