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Incidence of Cholecystectomy and Splenectomy in Sickle Cell Patient in VIMSAR Burla –A Prospective Study

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Introduction

Sickle Cell disease (SCD) is a disabling condition usually arising from inheritance, carries a turbulent lifespan with occult or overt manifestations of its complications. These complications may at times calls for an emergency management of its complications. Vaso occlusive crisis in sickle cell anemia is responsive for majority of surgical complications like chronic leg ulcer, priapism, osteomyelitis, splenomegaly, acute abdominal pain, cholecystitis. With these pathology the sickle cell disease patients present before the surgeons, it can affect any part of the body & one of the most common and early organ to be affected in SCD is Spleen .Spleen can over functions resulted in fall in all blood component[1], in its milder and usually more chronic form as hypersleenism. Spleenectomy is done for varities of reasons including acute spleenic sequestration crisis hyperspleenism, Spleenic abscess[2,5-8]. Spleenectomy may be done either open & laparascopic method[3,4]. Lap. splneectomy is safe & effective[10]. There is also quantitative improvement in blood indices such as Hb%, Plt. count, mcv etc in post spleenectomy [9] & with improvement of immunization & penicillin prophylaxis ,the relative risk from removal of spleen have probably decreased[11] .it is commonly in larged in 1st decade of life. spleenectomy in SCD patient with hyperspleenism to be beneficial in decreasing their transfusion requirement & discomfort from mechanical pressure of in larged spleen

Cholecystectomy is very common in patient with SCD in both symptomatic and asymptomatic gall stone disease.SCD is the most important cause of cholelithiasis in children[12]. pigment gall stones affects 15% of children with SCD in younger than 10 years of age & more than 80% of those older than 30 years[13]. Early diagnosis & appropriate treatment increase the survival rate, improve the quality of life of patient in SCD[14,15,16]., the surgical approaches used are laparotomy, laparoscopy, most auther considering later the safer for patient with less complications related to surgery, shorter operative time & faster the post operative recovery time, shorter hospital stay [17,18,19,20,21]. major post operative copmplication described as atelectesis, pneunonitis, pulmonary infection, infection.[15,22].

Materials and Method

- All the patients admitted to the General Surgery wards of VSSIMSAR, Burla over a period of 2 year from October, 2014 to October 2016 were screened by sickling test, by the simple cover slip method.
- Confirmatory test done by Electrophoresis or Liquid Chromatography. Those cases which were having sickling positive, were taken as the study group in a prospective trial
- Study Design: Hospital based clinical study.
- Source of Data: All the indoor patients who have sickle cell disease with gall stones and splenomegaly and other complications.
- Study Period: Nov 2014 to Oct 2016.
- Inclusion Criteria
  1. Sickle cell disease patient with gall stones and splenomegaly and other complications presenting to the OPD of VSSIMSAR, Burla
  2. Patients giving consent for investigation and operation.
- Exclusion Criteria
  1. Patient not giving consent
  2. Patients having gall stones and splenomegaly without sickle cell disease.

Aims and Objectives

- To know the actual incidence of gall stone diseases & splenomegaly in patients of sickle cell disease in VSSIMSAR, Burla.
- To analyze the predisposing factor developing for above complications in sickle cell patients.
- To study the aetiopathogenesis and management protocol with aim to evaluate different operative procedure and post op morbidity and mortality of patients of sickle cell disease.

Hbs and Genetics

- The conventional designation of α_2 β_2 indicates normal adult haemoglobin. Replacement of Glutamic acid at the sixth position of the β chain by valine leads to severe conformational change in the molecule of haemoglobin, known as haemoglobin-S.
- Sickle cell disease is determined by homozygosity of a mendalian gene, and sickle cell trait is heterozygous.

The Pathology of Sickle Cell Anaemia

- Reduced oxygen tension
- Sickle cell increased viscosity stasis occlusion of vessels.
- Sensitivity to trauma
- Haemolysis

Clinical Types of Sickle Crisis

- Four clinical types of sickle crisis have been noted which may occur either singly or in combination. In their order of frequency these are as follows:
- Vaso-occlusive crisis
- Hemolytic crisis
- Sequestration syndrome
- Aplastic or hypoplastic crisis.

Result and Discussion

The present study comprised of 60 cases of sickle cell disease admitted to General Surgery and wards of VSSIMSAR, Burla from Nov. 2014 to Oct. 2016.

The observations and discussions in this thesis works dealt with

- Incidence
- Surgical complications
- Management and results
- Follow up.

Table – 1 General Incidence

Total Nos. of cases admitted to the Surgery ward during the period of study	Total no. of cases with sickle cell disease	Percentage
10500	60	0.57%

Table – 2 Showing Age incidence

Age group in years	Number of Cases	Percentage
1-10	12	20%
11-20	25	41.6%
21-30	16	26.6%
31-40	5	8.3%
41-50	2	3.3%
>50	0	0
Total	60	100

Table – 3 Showing Sex Incidence

Sex	No. of cases	Percentage
Male	36	60
Female	24	40
Total	60	100

Table – 4 Incidence of Surgical Complications in different states of sickle cell disease

State of sickle cell disease	No. of cases	Percentage
Steady state	57	95%
Crisis	3	5%
Total	60	100

Table – 5 Gall bladder and Billary System Incidence of cholelithiasis in sickle cell anaemia

Authors	No. of cases	Age range	No. of cholelithiasis cases	Percentage	Diagnostic method
Diggs (1934)	18	-	3	16.7	Autopsy / X-ray
Green et al (1953	15	>12 years	5	33.0	X-ray/ Autopsy
Mintz (1955)	21	3 to 16 years	2	9.5	X-ray
Jordan (1957)	27	>14 years	10	37.0	X-ray, Surgery, Autopsy
Cameron et al (1971)	147	16-47	33	22.0	X-ray, Autopsy/ laparoscopy
Present series	60	5 to 45 (4.5)	15	25.0	USG/ Surgery

Observation

Age

Age incidence of 15 sickling positive cases with clinical radiological or operative evidence of Biliary tract disease.

Table – 6

Age group	Number of Patients	Percentage
5-10	3	20%
11-20	6	40%
21-30	4	27%
>30	2	13%

In the following study about nearly half the patients were of 20 years of age.

Table – 7 Sex

No. of patients having cholelithiasis	No. of males	No. of Females	Ratio (M:F)
15	11	4	3.1

All though the incidence of cholelithiasis is more seen in females, but in this series male were more affected than females.

Symptoms

The following table shows the symptoms presented by sickle cell patients with biliary disease.

Table – 8

Symptoms	No. of Patients	Percentage
Jaundice	7	46%
Abdominal pain	8	53%
Indigestion	3	20%

Operative management

These patients were managed surgically and cholecystectomy was done in 10 patients. Multiple small pigment stones were present inside the gall bladder. Rest patients were managed conservatively. One patient with cholelithiasis with CBD dilation was referred to higher centre for ERCP and management.

The patients were followed up to six months after operation. Post operative complication was present in only one patient who had presented with bleeding from the drain site which was controlled successfully.

Splenomegally

Table – 9 Age incidence of splenomegally in this present series

Age group	No. of patients	Percentage
Upto 10 years	12	20%
11 to 18 years	7	11.6%
19yrs and above	11	18.3%
All age groups included	30	50%

The youngest patient was 5 years old and the eldest was 30 yrs old. Splenomegaly in sickle cell disease was seen almost in each decade of life equally.

Table – 10 Incidence of Splenomegaly

Authors	Total No. of cases studied	Age group	Splenomegaly	Percentage
Grove (1947)	47	All age group	19	40.4
Watson et al (1956)	115	All age group	21	18.2
Serjent (1969)	89	Adults	17	20
Present series	60	All age group	30	50

In this series sickle cell anaemia cases an incidence of 50% was seen which is slightly more than the incidence of 40.4% as studied by Grover (1947). The incidence of splenomegaly in only adult population was seen to be 18.3% which is compatible with that of Serjeant (1969). But Watson et al (1956) reported the lowest incidence of splenomegaly in his series, which was not in accordance with the observations of others including my own observation.

Management

The criteria of operation for doing a splenectomy was recurrent acute splenic sequestration and chronic hypersplenism. In younger age group, the splenectomised patients were more susceptible to infections. Children who underwent splenectomy were given pneumococal vaccine (pneumovax) and antibiotics.

Results: All were relieved of symptoms and did well at the time of discharge.

Follow up: All cases were symptom free at 2nd month of post operative period and splenectomy cases were marked with dramatic clinical improvement. Both Hahn (1928) and London et al (1929) suggested that the best results were seen in patients with large spleens.

Table- 11 Operation performed in sickle cell disease patients during the period of study

Splnenectomy	25
Cholecystectomy	10
Laparotomy for splenic abscess	1

Of the 60 cases, 15 cases were treated conservatively. All other cases underwent different types of operations. There were neither problems nor complications during the administration of general anesthesia. All cases had uneventful recovery.

Table 12 Incidence of different surgical complications in Sickle Cell Disease

Surgical Compliations	No. of cases	Percentage
Chronic leg ulcer	1	1.66%
Gall bladder and billary tract disease	12	20%
Splenomegaly	30	50%
Osteomyelitis	2	3.3%
Aseptic necrosis of Femoral head	9	15%
Chronic duodenal ulcer	2	3.33%
Hepatomegaly	24	40%
Priapism	1	1.66%
Acute abdominal crises	3	5%
Total	60	100%

Summary

- In this present study of Spectrum of surgical complications of sickle cell disease and its management. 60 cases of sickle cell disease were taken into consideration.
- The incidence of surgical complications in sickle cell disease was 0.57%.
- The highest incidence of surgical complications in sickle cell disease occurred in the age group of 11 to 20 years (41.6%)
- The maximum number of cases were males (60%) with a Male:female ratio of 3:2.

- Maximum number of cases presented without the sickle cell crisis (95%).
- The incidence of surgical complications with maximum when the haemoglobin percentage in sickle cell disease patient was in the range of 8gm% to 10gm%.
- Relevant family history was traced in 33.3% of cases.
- The surgical complications of sickle cell disease like priapism chronic leg ulcer, cholecystitis with cholelithiasis, splenomegaly, ostemyelitis, aseptic necrosis of femoral head, chronic duodenal ulcer, acute abdomen were found in the present series in 1.6%, 1.6%, 20%, 50%, 3.3%, 15%, 3.3%, 5% of cases respectively.
- Hepatomegally was found in 40% of cases.

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